

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

It gives us a great pleasure to present our effort during our residency years in summarizing MOST of the ORL-HNS topics from the main textbooks and references in ORL-HNS. We have done our best in organizing the materials in each topic to be easy to understand and memorize.

We based our notes mainly on the following references:

- Bailey's Head and Neck Surgery, 5th Ed.
- Cummings Otolaryngology 6th Ed.
- Pediatric Airway Surgery, Philippe Monnier.
- Head and Neck Surgery, Clinical Reference Guide, Pasha 4th Ed.
- Diseases of ENT, Dhingra 5th Ed.
- Clinics of North America.
- Netter's Head and Neck Anatomy for Dentistry, 2nd Ed.

These Notes are not considered as a substitution to the textbooks, as some of the topics were not covered in these notes. Also, please refer to the above mentioned references in case of any conflict in the provided information, as this is a pure human work and amenable to mistakes. We do recommend to keep adding your own notes to these notes. Moreover, we strongly advise the final-year residents to read from Manitoba Notes and/or Hart Notes as a quick review in preparation for the final board exam.

Finally, we would like to thank all of our colleagues, senior registrars, consultants, and professors who have done a great effort in teaching us throughout the residency years. Also, we wish you all the best in your residency training program and your future career.

Regards,

**Riyadh et al.¹
2016**

1- Bedah, Obaid, Majed, and Feras

ولاتنسونا من خالص دعائكم (:

GENERAL:

- 1- Embryology of Pharyngeal Apparatus**
- 2- Skull and Facial Bones Anatomy**
- 3- Cervical Muscles Anatomy**
- 4- Vessels and Lymphatics Anatomy**
- 5- Cranial Nerves Anatomy**
- 6- Trigeminal Nerve Anatomy**
- 7- Facial Nerve Anatomy**
- 8- Anatomy of Pharynx**
- 9- Physiology of Pharynx**
- 10- Pharyngitis**
- 11- Tonsils**
- 12- Adenoid**
- 13- Velopharyngeal Insufficiency**
- 14- Snoring and Obstructive Sleep Apnea (OSA)**
- 15- Cervical Fascia and Deep Neck Infections**
- 16- Pharyngeal Pouch**
- 17- Esophageal Anatomy and Disorders**
- 18- Angioedema**
- 19- Neck Trauma**

Embryology

- **Head and Neck are formed by:**

1. Paraxial mesoderm
2. Lateral plate mesoderm
3. Neural crest
4. Ectodermal placodes

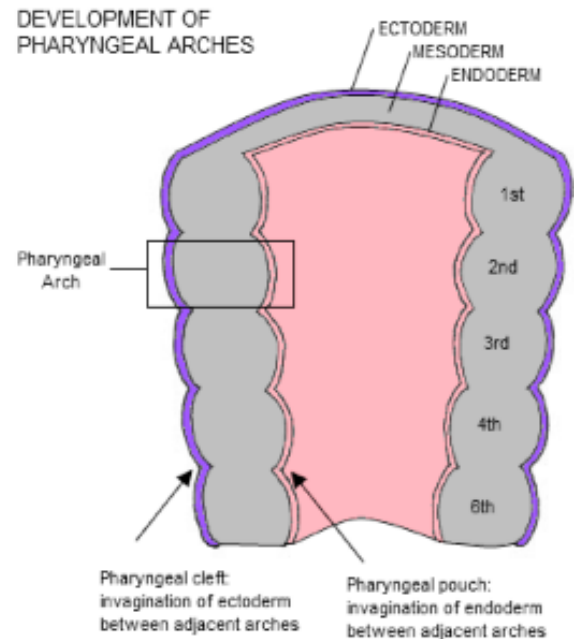
- Pharyngeal (branchial) apparatus appear in embryo by **4th week.**

- It consist of:

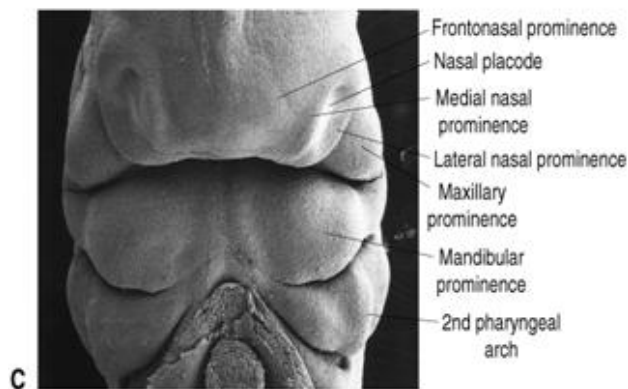
- 1- Pharyngeal Arches (6)
- 2- Pharyngeal Pouches (4)
- 3- Pharyngeal Groove (1)
- 4- Pharyngeal Membrane (1)

Pharyngeal Arches:

- Early in the 4th week, neural crest cells migrate to develop it.
- 1st pharyngeal arch pair (premordium jaws) elevates lateral to develop pharynx.
- Then other arches appear.
- By end of 4th week, 4 pairs of arches are visible externally.
- 5th and 6th pharyngeal arches are rudimentary and not visible.
- Pharyngeal arches separated by fissures (grooves).

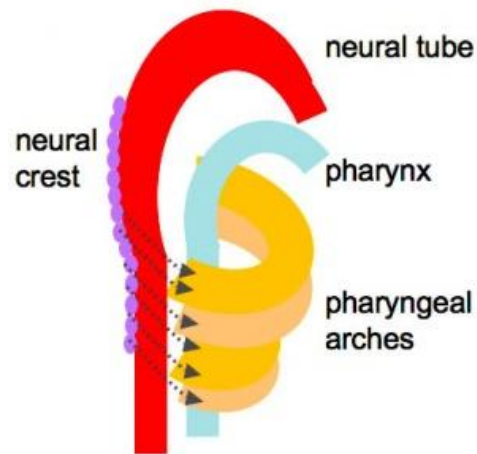


- **1st pharyngeal arch (Mandibular Arch)** Develop 2 prominences:
 - 1- Maxillary prominence.
 - 2- Mandibular prominence.

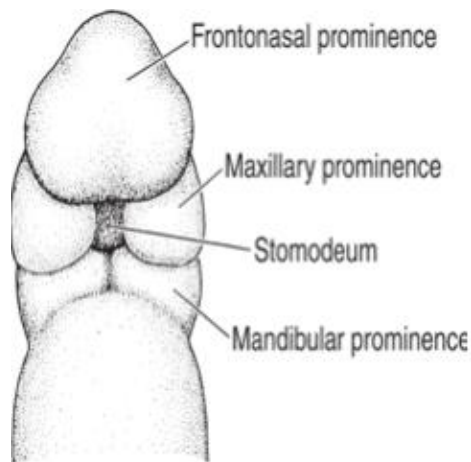


8-6C Face development

- **2nd pharyngeal arches (Hyoid Arch).**
- Arches support lateral wall of primordial pharynx which is derived from cranial part of the foregut.



- Primordial mouth or stomodeum appears as a slight depression of the surface ectoderm.
- It is separated from the cavity of primordial pharynx by a bilaminar membrane, **Oropharyngeal membrane** (composed of ectoderm and endoderm).
- Rupture at about 26 days and communicate pharynx and foregut with amniotic cavity



- **Components of Pharyngeal Arch:**

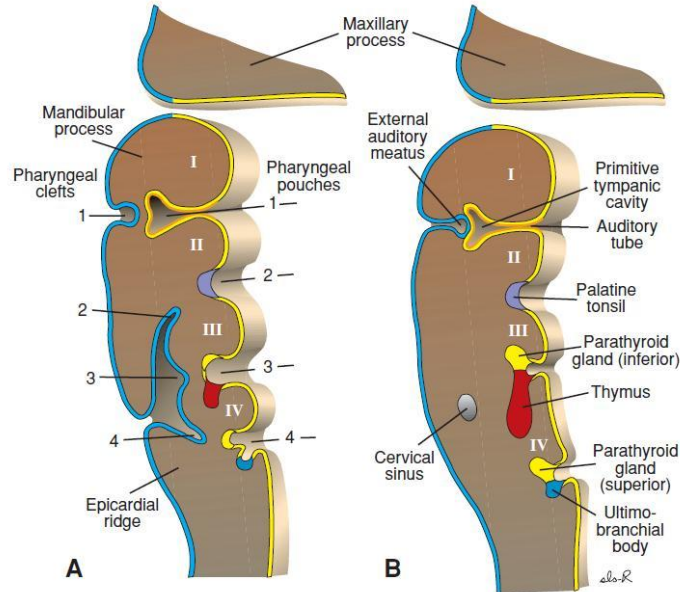
1. Externally ectoderm.
2. Core mesenchyme (embryonic connective tissue)
3. Internally endoderm.

Head and Neck mesenchyme is derived from:

1. Mesoderm (Original Mesenchyme) gives rise to:
 - Skeletal musculature
 - Vascular endothelia.
 2. Cranial neural crest mesenchyme (Ectomesenchyme) Gives rise to:
 - Most Head and neck mesenchyme.
- All the mesenchyme of head and neck derived from neural crest cells **EXCEPT** skeletal musculature and vascular endothelia which derived from original mesenchyme.

Fate of Pharyngeal Arches:

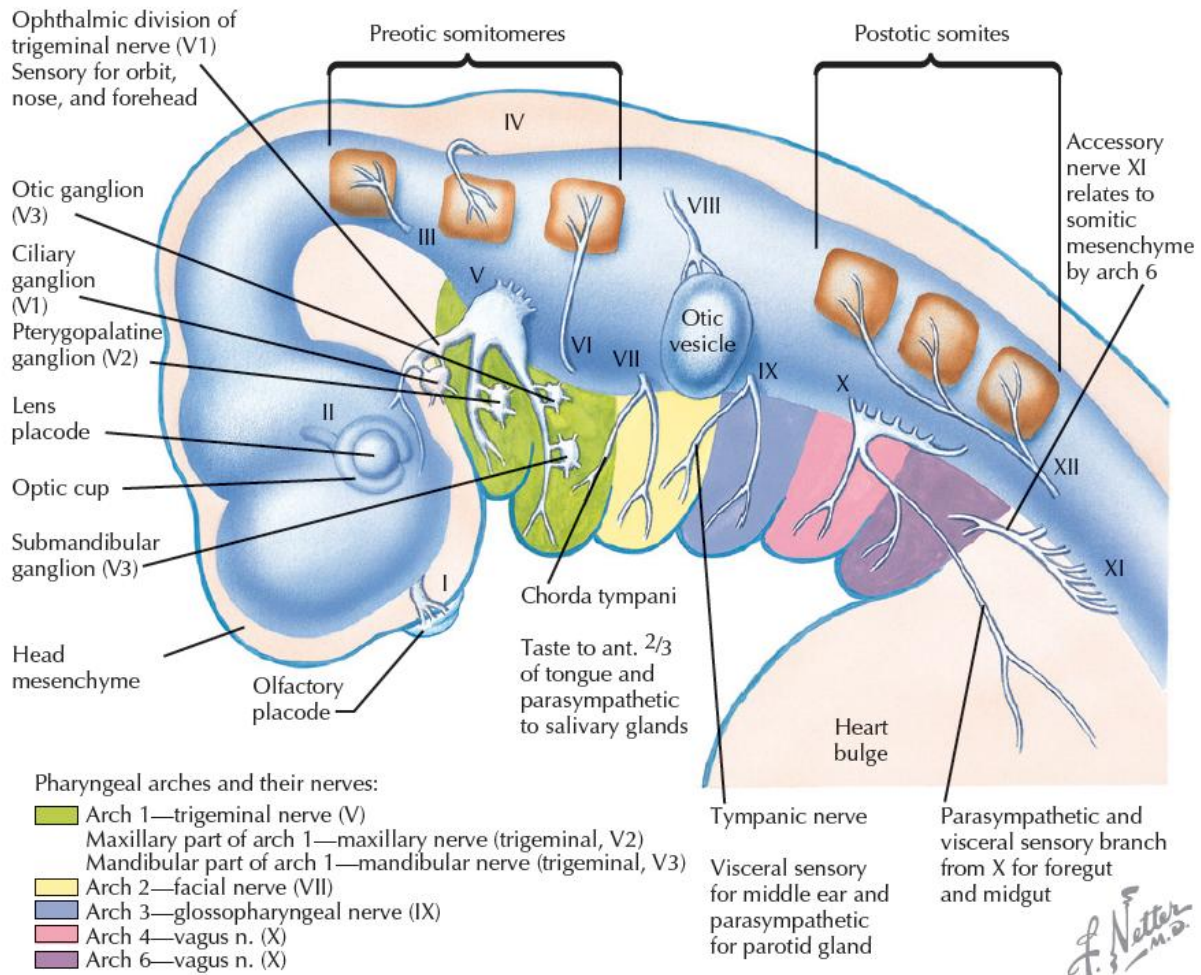
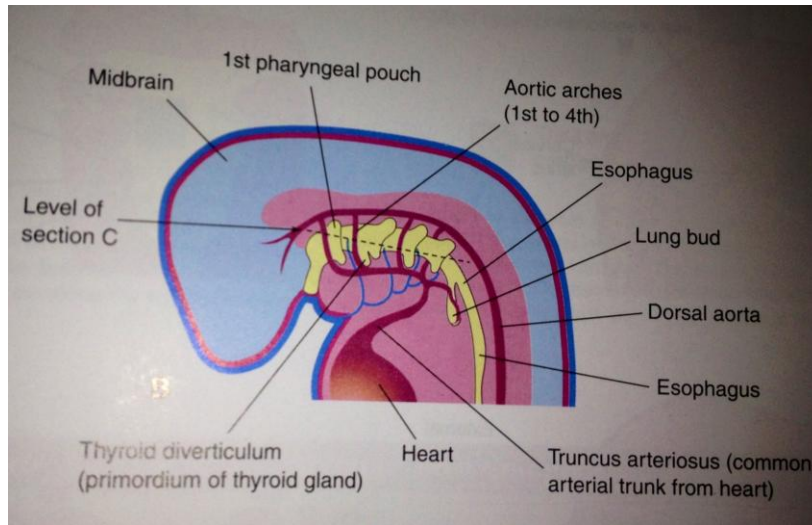
- Face, nasal cavity, mouth, pharynx, larynx and neck.
- **By 5th week:**
 - 2nd pharyngeal Arch Overgrows the 3rd and 4th arches, forming **Cervical sinus**.
- **By the 7th week:**
 - 2nd, 3rd, 4th Grooves + Cervical sinus have disappeared.



- **Typical Pharyngeal Arch contains:**

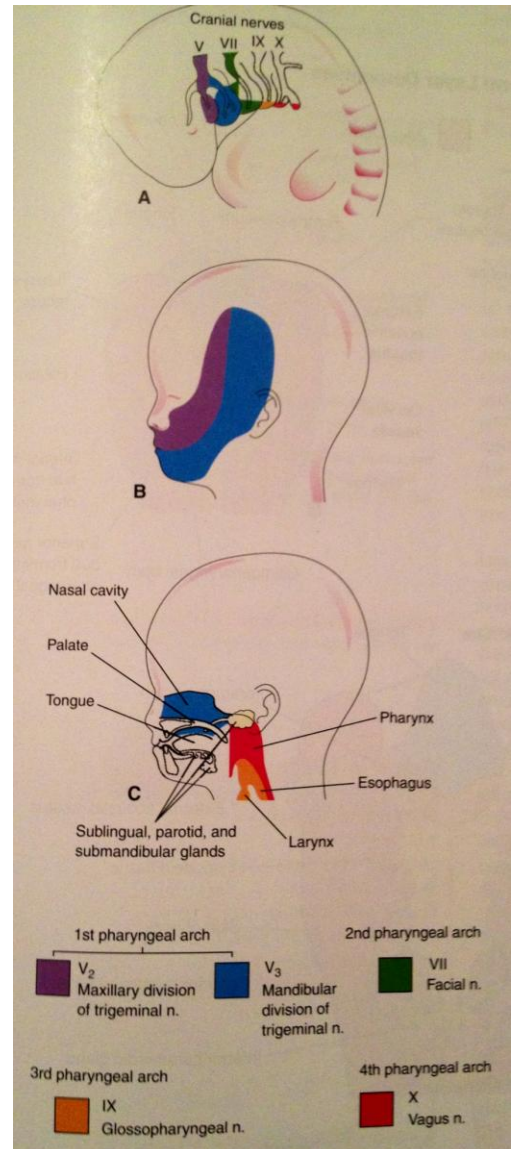
- 1- Nerve.**
- 2- Skeletal Structures.**
- 3- Muscular component .**
- 4- Aortic arch:**

- Arise from Truncus arteriosus pass around pharynx and enter Dorsal aorta



Derivatives of Pharyngeal Arch Nerves:

- **1st Arch:**
 - Trigeminal Nerve (CN-V):
 1. Maxillary Nerve (V₂)
 2. Mandibular Nerve (V₃)
- **2nd Arch:**
 - Facial Nerve (CN-VII)
- **3rd Arch:**
 - Glossopharyngeal Nerve (CN-IX)
- **4th Arch:**
 - Vagus Nerve (CN-X):
 - Superior Laryngeal Nerve
- **5th Arch:**
 - **No Derivatives.**
- **6th Arch:**
 - Vagus Nerve (CN-X):
 - Recurrent Laryngeal Nerve



- **Derivatives of Pharyngeal Arches Skeletal Structures:**

- **1st Arch (Meckel Cartilage):**

1. Malleus.
2. Incus.
3. Squamous portion of temporal bone.
4. Zygoma.
5. Maxilla.
6. Mandible.
7. Anterior ligament of malleus.
8. Sphenomandibular Ligament

- **2nd Arch (Reichert Cartilage):**

1. Stapes supra structures.
2. Styloid Process
3. Lesser cornu of Hyoid.
4. Superior part of body of Hyoid.
5. Stylohyoid ligament

- **3rd Arch:**

1. Greater cornu of Hyoid.
2. Inferior part of body of Hyoid.

- **4th Arch:**

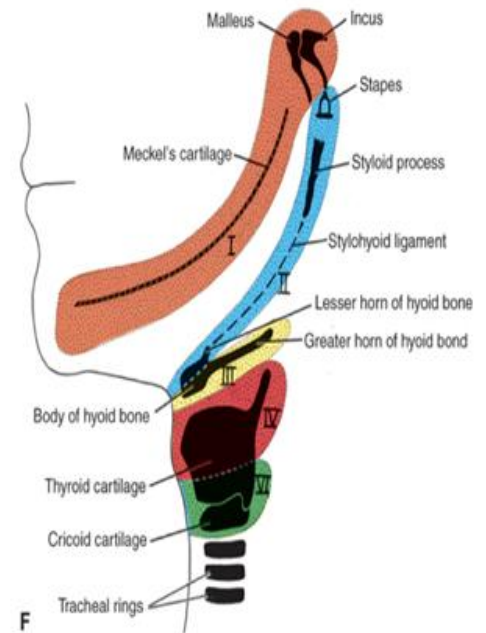
1. Epiglottis.
2. Thyroid Cartilage.

- **5th Arch:**

- **No Derivatives.**

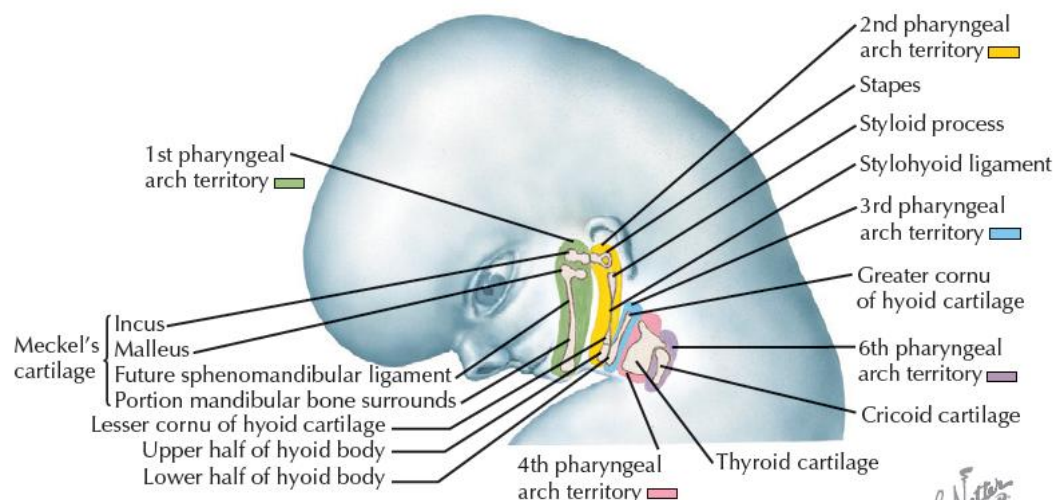
- **6th Arch:**

1. Arytenoid Cartilage.
2. Corniculate Cartilage.
3. Cuneiform Cartilage.
4. Cricoid Cartilage.

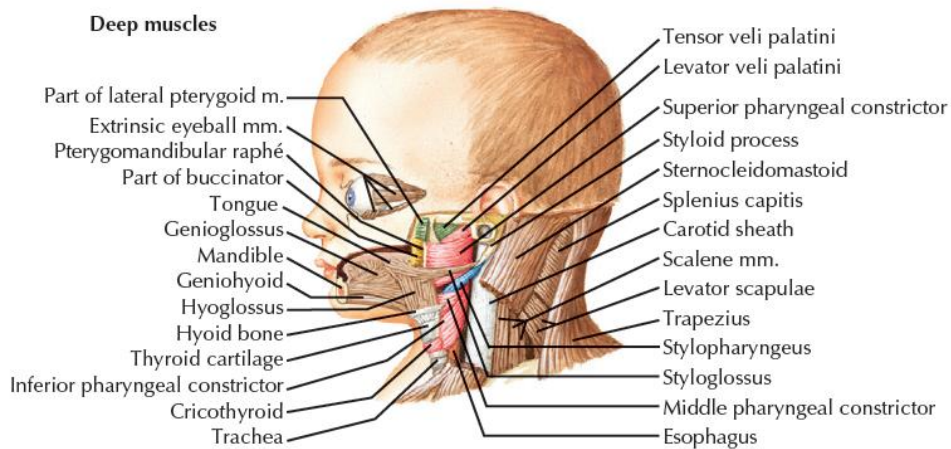
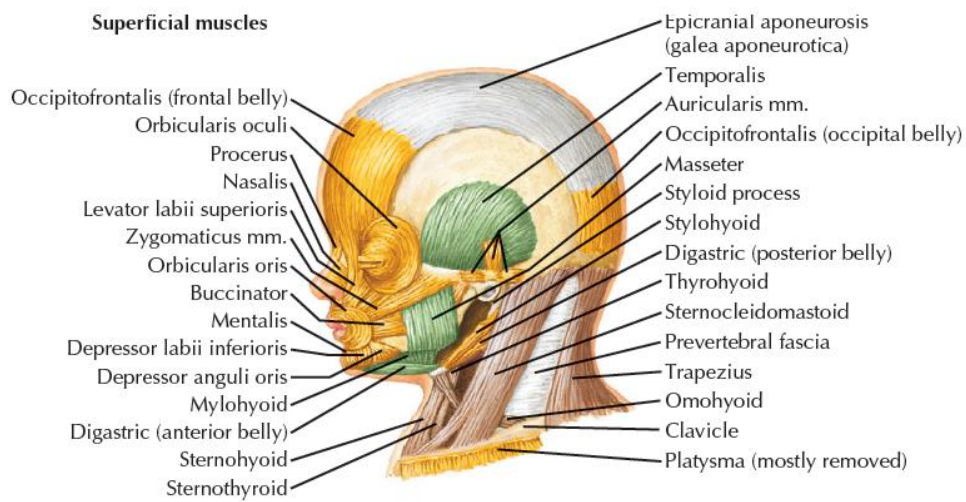
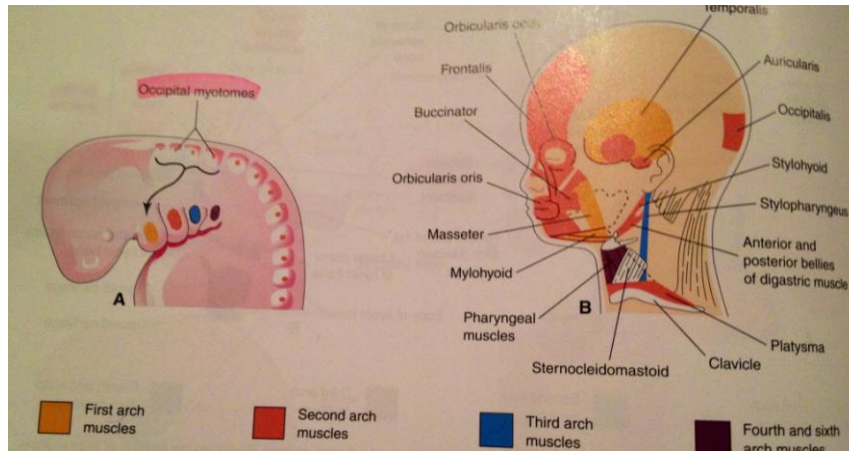


8-1F Pharyngeal arch formation and derivatives

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- **Derivatives of Pharyngeal Arches Muscles**
- **1st Arch: (Trigeminal Nerve)**
 - 1. Muscles of Mastication
 - 1. Temporalis.
 - 2. Masseter.
 - 3. Medial Pterygoid.
 - 4. Lateral Pterygoid.
 - 2. Tensor Tympani.
 - 3. Tensor Veli Palatini.
 - 4. MyloHyoid.
 - 5. Anterior belly of Digastric.
- **2nd Arch: (Facial Nerve)**
 - 1. Muscles of Facial Expression.
 - 2. Stapedius.
 - 3. StyloHyoid.
 - 4. Posterior belly of Digastric.
- **3rd Arch: (Glossopharyngeal Nerve)**
 - 1. StyloPharyngeus muscle.
- **4th Arch: (Superior Laryngeal Nerve)**
 - 1. CricoThyroid (Tensor).
 - 2. All Muscles of Pharynx (**EXCEPT** StyloPharyngeus):
 - 1. Superior Constrictor.
 - 2. Middle Constrictor.
 - 3. Inferior Constrictor.
 - 4. SalpingoPharyngeus.
 - 5. PalatoPharyngeus.
 - 3. All Muscles of Soft Palate (**EXCEPT** Tensor Veli Palatini):
 - 1. Levator Veli Palatini.
 - 2. Musculus uvulae.
 - 3. PalatoGlossus
 - 4. PalatoPharyngeus.
- **5th Arch:**
 - **No Derivatives.**
- **6th Arch: (Recurrent Laryngeal Nerve)**
 - 1. All Intrinsic Muscles of Larynx (**EXCEPT** CricoThyroid):
 - 1. Posteriore CricoArytenoid (Abductor)
 - 2. Lateral CricoArytenoid. (Adductor)
 - 3. InterArytenoid (Adductor)
 - 4. ThyroArytenoid (Adductor)
 - 5. Vocalis (Tensor)
 - 6. ThyroEpiglottis (Opens the laryngeal inlet)
 - 7. AryEpiglottis (Closes the laryngeal inlet)



PHARYNGEAL ARCH BONES AND CARTILAGE

Arch #	Derivatives of Arch Cartilages
1	Malleus, incus, sphenomandibular ligament
2	Stapes, styloid process, stylohyoid ligament, upper half of hyoid
3	Lower half and greater horns of hyoid
4	Thyroid and epiglottic cartilages of larynx
6	Cricoid, arytenoid, and corniculate cartilages of larynx

- **Derivatives of Pharyngeal Arches Arteries (Aortic Arches):**

- **1st Arch:**

- Disappear early in life, but the remnant forms:
1- Maxillary Artery.

- **2nd Arch:**

- 1. Stapedial Artery.
 - Disappear early in fetal life.
 - Originate from ICA.
 - Pass between Stapedial crura.

- **3rd Arch:**

- 1. Internal Carotid Artery.

- **4th Arch:**

- Right:
1- Right Subclavian Artery.
- Left:
1- Arch of Aorta.

- **5th Arch:**

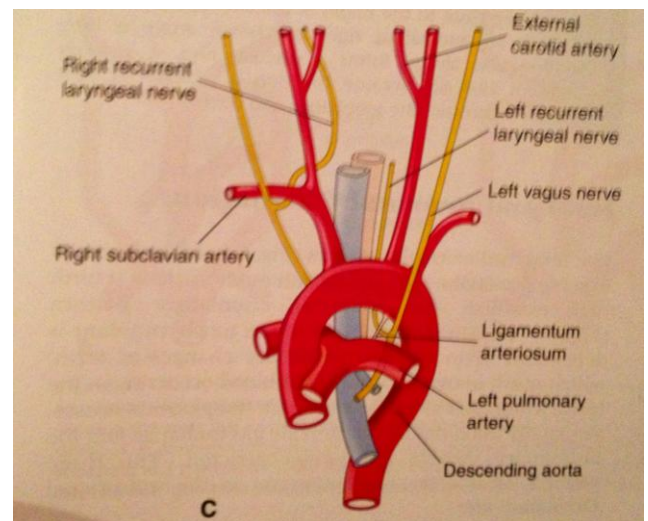
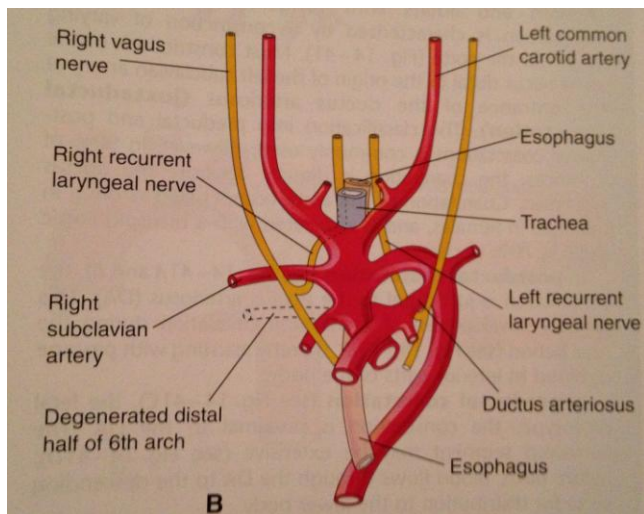
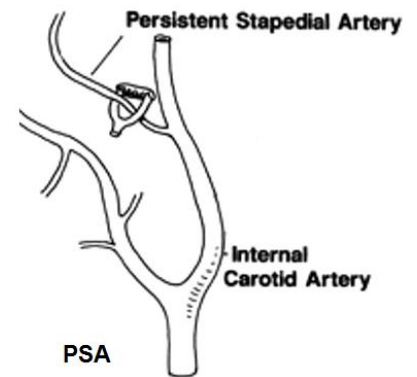
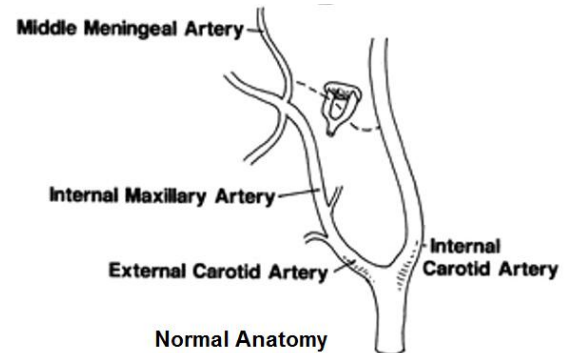
- **No Derivatives.**

- **6th Arch:**

- Right:
 1. Proximal: Right Pulmonary Artery
 2. Distal: **Degenerates**
- Left:
 1. Proximal: Left Pulmonary Artery
 2. Distal: Ductus Arteriosum.

- This explain why the course of the recurrent laryngeal nerves differ on the two sides:

- Right side: hooks around Right subclavian artery
- Left side: hooks around Ligamentum Arteriosum (Remenant of Ductus Arteriosum).



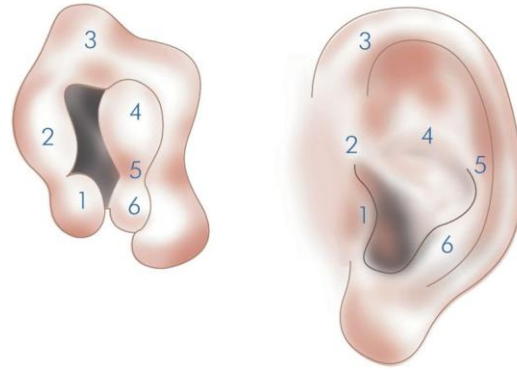
- **Hillock of His Embryology:**

- **1st Arch:**

1. Tragus
2. Helical crus
3. Helix

- **2nd Arch:**

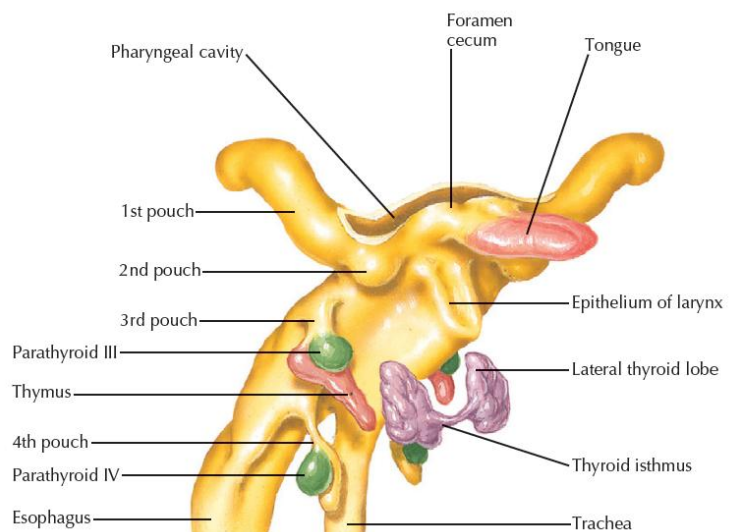
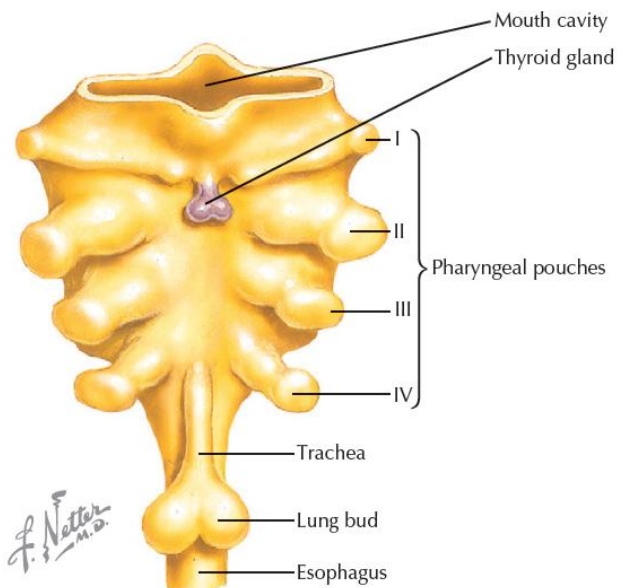
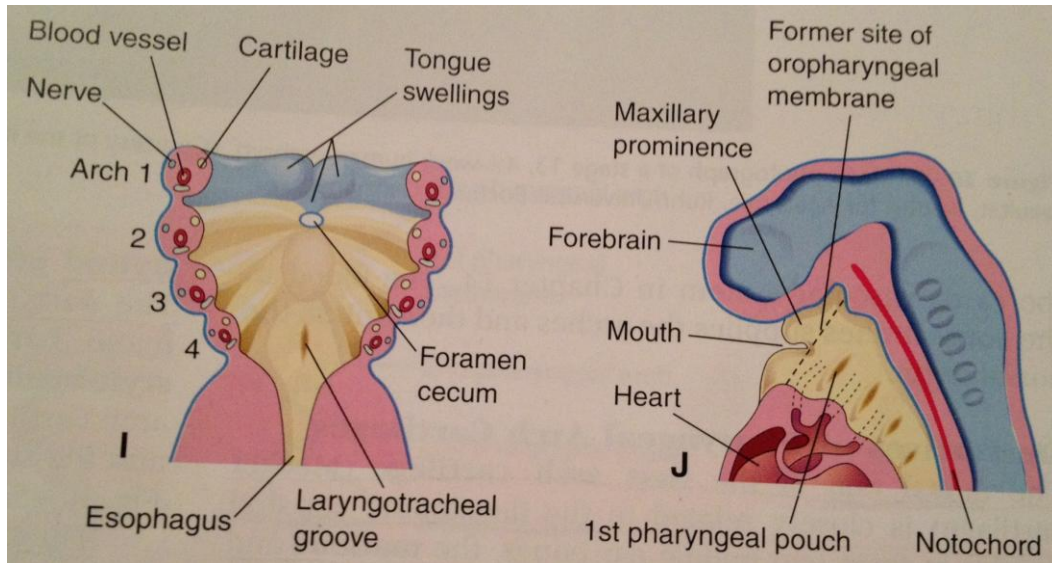
1. Anti-Helix.
2. Scapha.
3. Lobule



Pharyngeal Arch	Nerve	Muscles	Skeleton
1. Mandibular	V. Trigeminal: mandibular divisions	Mastication (temporal; masseter; medial, lateral pterygoids); mylohyoid; anterior belly of digastric; tensor palatine, tensor tympani	Premaxilla, maxilla, zygomatic bone, part of temporal bone, Meckel's cartilage, mandible, malleus, incus, anterior ligament of malleus, sphenomandibular ligament
2. Hyoid	VII. Facial	Facial expression (buccinator; auricularis; frontalis; platysma etc); posterior belly of digastric; stylohyoid; stapedius	Stapes; styloid process; stylohyoid ligament; lesser horn and upper portion of body of hyoid bone
3	IX. Glossopharyngeal	Stylopharyngeus	Greater horn and lower portion of body of hyoid
4-6	X. Vagus Superior laryngeal branch (nerve to fourth arch) Recurrent laryngeal branch (nerve to sixth arch)	Cricothyroid; levator palatine; constrictors of pharynx Intrinsic muscles of larynx	Laryngeal cartilages (thyroid cricoid, arytenoid, corniculate, cuneiform)

Arch	Muscles from Mesoderm	Skeletal Structures from Neural Crest	Cartilage Structures	Connective Tissue Structures	Nerve
1 Develops into: • Maxillary process • Mandibular process	Masseter Temporalis Lateral pterygoid Medial pterygoid Mylohyoid Anterior digastric Tensor tympani Tensor veli palatini	Maxilla Temporal (squamous portion) Zygoma Mandible Malleus Incus	Meckel's cartilage (degenerates in adulthood)	Sphenomandibular ligament Anterior ligament of the malleus	Trigeminal
2	Muscles of facial expression Posterior digastric Stylohyoid Stapedius	Lesser cornu of the hyoid Superior part of the hyoid body Styloid process Stapes	Reichert's cartilage	Stylohyoid ligament Connective tissue of the tonsil	Facial
3	Stylopharyngeus	Greater cornu of the hyoid Inferior part of the hyoid body		Connective tissue of the thymus and inferior parathyroid	Glossopharyngeal
4	Musculus uvulae Levator veli palatini Palatopharyngeus Palatoglossus Superior constrictor Middle constrictor Inferior constrictor Salpingopharyngeus Cricothyroid		Thyroid (from lateral plate mesoderm) Epiglottis	Connective tissue of the superior parathyroid and the thyroid	Vagus
6	Thyroarytenoid Vocalis Lateral cricoarytenoid Oblique arytenoids Transverse arytenoids Posterior cricoarytenoid Aryepiglottis Thyroepiglottis		Arytenoid Cricoid Cuneiform Corniculate (from lateral plate mesoderm)		Vagus

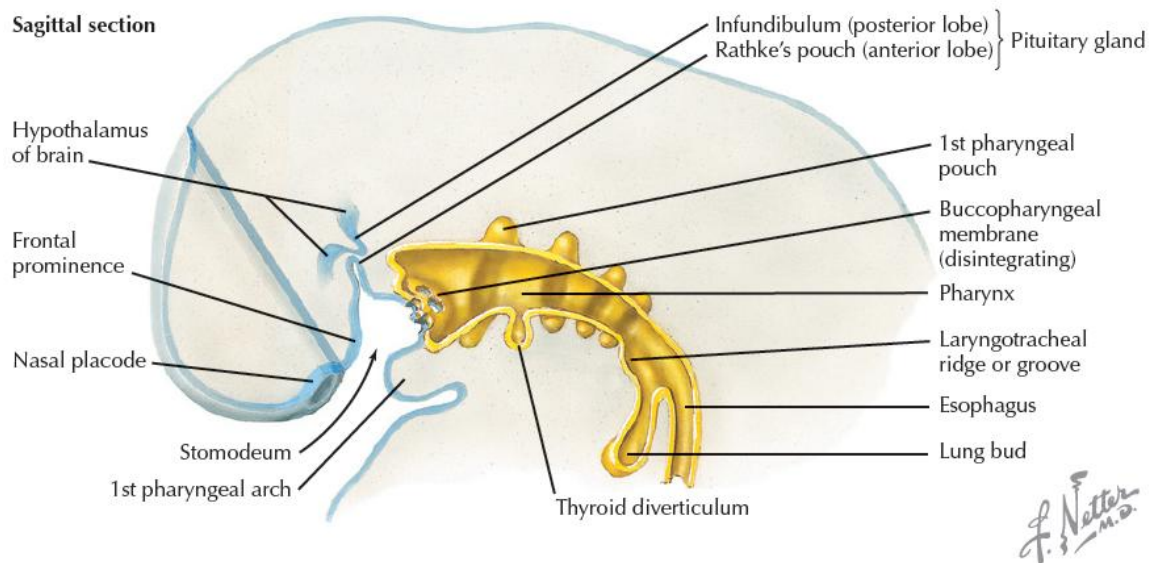
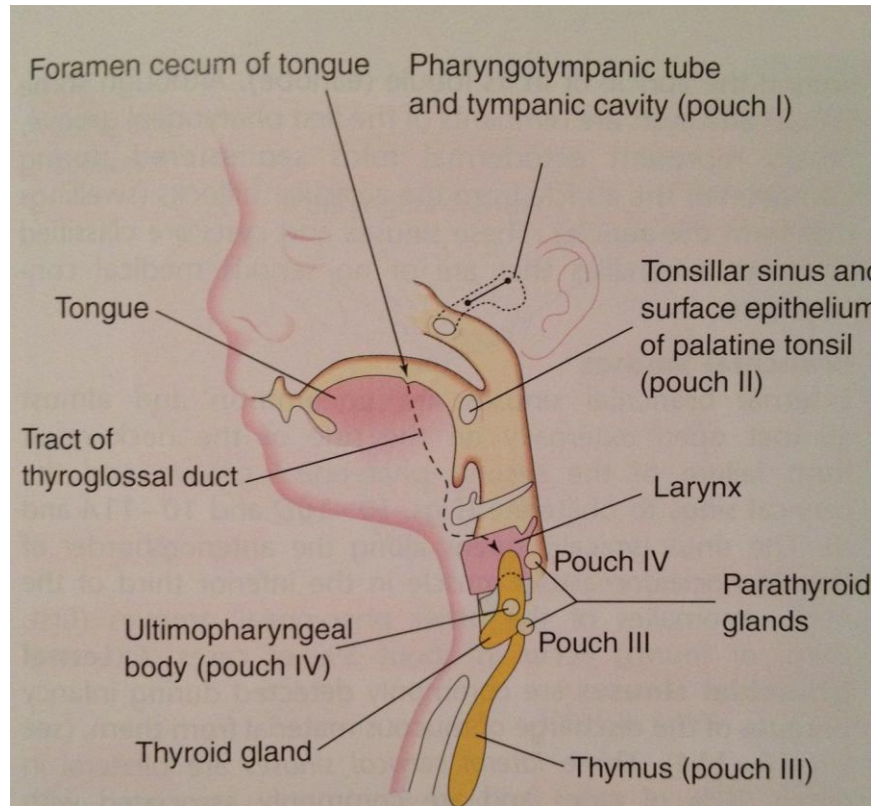
- **Pharyngeal Pouches:**
- Primordial Pharynx derived from **Foregut.**
- Widens cranially and joins Primordial mouth (Stomodeum)
- Narrows caudally and joins Esophagus.
- Endoderm of pharynx lines internal aspect of Pharyngeal Arches
- Pharyngeal Pouches forms from Endodermal invagination.



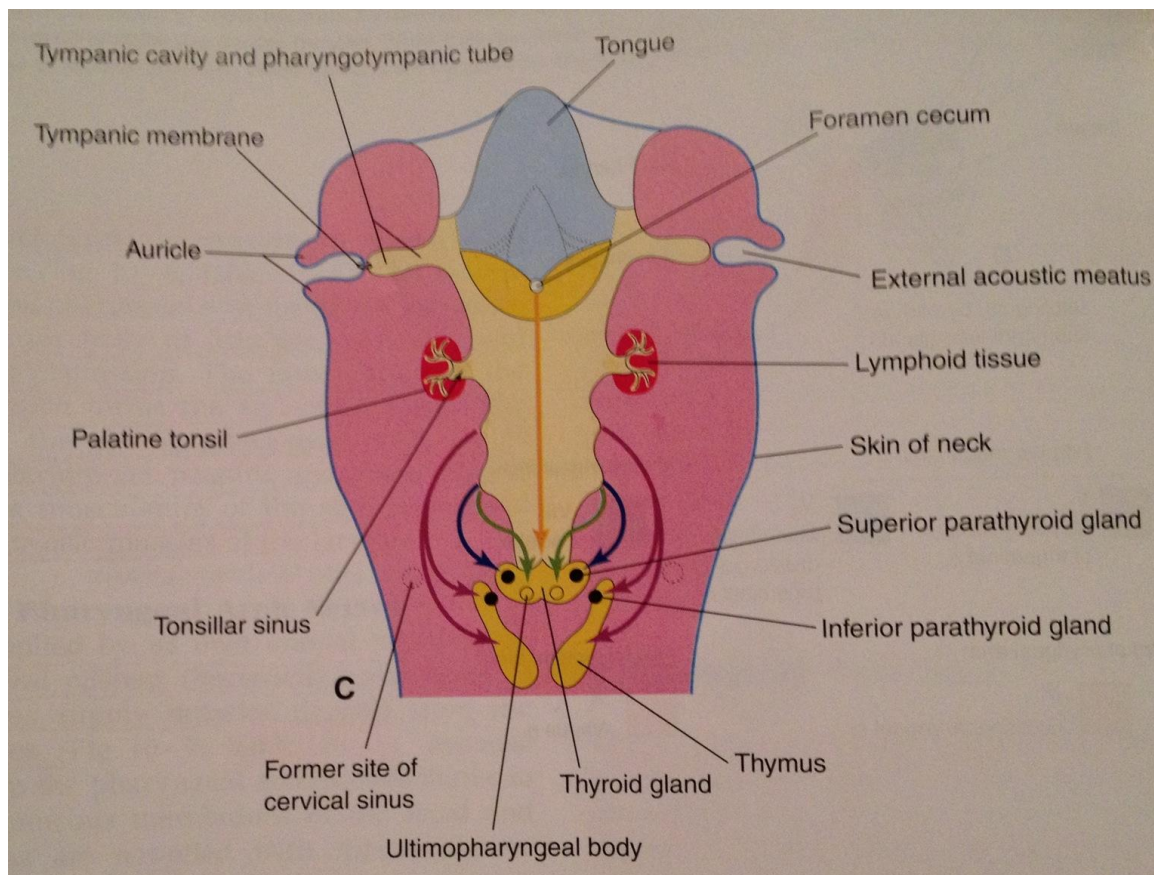
- **Pharyngeal Pouches:**
- **1st Pouch:**
 1. Tubotympanic Recess:
 1. Tympanic Cavity.
 2. Mastoid Antrum.
 2. Eustachian Tube.
- **2nd Pouch:**
 1. Tonsillar Fossa.
 2. Epithelium of Tonsils
- **3rd Pouch:**
 1. Ventral Part:
 1. Thymus.
 2. Dorsal Part:
 2. Inferior Parathyroid glands.
- **4th Pouch:**
 1. Ventral Part:
 1. Ultimopharyngeal Body → Parafollicular Cells.
 2. Dorsal Part:
 2. Superior Parathyroid glands.

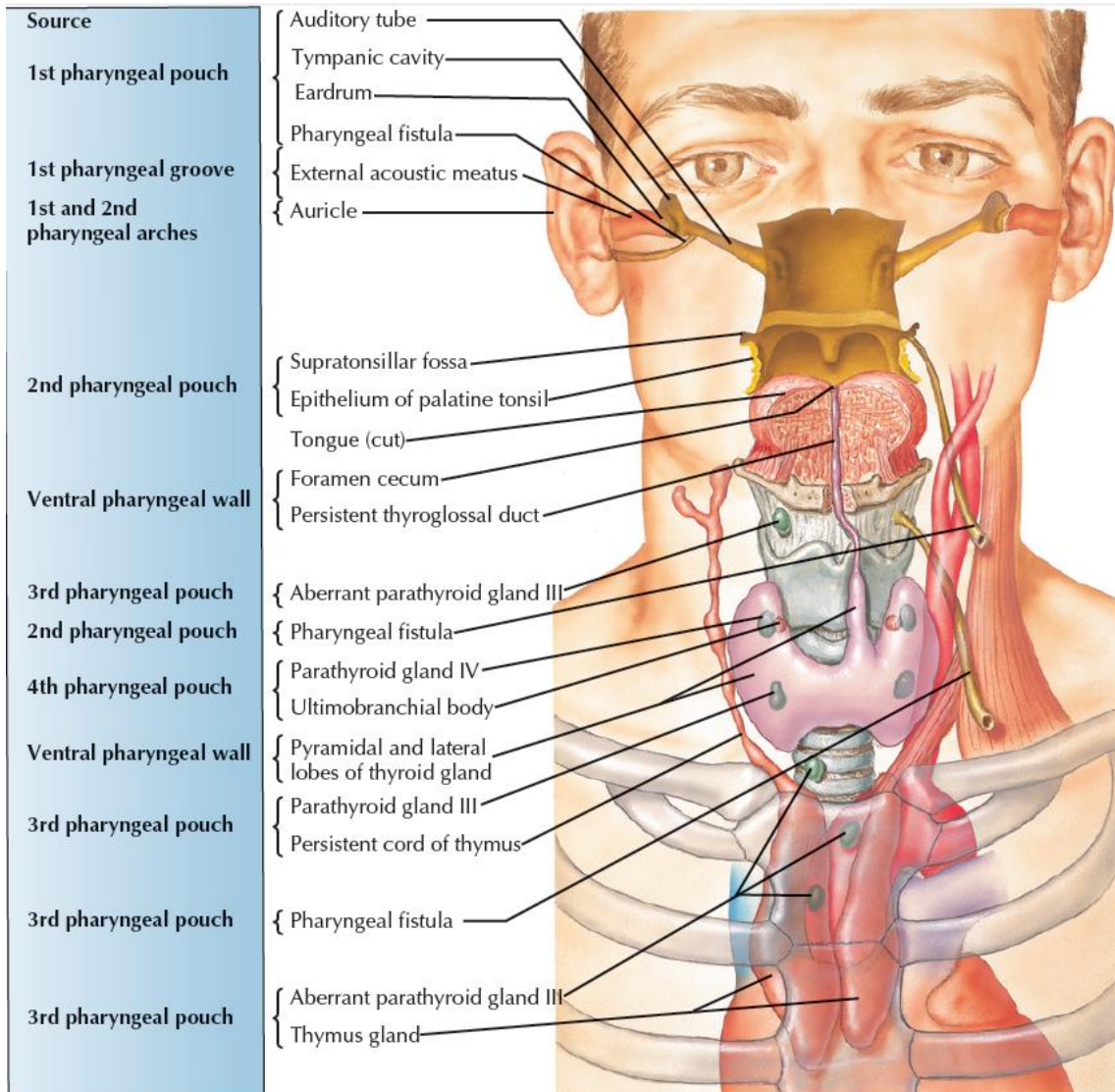
Pouch	Location	Embryonic Structure	Adult Structure
1	Opposite the 1st pharyngeal cleft, separated by the 1st pharyngeal membrane	Tubotympanic recess	Epithelium of the auditory tube and tympanic cavity
2	Opposite the 2nd pharyngeal cleft, separated by the 2nd pharyngeal membrane	Primitive palatine tonsils	Tonsillar fossa Epithelium of the palatine tonsil
3	Opposite the 3rd pharyngeal cleft, separated by the 3rd pharyngeal membrane	Divides into a dorsal and a ventral part Dorsal part migrates inferiorly toward the thorax	Inferior parathyroid gland (from the dorsal part) Thymus (from the ventral part)
4	Opposite the 4th pharyngeal cleft, separated by the 4th pharyngeal membrane	Divides into a dorsal and a ventral part Ventral part is invaded by neural crest to form the parafollicular cells	Superior parathyroid gland (from the dorsal part) Ultimobranchial body (from the ventral part)

- **Ultimopharyngeal Body:**
- Last of the series of structures from the pouches.
- From neural crest cells that migrate from 4th pouch (5th pouch which become part of 4th helps also to form ultimopharyngeal body)
- Gives **Parafollicular cells (C cells)** and fuses with thyroid gland
- Produce Calcitonin.



- **Pharyngeal Groove (Cleft):**
- Grooves formed from Ectoderm.
- Initially, 4 Pharyngeal Grooves.
- Later on 2nd, 3rd, 4th Pharyngeal Grooves normally obliterated by overgrowth of 2nd Pharyngeal Arch.
- Only One Pharyngeal Groove contributes to postnatal structure.
- **1st Pharyngeal Groove:**
 1. External Auditory Meatus.
- **Pharyngeal Membrane:**
- Composed of:
 1. External Ectoderm of Pharyngeal Grooves.
 2. Mesoderm and Neural crest in the core.
 3. Internal Endoderm of Pharyngeal Pouches.
- Only One Pharyngeal Membrane contributes to postnatal structure.
- **1st Pharyngeal Membrane:**
 1. Tympanic Membrane.





- **Embryology of Adenoid:**

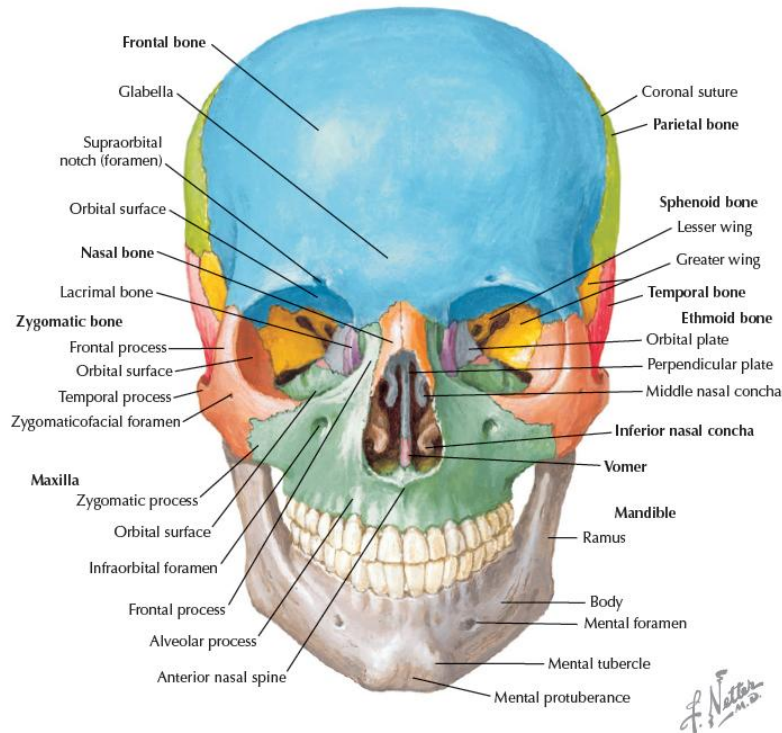
- Formed between 3rd and 7th month.
- Arise from subepithelial infiltrations of lymphocytes
- Present at birth

- **Anomalies of Head and Neck:**

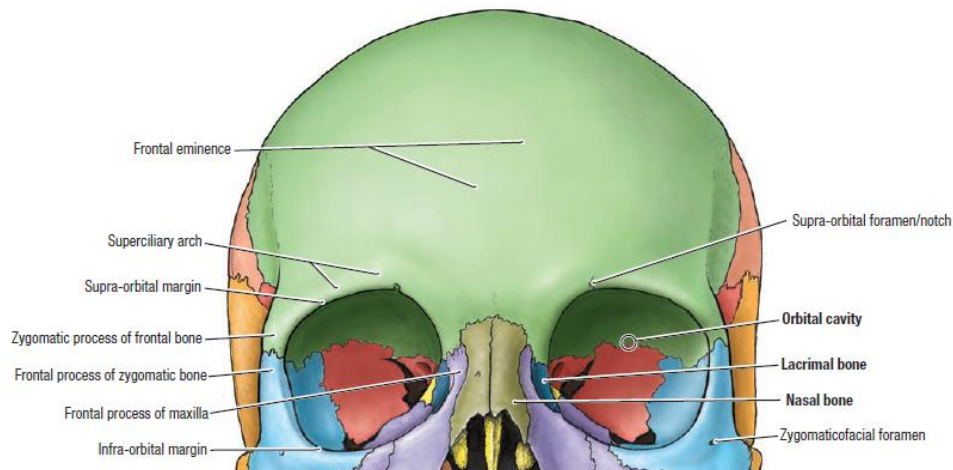
- Congenital auricular sinuses and cysts
- Branchial sinuses, fistula or cysts
- Branchial vestiges (cartilaginous and bony remnant which should have disappeared)
- 1st ach syndrome (Treacher collins syndrome, Pierre robin syndrome)
- Accessory thymic tissue and variation of thymus
- Ectopic parathyroid and abnormal number

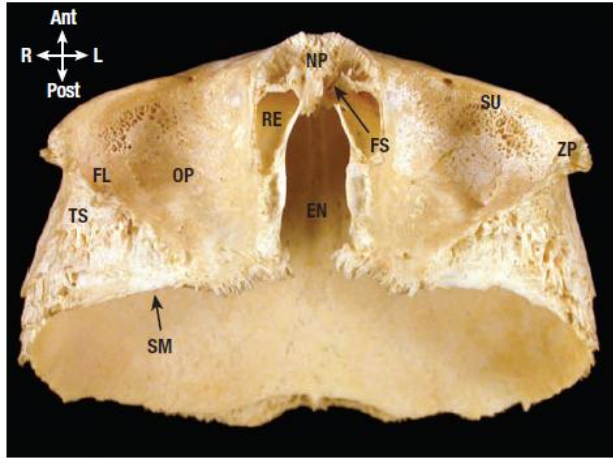
Skull and Facial Bones:

- Most complicated bony structure in the human body.
- 22 bones make up the skull:
 - **8 Paired bones:**
 1. Parietal
 2. Temporal
 3. Zygomatic
 4. Maxilla
 5. Palatine
 6. Nasal
 7. Lacrimal
 8. Inferior Turbinate
 - **6 Single bones:**
 1. Frontal
 2. Ethmoid
 3. Sphenoid
 4. Vomer
 5. Mandible
 6. Occipital
- Most important function: to protect the brain
- Also protects the 5 organs of special sense:
 - Olfaction
 - Vision
 - Taste
 - Vestibular function
 - Auditory function
- **Skull is divided into:**
 1. Mandible (lower jaw)
 2. Cranium (skull without the mandible)
 - Cranium is further divided into:
 - Cranial vault: Upper portion of skull.
 - Cranial base: Inferior portion of skull.
 - Cranial cavity: Interior of skull.
 - Facial skeleton: Bones that make up the face
 - Acoustic skeleton: Ear ossicles.
- **Cranial cavity divisions:**
 1. Anterior cranial fossa:
 - Contains Frontal lobe of the brain.
 2. Middle cranial fossa:
 - Contains Temporal lobe of the brain.
 3. Posterior cranial fossa:
 - Contains Cerebellum.

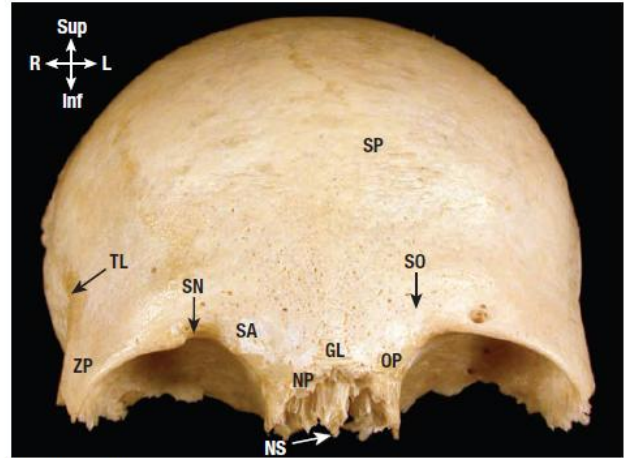


- **FRONTAL BONE**
- Single bone.
- Contains Frontal Sinuses
- **3 Parts:**
 - 1. Squamous Portion:**
 - Largest part.
 - Forms majority of forehead.
 - Forms supra-orbital margin and superciliary arch.
 - Zygomatic process of frontal bone extends from posterior part of supra-orbital margin.
 - 2. Orbital Portion:**
 - Forms roof of the orbit and floor of Anterior cranial fossa.
 - 3. Nasal Portion:**
 - Trochlea of orbit articulates with the orbital portion.
 - Articulates with Nasal bones and Frontal process of Maxilla to form Root of the nose.





A. Inferior View

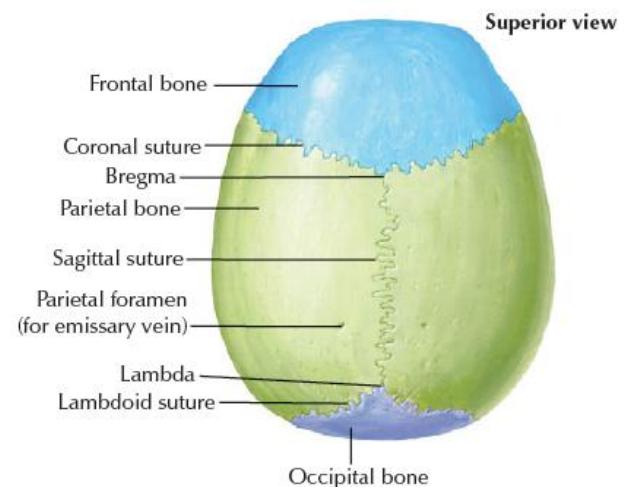
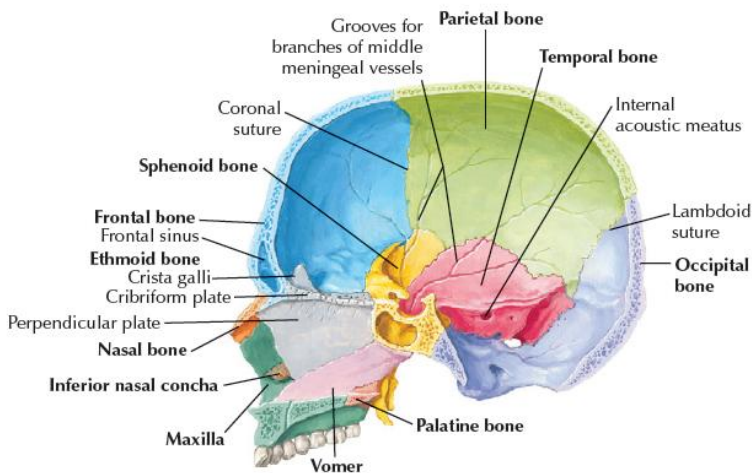


B. Anterior View

Key for A and B: Frontal Bone

EN	Ethmoidal notch	NP	Nasal part	SA	Superciliary arch	SU	Supra-orbital margin
FL	Fossa for lacrimal gland	NS	Nasal spine	SM	Sphenoidal margin	TL	Temporal line
FS	Opening of frontal sinus	OP	Orbital part	SN	Supra-orbital notch	TS	Temporal surface
GL	Glabella	RE	Root of ethmoid cells	SO	Supra-orbital foramen	ZP	Zygomatic process
				SP	Squamous part		

- **PARIETAL BONE**
- Paired bone.
- Forms majority of Cranial vault (roof and sides).
- Provides attachment of Temporalis muscle.



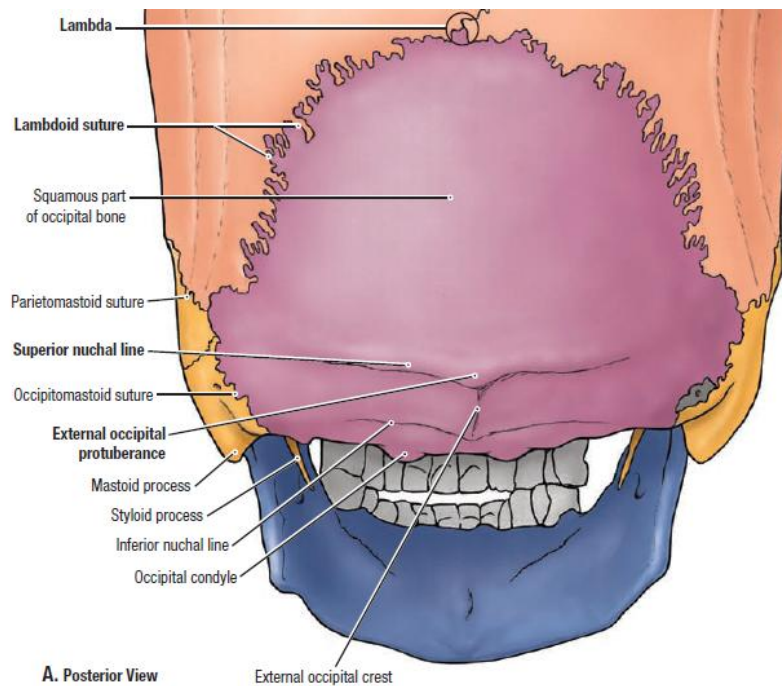
- **OCCIPITAL BONE**

- Single bone.
- Forms posterior part of Cranial vault.
- Articulates with Atlas.

- **3 Parts:**

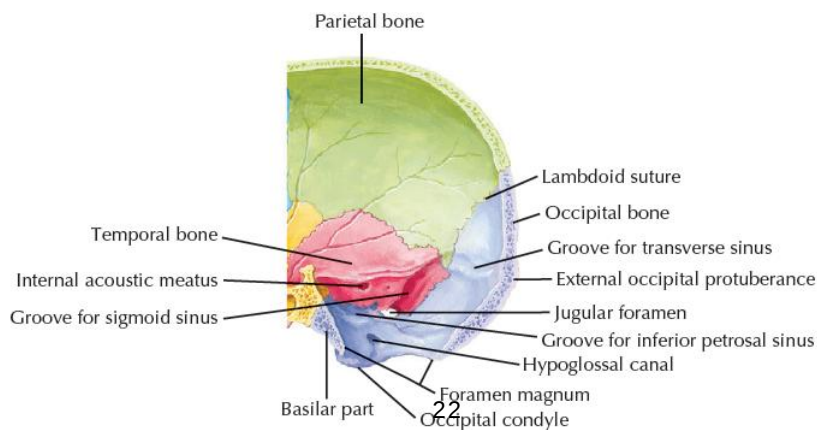
1. Squamous portion:

- Largest portion of Occipital bone.
- Posterior and superior to Foramen magnum.
- Articulates with Temporal and Parietal bones.
- External occipital protuberance (more pronounced in males).
- Superior and Inferior nuchal lines.



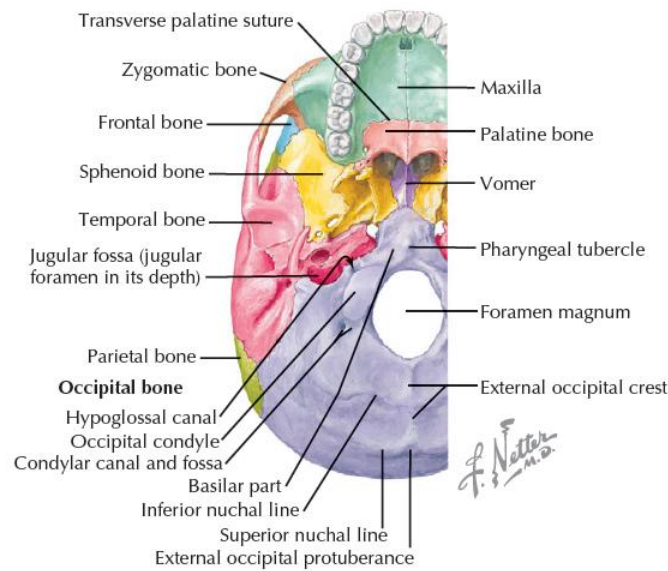
2. Lateral portion:

- Lateral to Foramen magnum.
- Articulates with Temporal bone.
- Occipital condyle articulates with Atlas.
- Contains Hypoglossal canal.
- Forms a portion of Jugular foramen.



3. Basilar portion:

- Immediately Anterior to Foramen magnum.
- Articulates with Petrous and sphenoid bones.
- Pharyngeal tubercle provides attachment for superior constrictor.
- Internal surface of Basilar portion is called Clivus, and part of the brainstem lies against it.



- **TEMPORAL BONE**

- Paired bone.
- Form part of base and lateral walls of Skull.
- House Auditory and Vestibular apparatuses.
- Contain Mastoid air cells.

- **4 parts:**

1. Squamous part of Temporal bone:

- Largest Part.
- 3 portions:

1. Temporal:

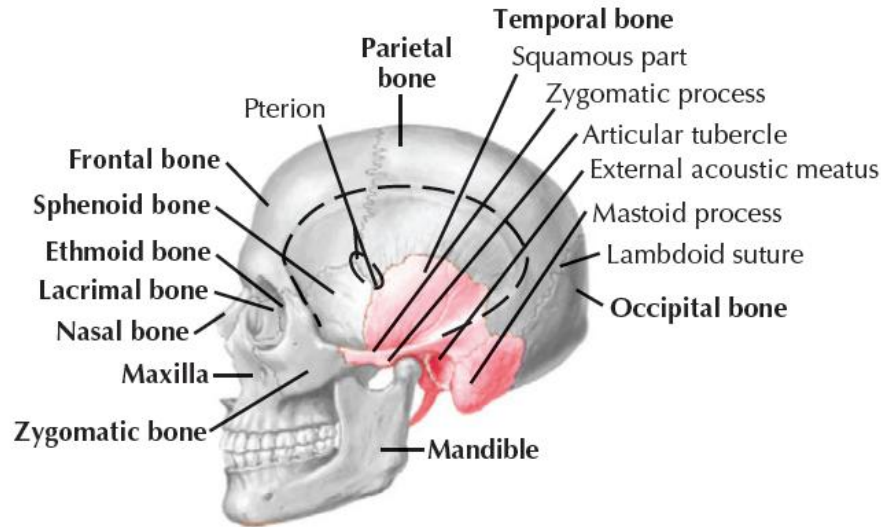
- Thin large area on Squamous part of temporal bone.
- On the internal surface of temporal portion lies a groove for Middle meningeal Artery.

2. Zygomatic process:

- Extends laterally and anteriorly from the squamous portion
- Articulates with Temporal process of zygomatic bone to make Zygomatic arch.

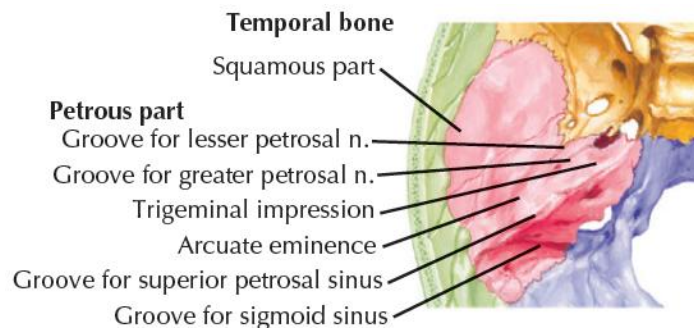
3. Glenoid fossa:

- Inferior and medial to Zygomatic process.
- Articulates with Mandibular condyle forming Temporomandibular joint.



2. Petrous part of Temporal bone:

- The solid portion of Temporal bone.
- Extends Anteriorly and Medially.
- Auditory and Vestibular apparatuses are located within petrous part.
- Separate Temporal and Occipital lobes of the brain.
- Medial part articulates with sphenoid bone to form Foramen lacerum.
- Internal Acoustic Meatus (IAM) is observed on the medial side of petrous part.
- Carotid canal lies on Inferior part of Petrous part.
- Petrotympenic fissure lies between Petrous and Tympanic parts of Temporal bone.
- On the posterior inferior surface lies Jugular fossa.
- Between Jugular fossa and Carotid canal is Tympanic canaliculus.
- Mastoid process extends posteriorly and has large mastoid air cells.

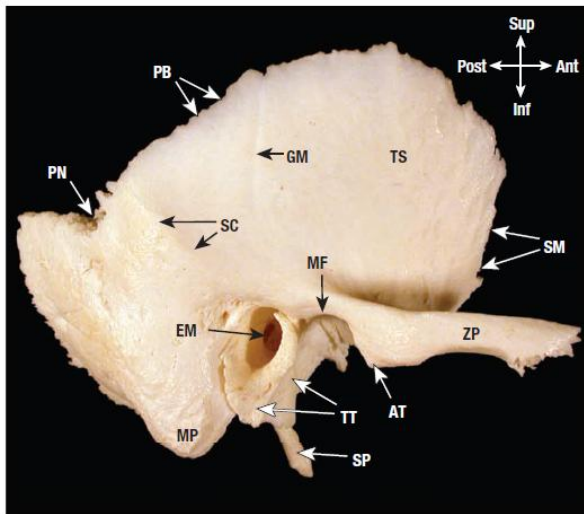


3. Tympanic part of Temporal bone:

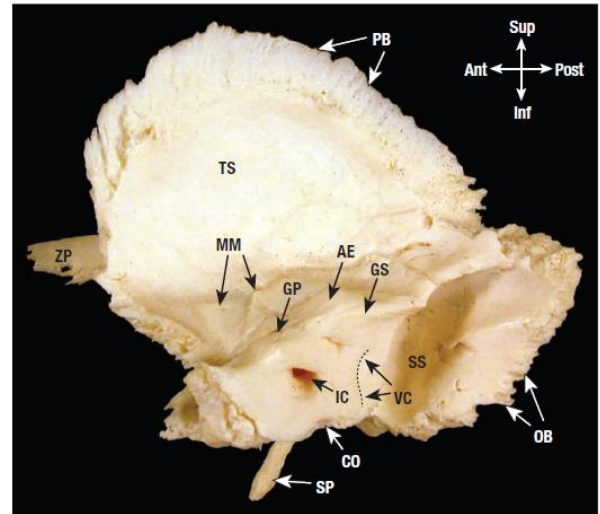
- A plate of bone forming Anterior, Posterior and Inferior portions of External acoustic meatus.
- Anterior part forms Posterior portion of Glenoid fossa.

4. Styloid process:

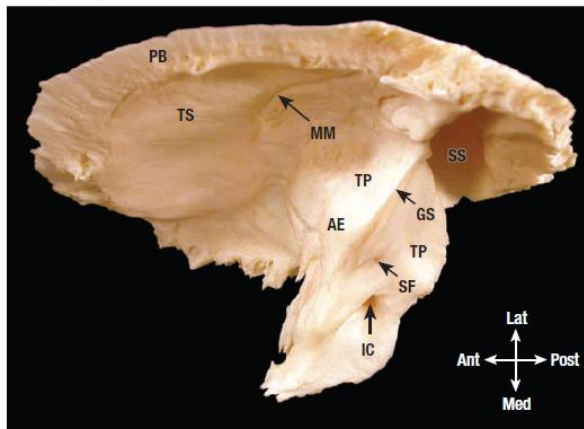
- A projection from the temporal bone.
- Stylomastoid foramen lies posterior to this process.



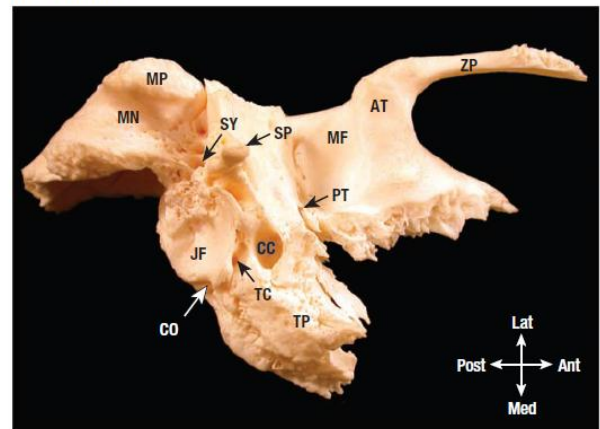
A. Lateral View



B. Medial View



C. Superior View



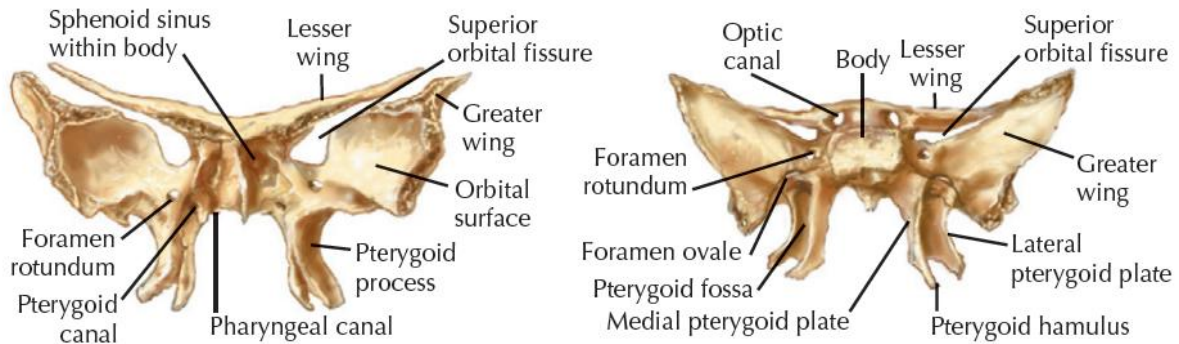
D. Inferior View

Key for A-D: Temporal Bone

AE Arcuate eminence	MF Mandibular fossa	SM Sphenoid margin
AT Articular tubercle	MM Groove for middle meningeal artery	SP Styloid process
CC Carotid canal	MN Mastoid notch	SS Groove for sigmoid sinus
CO Cochlear canaliculus	MP Mastoid process	SY Stylomastoid foramen
EM External acoustic meatus	OB Occipital border	TC Tympanic canaliculus
GM Groove for middle temporal artery	PN Parietal border	TP Temporal bone (petrous part)
GP Hiatus for greater petrosal nerve	PT Petrotympanic fissure	TS Temporal bone (squamous part)
GS Groove for superior petrosal sinus	SC Supramastoid crest	TT Temporal bone (tympanic part)
IC Internal acoustic meatus	SF Subarcuate fossa	VC Vestibular canaliculus
JF Jugular fossa		ZP Zygomatic process

- **SPHENOID BONE**

- Single bone.
- Forms majority of Middle portion of Cranial base.
- Forms majority of Middle Cranial fossa.
- Contains Sphenoid Sinus.



- **4 Parts:**

1. Body of Sphenoid:

- Center of sphenoid.
- Anterior portion forms part of nasal cavity.
- Sella turcica:
 - Superior part of sphenoid body.
 - Saddle shaped and possesses the Anterior and posterior clinoid processes.
 - Hypophyseal fossa, the deepest part of Sella turcica, houses Pituitary gland.
- Dorsum sellae is a square-shaped part of bone that lies posterior to sella turcica.
- Clivus is the portion that slopes posterior to the body.
- Body contains sphenoid paranasal sinuses.
- Lateral portion of the body is covered by Cavernous sinus.
- Optic canal is found in the body of the sphenoid.

2. Greater wing:

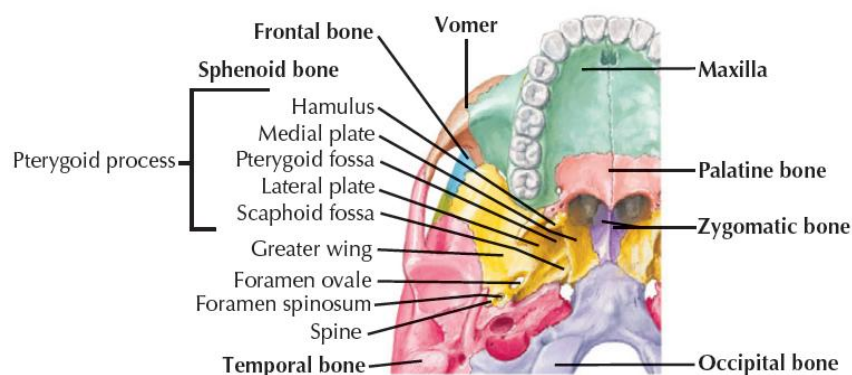
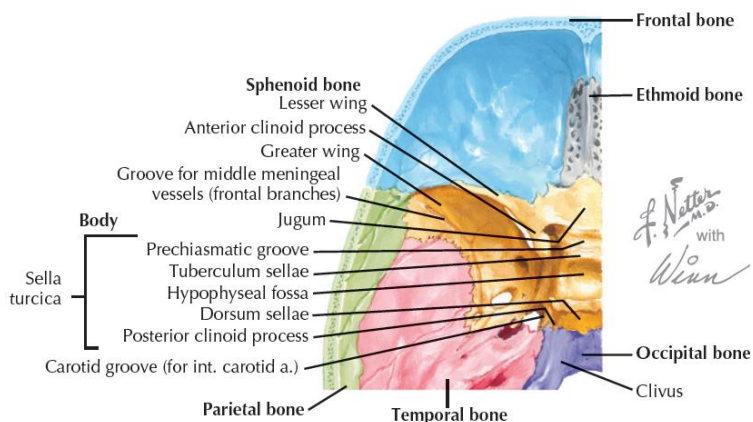
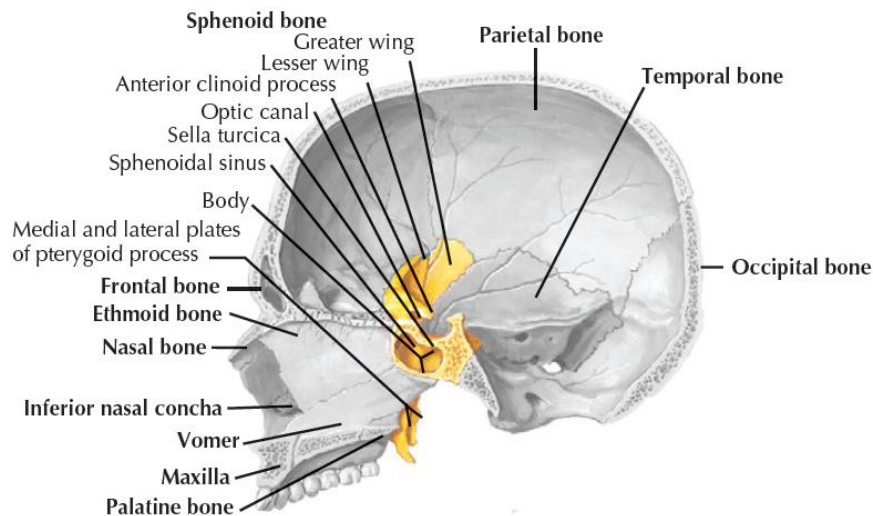
- Extends Laterally and Anteriorly from posterior portion of the body.
- Endocranial portion helps form a large part of the middle cranial fossa.
- Lateral portion is the infratemporal surface.
- Anterior portion lies in the orbit.
- Contains 3 foramina:
 - 1.** Foramen spinosum
 - 2.** Foramen rotundum
 - 3.** Foramen ovale

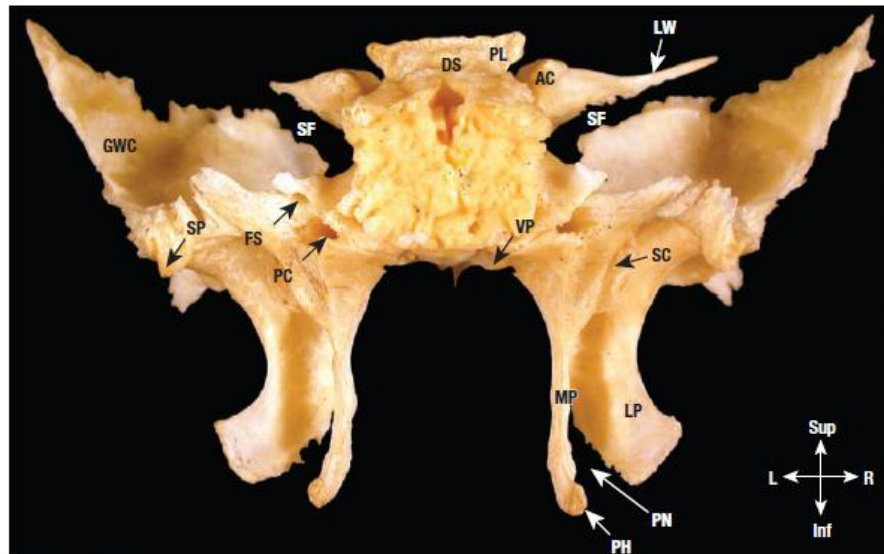
3. Lesser wing:

- Extends Laterally and Anteriorly from the superior portion of the body.
- Separated from the greater wing by Superior orbital fissure.

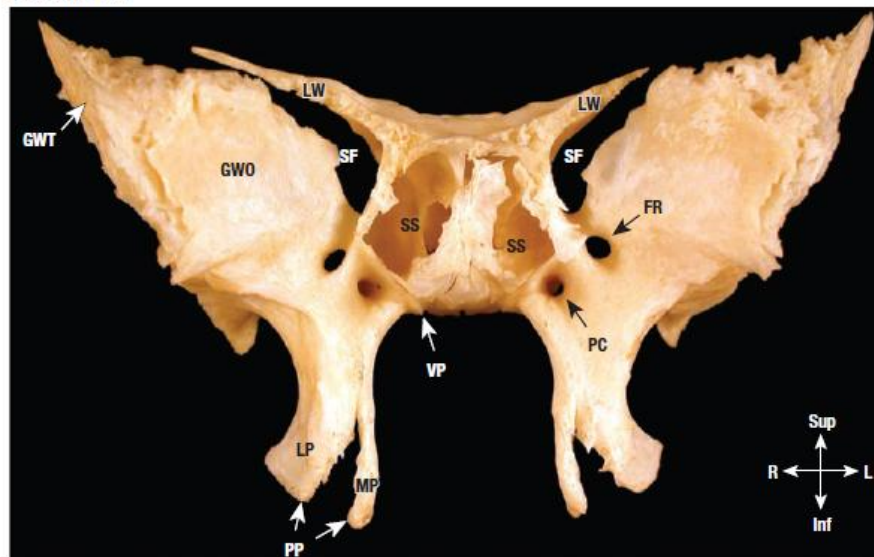
4. Pterygoid process:

- Arises from Inferior surface of the body.
- 2 Pterygoid processes, each has:
 1. Lateral pterygoid plate
 2. Medial pterygoid plate
- Pterygoid hamulus extends from Medial pterygoid plate.
- Two canals are associated with Pterygoid process:
 1. Pterygoid canal
 2. Pharyngeal canal





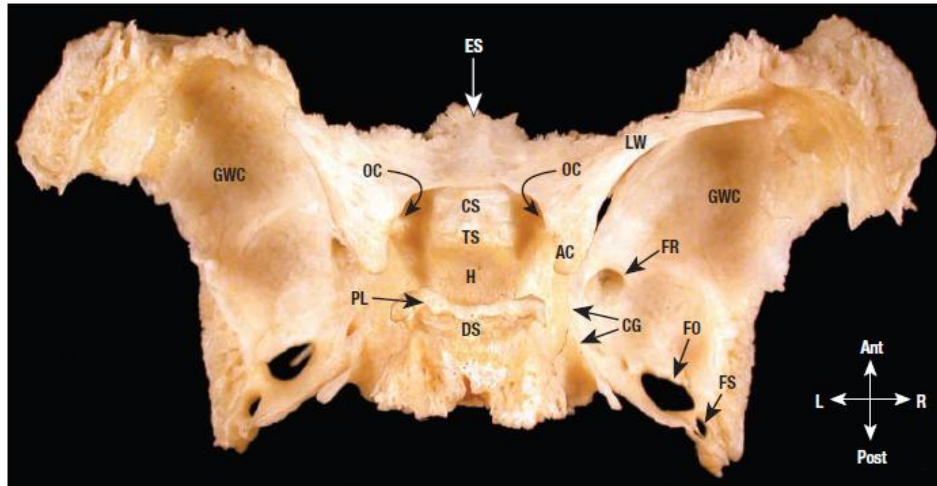
A. Posterior View



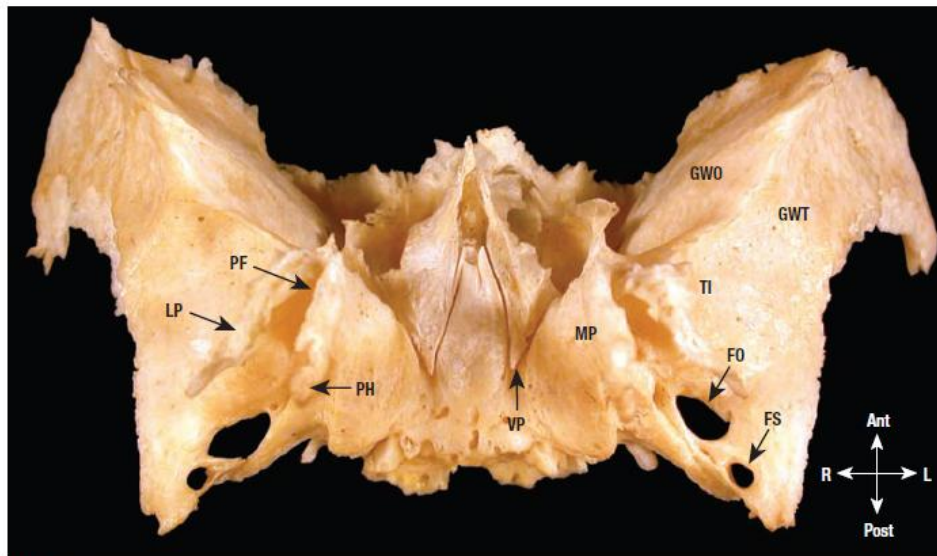
B. Anterior View

Key for A-D: Sphenoid Bone

AC	Anterior clinoid process	FO	Foramen ovale	GWO	Greater wing (orbital surface)
CG	Carotid sulcus	FR	Foramen rotundum	GWT	Greater wing (temporal surface)
CS	Prechiasmatic sulcus	FS	Foramen spinosum	H	Hypophysial fossa
DS	Dorsum sellae	GWC	Greater wing (cerebral surface)	LP	Lateral pterygoid plate
ES	Ethmoidal spine	GWI	Greater wing (infratemporal surface)	LW	Lesser wing



C. Superior View



D. Inferior View

Key for A-D: Sphenoid Bone (Continued)

MP	Medial pterygoid plate	PL	Posterior clinoid process	SP	Spine of sphenoid bone
OC	Optic canal	PN	Pterygoid notch	SS	Sphenoidal sinus (In body of sphenoid)
PC	Pterygoid canal	PP	Pterygoid process	TS	Tuberculum sellae
PF	Pterygoid fossa	SC	Scaphoid fossa	VP	Vaginal process
PH	Pterygoid hamulus	SF	Superior orbital fissure		

- **ETHMOID BONE**

- Single porous bone.
- Forms the major portion of middle part of Face between the orbits.
- Helps form Orbit, Nasal cavity, Septum and Anterior cranial fossa.

- **3 Parts:**

1. Perpendicular plate:

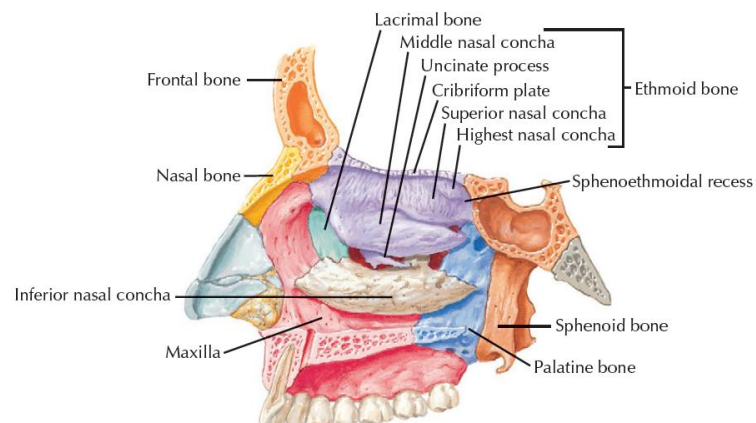
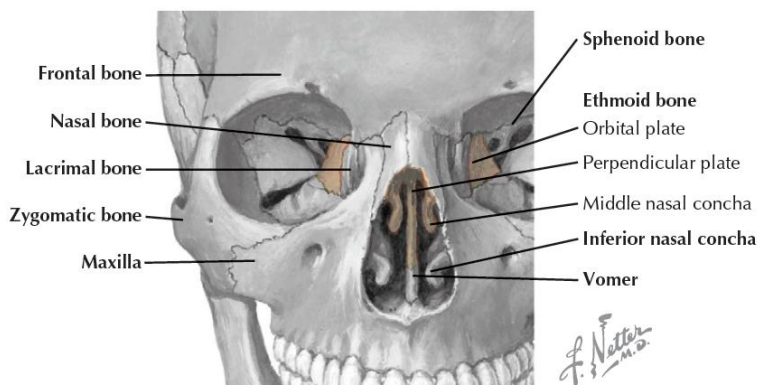
- Flat plate descends from Cribriform plate to form part of Nasal septum.
- Articulates with the vomer inferiorly.

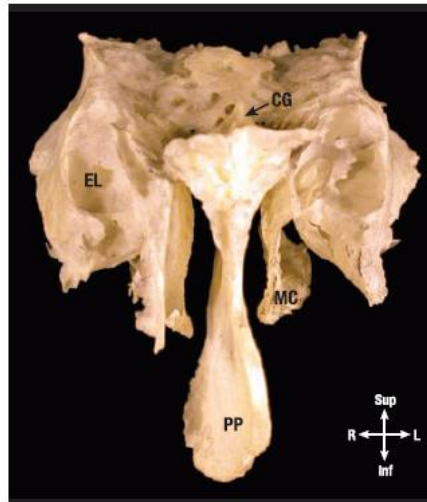
2. Cribriform plate:

- Horizontal bone forms Superior surface of Ethmoid.
- Contains numerous foramina for Olfactory nerve.
- Crista galli is a vertical plate extends superiorly from cribriform plate providing attachment for Falx cerebri of meninges.
- Associated with a small foramen cecum.

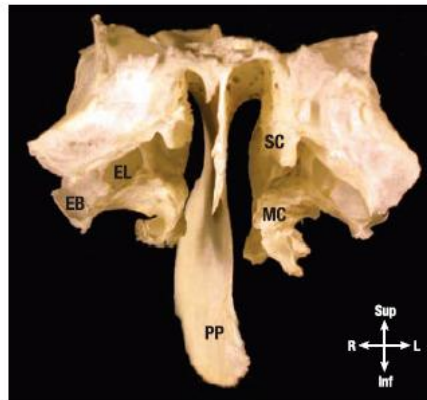
3. Ethmoid labyrinth:

- Largest part of Ethmoid bone.
- Descends inferiorly from the cribriform plate.
- Ethmoid sinuses are located within the ethmoid labyrinth.
- Ethmoid labyrinth forms 2 major structures within Nasal cavity:
 - 1. Superior Nasal Turbinate**
 - 2. Middle Nasal Turbinate**
- Ethmoid bulla is the large elevation of bone located by the middle ethmoid paranasal sinuses
- Uncinate process is a curved piece of bone.
- Between Uncinate process and Ethmoid bulla is the Hiatus semilunaris.

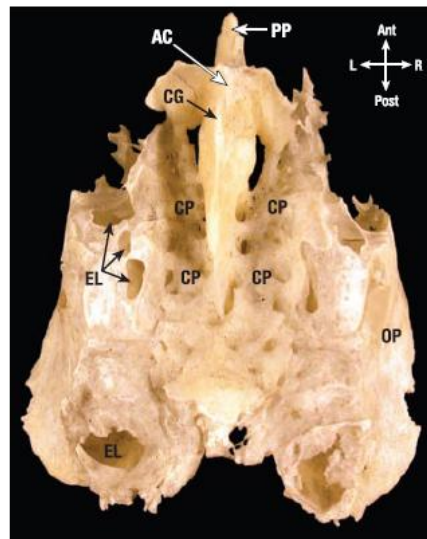




E. Anterior View



F. Posterior View

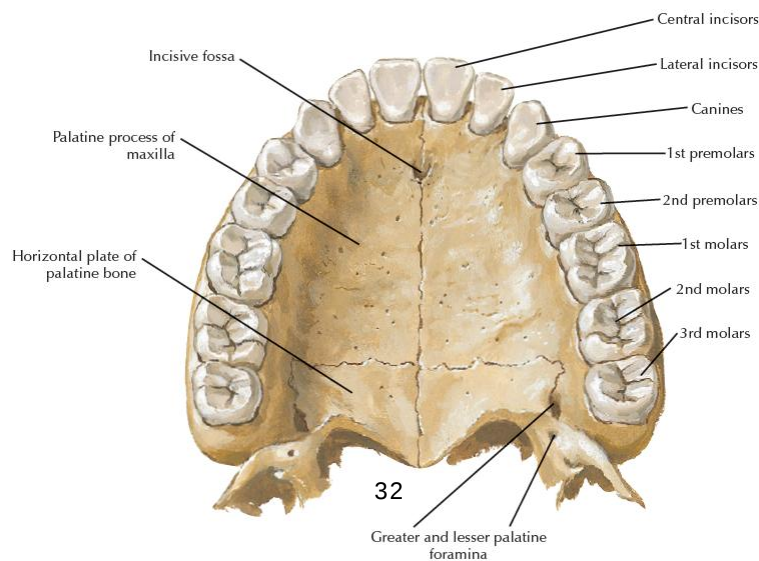
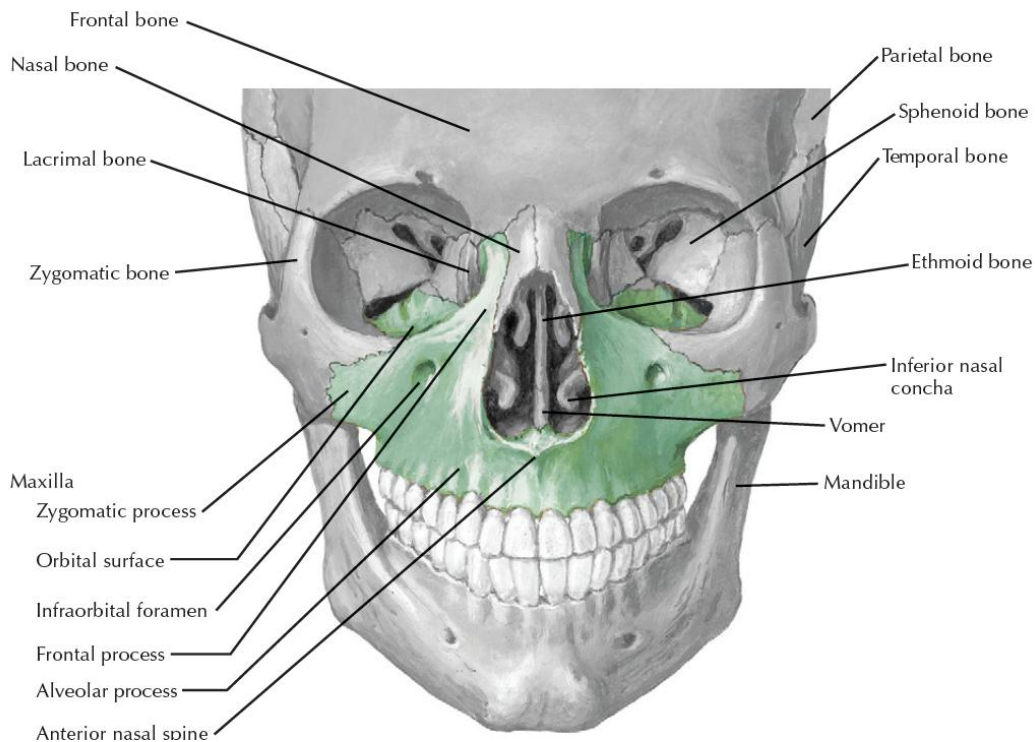


G. Superior View

Key for E-G: Ethmoid Bone

AC	Ala of crista galli	EB	Ethmoidal bulla	OP	Orbital plate
CG	Crista galli	EL	Ethmoidal labyrinth (cells)	PP	Perpendicular plate
CP	Cribriform plate	MC	Middle nasal concha	SC	Superior nasal concha

- **MAXILLA**
- Paired bone.
- Forms majority of skeleton of Face and upper jaw.
- Contains Maxillary sinus.
- Articulates with:
 - Opposite Maxilla
 - Frontal bone
 - Sphenoid bone
 - Nasal bone
 - Vomer bone
 - Ethmoid bone
 - Inferior Nasal Turbinate
 - Palatine bone
 - Lacrimal bone
 - Zygomatic bone
 - Septal cartilages



- **5 Parts:**

1. Body of Maxilla:

- Major part of the bone.
- Shaped like a pyramid.
- Contains Maxillary sinus.
- 4 different regions:
 - Orbit
 - Nasal cavity
 - Infratemporal fossa
 - Face
- Infraorbital canal and foramen pass from the orbit region to the face region.

2. Frontal Process:

- Extends Superiorly.
- Articulate with:
 - Nasal bone
 - Frontal bone
 - Ethmoid bone
 - Lacrimal bone
- Forms the posterior boundary of Lacrimal fossa

3. Zygomatic Process:

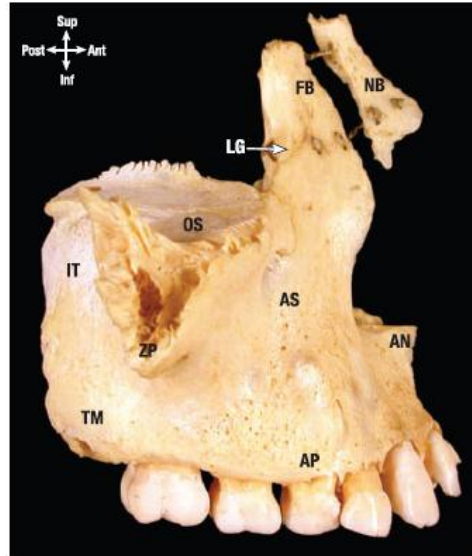
- Extends Laterally.
- Articulates with Maxillary process of Zygomatic bone.

4. Palatine Process:

- Extends Medially.
- Form Majority of Hard palate.
- Articulates with:
 - Palatine process of opposite Maxilla
 - Horizontal plate of Palatine bone
- Incisive foramen is located in the Anterior portion.

5. Alveolar Process:

- Extends Inferiorly.
- Supports all Maxillary teeth.
- Each maxilla contains:
 - 5 primary teeth.
 - 8 permanent teeth.
- Alveolar bone is resorbed when a tooth is lost.



I. Lateral View

Key for I: Maxilla and Nasal Bone

AN	Anterior nasal spine	LG	Lacrimal groove
AP	Alveolar part	NB	Nasal bone
AS	Anterior surface	OS	Orbital surface
FP	Frontal process	TM	Tuberosity
IT	Infratemporal surface	ZP	Zygomatic process

- **PALATINE BONE**

- Paired L-shaped bone
- Forms part of Lateral wall of Nasal cavity and Hard palate.

- **3 Parts:**

1. Perpendicular Plate:

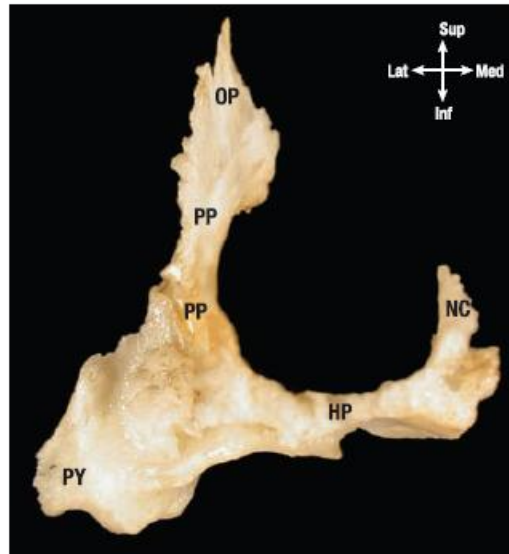
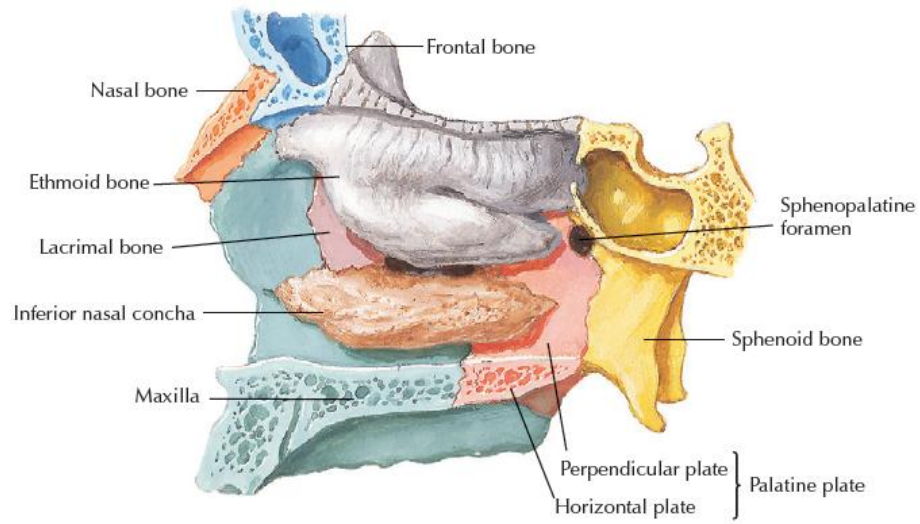
- Vertical rectangle shape.
- On the superior border is a notch articulates with the sphenoid bone, forming Sphenopalatine foramen.
- Small orbital process helps form part of the orbit.
- Forms part of wall of the Pterygopalatine fossa and Lateral wall of Nasal cavity.
- Lateral wall articulates with Maxilla to form palatine canal.

2. Horizontal Plate:

- Forms Posterior portion of Hard palate.
- Superior to horizontal plate is Nasal cavity.
- Posterior nasal spine is located on the medial part and formed by both of the horizontal plates.
- Greater palatine foramen is located on this plate.

3. Pyramidal Process:

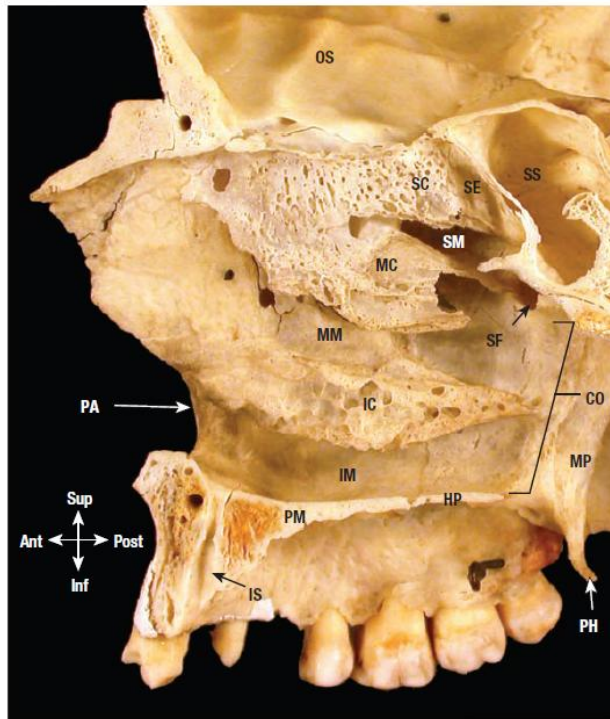
- Extends posteriorly and inferiorly from junction of Perpendicular and Horizontal plates of palatine bone.
- Lesser palatine foramina are located here.



H. Anterior View

Key for H: Palatine Bone

HP	Horizontal plate	PP	Perpendicular plate
NC	Nasal crest	PY	Pyramidal process
OP	Orbital process		



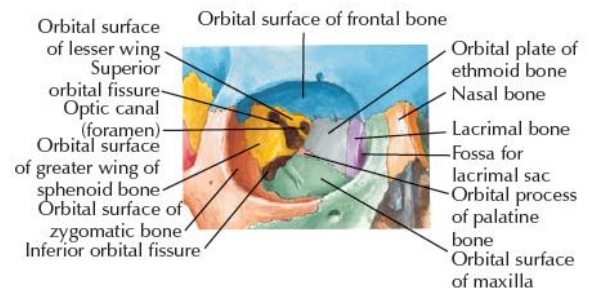
A. Lateral Wall of Nose, Medial View

Key for A: Lateral Wall of Nose

CO	Choana (posterior nasal aperture)
HP	Horizontal plate of palatine bone
IC	Inferior nasal concha
IS	Incisive canal
IM	Inferior nasal meatus
MC	Middle nasal concha
MM	Middle nasal meatus
PH	Pterygoid hamulus
PM	Palatine process of maxilla
OS	Orbital surface of frontal bone
PA	Piriform aperture
PM	Palatine process of maxilla
SC	Superior nasal concha
SE	Spheno-ethmoidal recess
SF	Sphenopalatine foramen
SM	Superior nasal meatus
SS	Sphenoidal sinus

- LACRIMAL BONE

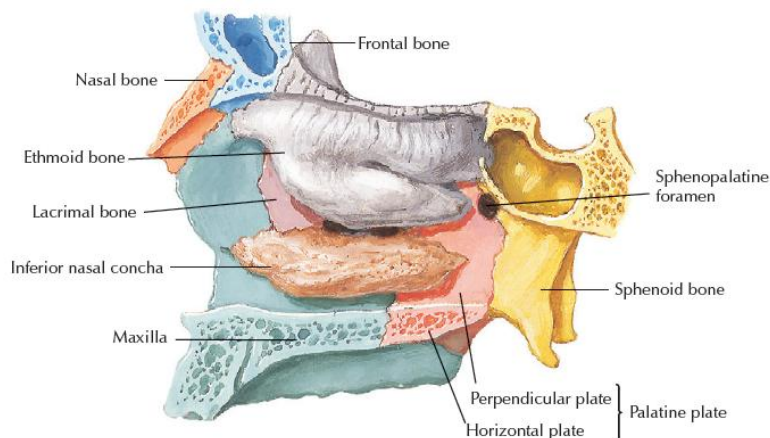
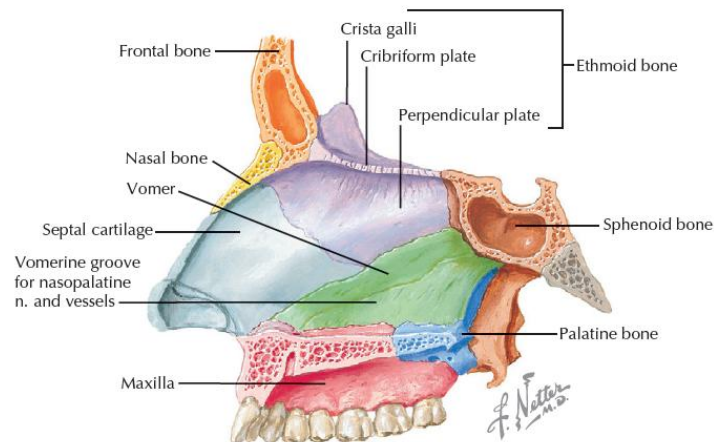
- Paired bone.
- Small and rectangular in shape.
- Very thin and fragile.
- Forms a small portion of:
 - o Medial wall of Orbit.
 - o Lateral wall of Nasal cavity.
- Articulates with:
 - o Frontal process of Maxilla
 - o Orbital plate of Ethmoid bone
 - o Frontal bone
 - o Inferior Nasal Turbinate
- The region that articulates with Frontal process of Maxilla forms Lacrimal fossa, the location of the Lacrimal Sac.



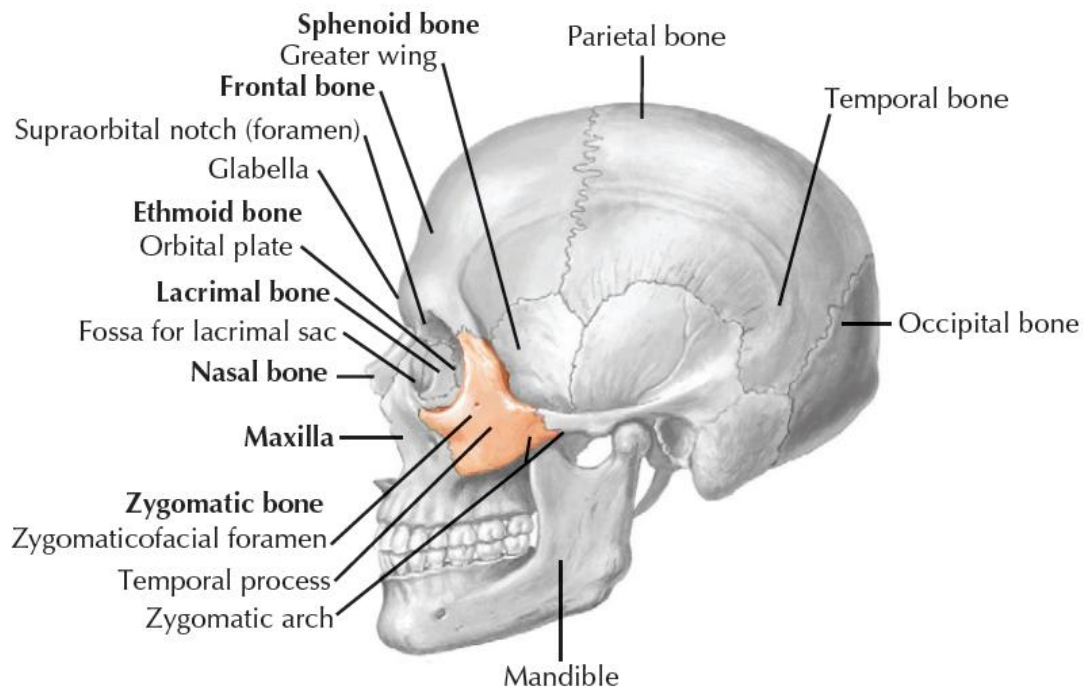
- NASAL BONE

- Paired bone.
- Inferior portion forms Superior margin of Nasal aperture.
- Forms bridge of Nose.
- Articulates with:
 - o Nasal bone of opposite side
 - o Nasal portion of Frontal bone
 - o Frontal process of Maxilla
 - o Perpendicular plate of Ethmoid
- Inferior portion of the nasal bones attaches with Lateral nasal cartilages and septal cartilage.

- **VOMER**
 - Single bone.
 - Shaped like a "plough".
 - Forms Posterior inferior part of Nasal septum.
 - Articulates with:
 - o Perpendicular plate of Ethmoid
 - o Maxilla
 - o Palatine
 - o Sphenoid bone
 - o Septal cartilage
 - Posterior border does not articulate with any other bone.
-
- **INFERIOR NASAL CONCHA**
 - Paired curved bone.
 - Forms part of Lateral wall of Nasal cavity.
 - Lies within a curve in Lateral wall of Nasal cavity.
 - Articulates with:
 - o Maxilla
 - o Perpendicular plate of Palatine
 - o Lacrimal bone
 - o Ethmoid bone



- **ZYGOMATIC BONE (ZYGOMA)**
- Paired bone.
- Forms Majority of skeleton of Cheek.
- Provides for attachment of Masseter.
- Three foramina in Zygoma:
 1. Zygomatico-orbital foramen
 2. Zygomaticofacial foramen
 3. Zygomaticotemporal foramen
- **3 Parts:**
 1. **Frontal Process:**
 - Articulates with Zygomatic process of Frontal bone to help form Orbit.
 2. **Temporal Process:**
 - Articulates with Zygomatic process of Temporal bone to form Zygomatic Arch.
 3. **Maxillary Process:**
 - Articulates with Zygomatic process of Maxillary bone to help form Orbit.

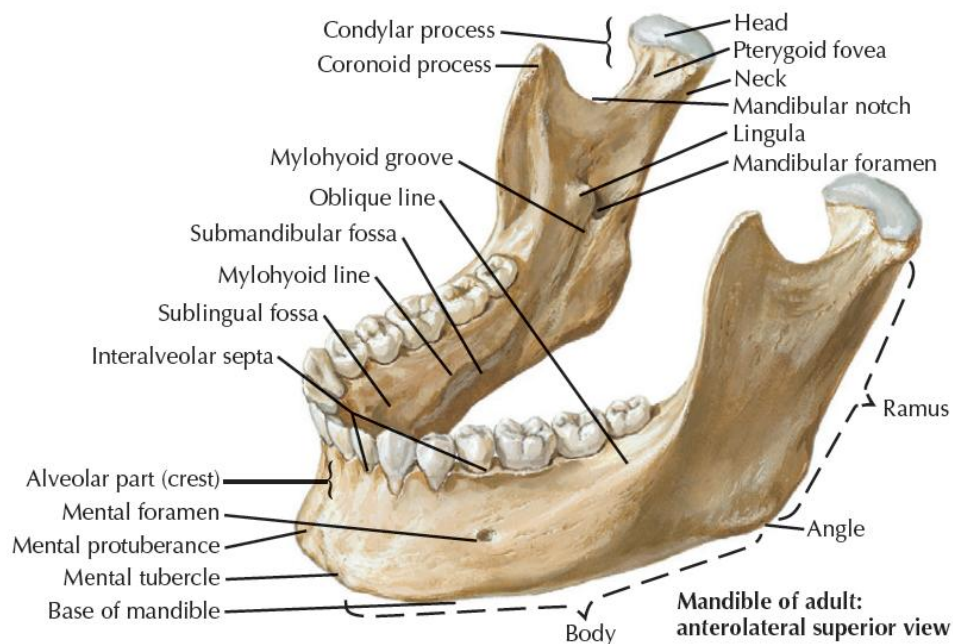


- **MANDIBLE**
- Single horseshoe shaped bone.
- Forms Lower jaw.
- All muscles of Mastication attach to Mandible.

- **5 Parts:**

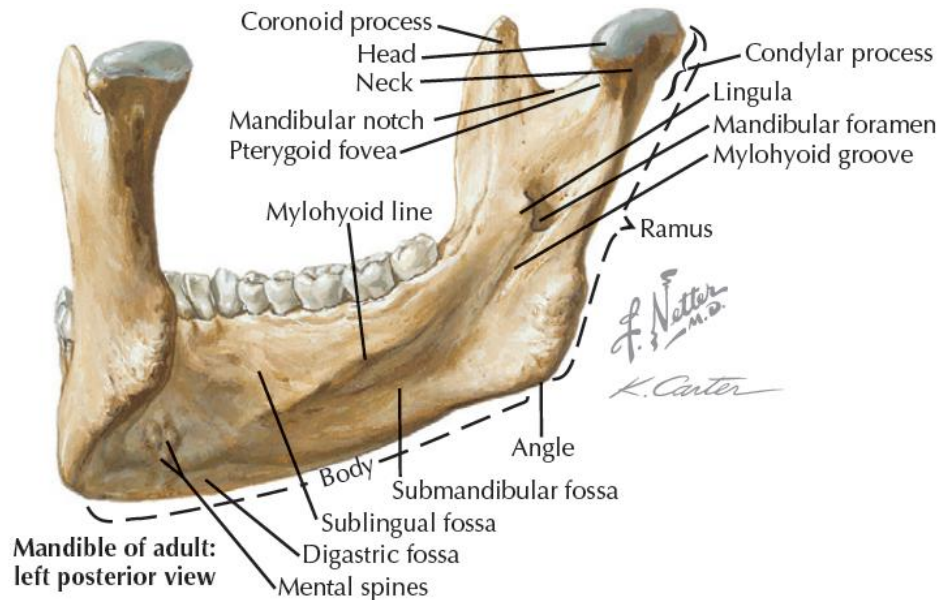
1. Body of Mandible:

- Mental foramen lies on Anterior part of Lateral surface of the body.
- External oblique line located on Lateral side of Body.
- Mylohyoid line located on Medial side of Body
- Mylohyoid line helps divide Sublingual from Submandibular fossa.
- Posterior border of Mylohyoid line provides attachment of Pterygomandibular raphe.
- At the midline on the medial side are the superior and inferior genial tubercles, as well as Digastric fossa.



2. Ramus of Mandible:

- Meets body of Mandible at the Angle on each side.
- Lateral side of Ramus:
 - Attachment of Masseter muscle.
- Medial side of Ramus:
 - Attachment of Medial pterygoid muscle.
 - Mandibular foramen.
- Superior part of Ramus divides into:
 - Coronoid Process (Anteriorly)
 - Condylar Process (Posteriorly)
 - Separated by Mandibular notch.



3. Coronoid Process:

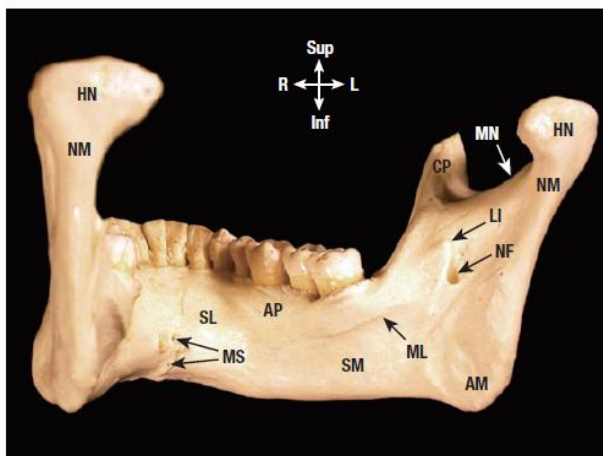
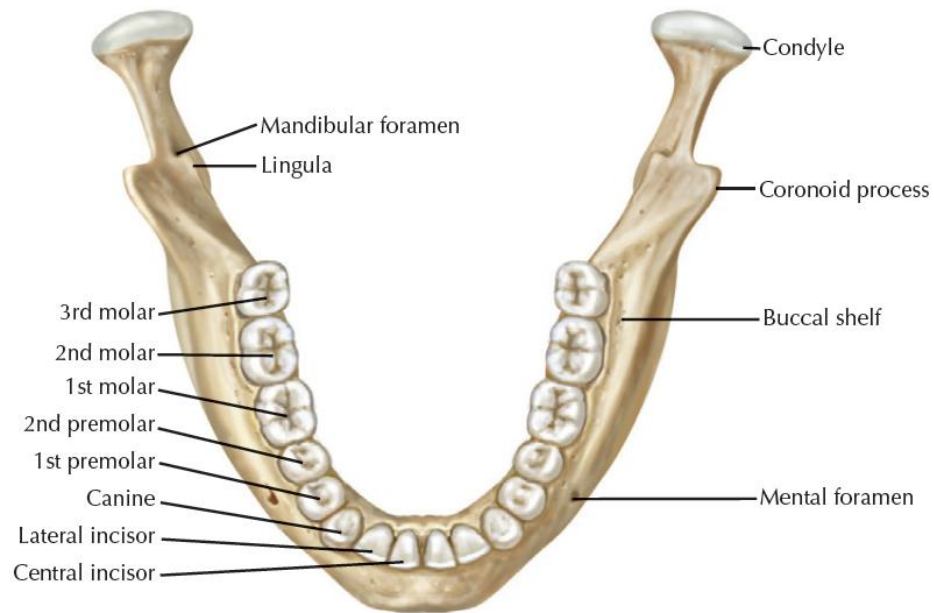
- Anterior superior extension of each Ramus.
- Attachment of Temporalis muscle.

4. Condylar Process:

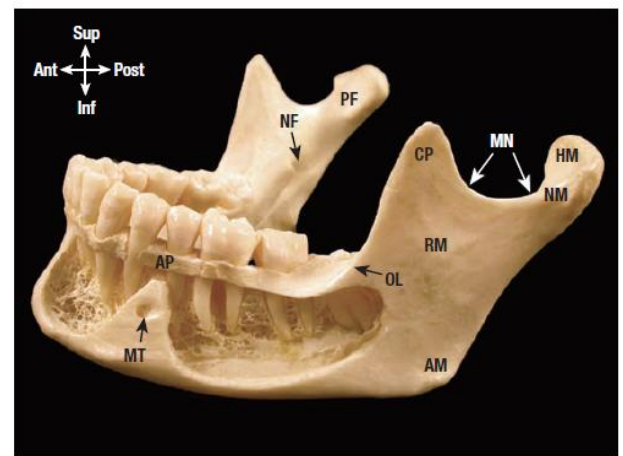
- Posterior superior extension of each Ramus.
- Articulates with Temporal bone in TM joint.
- Has a neck that forms a condyle superiorly.
- Attachment of Lateral pterygoid muscle into pterygoid fovea on the Neck.

5. Alveolar Process:

- Extends superiorly from the body.
- Created by a thick buccal and a thin lingual plate of bone
- Supports the mandibular teeth
- Each side of the mandible contains:
 - 5 primary teeth.
 - 8 permanent teeth.
- Alveolar bone is resorbed when a tooth is lost.



C. Posteromedial View



D. Lateral View

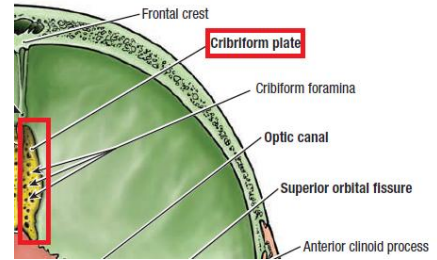
Key for C and D: Mandible

AM	Angle of mandible	ML	Mylöhoid groove	NM	Neck of mandible
AP	Alveolar part	MN	Mandibular notch	PF	Pterygoid fovea
CP	Coronoid process	MS	Mental (genial) spines	RM	Ramus of mandible
HM	Head of mandible	MT	Mental foramen	SL	Sublingual fossa
LI	Lingula	NF	Mandibular foramen	SM	Submandibular fossa

- **Major Skull Foramina and Fissures:**

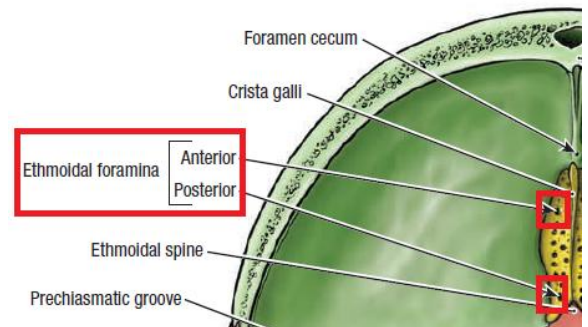
○ **Cribriform Plate:**

- Formed by:
 - Ethmoid bone.
- Contents:
 1. Olfactory Nerve (**CN-I**).



○ **Anterior Ethmoid Foramen:**

- Formed by:
 - Between Frontal and Ethmoid bone
- Contents:
 1. Anterior Ethmoid Nerve.
 2. Anterior Ethmoid Vessels.

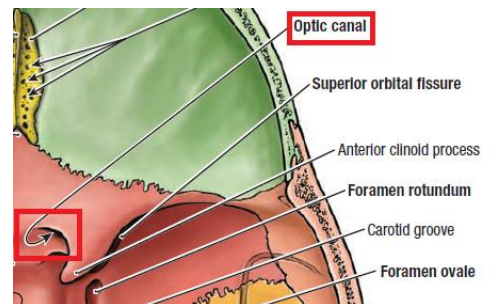


○ **Posterior Ethmoid Foramen:**

- Formed by:
 - Between Frontal and Ethmoid bone
- Contents:
 1. Posterior Ethmoid Nerve.
 2. Posterior Ethmoid Vessels.

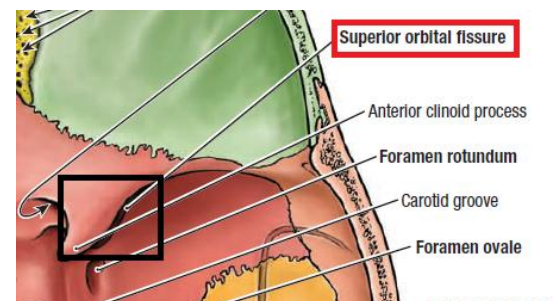
○ **Optic Canal:**

- Formed by:
 - Sphenoid bone
- Contents:
 1. Optic Nerve (**CN-II**)
 2. Ophthalmic Artery.



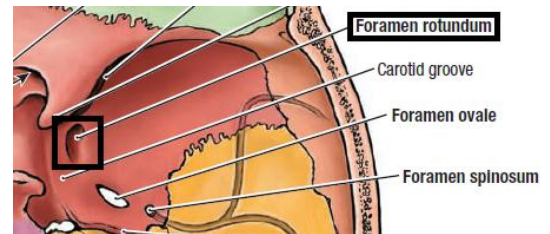
○ **Superior Orbital Fissure:**

- Formed by:
 - Between Greater and Lesser wings of Sphenoid.
- Contents:
 1. Oculomotor (**CN-III**) (Upper+Lower)
 2. Trochlear Nerve (**CN-IV**).
 3. Ophthalmic Nerve (**V1**) (Frontal+Lacrimal+Nasocilliary)
 4. Abducens Nerve (**CN-VI**)
 5. Ophthalmic vein (Superior+Inferior).



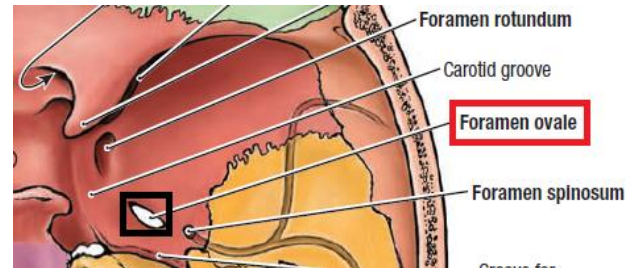
○ **Foramen Rotundum:**

- Formed by:
 - Sphenoid.
- Contents:
 1. Maxillary Nerve (**V2**)



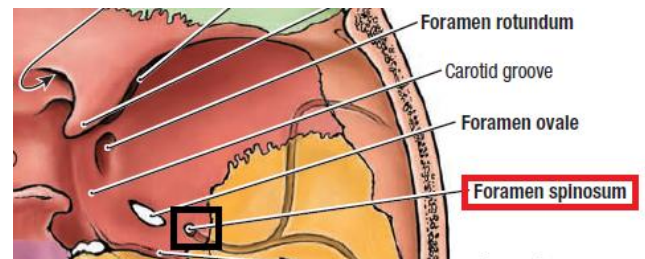
○ **Foramen Ovale:**

- Formed by:
 - Sphenoid.
- Contents:
 1. Mandibular Nerve (**V1**)
 2. Accessory Meningeal Artery.
 3. Lesser Petrosal Nerve.
 4. Emissary Veins.



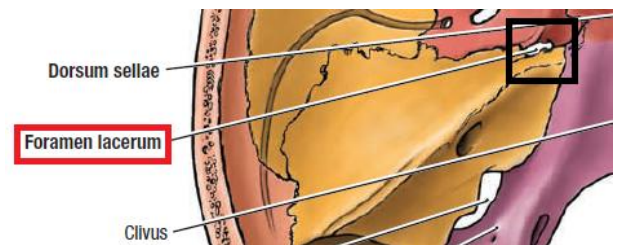
○ **Foramen Spinosum:**

- Formed by:
 - Sphenoid.
- Contents:
 1. Middle Meningeal Vessels.
 2. Meningeal branch of Mandibular Nerve (**V1**)



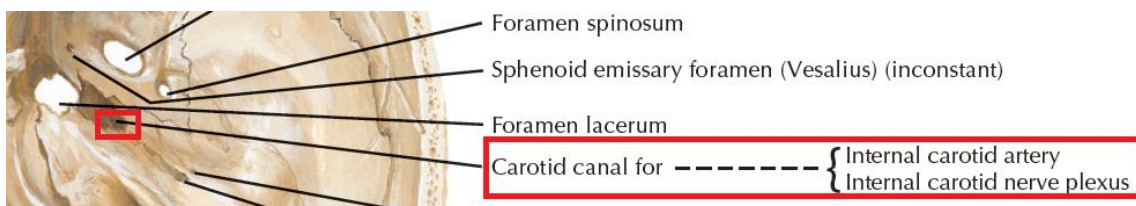
○ **Foramen Lacerum:**

- Formed by:
 - Articulation of Greater wing and body of Sphenoid + Petrous portion of Temporal + Basilar portion of Occipital bone
- **No Contents**



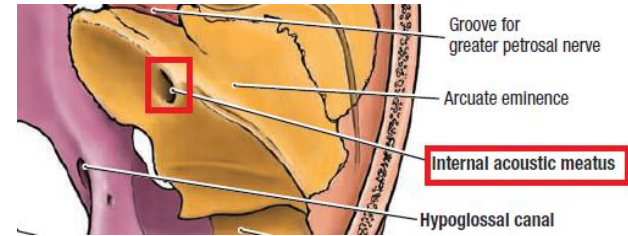
○ **Carotid Canal:**

- Formed by:
 - Petrous portion of Temporal Bone.
- Contents:
 1. Internal Carotid Artery.
 2. Internal Carotid Plexus (Sympathetics).



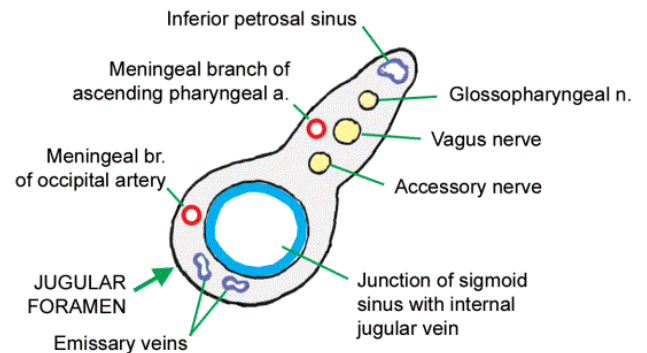
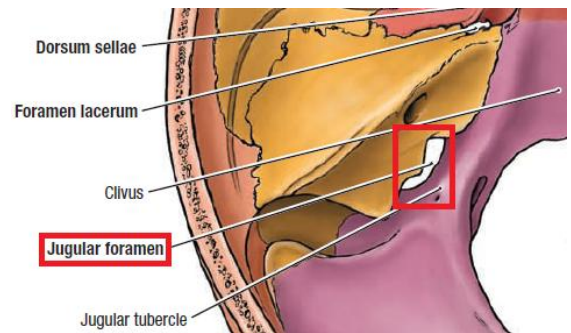
○ **Internal Acoustic Meatus:**

- Formed by:
 - Petrous portion of Temporal Bone.
- Contents:
 1. Facial Nerve (**CN-VII**)
 2. Vestibulocochlear Nerve (**CN-VIII**)
 3. Labyrinthine Artery.



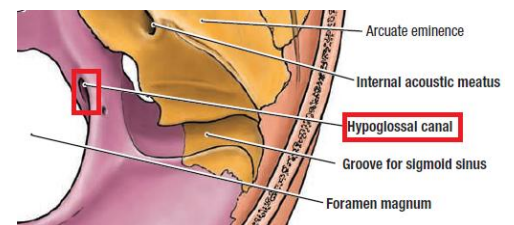
○ **Jugular Foramen:**

- Formed by:
 - Petrous portion of Temporal Bone + Occipital.
- **Anterior-medial compartment:**
 1. Inferior Petrosal Sinus
- **Intermediate compartment:**
 1. Glossopharyngeal Nerve (**CN-IX**)
 2. Vagus Nerve (**CN-X**)
 3. Spinal Accessory Nerve (**CN-XI**)
- **Posterior-lateral compartment:**
 1. Sigmoid Sinus
 2. Posterior Meningeal Artery.



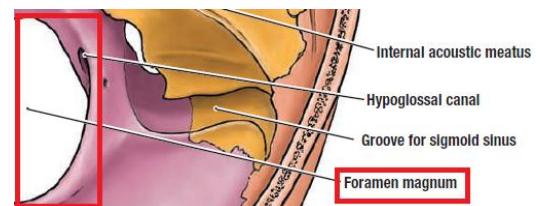
○ **Hypoglossal Canal:**

- Formed by:
 - Occipital Bone.
- Contents:
 1. Hypoglossal Nerve (**CN-XII**)



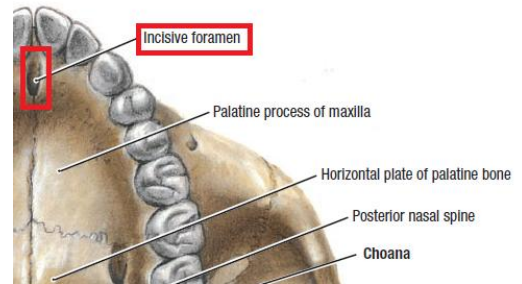
○ **Foramen Magnum:**

- Formed by:
 - Occipital Bone.
- Contents:
 1. Medulla oblongata
 2. Vertebral Arteries
 3. Spinal roots of Spinal Accessory Nerve (**CN-XI**)



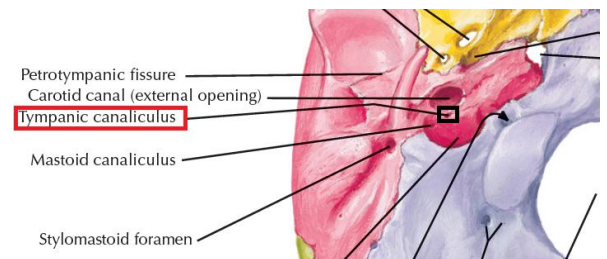
○ **Incisive Foramen:**

- Formed by:
 - Palatine Process of Maxilla.
- Contents:
 1. Nasopalatine Nerve.
 2. Sphenopalatine Artery.



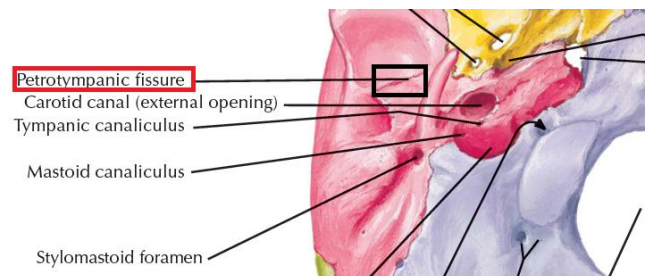
○ **Tympanic Canaliculus:**

- Formed by:
 - Temporal bone.
- Contents:
 1. Tympanic branch of Glossopharyngeal Nerve (**CN-IX**)



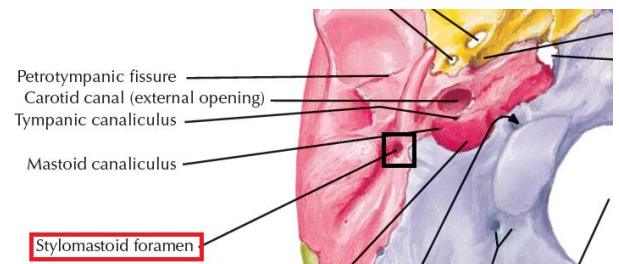
○ **Petrotympanic Fissure:**

- Formed by:
 - Temporal bone.
- Contents:
 1. Chorda Tympani Nerve.
 2. Anterior Tympanic Artery.



○ **Stylomastoid Foramen:**

- Formed by:
 - Temporal bone.
- Contents:
 1. Facial Nerve (**CN-VII**)
 2. Stylomastoid Artery.



- **Supraorbital Foramen:**

- Formed by:
 - Frontal bone.
- Contents:
 1. Supraorbital Nerve.
 2. Supraorbital Vessels

- **Infraorbital Fissure:**

- Formed by:
 - Between Greater wing of sphenoid and Maxilla and Orbital postion of Palatine bone.
- Contents:
 1. Maxillary Nerve (**V2**).
 2. Zygomatic Nerve.
 3. Infraorbital Vessels.

- **Zygomaticofacial Foramen:**

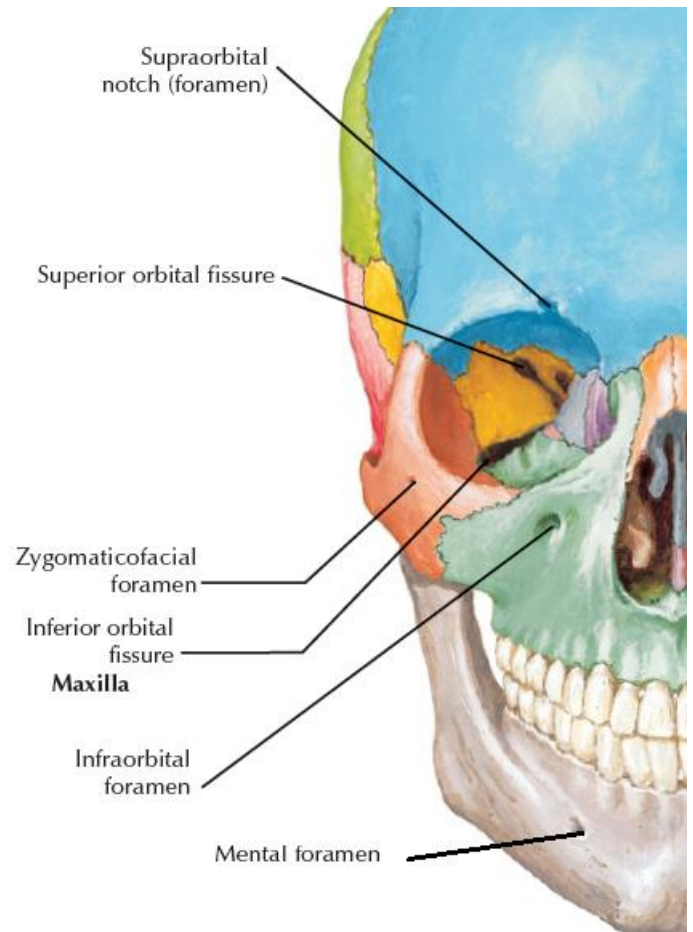
- Formed by:
 - Zygomatic bone.
- Contents:
 1. Zygomaticofacial Nerve.
 2. Zygomaticofacial Vessels.

- **Infraorbital Foramen:**

- Formed by:
 - Maxilla bone.
- Contents:
 1. Infraorbital Nerve
 2. Infraorbital Vessels

- **Mental Foramen:**

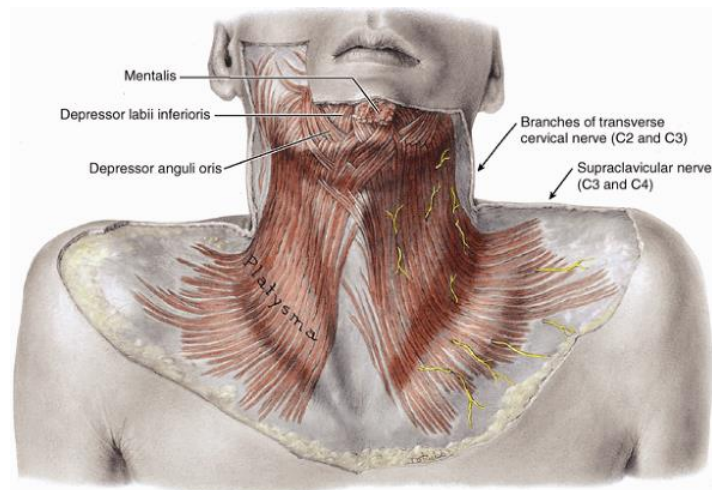
- Formed by:
 - Mandible bone.
- Contents:
 1. Mental Nerve.
 2. Mental Vessels.



Anatomy of Cervical Muscles:

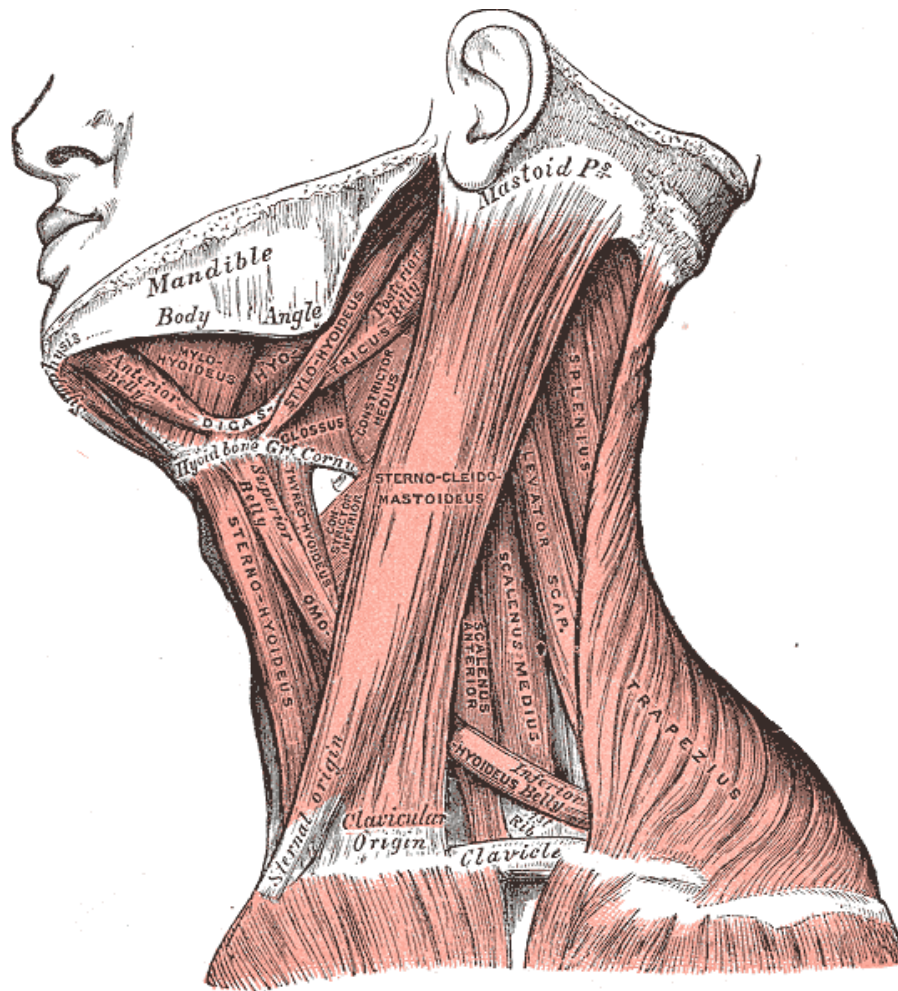
Platysma:

- Platysma is a broad sheet arising from the fascia covering the upper parts of the Pectoralis major and Deltoid.
- Superficial Fascia of the neck is a thin lamina investing the Platysma.
- Platysma fibers cross the clavicle, and proceed obliquely upward and medialward along the side of the neck.
- Anterior fibers interlace, below and behind the symphysis menti, with the fibers of the opposite side; the posterior fibers cross the mandible, inserted into the bone below the oblique line, into the skin and subcutaneous tissue of the lower part of the face.
- Under Platysma, the external jugular vein descends from the angle of the mandible to the clavicle.
- Variations occur in the extension over the face and over the clavicle and shoulder; it may be.
- **Nerve:** supplied by the cervical branch of the facial nerve.
- **Actions:** When the entire Platysma is in action it produces a slight wrinkling of the surface of the skin of the neck in an oblique direction. Its anterior portion, the thickest part of the muscle, depresses the lower jaw; it also serves to draw down the lower lip and angle of the mouth in the expression of sadness.



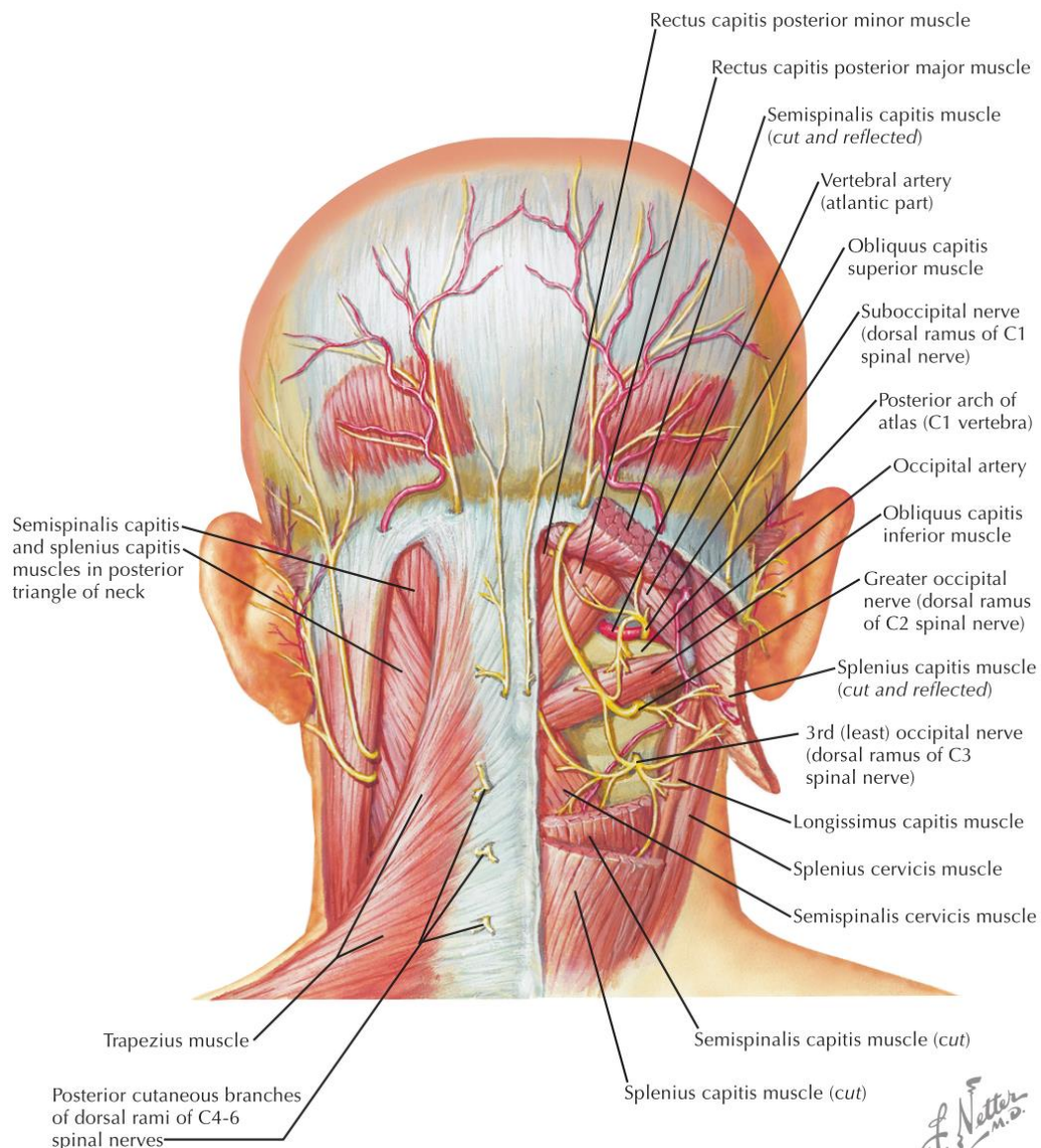
Sternocleidomastoid muscle:

- Medial (sternal) head, tendinous, arises from the upper part of the anterior surface of the manubrium sterni. Lateral (clavicular) head, fleshy, arises from the superior border and anterior surface of the medial third of the clavicle.
- Inserted, by a strong tendon, into the lateral surface of the mastoid process, and by a thin aponeurosis into the lateral half of the superior nuchal line of the occipital bone.
- **Nerves:** Spinal accessory nerve (CN XI) and branches from C2 and C3.
- **Actions:** it draws the head toward the shoulder of the same side, rotates the face toward the opposite side. Acting together, the muscles will flex the neck. If the head be fixed, the two muscles assist in elevating the thorax in forced inspiration.



Trapezius:

- It arises from: external occipital protuberance and medial third of the superior nuchal line of the occipital bone, ligamentum nuchæ, spinous process of the seventh cervical, and spinous processes of all the thoracic vertebræ.
- Inserted into posterior border of the lateral third of the clavicle; acromion, and spine of scapula.
- **Nerve:** Spinal accessory nerve (CN XI) and branches from C3 and C4.
- **Action:** it elevates shoulder, rotates scapula, and draws head to one side .



Suprahyoid muscles:

1. Digastric muscle:

- Posterior belly, longer than the anterior, arises from the mastoid notch and passes downward and forward.
- Anterior belly arises from a depression on the inner side of the lower border of the mandible, close to the symphysis, and passes downward and backward.
- The two bellies end in an intermediate tendon which is connected with body of the hyoid bone by a broad aponeurotic layer (suprahyoid aponeurosis).
- **Nerves:**
- **Posterior belly:** (Facial Nerve)
- **Anterior belly:** (Myelohyoid branch of inferior alveolar N, of V3).

2. Stylohyoid muscle:

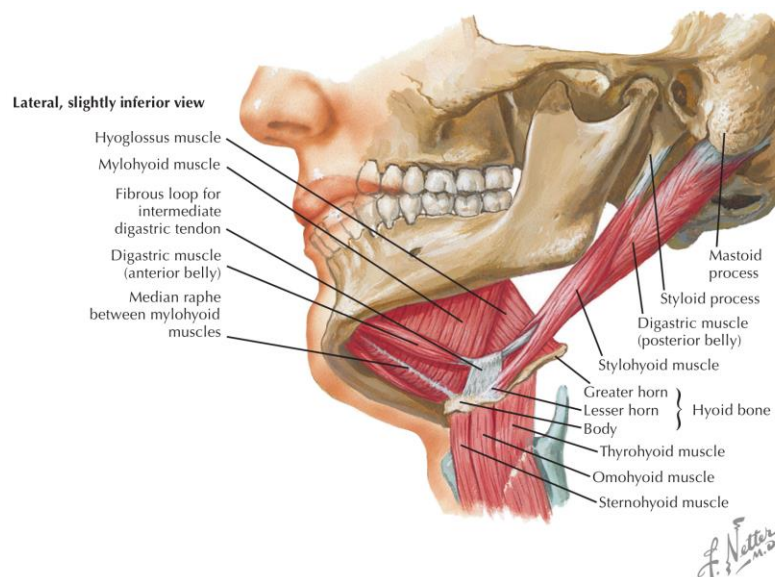
- It arises from lateral surface of styloid process, near the base.
- Passing downward and forward, in front of, and above the posterior belly of the Digastric.
- Inserted into body of the hyoid bone.
- **Stylohyoid Ligament** is a fibrous cord, which is attached to the tip of the styloid process and the lesser cornu of the hyoid bone. It frequently contains a little cartilage in its center, which is often partially ossified.
- **Nerves:** Facial Nerve.

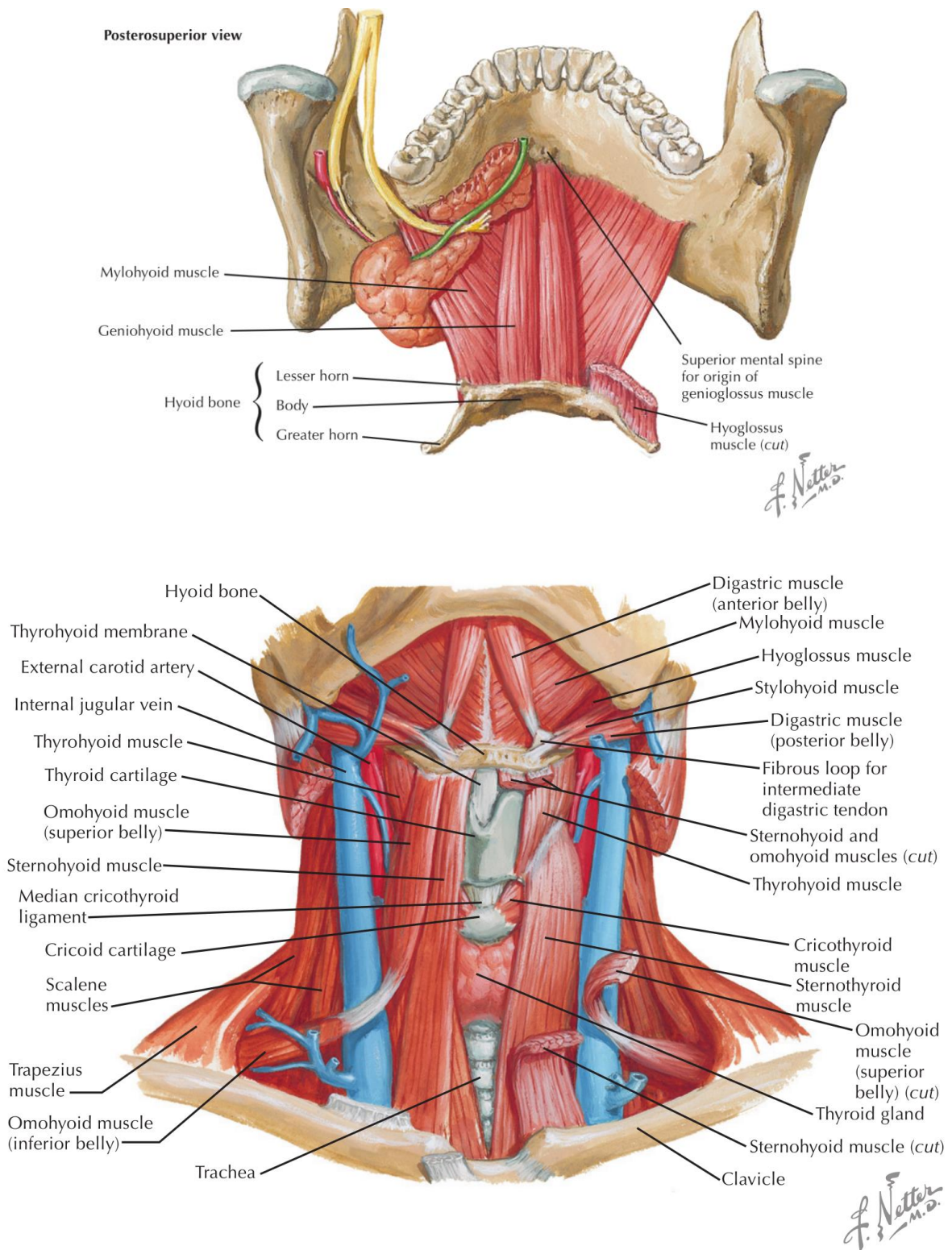
3. **Mylohyoid muscle:**

- It arises from the mylohyoid line of the mandible (extending from the symphysis in front to the last molar tooth behind).
- Forms, with its fellow of the opposite side, a muscular floor (diaphragm) for the oral cavity.
- Posterior fibers inserted into body of the hyoid bone, while anterior fibers inserted into a median fibrous raphé extending from the symphysis menti to the hyoid bone.
- **Nerves:** Mylohyoid branch of inferior alveolar N, of V3.

4. **Geniohyoid muscle:**

- Narrow muscle, situated above the medial border of the Mylohyoid.
- It arises from the back of the symphysis menti (inferior mental spine).
- Inserted into the anterior surface of the body of the hyoid bone.
- **Nerves:** Hypoglossal N. (CN XII).
- **Actions:** Suprahyoid muscles arise the hyoid (with base of the tongue) during the act of deglutition. When the hyoid bone is fixed by its depressors and those of the larynx, they depress the mandible.





Infrahyoid muscles (Strap muscles):

1. Sternohyoid muscle:

- Arises from posterior sternoclavicular ligament, and upper and posterior part of manubrium.
- Inserted into the lower border of the body of the hyoid bone.

2. Sternothyroid muscle:

- Arises from the posterior surface of the manubrium.
- Inserted into the oblique line on the lamina of the thyroid cartilage.

3. Thyrohyoid muscle:

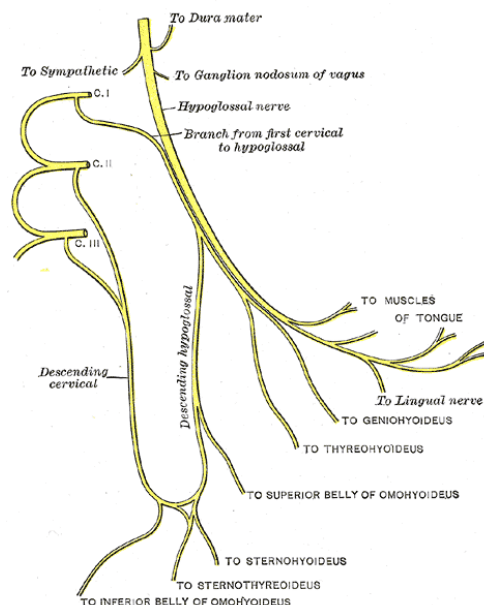
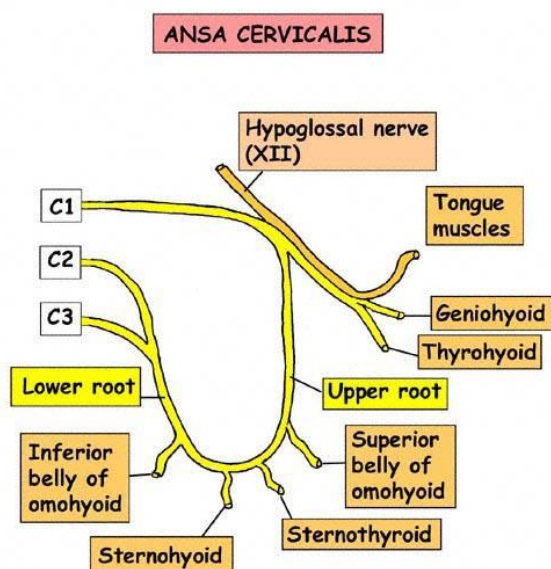
- Arises from the oblique line on the lamina of the thyroid cartilage.
- Inserted into the lower border of the greater cornu of the hyoid bone.

4. Omohyoid muscle:

- Inferior belly arises from upper border of the scapula, runs forward and upward. Behind the Sternocleidomastoid, it becomes tendinous and changes its direction, forming an obtuse angle with superior belly, which passes almost vertically upward, to be inserted into the lower border of the body of the hyoid bone.
- Central tendon varies much in length, and is held in position by a process of the deep cervical fascia, which sheaths it.

Nerve supply to strap muscles: Ansa Hypoglossi:

- C1, and C2 give a branch that joins the hypoglossal (CN XII) trunk, runs with it some distance, and sends off a branch to the Thyrohyoid; it then leaves the hypoglossal to form the descendens hypoglossi and unites with the communicantes cervicales from C3 to form ansa hypoglossi from which nerves pass to the other infrahyoid muscles.



Anterior Vertebral Muscles:

1. Longus colli:

- Between the atlas and the third thoracic vertebra.

2. Longus capitis:

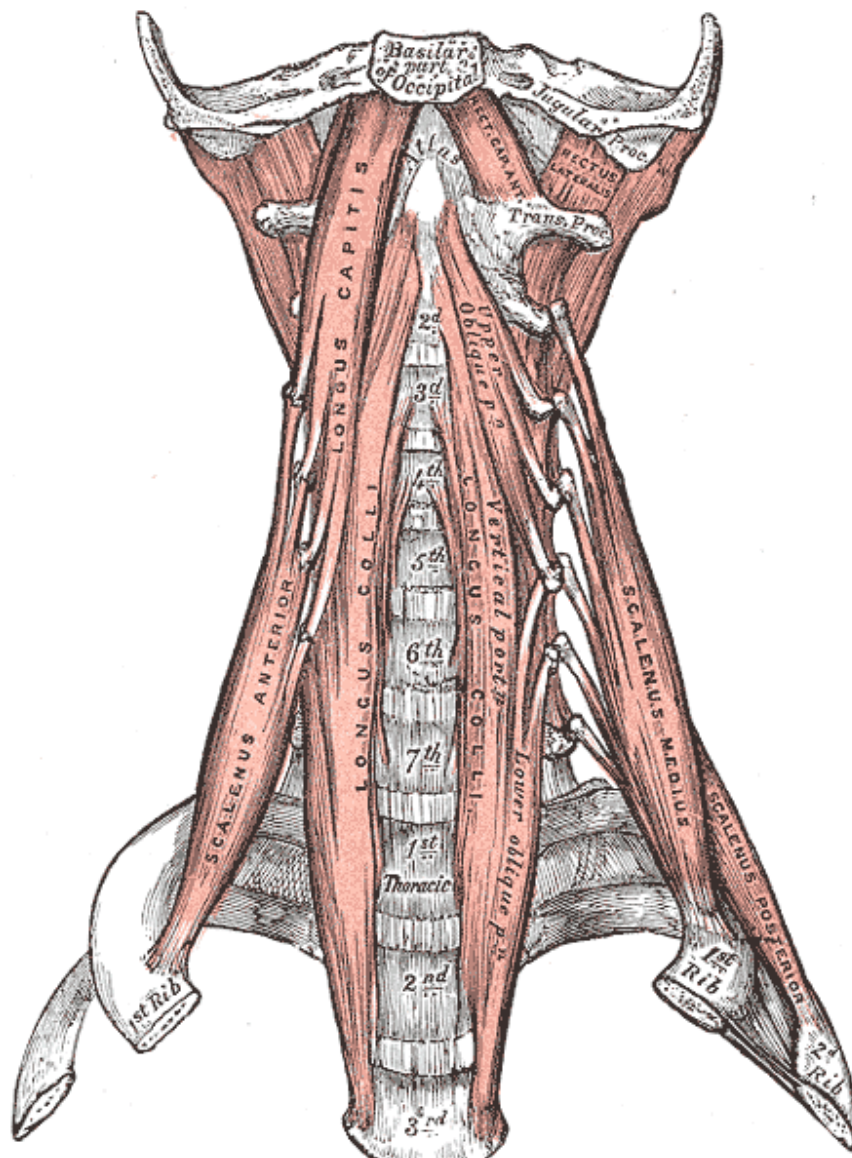
- Between occipital bone and sixth cervical vertebra.

3. Rectus capitis anterior & lateralis:

- Short muscles, lies behind longus capitis, runs from atlas to occipital bone.

- Nerves: C1, C2, C3.

- **Actions:** They act as direct antagonists of the muscles at the back of the neck, serving to restore the head to its natural position. These muscles also flex the head.



Lateral Vertebral Muscles:

1. Scalenus anterior, and medium, and posterior:

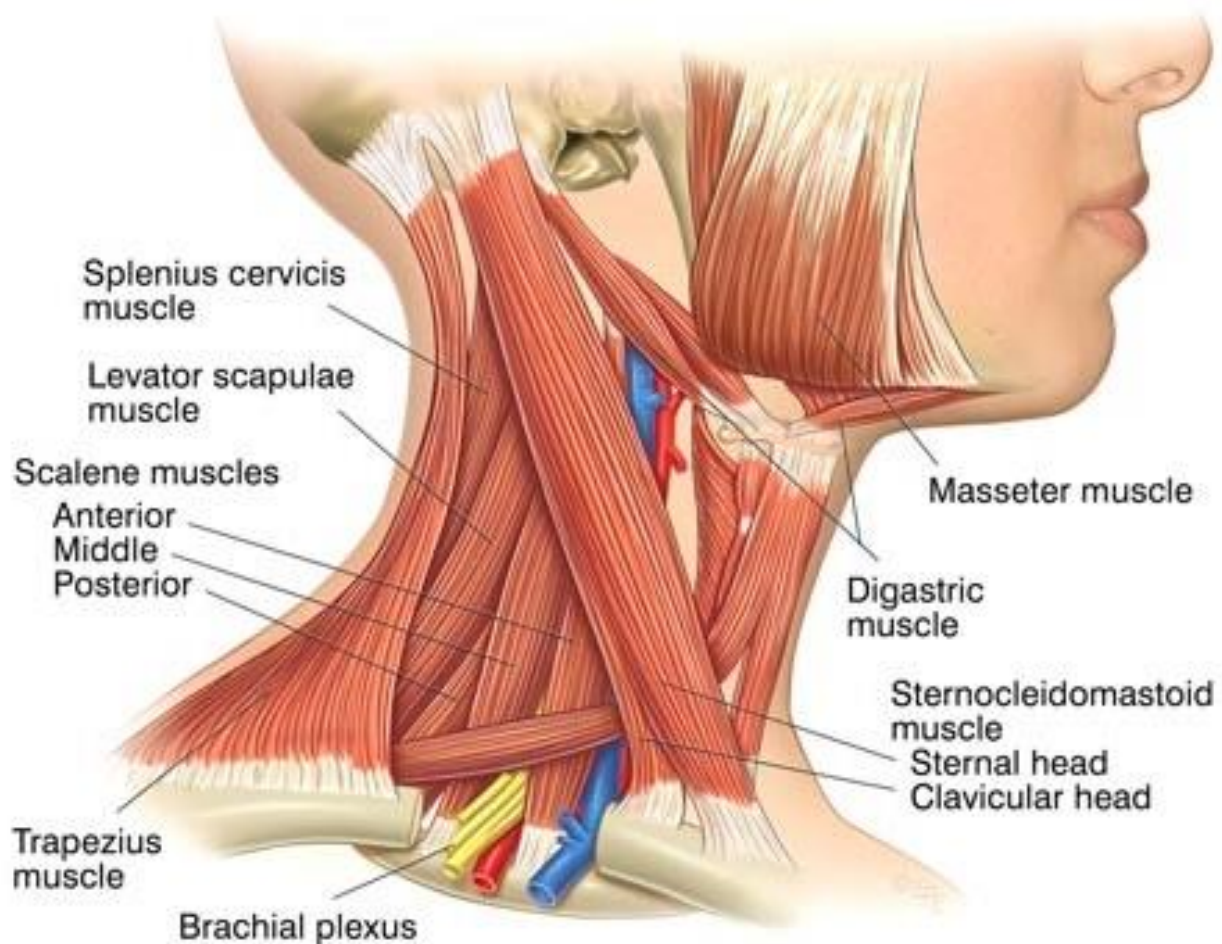
- From transverse processes of cervical vertebræ (3-6) to the scalene tubercle on the inner border of the first rib.

2. Splenius capitis.

3. Levator scapulæ

- **Nerves: C2 – C7.**

- **Actions:** elevate the first rib, and are, therefore, inspiratory muscles.



Muscles of Mastication:

1. Masseter:

- Superficial portion, arises by a thick, tendinous aponeurosis from the zygomatic process of the maxilla, and the zygomatic arch; and it is inserted into the lower part of lateral surface of the ramus of the mandible.
- Deep portion, is inserted into the upper half of the ramus and the lateral surface of the coronoid process.

2. Temporalis:

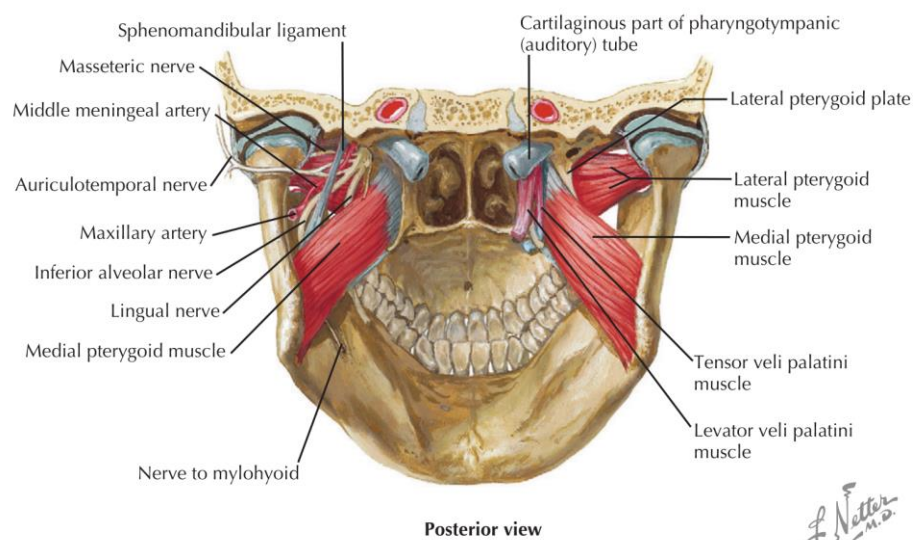
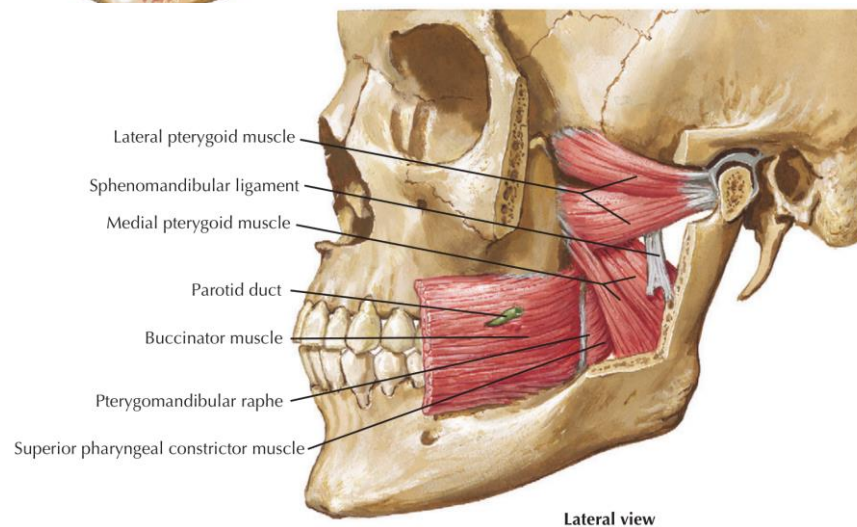
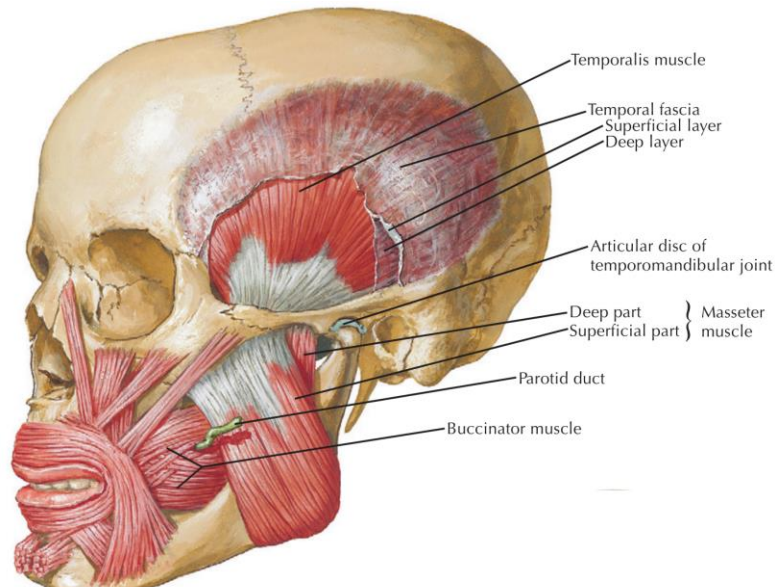
- It arises from the whole of the temporal fossa.
- Its fibers end in a tendon, which passes deep to the zygomatic arch and is inserted into the coronoid process, and the anterior border of the ramus of the mandible.

3. Lateral pterygoid:

- Upper head: arises from lower part of the lateral surface of the great wing of the sphenoid, and lower head from the lateral surface of the lateral pterygoid plate.
- Its fibers pass backward and lateralward, to be inserted in front of neck of the mandibular condyle.

4. Medial pterygoid:

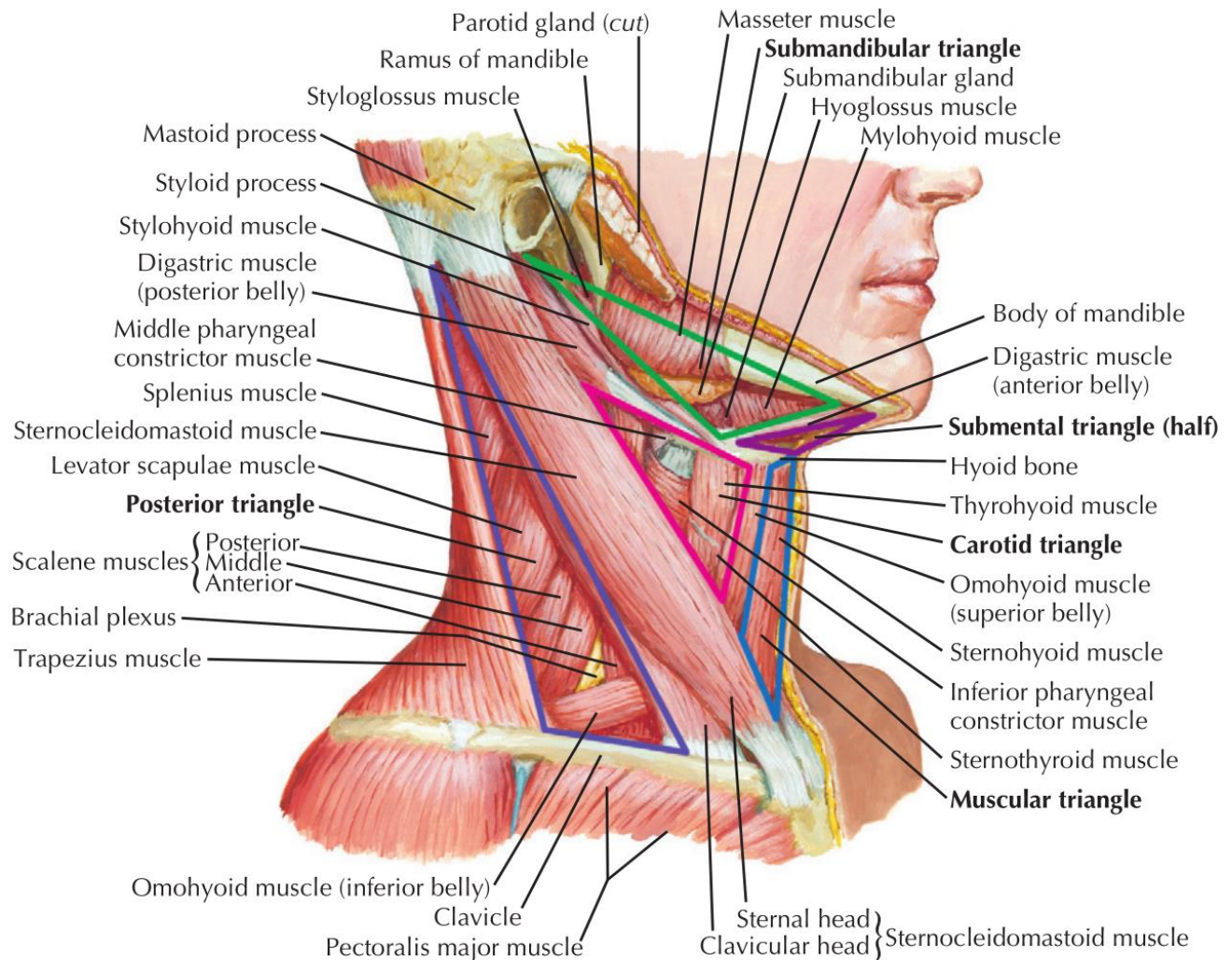
- It arises from the medial surface of the lateral pterygoid plate.
- Its fibers pass downward, lateralward, and backward, and are inserted into the lower medial surface of the ramus (it forms, with its facial envelop and the ramus of mandible, the lateral boundary of the parapharyngeal space).
- **Nerves:** V3.
- **Actions:** raise the mandible against the maxilla, and draw forward the mandible.



Triangles of the Neck

Anterior Triangle:

- Bounded, in front, by the midline of the neck; behind, by the anterior margin of the Sternocleidomast; its base, directed upward, is formed by the lower border of the body of the mandible, and a line extending from the angle of the mandible to the mastoid process; its apex is below, at the sternum.
- This space is subdivided into four smaller triangles by the digastric above, and the superior belly of the Omohyoid below.



1. Carotid Triangle:

- Bounded, behind by the Sternocleidomastoid; below, by the superior belly of the Omohyoideus; and above, by the Stylohyoid and the posterior belly of the Digastric.
- Its floor is formed by parts of the Thyrohyoid, Hyoglossus, and the middle and inferior constrictores.
- Contains:
 - o Arteries: common carotid artery, which bifurcates opposite the upper border of the thyroid cartilage into the external, internal carotid (external being the more anterior), branches of external carotid (superior thyroid, lingual, external maxillary, occipital, ascending pharyngeal).
 - o Veins: internal jugular, (lateral to common and internal carotid arteries), and veins corresponding to branches of the external carotid.
 - o Nerves: descendens hypoglossi (in front of carotid sheath), hypoglossal nerve (crosses both the internal and external carotids), vagus nerve (within the sheath, between the artery and vein, and behind both), accessory nerve (on lateral side of sheath, before it pierces the Sternocleidomast), internal branch of superior laryngeal nerve (on the medial side of the external carotid, just below the hyoid bone), and external branch of superior laryngeal nerve (more inferiorly).
 - o Viscera: upper portion of the larynx and lower portion of the pharynx.

2. Muscular Triangle:

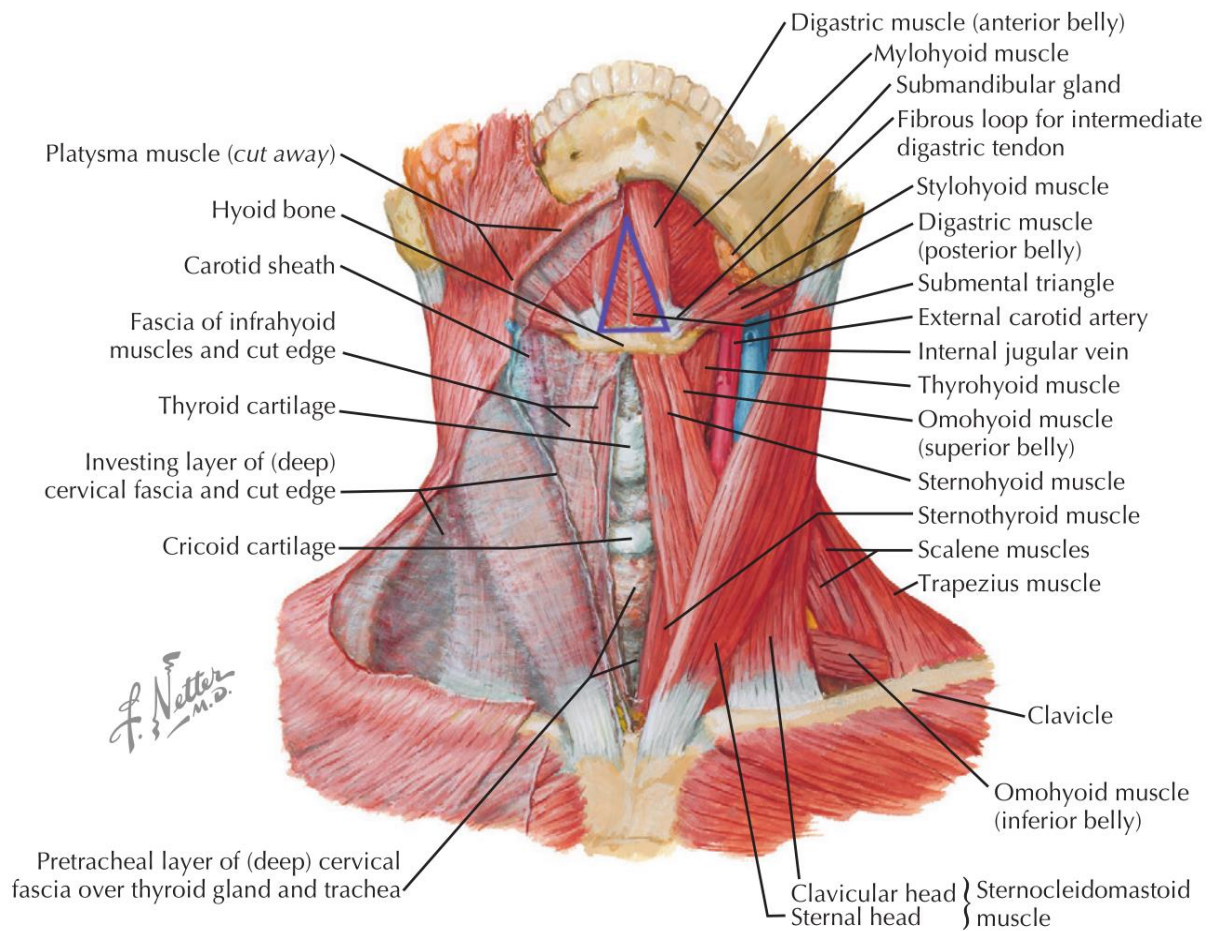
- Bounded, in front, by the midline of the neck; behind, by the anterior margin of the Sternocleidomastoid; above, by the superior belly of the Omohyoid.
- Floor is formed by Sternohyoid and Sternothyroid,
- Contains:
 - o Carotid sheath (covered by sternomastoid) ansa hypoglossi (in front of sheath); inferior thyroid artery, recurrent laryngeal nerve, and sympathetic trunk(all behind sheath).

3. Submandibular (Digastric) Triangle:

- Bounded, above, by the lower border of the body of the mandible, and a line drawn from its angle to the mastoid process; below, by the posterior belly of the Digastric and the Stylohyoid; in front, by the anterior belly of the Digastric.
- Its floor is formed by the Mylohyoid, Hyoglossus, and superior constrictor.
- It is divided by stylomandibular ligament into:
 - o **Anterior part:**
 - Contains submandibular gland, superficial to which is anterior facial vein, beneath the gland, facial (external maxillary) artery and its glandular branches; on the surface of the Mylohyoid, are the submental artery and mylohyoid artery and nerve.
 - o **Posterior part:**
 - Related to the tail of parotid gland with its piercing structures.

4. Submental (Suprahyoid) Triangle:

- Limited behind by the anterior belly of the Digastric, in front by the middle line of the neck between the mandible and the hyoid bone; below, by the body of the hyoid bone; its floor is formed by the Mylohyoideus. It contains one or two submental lymph glands and some small veins; the latter unite to form the anterior jugular vein.



Posterior Triangle:

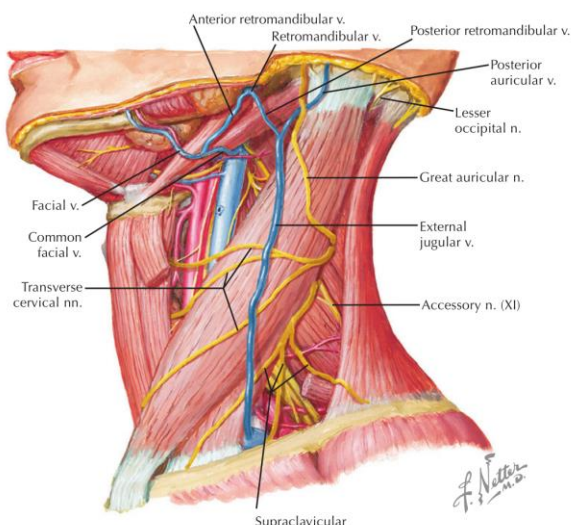
- Bounded, in front, by the Sternocleidomastoid; behind, by the anterior margin of the Trapezius; its base is formed by the middle third of the clavicle; its apex, by the occipital bone.
- Crossed, about 2.5 cm. above the clavicle, by the **inferior belly of the Omohyoideus**, which divides it into two triangles, an upper or occipital, and a lower or subclavian.

1. Occipital Triangle,

- Its floor is formed from above downward by the Splenius capitis, Levator scapulæ, and the Scaleni medius and posterior.
- Accessory nerve is directed obliquely across the space from the substance of Sternocleidomastoid, to the under surface of the Trapezius. Below, transverse cervical vessels and the upper part of the brachial plexus cross the space. A chain of lymph glands is also found running along the posterior border of the Sternocleidomastoideus, from the mastoid process to the root of the neck.

2. Subclavian Triangle:

- Its floor is formed by the first rib and Serratus anterior.
- Crossed by supraclavicular nerves, third portion of the subclavian artery across the first rib, to the axilla.



- **Blood Supply of Head and Neck:**

- Major arteries of Neck are:
 - o Common Carotid Artery
 - o Subclavian Artery

- **Common Carotid Artery:**

- **Right Common Carotid Artery:**

- o Begins at bifurcation of Brachiocephalic (innominate) Artery behind Sternoclavicular joint.

- **Left Common Carotid Artery:**

- o Arises directly from Arch of Aorta.

- Ascend obliquely upward within Carotid Sheath to **Upper border of Thyroid cartilage**, where it divides into External and Internal Carotid Arteries.

- **Carotid Sinus:**

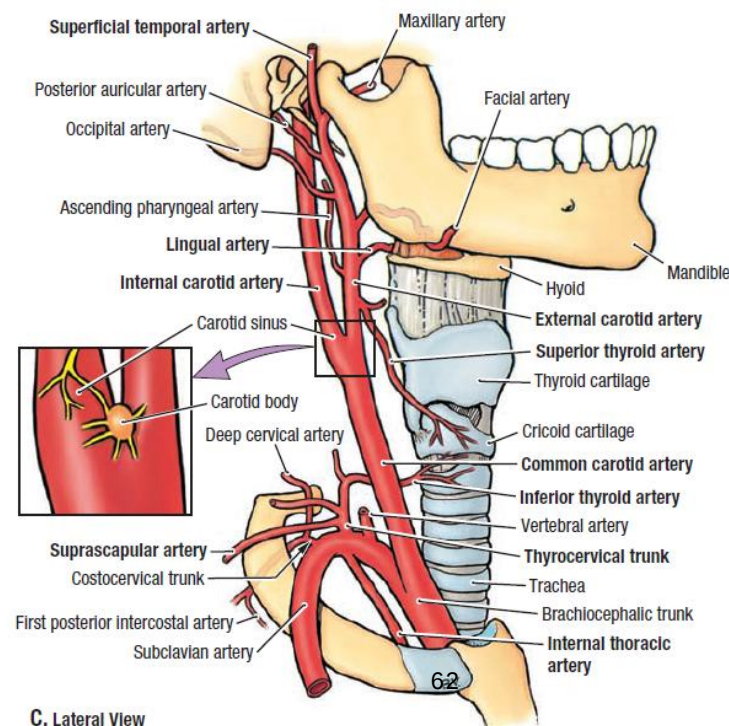
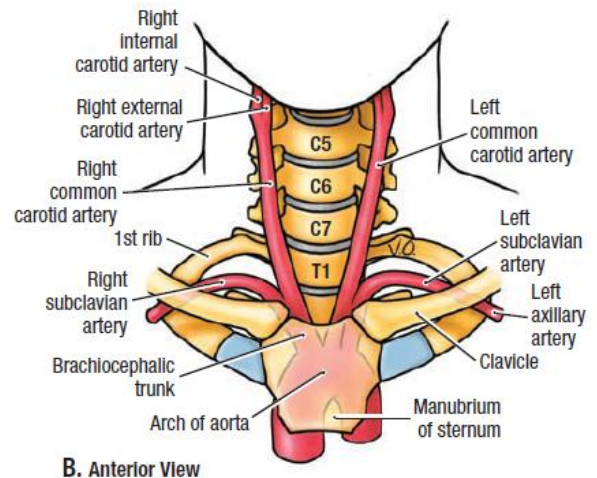
- o Baroreceptor located in Dilation of proximal part of Internal carotid.
- o receives its sensory innervations from Glossopharyngeal nerve (**CN-IX**)
- o Reacts to changes in Arterial blood pressure.

- **Carotid Body:**

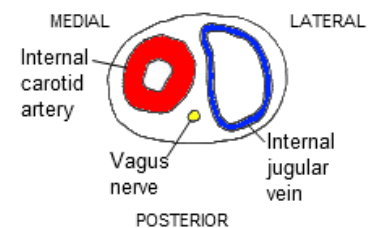
- o Chemoreceptor lies at Bifurcation of Common carotid.
- o Monitors level of Oxygen in blood

- **Carotid sheath encloses:**

1. Common Carotid artery (**Medial**)
2. Vagus nerve (**CN-X**) (**Middle + Posterior**)
3. Internal Jugular vein. (**Lateral**)

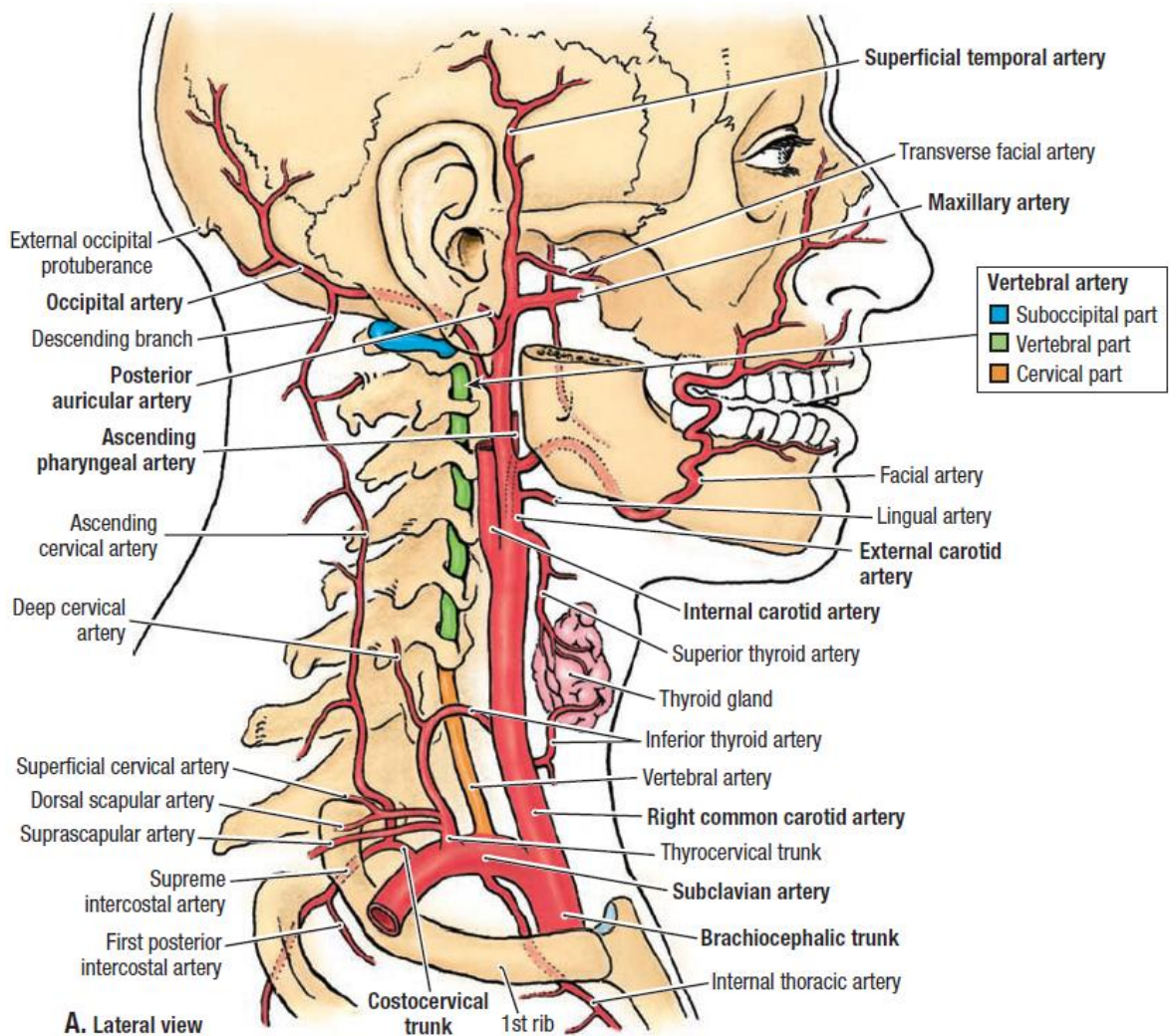


CAROTID SHEATH

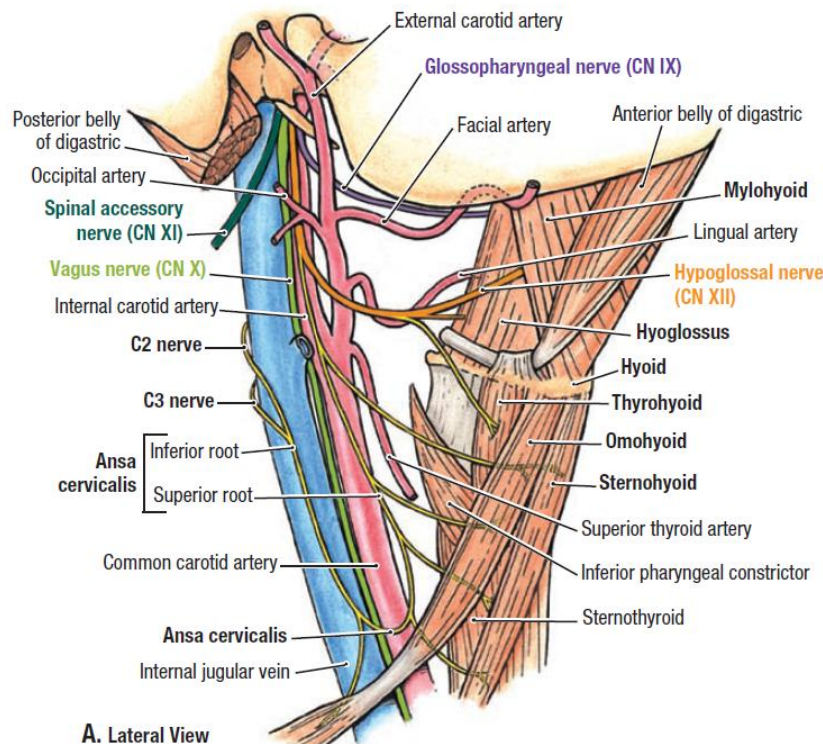


C. Lateral View

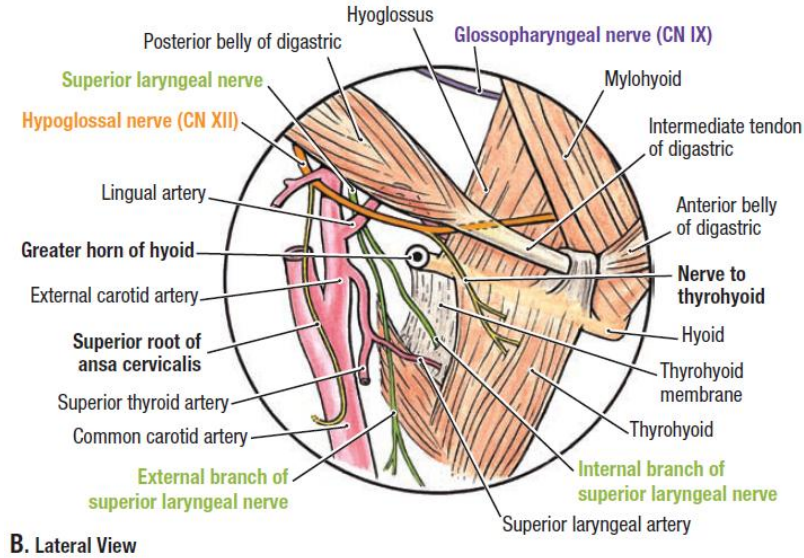
- **Relations of Common Carotid Artery in Neck:**
- Crossed by:
 1. Sternocleidomastoid branch of Superior Thyroid Artery.
 2. Superior and Middle thyroid veins which end in the internal jugular.
 3. Anterior jugular vein just above the clavicle.
 4. Inferior thyroid artery crosses behind the lower part of the vessel.
- Sympathetic trunk interposed **posteriorly** between the artery and anterior vertebral muscles.
- Superior root of Ansa Cervicalis nerve descends **in front** of its sheath.
- Branches of Common Carotid Artery in Neck:
 - **NO Branches.**



- **External Carotid Artery:**
- One of terminal branches of Common Carotid Artery.
- Begins:
 - o At level of **Upper thyroid Cartilage**.
- Course:
 - o Passes upward and forward.
 - o **Medial to Internal Carotid artery.**
 - o External to the carotid sheath
- Terminates:
 - o **Behind Neck of mandible**
 - o Dividing into Superficial temporal artery and Maxillary Artery.
- Supplies:
 - o Supplies most structures External to cranium **Except** (Orbit, Part of forehead and scalp supplied by Ophthalmic artery of internal carotid)

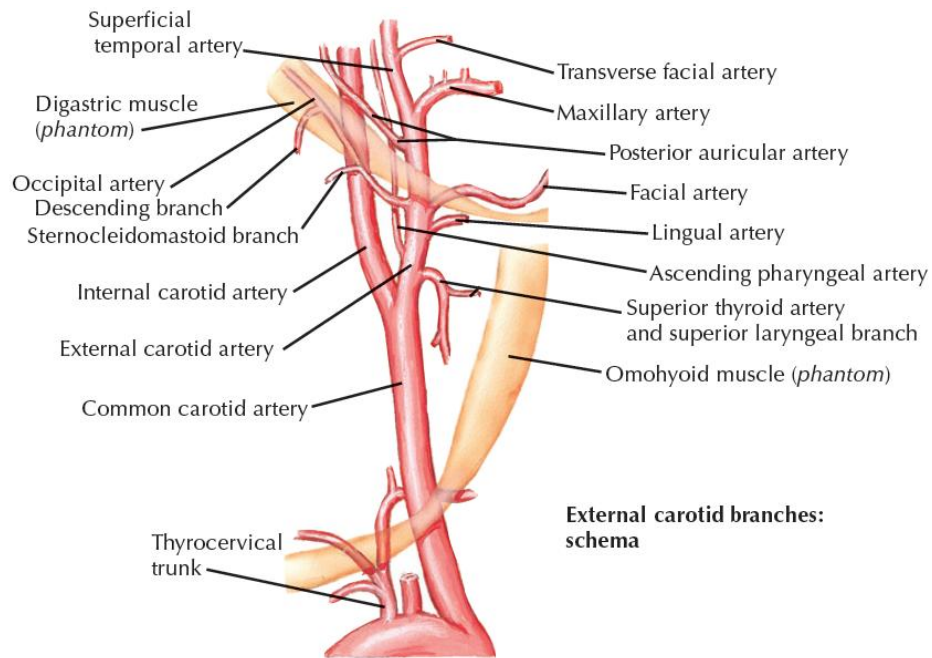


- **Relations of External Carotid Artery:**
- Crossed by:
 1. Lingual vein
 2. Common facial vein
 3. Superior thyroid vein
 4. Hypoglossal nerve
 5. Digastric and Stylohyoid.
- Lies deep to:
 1. Retromandibular vein
 2. Facial nerve
 3. External jugular vein.

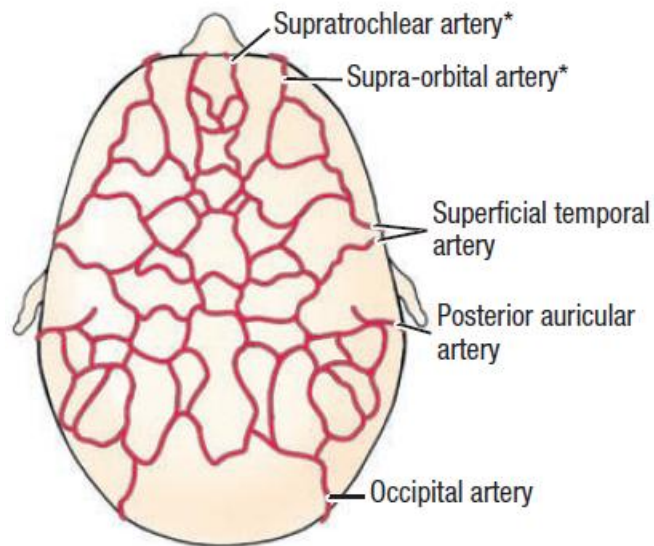


- Braches of External Carotid Artery:

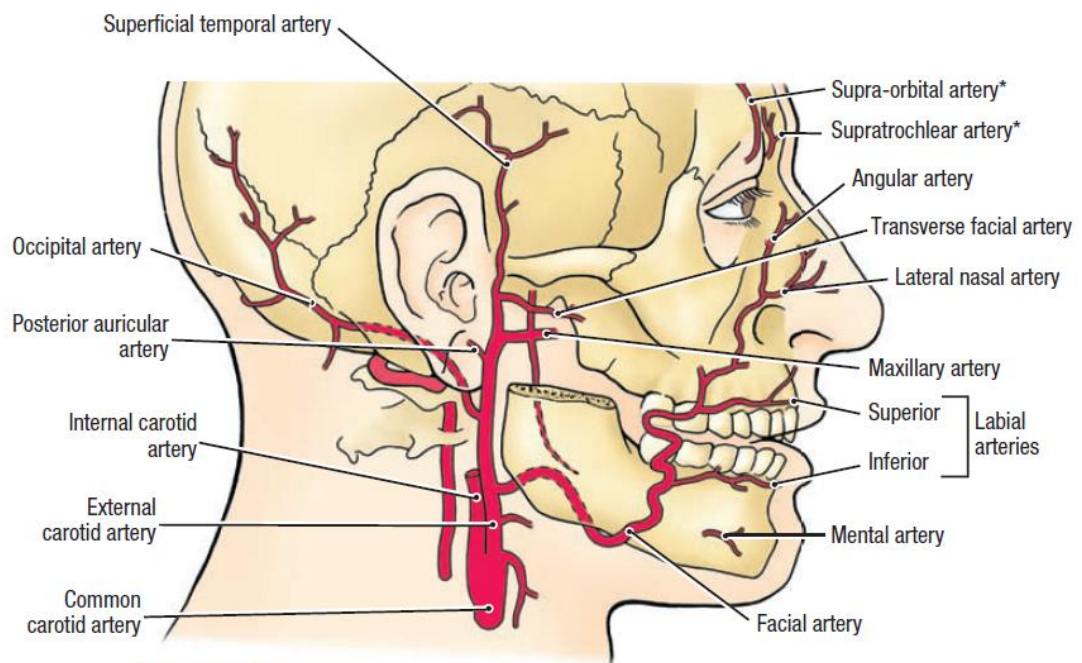
1. Superior Thyroid Artery.
2. Ascending Pharyngeal Artery.
3. Lingual Artery.
4. Facial Artery.
5. Occipital Artery.
6. Posterior Auricular Artery.
7. Maxillary Artery.
8. Superficial Temporal Artery.



- **Superior Thyroid Artery:**
- 1st branch of External Carotid Artery.
- Arises just below Greater cornu of Hyoid
- Passes inferiorly along Inferior constrictor muscle to reach upper pole of Thyroid gland.
- Branches:
- 1. **Superior Laryngeal Artery:**
 - o Perforates Thyrohyoid membrane to supply the larynx.
 - o Accompanied with Internal branch of Superior laryngeal nerve.
- **Ascending Pharyngeal Artery:**
- Smallest branch of External carotid Artery.
- Arises from Posterior portion of External carotid Artery.
- Ascends superiorly between Pharynx and Internal Carotid Artery.
- Supplies Pharyngeal Muscles.
- **Lingual Artery:**
- Arises in Carotid Triangle.
- Passes superiorly and medially toward Greater cornu of Hyoid.
- Loops by passing anteriorly and inferiorly while traveling superficial to Middle constrictor muscle.
- Crossed superficially by Hypoglossal nerve (**CN-XII**).
- Supplies the Tongue.
- **Facial Artery:**
- Arises in Carotid Triangle.
- Passes superiorly immediately deep to posterior belly of Digastric and Stylohyoid muscles.
- Passes along Submandibular gland.
- Passes superiorly over Body of Mandible at Masseter muscle in a tortuous pattern.
- Passes forward and upward, immediately beneath Platysma (deep to SMAS).
- Cross Cheek to Angle of mouth and ascends along side of nose.
- Ends at Medial commissure of the eye as **Angular artery**.
- Supplies the Face.
- Branches in Neck:
- **Tonsillar (perforating superior constrictor) – Glandular – Submental.**
- Branches in Face:
- **Inferior Labial – Superior Labial – Lateral Nasal – Angular – Muscular.**
- Branches:
- **Occipital Artery:**
- Arises in Carotid Triangle.
- Opposite to Facial Artery.
- Passes upward and reaches Back of Scalp.
- The terminal part of the artery is accompanied by Greater occipital nerve.



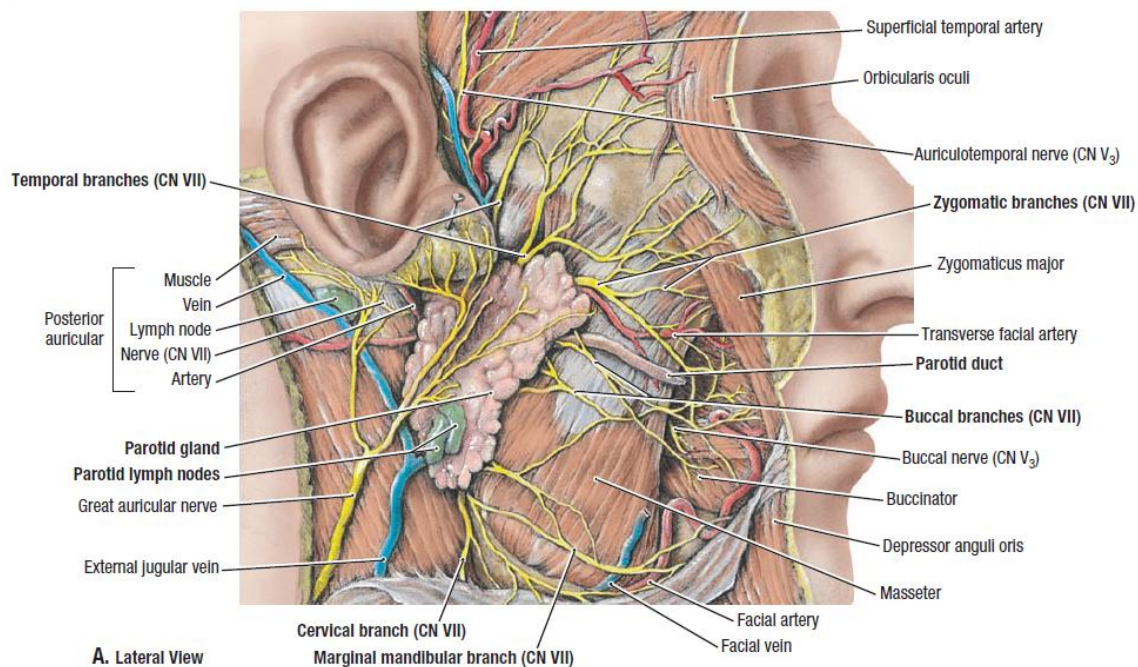
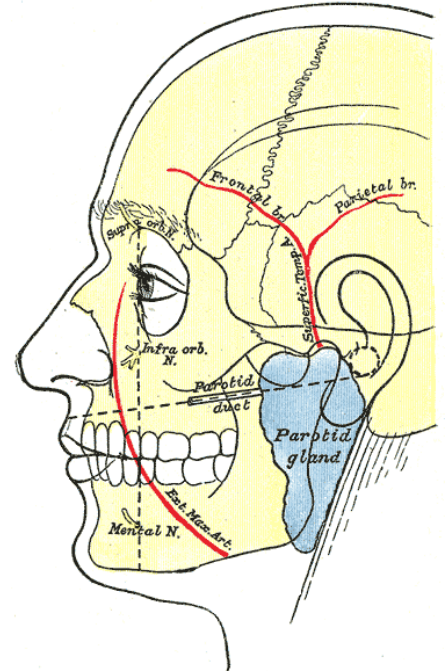
A. Superior View



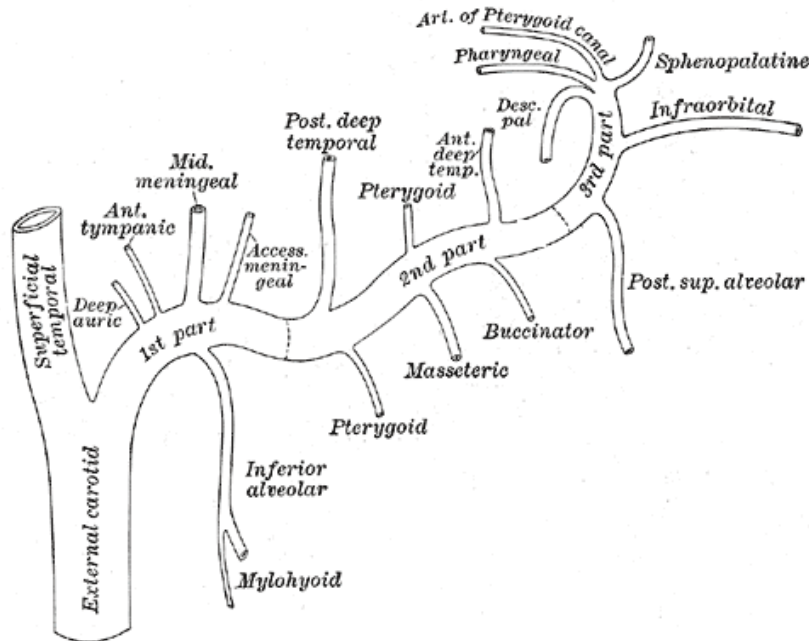
B. Lateral View

*Source= internal carotid artery (ophthalmic artery); all other labeled arteries are from external carotid

- **Posterior Auricular Artery:**
 - Arises high above the Digastric.
 - Ascends posterior to the parotid gland.
 - Between Auricle and Mastoid process
 - Divides into its auricular and occipital branches.
- **Superficial Temporal Artery:**
 - **Smallest** Terminal branch of External Carotid Artery.
 - Begins in Parotid gland, behind Neck of Mandible.
 - Runs in front of Auricle.
 - Crosses over root of Zygomatic process of Temporal bone.
 - Crossed by Temporal and Zygomatic branches of Facial nerve (**CN-VII**).
 - Accompanied by **Auriculotemporal nerve (V3)**
 - About 5 cm above Zygomatic process, it divides into two terminal branches:
 1. **Frontal branch.**
 2. **Parietal branch.**
 - Branches:
 1. **Transverse Facial Artery:**
 - o Arises within the parotid gland
 - o Passes transversely over Masseter muscle across the side of the face.
 - o Accompanied by one or two branches of the facial nerve.
 - o Passes between Parotid (Stenone) duct and Zygomatic Arch.

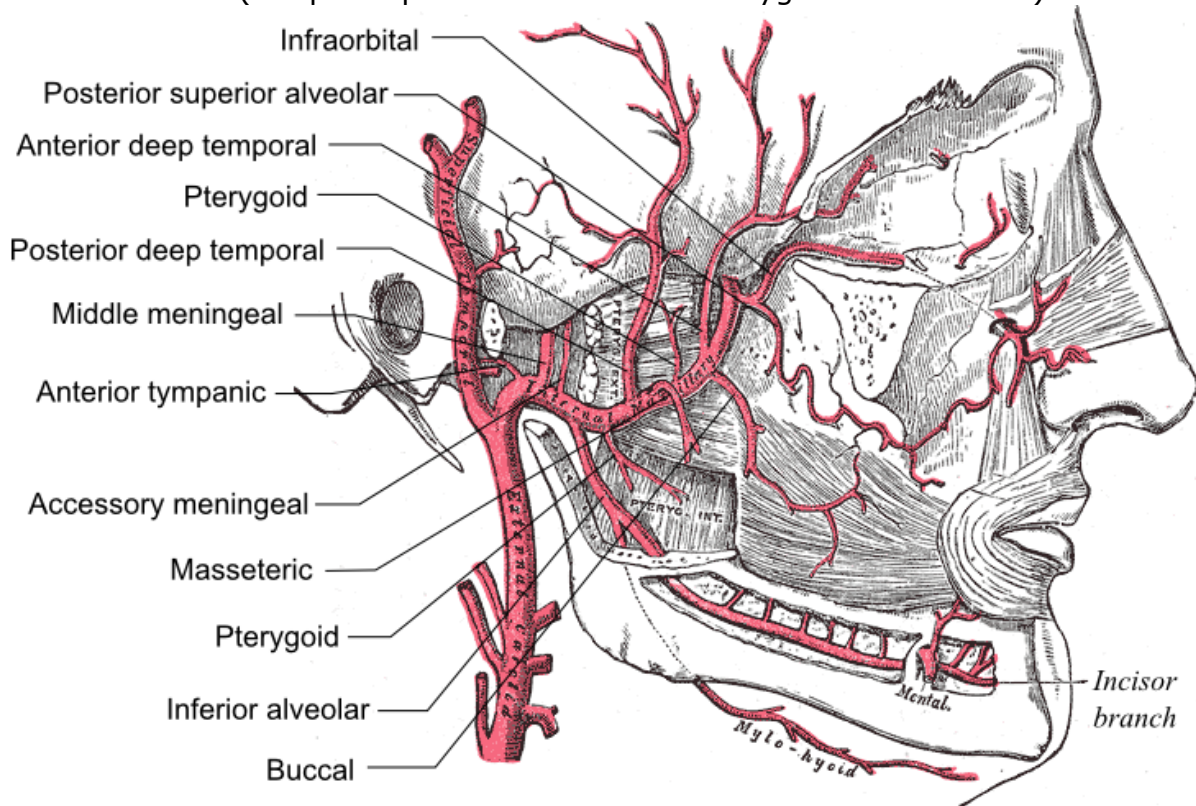


- **Maxillary Artery:**
- **Largest** Terminal branch of External Carotid Artery.
- Begins in Parotid gland, behind Neck of Mandible.



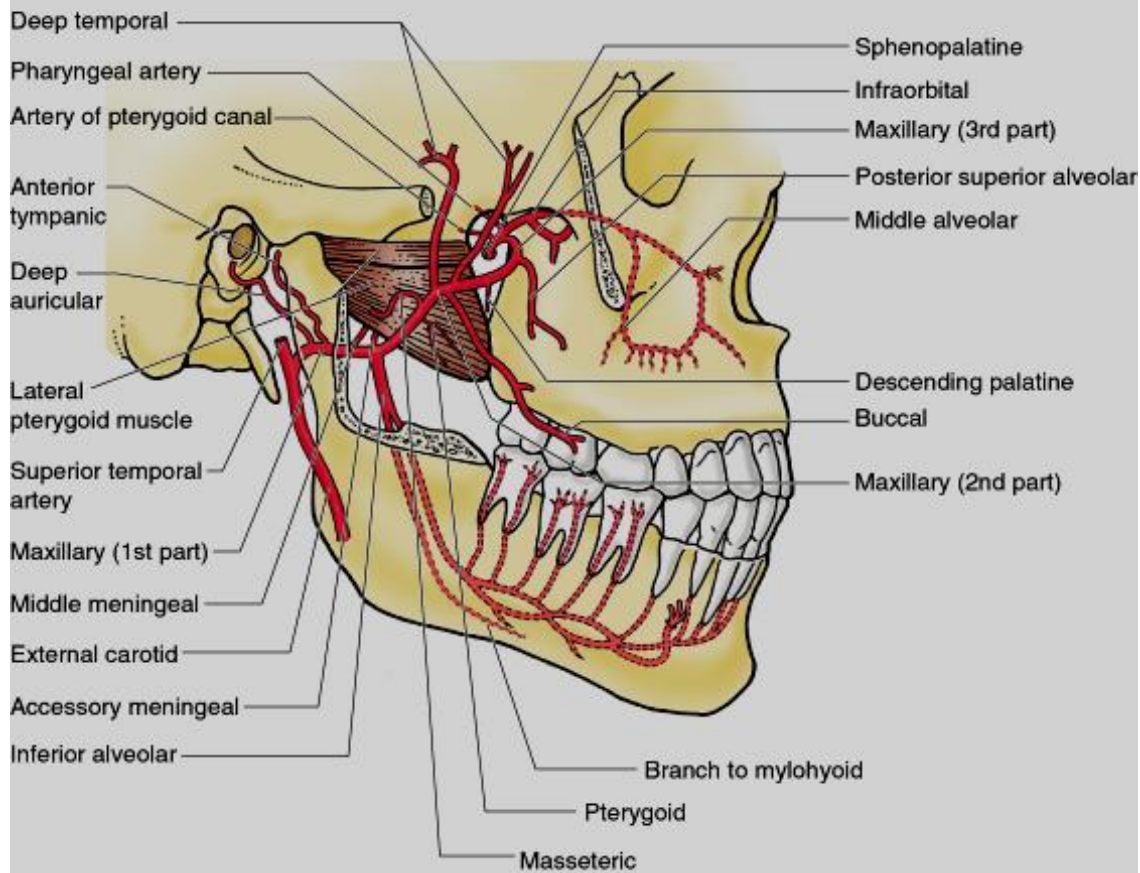
- **1st Portion (Mandibular):**
- Passes horizontally forward between Ramus of Mandible and Sphenomandibular ligament
- Runs along lower border of Lateral Pterygoid Muscle.
- Branches:
- **Anterior Tympanic Artery:**
 - o Enters tympanic cavity via petro-tympanic fissure.
 - o Anastomose with Inferior tympanic branch of Ascending Pharyngeal Artery, Stylomastoid branch of Posterior Auricular Artery, Pterygoid canal Artery and with Caroticotympanic branch from Internal carotid artery.
- **Deep Auricular Artery:**
 - o Pierces cartilaginous or bony wall of External Acoustic Meatus.
 - o Supplies its cuticular lining and the outer surface of the tympanic membrane.
 - o Gives a branch to TMJ.
- **Inferior Alveolar Artery:**
 - o Descends with Inferior Alveolar Nerve along Mandibular canal
 - o Opposite 1st Premolar tooth divides into two branches, incisor and mental.

- **2nd Portion (Pterygoid):**
- Runs forward and upward under Ramus of the mandible
- Passes between two heads of Temporalis
- Passes through Pterygo-maxillary fissure to enter Pterygopalatine Fossa.
- Branches:
- Muscular (Deep Temporal - Masseteric - Pterygoid - Buccinator).



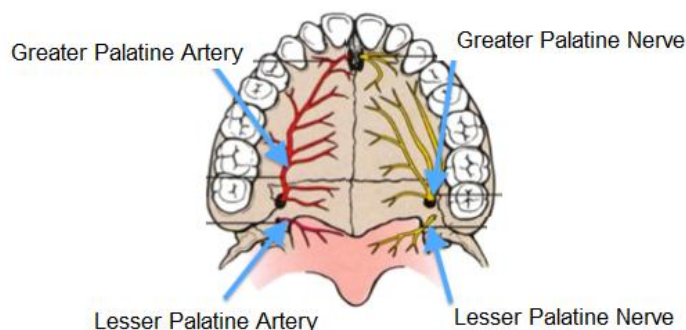
- **3rd Portion (Pterygopalatine):**
- Lies in Pterygopalatine Fossa.
- Related to Sphenopalatine ganglion.
- Branches:
- Posterior Superior Alveolar - Artery of the Pterygoid Canal - Infraorbital - Descending Palatine - Sphenopalatine.
- **Posterior Superior Alveolar Artery:**
 - Enters alveolar canals (begin on posterior surface of maxilla)
 - Supply the molar and premolar teeth and the lining of the maxillary sinus.
- **Infraorbital Artery:**
 - Runs along Inferior orbital Fissure with Infraorbital Nerve
 - Emerges on face through Infraorbital foramen
 - While in the canal, it gives off:
 - (a) Orbital branches
 - (b) Anterior superior alveolar branches.

7.42. Maxillary artery and its branches.

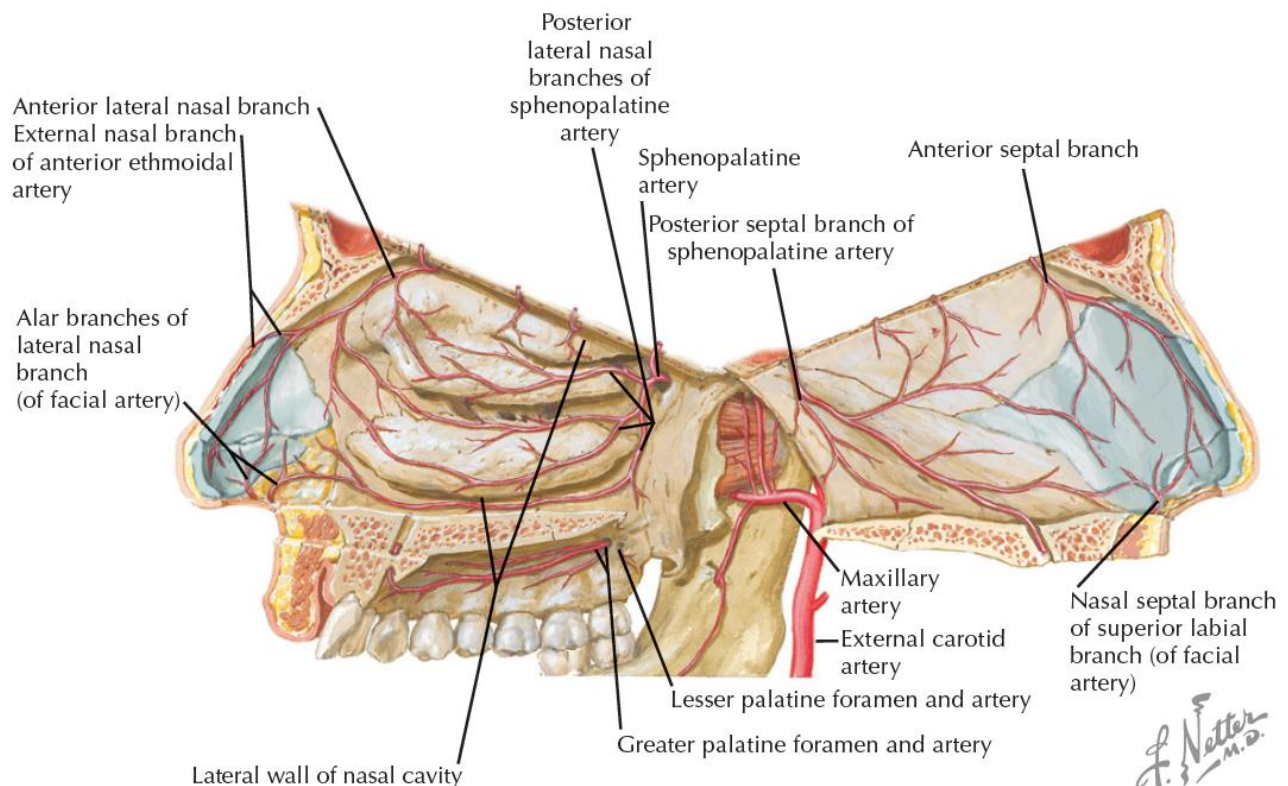


- **Descending (Greater) Palatine Artery:**

- Descends through Pterygopalatine canal with Anterior Palatine branch of Sphenopalatine ganglion.
- Emerges from Greater palatine foramen.
- Runs forward in a groove on medial side of alveolar border of Hard palate to Incisive canal.
- Passes upward through Incisive canal to anastomose with Sphenopalatine artery.
- In Pterygopalatine canal, gives off Lesser Palatine Artery which descend in lesser palatine canals to supply the soft palate and palatine tonsil.

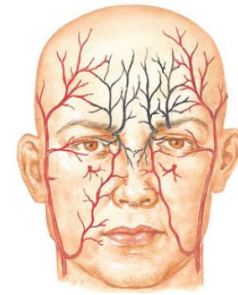


- **Artery of the Pterygoid Canal (Vidian Artery):**
 - Passes backward along pterygoid (Vidian) canal
 - Gives branches to ET and Tympanic cavity.
- **Sphenopalatine Artery (Nasopalatine Artery):**
 - Terminal branch of Maxillary artery.
 - Passes through Sphenopalatine Foramen into Cavity of nose, at Posterior part of Superior meatus.
 - Gives off its posterior lateral nasal branches which spread forward over the conchæ and meatuses, anastomose with Ethmoidal arteries and Nasal branches of Descending palatine Artery.
 - Crosses face of sphenoid.
 - Ends on nasal septum as the posterior septal branches which anastomose with Ethmoidal arteries and Septal branch of Superior labial.
 - One branch descends in a groove on the vomer to the incisive canal and anastomoses with Descending palatine artery.



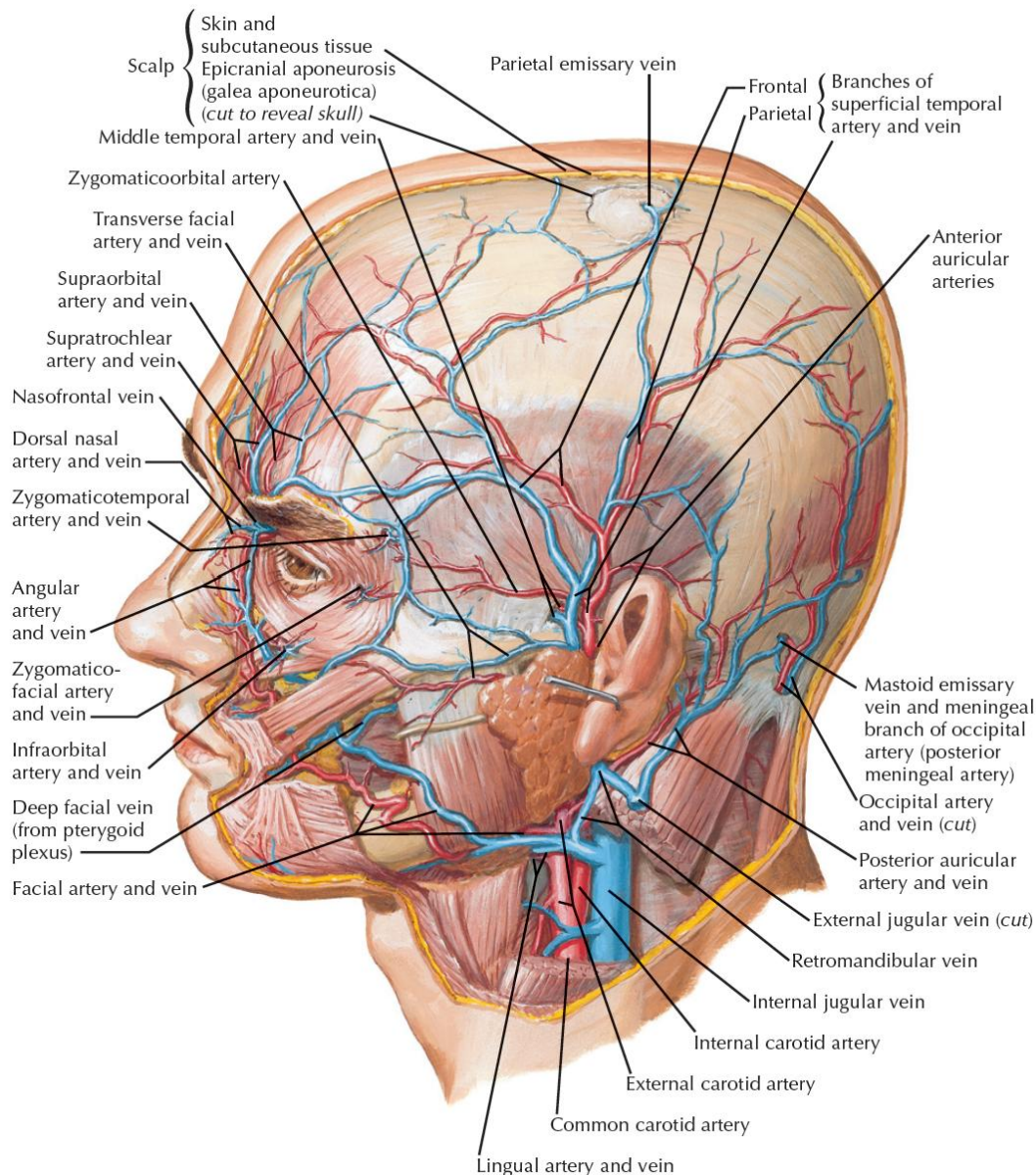
- **Blood supply of Face:**

1. **Facial Artery.**
2. **Transverse facial artery** (Superficial Temporal Artery).
3. **Frontal artery** (Superficial Temporal Artery).
4. **Supraorbital Artery** (Ophthalmic Artery).
5. **Supratrochlear artery** (Ophthalmic Artery).
6. **Dorsal nasal artery** (Ophthalmic Artery).
7. **Zygomatico-facial & Zygomatico-temporal artery** (Ophthalmic Artery).
8. **External nasal artery** (Anterior Ethmoid Artery).
9. **Infraorbital artery** (Maxillary artery).

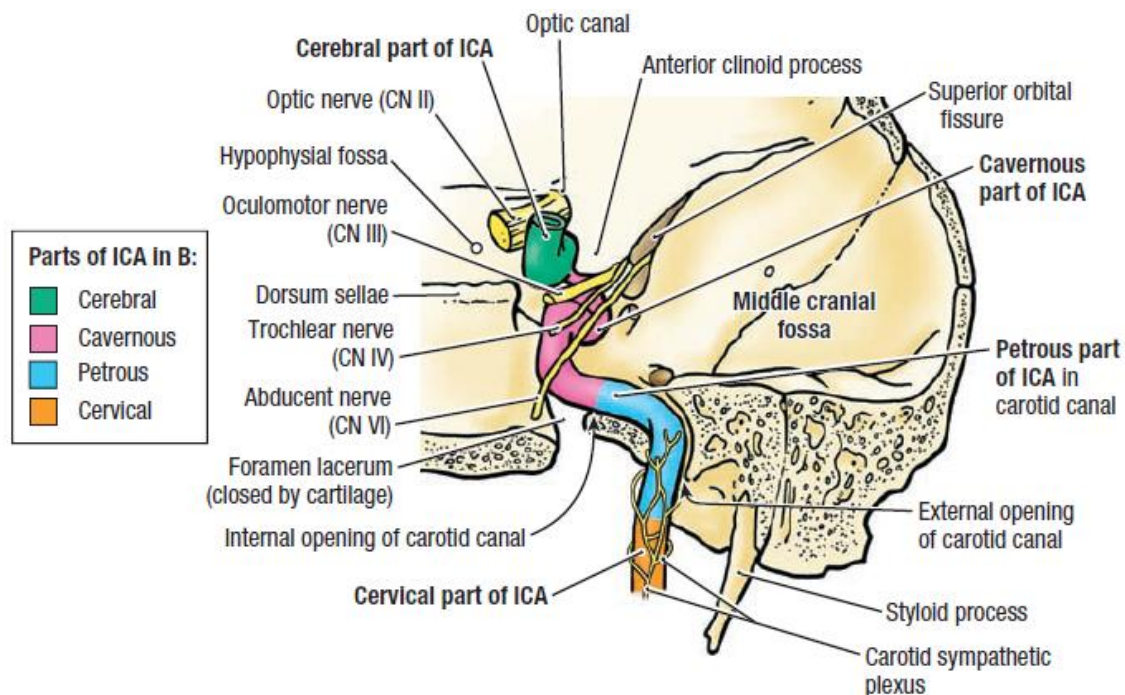


Sources of arterial supply of face
Black: from internal carotid artery (via ophthalmic artery)
Red: from external carotid artery

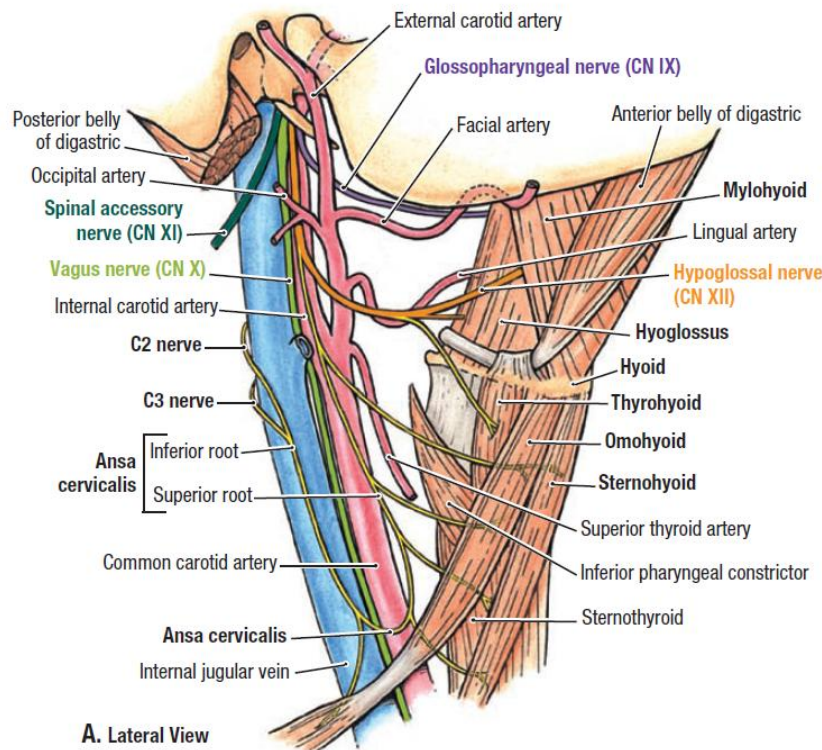
- Skin of midface is supplied by muscular cutaneous branches from these arteries that forms subdermal plexus.



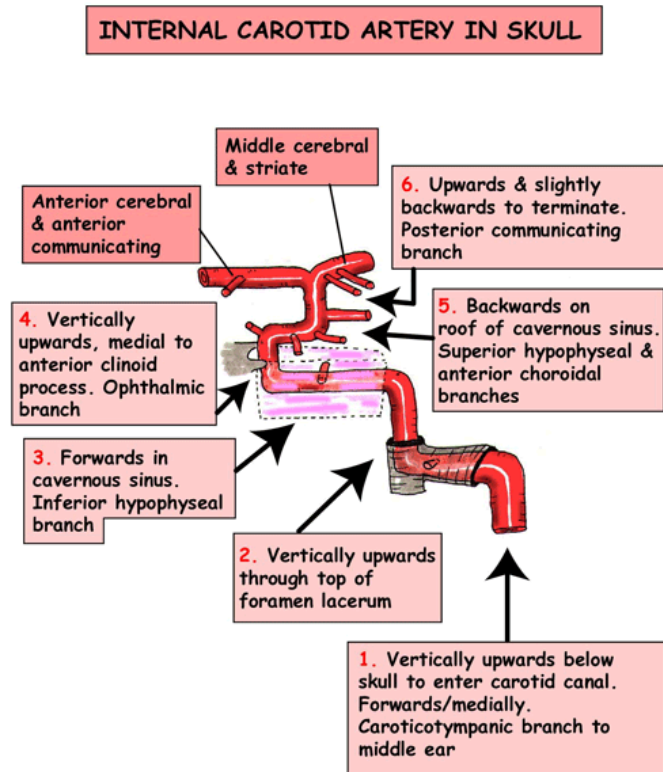
- **Internal Carotid Artery**
- One of terminal branches of Common Carotid Artery.
- Supplies Cranium, Orbit and Part of Forehead and scalp.
-
- Begins:
 - o At level of **Upper thyroid Cartilage**.
- Course:
 - o Ascends in the neck to base of skull.
 - o Enclosed within Carotid sheath.
 - o Gives NO branches in the Neck
 - o Enters Cranial cavity through **Carotid Canal** in Petrous portion of Temporal bone.
- Supplies:
 - o Supplies Cranium, Orbit and Part of Forehead and scalp.
- Divided into 4 Portions:
- **1st Portion of ICA (Cervical):**
- Begins at level of **Upper thyroid Cartilage**.
- Ascends in the neck to base of skull.
- Enclosed within Carotid sheath.
- Gives NO branches in the Neck.
- Enters Cranial cavity through **Carotid Canal** in Petrous portion of Temporal bone.



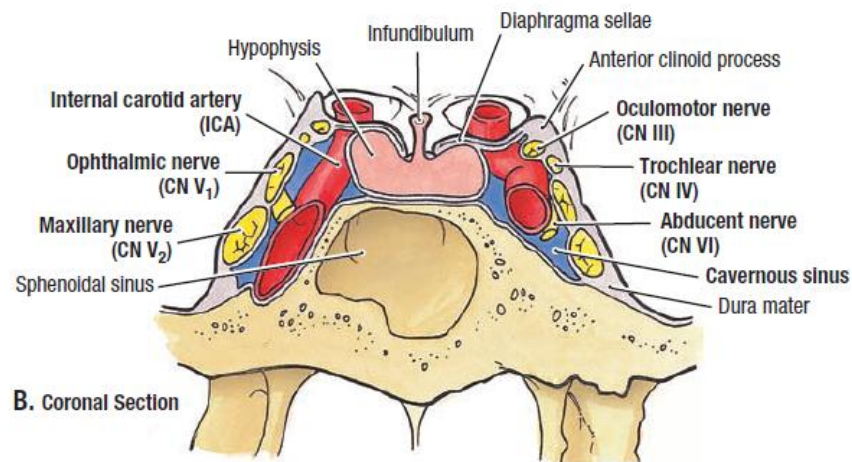
- **Relations:**
- Posteriorly:
 - Superior cervical ganglion of the sympathetic trunk.
- Laterally:
 - Internal jugular vein
 - Vagus nerve.
- Medially:
 - Pharynx.
- At the base of the skull:
 - **CNs-IX ,X , XI, XII** lie between Internal Carotid Artery and Internal jugular vein.
 - Separated from External carotid Artery by styloid apparatus.



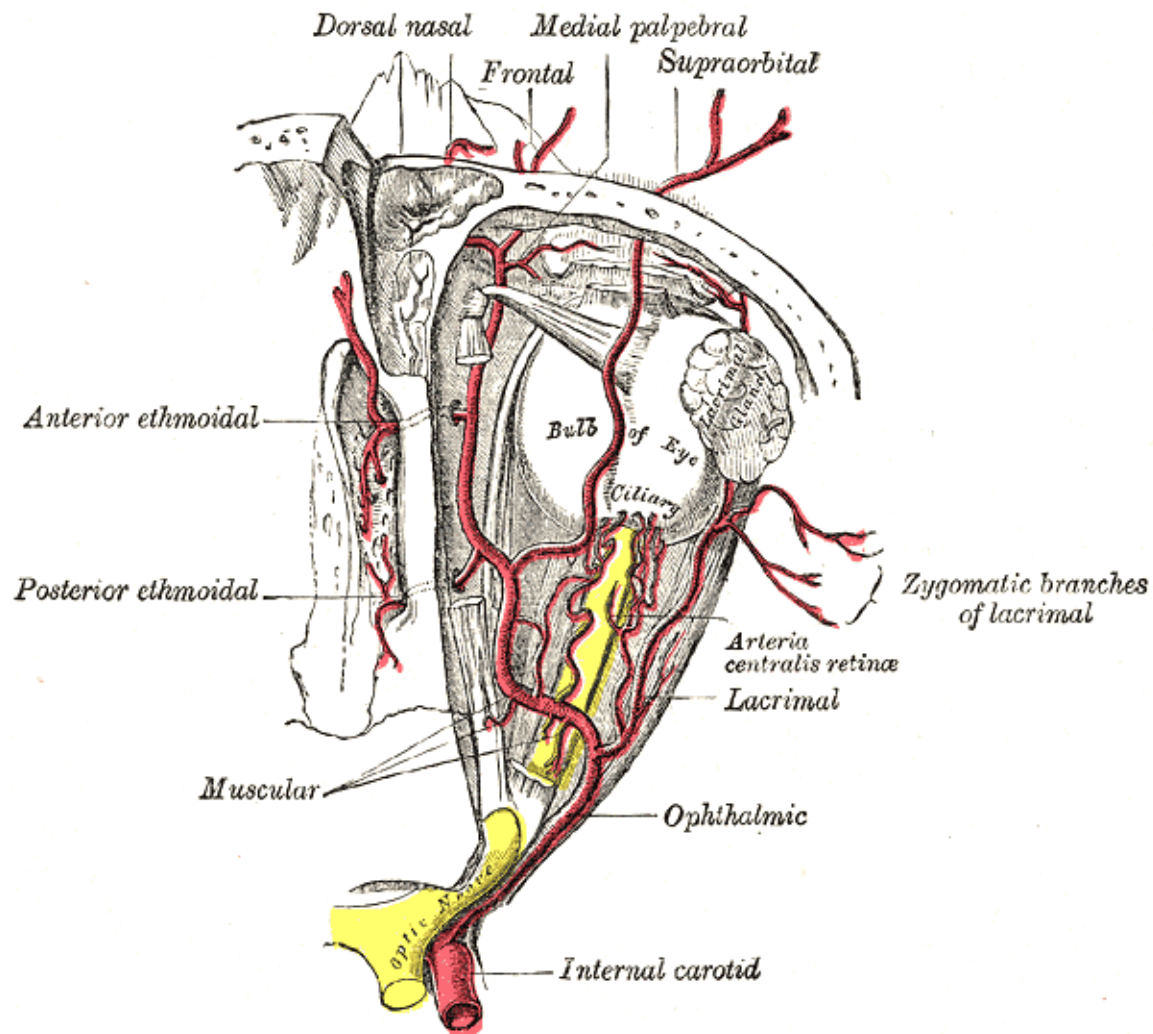
- **2nd Portion of ICA (Petrus):**
- Starts from inferior aperture of the carotid canal located in the temporal bone.
- Ascends a short distance, then curves forward and medialward, and again ascends through top of Foramen Lacerum to enter Skull.
- Lies in front of Cochlea and Tympanic cavity.
- **Branch:**
- 1. Caroticotympanic artery.**
 - Enters Tympanic cavity through a minute foramen in the carotid canal, and anastomoses with other tympanic vessels.



- **3rd Portion of ICA (Cavernous):**
- Artery is situated between the layers of the dura mater forming the cavernous sinus, but covered by the lining membrane of the sinus.
- First ascends toward Posterior clinoid process.
- Passes forward by the side of Body of Sphenoid bone.
- Curves upward on Medial side of Anterior clinoid process.
- Perforates Dura mater forming Roof of the sinus.
- On its lateral side is Abducent nerve.
- Branches:
- **Hypophyseal - Anterior Meningeal - Ophthalmic.**



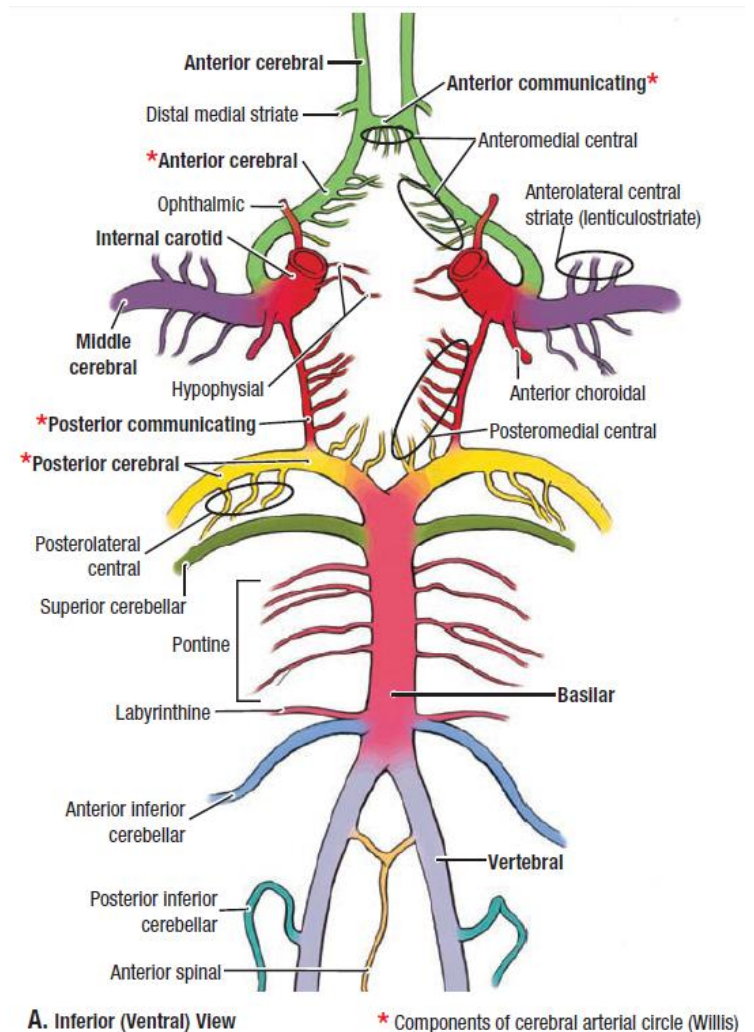
- **Ophthalmic Artery:**
- Arises from Internal carotid Artery, just as that vessel is emerging from the cavernous sinus.
- Enters Orbital cavity through Optic foramen (below and lateral to the optic nerve).
- Passes over Optic nerve to reach Medial wall of Orbit.
- Divides into two terminal branches,
 1. Frontal (supratrochlear) Artery
 2. Dorsal nasal Artery
- Orbital branches:
- Lacrimal, Supraorbital, Anterior & Posterior Ethmoidal, Medial Palpebral, Frontal, Dorsal Nasal.
- Ocular branches:
- Central Artery of the Retina, Short Posterior Ciliary, Long Posterior Ciliary, Anterior Ciliary, Muscular.



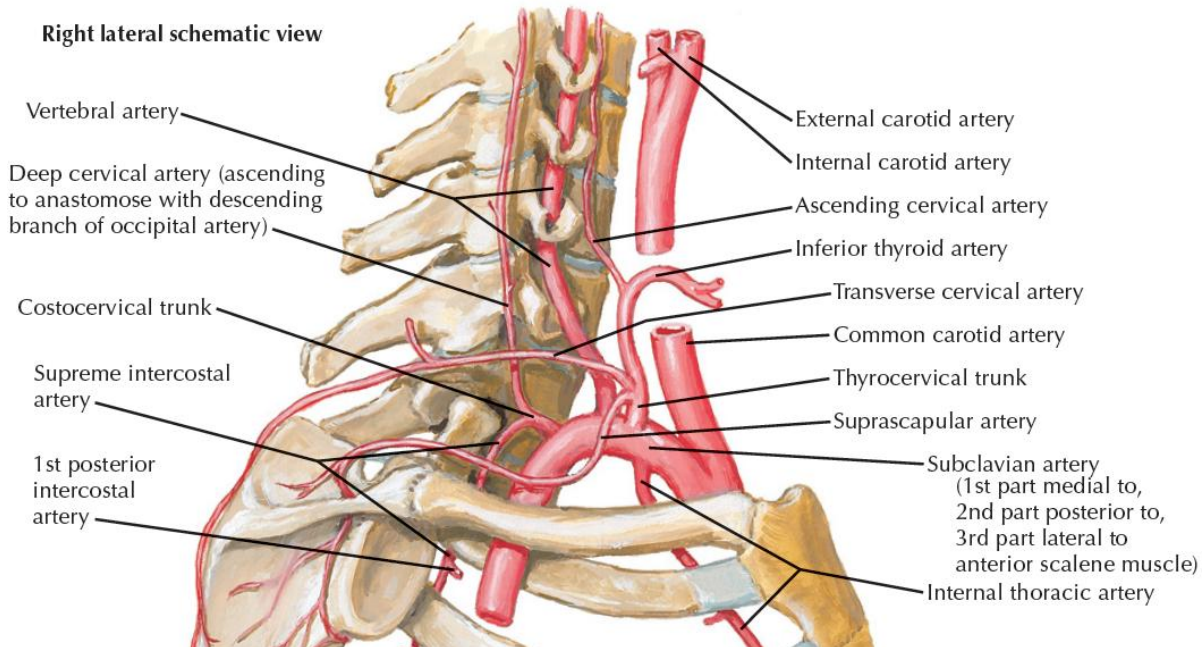
- **4th Portion of ICA (Cerebral):**
- Starts after emergence through Dural roof.
- Passes between Optic (**CN-II**) and Oculomotor (**CN-III**) Nerves.
- Gives off its terminal cerebral branches.
- Branches:
- Anterior Cerebral - Middle Cerebral - Posterior Communicating - Choroidal Artery.

- **Circle of Willis:**
- Circulatory anastomosis that supplies brain and surrounding structures.

- Anterior circulation:
 1. Internal Carotid Artery (**ICA**)
 2. Anterior cerebral Artery (**ACA**)
 3. Anterior communicating Artery (**ACOM**).
- Posterior circulation:
 1. Basilar artery
 2. Posterior cerebral artery (**PCA**)
 3. Posterior communicating Artery (**PCOM**).



- **Subclavian Artery:**
- **Right Subclavian Artery:**
 - o Branch of Right Brachiocephalic (innominate) Artery.
- **Left Subclavian Artery:**
 - o Arises directly from Arch of Aorta.
- Travel lateral to trachea into the root of Neck
- Passes between Anterior and Middle Scalene Muscles.



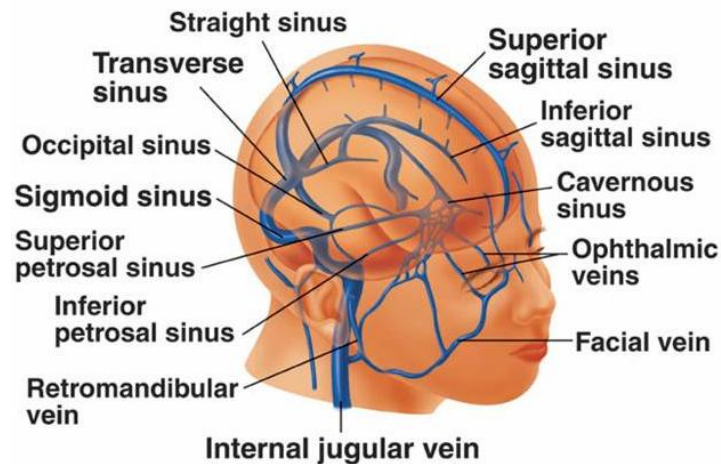
- Divides into 3 parts:
- **1st Part of Subclavian Artery:**
 - Extends from beginning of Subclavian to Medial border of Anterior scalene Muscle.
 - Most of branches of Subclavian Artery arise from the 1st part
- **2nd Part of Subclavian Artery:**
 - Located posterior to Anterior scalene Muscle.
 - Gives rise to Left Costocervical trunk.
- **3rd Part of Subclavian Artery:**
 - Starting from Lateral border of Anterior scalene Muscle to Lateral border of 1st rib, where it becomes Axillary Artery.

- **Branches of Subclavian Artery in Neck:**
 1. Thyrocervical Trunk.
 2. Vertebral Artery.
 3. Costocervical Trunk.
 4. Dorsal scapular Artery.
- **Thyrocervical Trunk:**
- Branch of 1st part of Subclavian Artery.
- Immediately divides into 3 branches:
- **Inferior thyroid Artery:**
 - o Travels along Medial border of Anterior Scalene Muscle to Thyroid gland.
 - o Runs Posterior to Carotid sheath and Anterior to Vertebral Artery.
 - o Accompanied by Recurrent laryngeal Nerve
- **Suprascapular Artery:**
 - o Travels inferior and laterally across Anterior Scalene Muscle and Phrenic nerve deep to SCM muscle.
 - o Crosses Posterior triangle of Neck to reach Scapula.
- **Transverse cervical Artery:**
 - o Travels across Posterior triangle of neck to reach Anterior border of Trapezius muscle.
- **Vertebral Artery:**
- Branch of 1st part of Subclavian Artery.
- Ascends to enter Foramen transversarium transverse process of C1-6
- Passes through **Foramen magnum** to enter the skull.
- Unites with the opposite vertebral artery to form Basilar Artery along the ventral surface of Pons.
- **Costocervical Trunk:**
- Branch of 1st part of Right Subclavian and 2nd part of Left Subclavian.
- Divides into 2 branches:
- Deep cervical:
 - o Supplies Posterior deep cervical muscles.
- Supreme intercostal:
 - o Supply the 1st and 2nd intercostal spaces.
- **Dorsal scapular Artery:**
- Branch of 2nd or 3rd part of Subclavian Artery.
- Travel across Posterior triangle of neck to reach the anterior border of trapezius muscle.

- Venous Drainage of Brain:

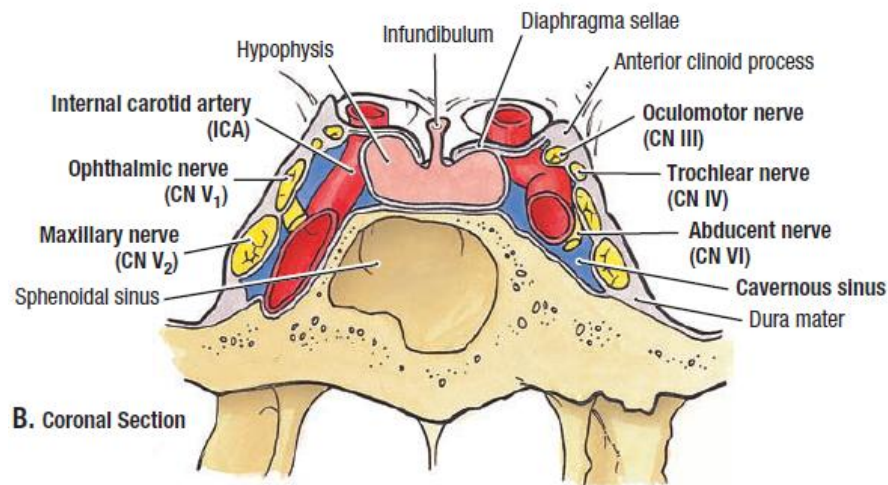
- Dural venous sinuses:

- Venous channels found between layers of dura mater in the brain.
- Receive blood from internal and external veins of the brain.
- Receive cerebrospinal fluid (CSF) from the subarachnoid space.
- Ultimately empty into Internal jugular vein through Sigmoid Sinus.



- Cavernous Sinus:

- Large paired reticulated venous structure on the lateral body of the sphenoid bone.
- Receives blood via Superior and Inferior Ophthalmic veins through the Superior orbital fissure.
- Infection from face reach Cavernous sinus through its many anastomotic connections, with severe consequences.
- Drains by two channels: Superior and Inferior Petrosal Sinuses, ultimately into Internal jugular vein via Sigmoid sinus.

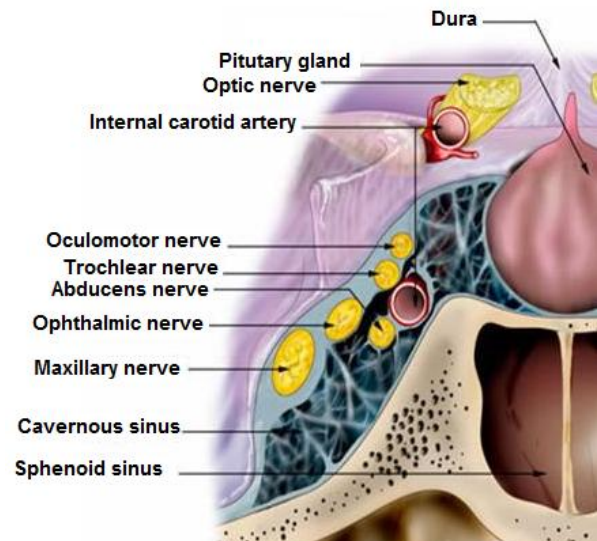


- **Lateral wall of Cavernous sinus contains:**

1. Oculomotor Nerve (**CN-III**)
2. Trochlear Nerve (**CN-IV**)
3. Ophthalmic Nerve (**V1**)
4. Maxillary Nerve (**V2**)

- **Middle of Cavernous sinus contains:**

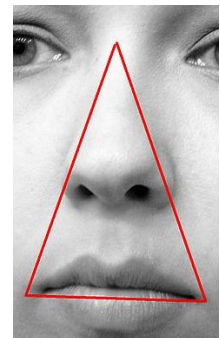
1. Internal Carotid Artery.
2. Abducens Nerve (**CN-VI**).



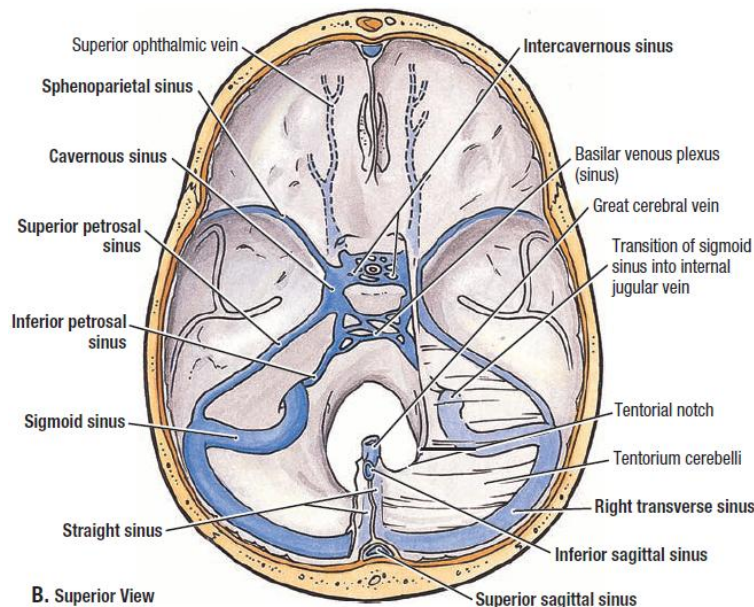
- All these nerves pass through Cavernous sinus to enter Orbital Apex through Superior Orbital Fissure **Except** Maxillary Nerve (V2) which travels through Lower portion of sinus and exits via Foramen Rotundum.
- Optic Nerve (**CN-II**) lies just above and outside Cavernous sinus, Superior and lateral to Pituitary gland on each side, and enters the Orbital Apex via Optic canal.
- Cavernous sinus receives tributaries from:
 1. Superior and inferior ophthalmic veins.
 2. Sphenoparietal sinus.
 3. Superficial middle cerebral veins.
- Cavernous sinus drains into:
 1. Superior and Inferior Petrosal Sinuses to Sigmoid sinus
 2. Emissary veins to veins outside skull.

- **Danger triangle of Face:**

- Area from corners of mouth to bridge of nose, including nose and maxilla.
- facial vein is connected to Cavernous sinus via Superior Ophthalmic vein.
- Risk of retrograde spread of infections from nasal area to cavernous sinus from an external facial injury, especially likely as facial vein has no valves, allowing blood to pass in both directions.
- In patients with thrombophlebitis of the facial vein, pieces of the clot may break off and enter the cavernous sinus, forming a cavernous sinus thrombosis.
- From there the infection may spread to the dural venous sinuses.

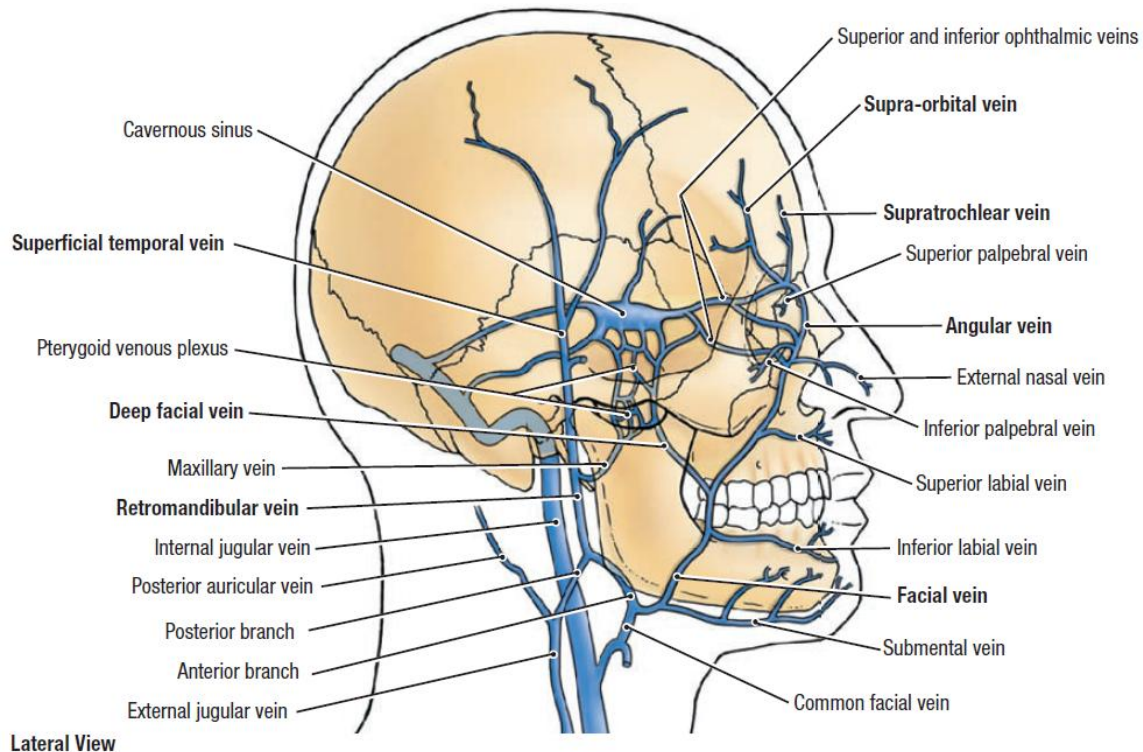


- **Sigmoid Sinus:**
- Two areas beneath the brain which allow blood to drain inferiorly from the posterior center of the head.
- Continuation of Transverse sinuses on the same side.
- Follows an S-shaped course to Jugular foramen.
- Sigmoid sinus becomes continuous with Internal jugular vein .



- **Venous Drainage of Head and Neck:**

- **Supratrochlear vein:**
- Superficial vein.
- Begins on the forehead.
- Communicates with Frontal tributary of Superficial temporal vein.
- Passes inferiorly along the forehead parallel with the vein of the opposite side
- Joins **Supraorbital vein** at Medial angle of Orbit to form **Angular vein.**
- Drain Anterior part of scalp and forehead.
- **Supraorbital vein:**
- Superficial vein.
- Begins on the forehead.
- Communicates with Frontal tributary of Superficial temporal vein.
- Passes inferiorly superficial to the frontalis muscle.
- Joins **Supratrochlear vein** at Medial angle of Orbit to form **Angular vein.**
- Drain Anterior part of scalp and forehead.

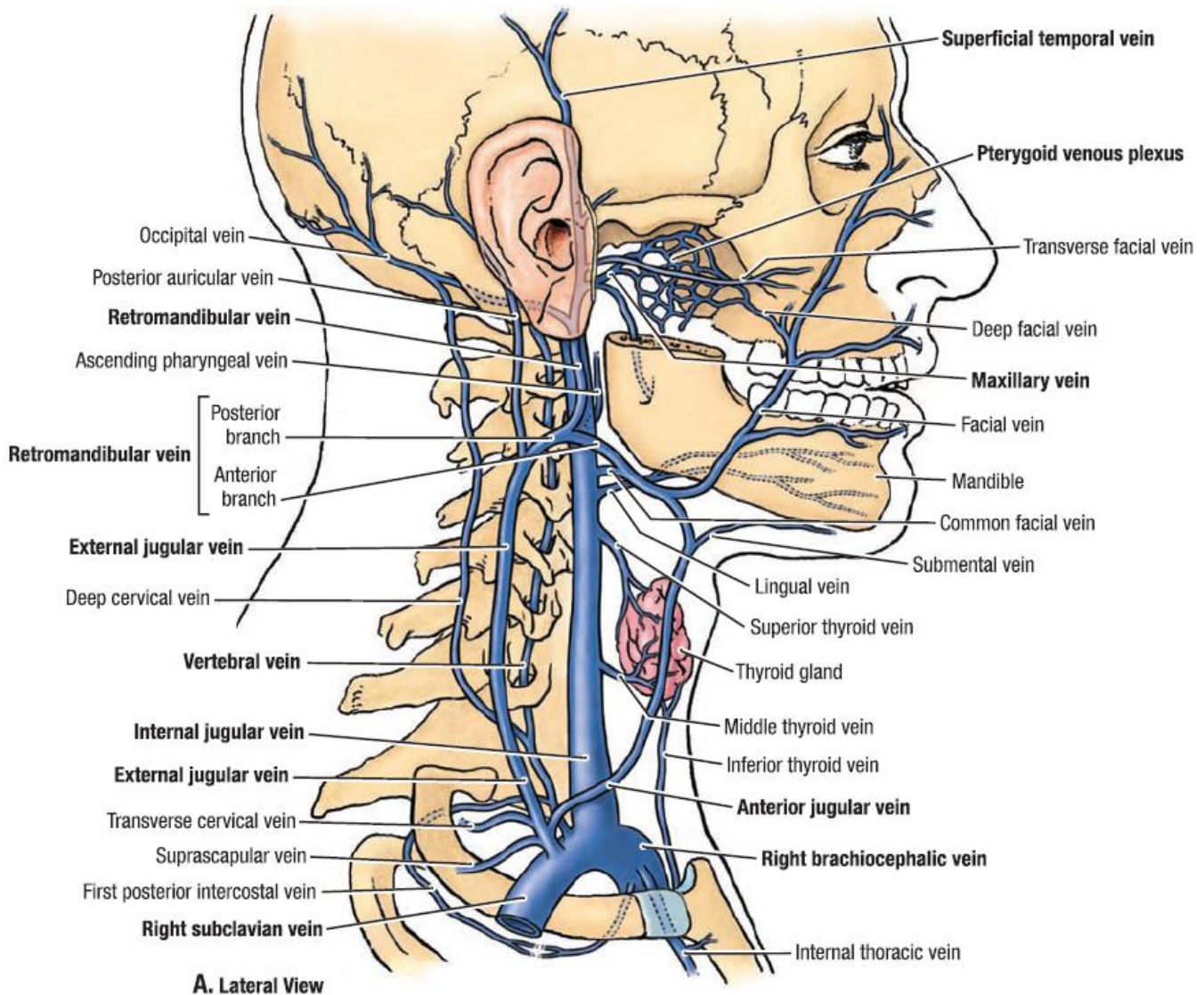


- **Superior Ophthalmic vein:**
 - Communicating vein.
 - Receives blood from roof of orbit and scalp.
 - Travels posteriorly to communicate with **Pterygoid plexus** and **cavernous sinus**.
- **Inferior Ophthalmic vein:**
 - Communicating vein.
 - Receives blood from floor of orbit.
 - Travels posteriorly with Infraorbital vein and passes through the **Inferior Orbital Fissure**.
 - Communicate with **Pterygoid plexus** and **cavernous sinus**.
- **Infraorbital vein:**
 - Communicating vein.
 - Receives blood from Midface via the lower eyelid, lateral aspect of the nose, and the upper lip.
 - Travels posteriorly with Inferior ophthalmic vein and passes through the **Inferior Orbital Fissure**.
 - Communicates with **Pterygoid plexus**.

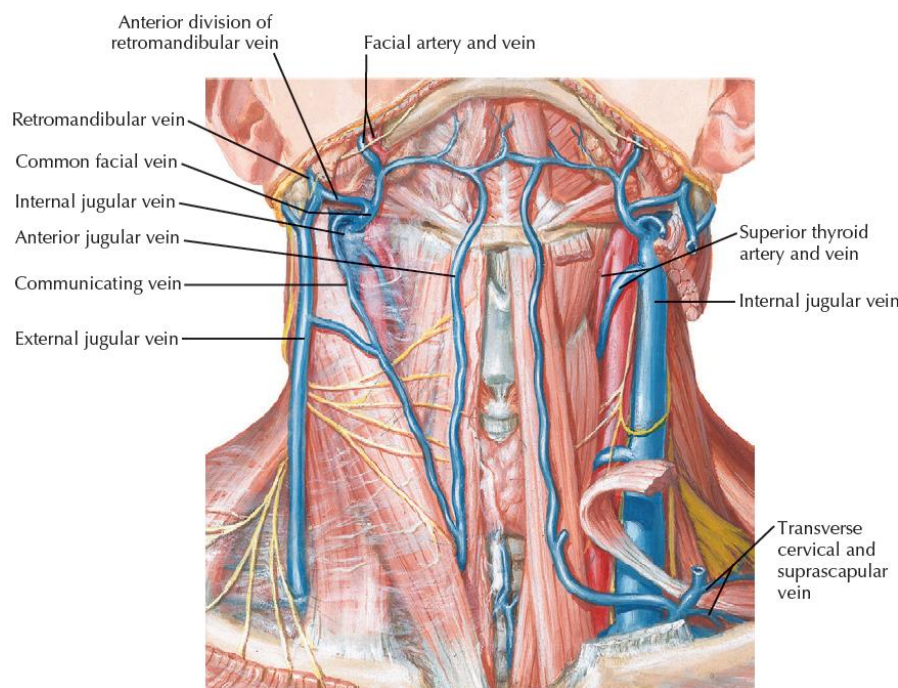
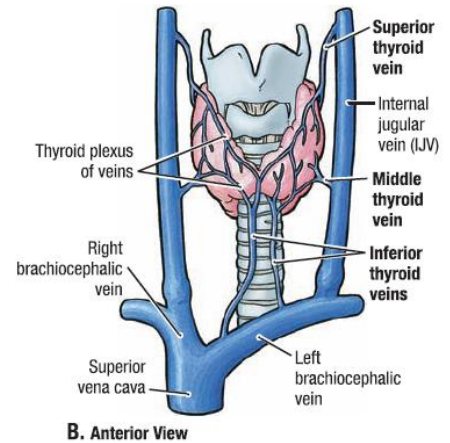
- **Angular vein:**
 - Superficial vein.
 - Begins at root of nose.
 - Forms from union of **Supraorbital** and **Supratrochlear veins** along medial part of the eye
 - Travels along Lateral aspect of the nose to become **Facial vein** at inferior orbital margin.
 - Drain Anterior part of scalp and forehead and eye lids.
- **Facial vein:**
 - Superficial vein.
 - Begins at lower orbital margin as continuation of **Angular vein**.
 - Passes inferiorly along side of Nose, receiving the lateral nasal vein.
 - Continues posteroinferiorly across the angle of the mouth to the cheek, receiving Superior and Inferior labial veins.
 - While passing toward the mandible, the deep facial vein connects it to the pterygoid plexus
 - Joins **Anterior branch of Retromandibular vein** in Submandibular triangle to form **Common Facial vein** which terminates in **Internal Jugular Vein** at level of hyoid.
 - Drains Anterior part of scalp and forehead, eye lids, external nose, anterior cheek, lips, chin and submandibular gland.
 - Has no valves that can allow blood to backflow.
- **Superficial temporal vein:**
 - Superficial vein.
 - Begins at sides of scalp on along zygomatic arch.
 - Descends posterior to the zygomatic root of temporal bone alongside Auriculotemporal nerve.
 - Enter parotid gland
 - Joins **Maxillary vein** posterior to Neck of Mandible to form **Retromandibular vein**.
 - Drains side of scalp and External ear.
- **Retromandibular vein:**
 - Superficial vein.
 - Begins Anterior to ear by union of **Superficial temporal** and **Maxillary veins**.
 - Runs posterior deep to Ramus of Mandible through Parotid gland.
 - Divides into two branches:
 - o **Anterior branch** joins **Facial vein** to form **Common Facial vein**.
 - o **Posterior branch** joins **Posterior Auricular vein** to form **External jugular vein**.
 - Drains Parotid gland and Masseter muscle.

- **Pterygoid plexus:**

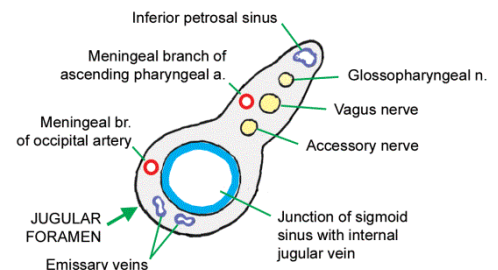
- Extensive network of veins that parallels 2nd and 3rd parts of maxillary Artery.
- Receives branches that correspond to the maxillary artery's branches
- Tributaries of the pterygoid plexus eventually converge to form **Maxillary vein.**
- Communicates with cavernous sinus, pharyngeal venous plexus, Facial vein via Deep facial vein and ophthalmic vein.



- **Lingual vein:**
 - Passes with Lingual artery deep to the Hyoglossus muscle.
 - Ends in **Internal Jugular vein.**
- **Superior and Middle thyroid vein:**
 - Forms a venous plexus on the thyroid gland with the middle and inferior thyroid veins.
 - Drains into **Internal jugular vein.**
- **Inferior thyroid vein:**
 - Forms a venous plexus on the thyroid gland with the middle and inferior thyroid veins.
 - Drains into **Left Brachiocephalic vein.**
- **Occipital vein:**
 - Begins on Posterior portion of Scalp at vertex.
 - Passes from superficial to deep by passing through the attachment of SCM muscle.
 - Termination is variable, joins Posterior Auricular vein to reach External Jugular vein or join Internal Jugular vein.
- **Anterior jugular vein:**
 - Begins near Hyoid bone.
 - Arises by joining of a series of superficial veins in Submental region.
 - Descends anterior to SCM muscle then deep to it.
 - Drains into **External Jugular vein** or directly into **Subclavian vein.**
 - Venous jugular arch:
 - o Transverse trunk communicate both Anterior jugular veins just above the sternum
 - o Receive tributaries from the inferior thyroid veins.



- **External jugular vein:**
- Formed by Combination of **Posterior branch of Retromandibular** and **Posterior Auricular veins** in Parotid gland just behind Angle of Mandible.
- Descends Obliquely.
- Superficial to SCM muscle.
- Deep to Platysma muscle.
- Passes into Posterior triangle of Neck.
- Pierces deep fascia.
- Drains into **Subclavian vein** immediately lateral to Anterior scalene muscle.
- Tributaries:
 - Posterior Auricular vein.
 - Posterior branch of Retromandibular vein.
 - Anterior Jugular vein.
 - Transverse cervical vein.
 - Suprascapular vein.



- **Internal Jugular vein:**
- Begins in at a dilation (superior bulb) in **Postero-lateral compartment of Jugular foramen.**
- Continuation of Sigmoid sinus.
- Runs down the side of Neck in a vertical direction.
- Lying Postero-lateral to Internal Carotid Artery (and common carotid) and behind **CN-IX, X, XI.**
- Travels within Carotid sheath lateral to Internal carotid artery and Anterior to Vagus nerve (**CN-X**).
- Lower down, **CN-IX** and **CN-XII** pass forward between Internal Jugular Vein and Internal Carotid Artery.
- 2nd dilation (Inferior bulb) located above its termination.
- Joins **Subclavian vein** to form **Brachiocephalic (innominate) vein** at root of Neck.
- Tributaries:
 - Inferior petrosal sinus.
 - Facial vein.
 - Pharyngeal vein.
 - Lingual vein.
 - Superior and Middle thyroid vein.

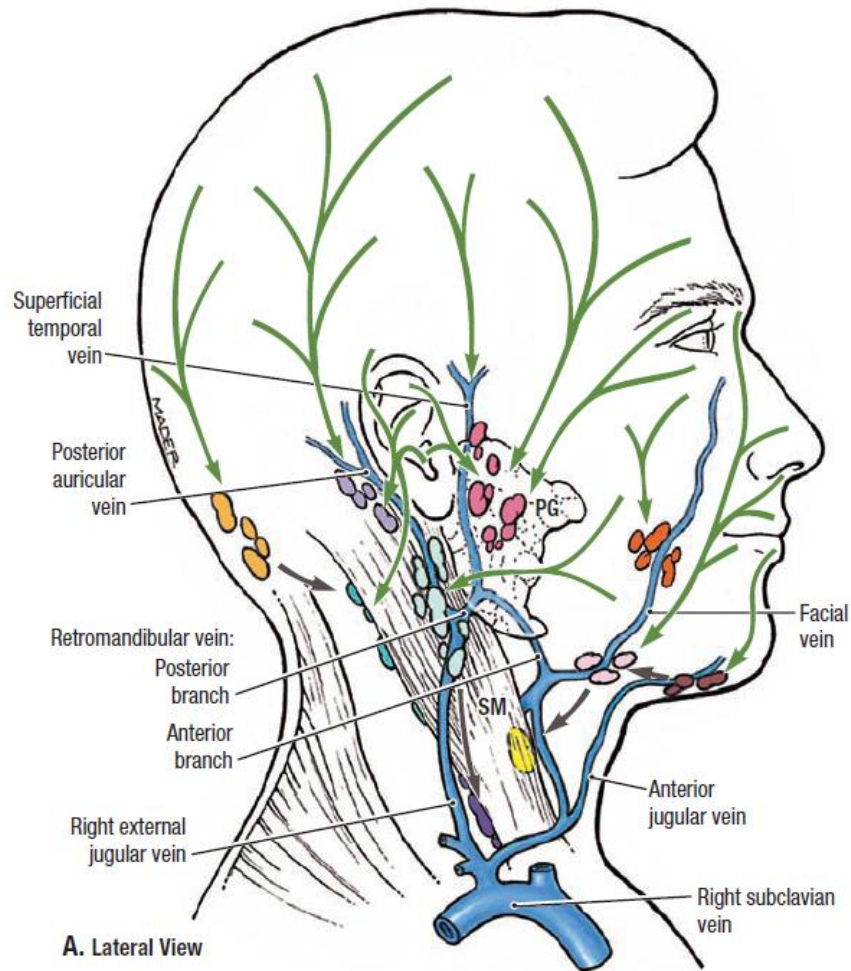
- **Vertebral vein:**
- Begins as a plexus in Suboccipital triangle.
- Descends through the **Foramen transversarium** of all of Cervical vertebrae.
- Drains into Subclavian or more commonly, **Brachiocephalic vein.**

- **Lymphatics of Head and Neck:**

- **Superficial Lymph Node of Head:**

1. Occipital LN:

- Located at back of Head close to margin of Trapezius.
- **Afferents:** from Occipital region of scalp
- **Efferents:** to Superior deep cervical LN.



Lymph nodes:			
■ Buccinator	■ Paratracheal	■ Superficial cervical	SM Sternocleidomastoid
■ Inferior deep cervical	■ Parotid	■ Superior deep cervical	T Trachea
■ Infrahyoid	■ Prelaryngeal	Structures:	
■ Jugulodigastric	■ Pretracheal	➔ Initial drainage	TC Thyroid cartilage
■ Jugulo-omohyoid	■ Retropharyngeal	➔ Secondary (subsequent) drainage	TG Thyroid gland
■ Mastoid (retro-auricular)	■ Submandibular	H Hyoid	P Palatine tonsil
■ Occipital	■ Submental		PG Parotid gland
			Ph Pharyngeal tonsil

2. Posterior Auricular LN:

- Located at Mastoid insertion of SCM muscle.
- **Afferents:** from posterior part of Temporoparietal region, Upper part of cranial surface of Auricle and back of EAC.
- **Efferents:** to Superior deep cervical LN.

3. Preauricular (superficial parotid) LN:

- Located immediately in front of the tragus.
- **Afferents:** from lateral surface of Auricle and skin of adjacent part of Temporal region.
- **Efferents:** to Superior deep cervical LN.

4. Parotid LN:

- Imbedded in the substance of the gland.
- **Afferents:** from Root of the nose, Eyelids, Fronto-temporal region, EAC and Tympanic cavity
- **Efferents:** to Superior deep cervical LN.

5. Facial LN:

- Three Groups:
 - i. Infraorbital LN
 - ii. Buccinator LN
 - iii. Supramandibular LN

- Waldeyer`s Ring:

1. Pharyngeal Tonsil (Adenoid):

- Located in the Nasopharynx.
- **No Afferents.**
- **Efferents:** Retropharyngeal and Parapharyngeal LN.

2. Tubal Tonsils:

- Located posterior to opening of ET in Nasopharynx.

3. Palatine Tonsil:

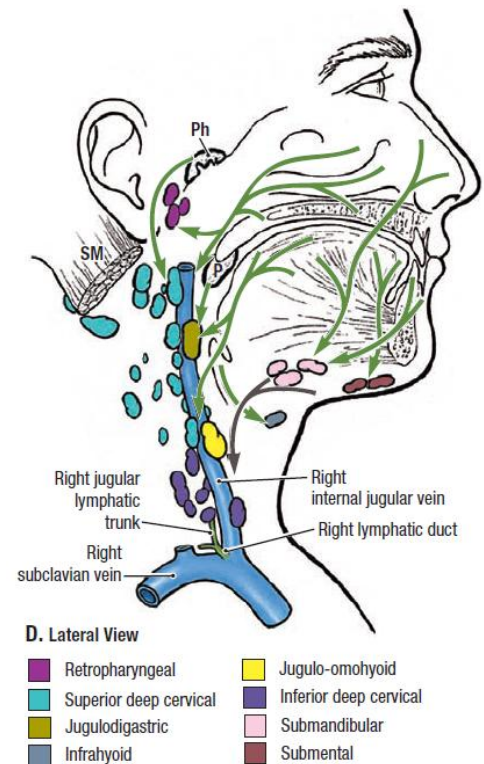
- Located in the Oropharynx.
- **No Afferents.**
- **Efferents:** to Jugulodigastric and Superior deep cervical LN.

4. Lingual Tonsil:

- Located in dorsal surface at base of tongue.

• Retropharyngeal LN:

- Deep LN (Not part of Waldeyer`s Ring)
- Located in the buccopharyngeal fascia, behind the upper part of the pharynx.
- **Afferents:** from Posterior Nasal cavity, Nasopharynx and ET.
- **Efferents:** to Superior deep cervical LN.



- **Regions of Lymph Nodes of the Neck:**

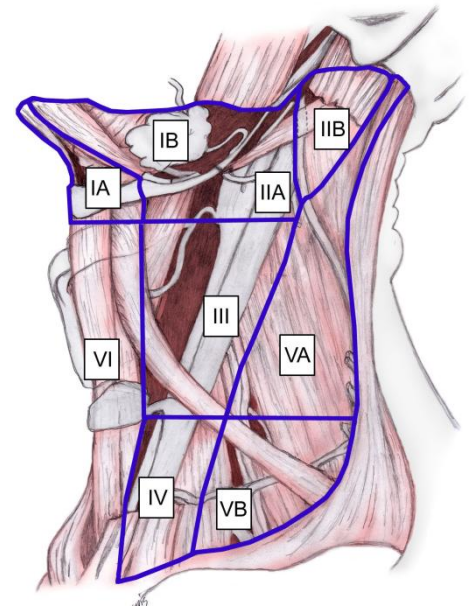
1. Level I:

• **Level Ia: Submental LN**

- Bounded by bilateral Anterior bellies of digastric muscles and Hyoid bone.
- Drains Floor of mouth, Anterior tongue, Anterior mandibular alveolar ridge, and Lower Lip.

• **Level Ib: Submental LN**

- Bounded by Anterior and Posterior bellies of digastric and body of Mandible.
- Drains oral cavity, Anterior Nasal Cavity, soft tissue structures of the mid face, and Submandibular gland.



2. Level II: Upper Jugular LN (Jugulodigastric)

- Related to upper third of Internal jugular vein.
- **Clinical Landmark:** From Skull base to Hyoid bone.
- **Surgical Landmark:** From Skull base to Carotid bifurcation.
- Drains the oral cavity, nasal cavity, nasopharynx, oropharynx, hypopharynx, larynx, and parotid gland.
- Spinal Accessory Nerve (**CN-XI**) travels obliquely across this area.

• **Level IIA: LN Anterior to Spinal Accessory Nerve (CN-XI)**

• **Level IIB: LN Posterior to Spinal Accessory Nerve (CN-XI)**

3. Level III: Middle Jugular LN

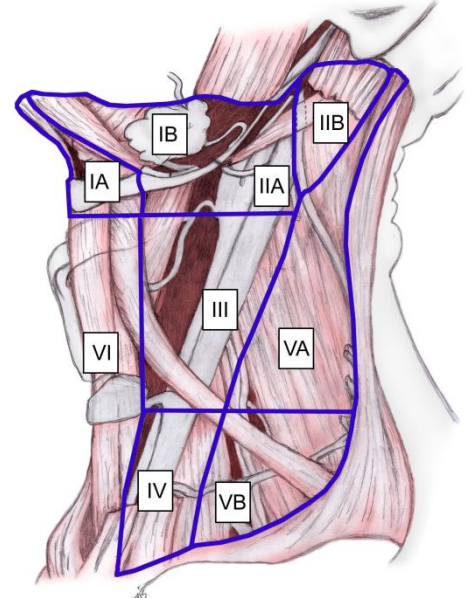
- Related to Middle third of Internal jugular vein.
- **Clinical Landmark:** From Hyoid bone to Cricothyroid notch.
- **Surgical Landmark:** From Carotid bifurcation to Omohyoid.
- Drains the oral cavity, nasopharynx, oropharynx, hypopharynx, and larynx.

4. Level IV: Lower Jugular LN

- Related to Lower third of Internal jugular vein.
- From Omohyoid muscle to Clavicle.
- Drains larynx, hypopharynx, thyroid, and cervical esophagus.

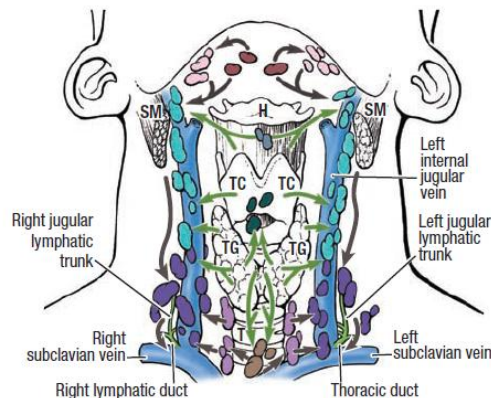
5. Level V: Posterior Triangle LN

- Located along lower half of the spinal accessory nerve and the transverse cervical artery.
- Extends from Apex of Convergence of SCM and trapezius muscle to Clavicle.
- Drains nasopharynx, oropharynx, and skin of the posterior scalp and neck.
- **Level Va: LN Associated with Spinal accessory nerve (CN-XI)**
- **Level Vb: LN Associated with Transverse Cervical and supraclavicular nodes.**



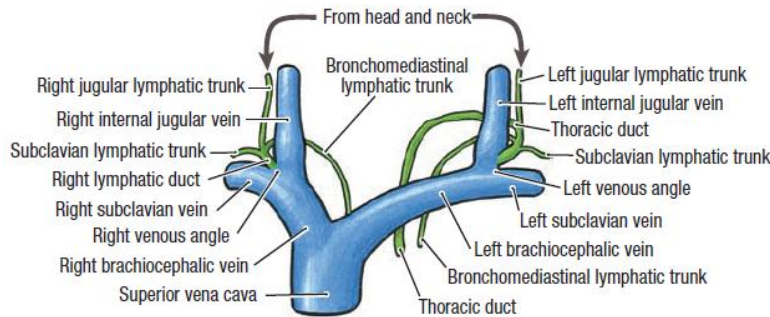
6. Level VI: Central Compartment LN

- Extend in midline from hyoid bone superiorly to Suprasternal notch inferiorly.
- Drains Thyroid gland, subglottic larynx, cervical trachea, hypopharynx, and cervical esophagus.
- **Paratracheal LN:** Located in Tracheoesophageal groove.
- **Pretracheal LN:** Located in front of the trachea
- **Parathyroidal LN:** Located around the thyroid gland
- **Precricoid or Delphian LN:** Located on Cricothyroid membrane.



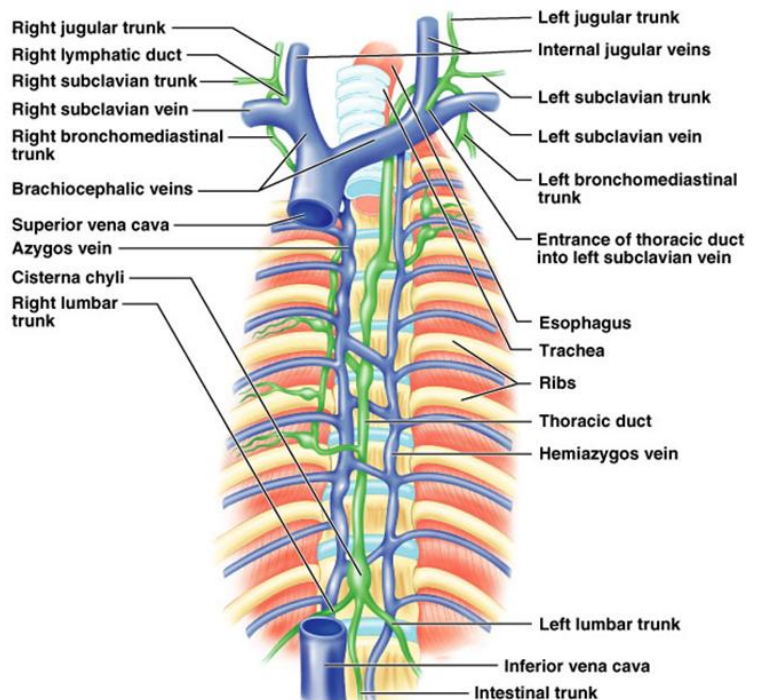
B. Anterior View

Lymph nodes:			
Buccinator	Paratracheal	Superficial cervical	SM Sternocleidomastoid
Inferior deep cervical	Parotid	Superior deep cervical	T Trachea
Infrahyoid	Prelaryngeal	Structures:	
Jugulodigastric	Pretracheal	Initial drainage	TC Thyroid cartilage
Jugulo-omohyoid	Retropharyngeal	Secondary (subsequent) drainage	TG Thyroid gland
Mastoid (retro-auricular)	Submandibular	H Hyoid	P Palatine tonsil
Occipital	Submental		PG Parotid gland
			Ph Pharyngeal tonsil



- **Thoracic Duct:**
- Left lymphatic duct.
- Common trunk of all lymphatic vessels of the body **Except:**
 - o Right side of Head, Neck, thorax, Right upper limb, Right lung, Right side of Heart.
 - o Drained by **Right lymphatic duct.**
- Length in adult: 40-45 cm
- Begins in Abdomen by a dilatation (**Cisterna chili**) in front of **L2**.
- Extends to the root of the neck posterior to Left Carotid artery and left Internal jugular vein.
- Passing into the neck it forms an arch above clavicle and crosses Anterior to Subclavian artery.
- Passes in front Scalenus Anterior.
- Ends in **Left Subclavian vein.**

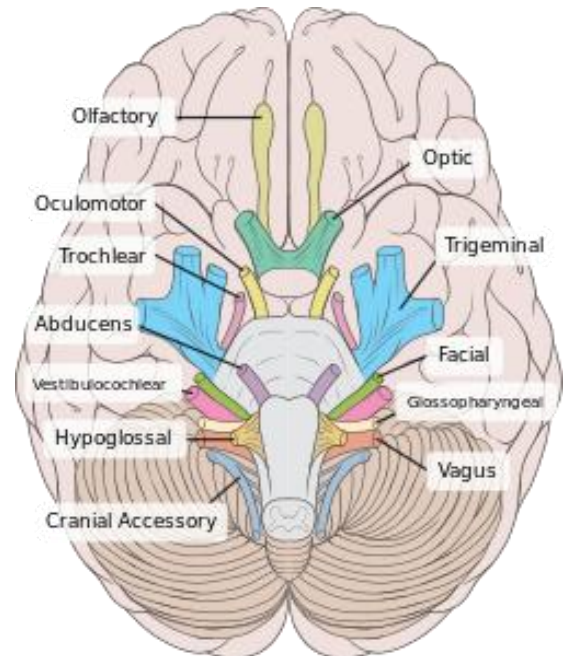
- **Right lymphatic duct:**
- Length in adult: 40-45 cm
- Courses along Medial border of Scalenus Anterior at Root of Neck.
- Ends in **Right Subclavian vein.**
- Drains Right side of Head, Neck, thorax, Right upper limb, Right lung, Right side of Heart.



- **CRANIAL NERVES (CN)**

- Peripheral nerves that exit from the cranial cavity through openings in the cranium.
- All CN are distributed in the Head and neck Except Vagus Nerve which also supplies structures in the thorax and abdomen.
- 12 pairs of cranial nerves that are named and numbered in rostrocaudal sequence of their superficial origins from the brain, brainstem, and superior spinal cord.

- I. Olfactory Nerve.**
- II. Optic Nerve.**
- III. Oculomotor Nerve.**
- IV. Trochlear Nerve.**
- V. Trigeminal Nerve.**
- VI. Abducens Nerve.**
- VII. Facial Nerve.**
- VIII. Vestibulocochlear Nerve.**
- IX. Glossopharyngeal Nerve.**
- X. Vagus Nerve.**
- XI. Spinal Accessory Nerve.**
- XII. Hypoglossal Nerve.**



- **Pure Sensory:**

1. Olfactory Nerve.
2. Optic Nerve.
3. Vestibulocochlear Nerve.

- **Pure Motor:**

1. Oculomotor Nerve.
2. Trochlear Nerve.
3. Abducens Nerve.
4. Spinal Accessory Nerve.
5. Hypoglossal Nerve.

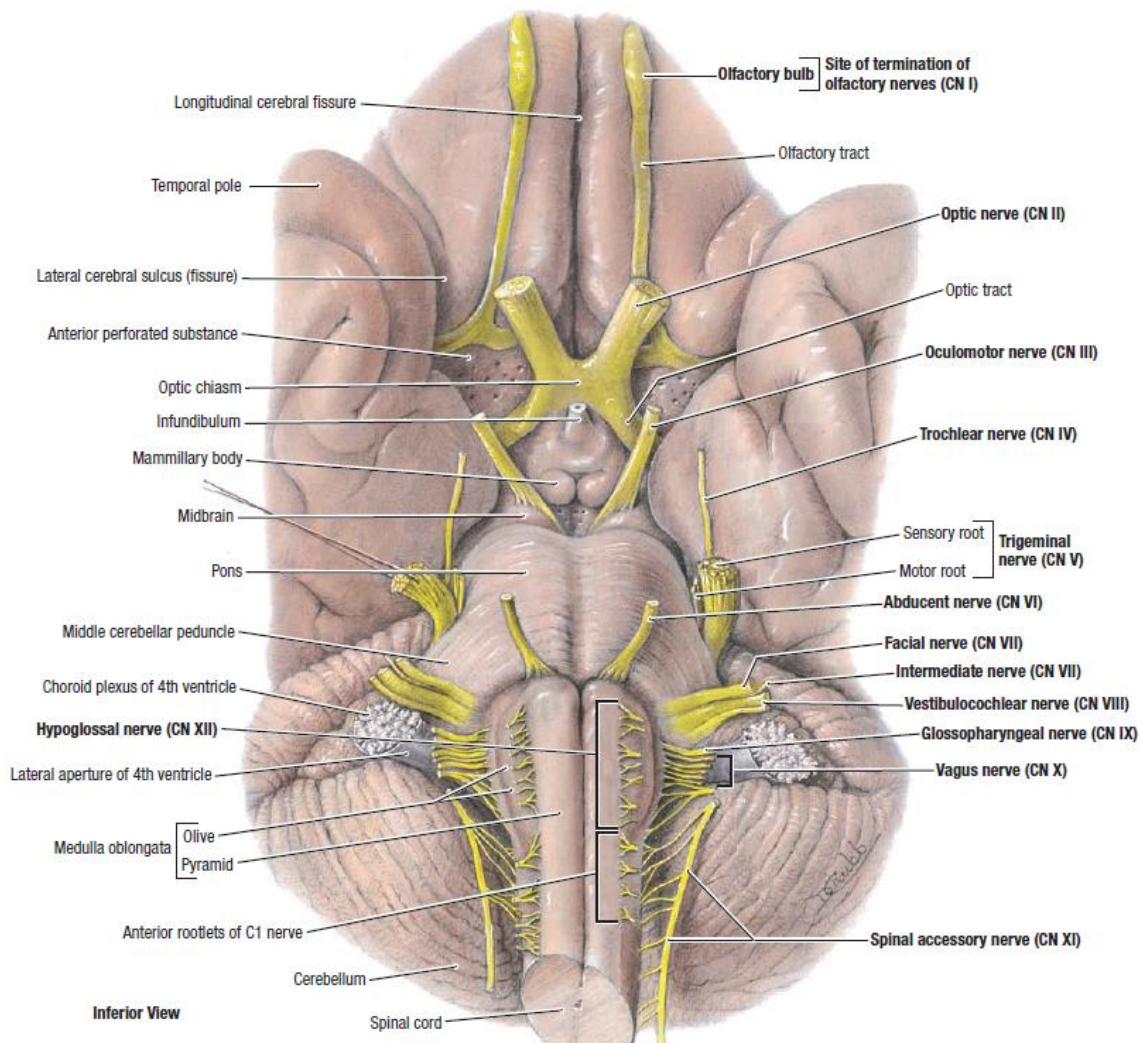
- **Mixed Sensory and Motor:**

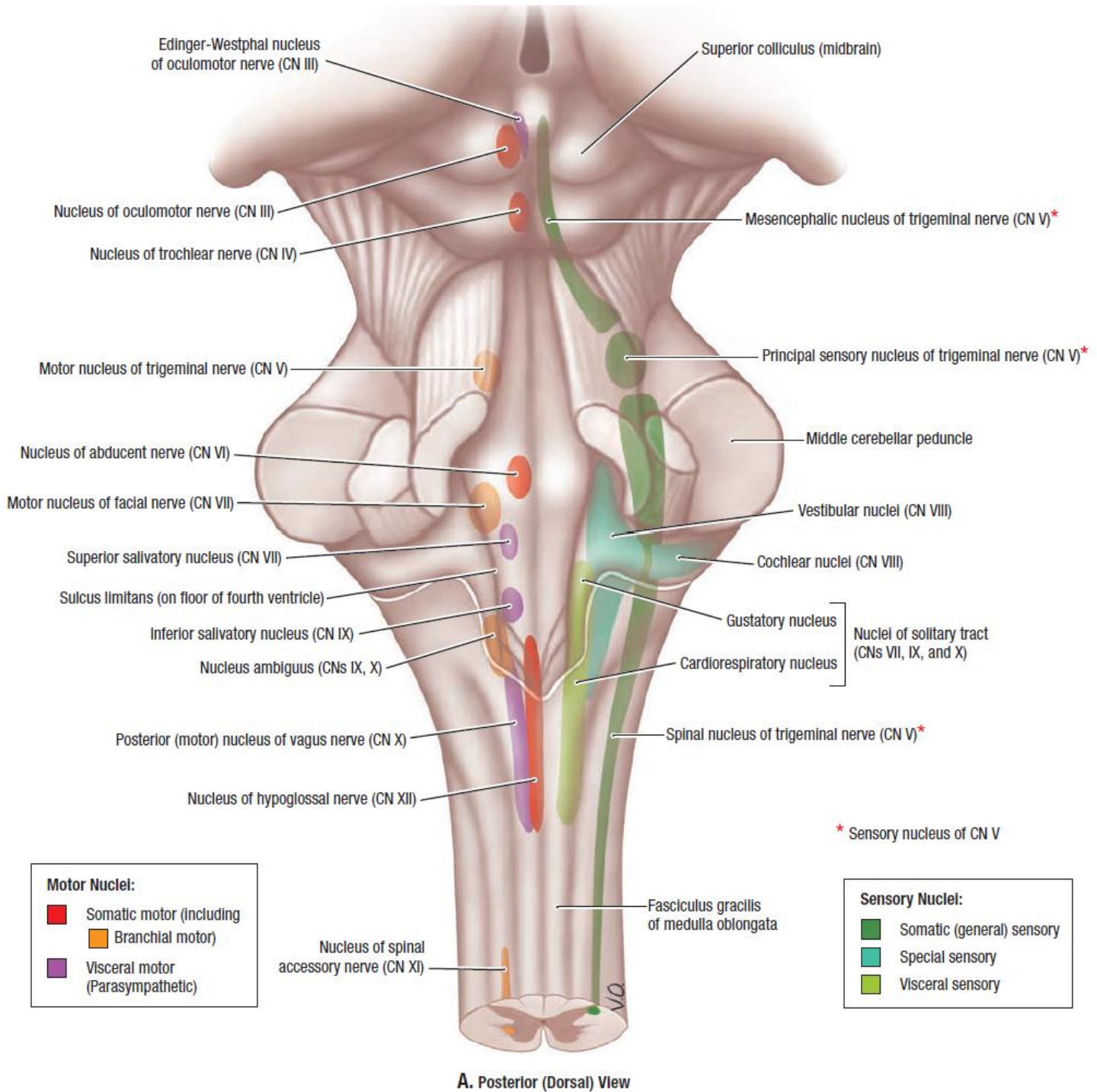
1. Trigeminal Nerve.
2. Facial Nerve.
3. Glossopharyngeal Nerve.
4. Vagus Nerve.

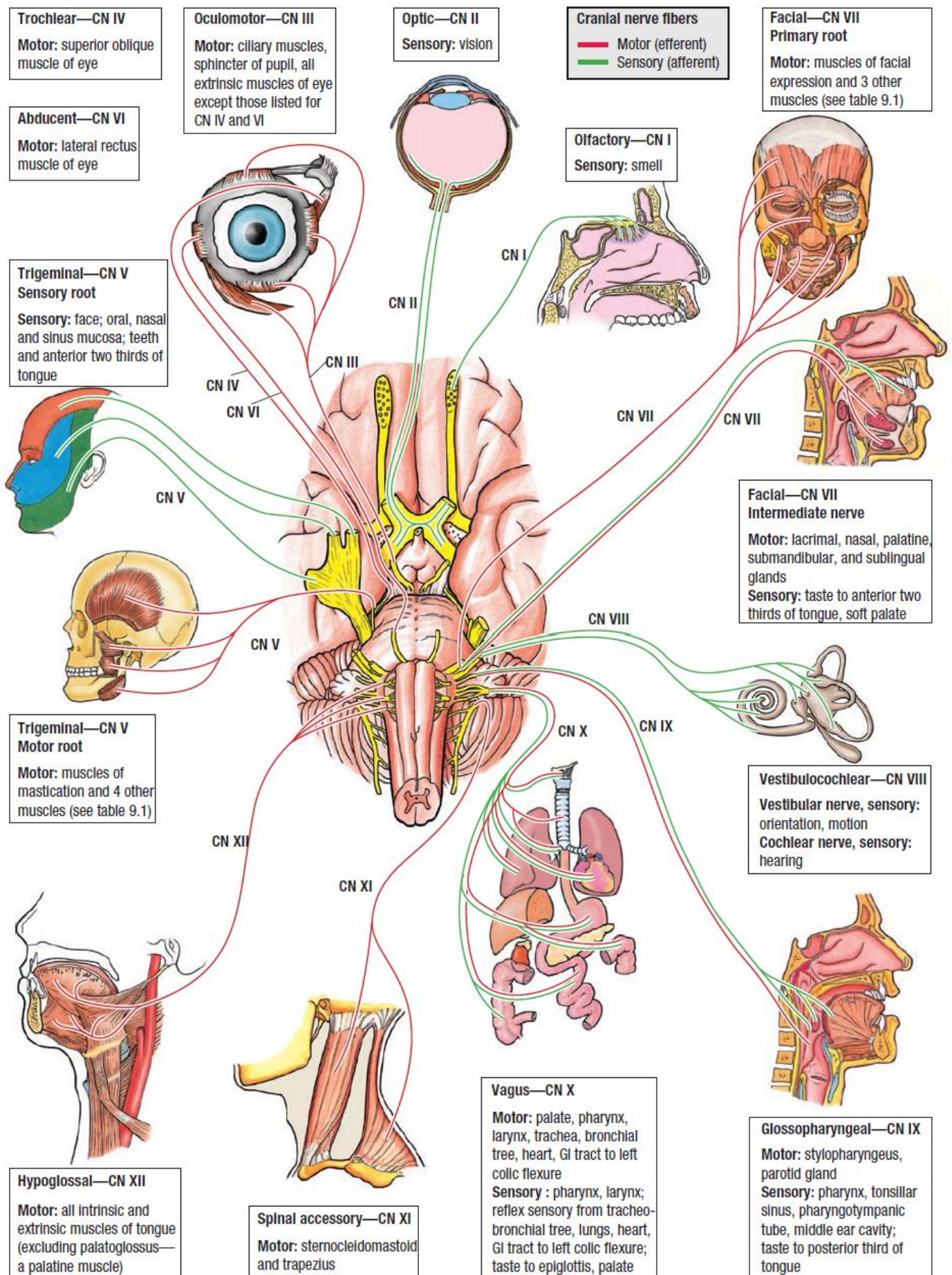
- **Nucleus:** A collection of nerve cell bodies located in the central nervous system (e.g., Chief sensory nucleus of CN-V, motor nucleus of CN-VII)
- **Ganglion:** A collection of nerve cell bodies located in the peripheral nervous system. (e.g., Trigeminal ganglion, ciliary ganglion)
- **Afferent (Sensory)** fibers terminates in the Nuclei.
- **Efferent (Motor)** fibers originate in the Nuclei.

- **Functional components (Columns) of CN:**

1. **General Somatic Afferents (GSA):**
 - General Sensation from skin and membranes (e.g., touch, pressure, heat, cold).
2. **General Visceral Afferents (GVA):**
 - General Sensation from viscera (e.g., Gut).
3. **Special Somatic Afferents (SSA):**
 - Vision, Hearing and Equilibrium.
4. **Special Visceral Afferents (SVA):**
 - Olfaction and Taste.
5. **General Somatic Efferents (GSE):**
 - Motor fibers innervating voluntary (striated muscle).
6. **General Visceral Efferents (GVE):**
 - Motor fibers innervating involuntary (smooth muscle) and Glands.
7. **Special Visceral Efferents (SVE):**
 - Motor fibers innervating voluntary Skeletal muscle which develops from Pharyngeal (branchial) Arches.

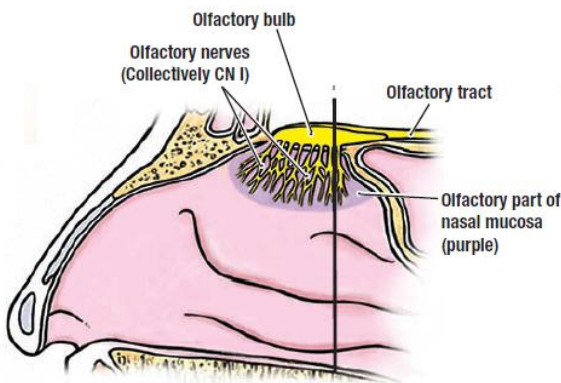
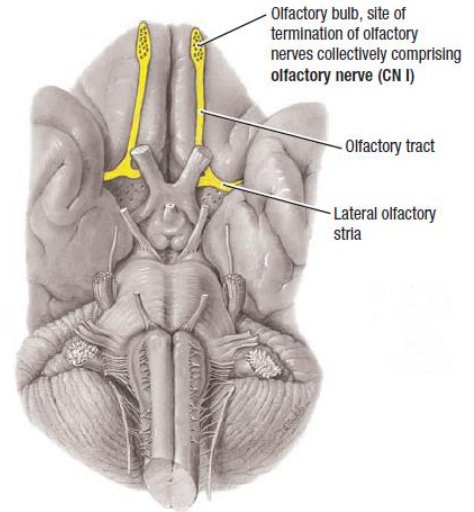




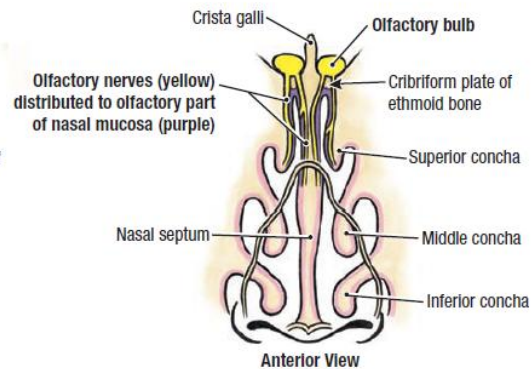


Nerve	Riyadh	Cr. p. Notes	Location of Nerve Cell Bodies	Cranial Exit	Function	
Olfactory (CN I)		Special sensory	Olfactory epithelium (olfactory cells)	Foramina in cribriform plate of ethmoid bone	Smell from nasal mucosa of roof of each nasal cavity, superior sides of nasal septum and superior concha	
Optic (CN II)		Special sensory	Retina (ganglion cells)	Optic canal	Vision from retina	
Oculomotor (CN III)		Somatic motor	Midbrain (nucleus of CN III)	Superior orbital fissure	Motor to superior, inferior, and medial rectus, inferior oblique, and levator palpebrae superioris that raise upper eyelid and direct gaze superiorly, inferiorly, and medially	
		Visceral motor	Presynaptic: midbrain (Edinger-Westphal nucleus); Postsynaptic: ciliary ganglion		Parasympathetic innervation to sphincter pupillae and ciliary muscles that constrict pupil and accommodate lens of eye	
Trochlear (CN IV)		Somatic motor	Midbrain (nucleus of CN IV)		Motor to superior oblique that assists in directing gaze inferolaterally	
Trigeminal (CN V) Ophthalmic division (CN V ¹)		Somatic (general) sensory	Trigeminal ganglion Synapse: sensory nucleus of CN V			Sensation from cornea, skin of forehead, scalp, eyelids, nose, and mucosa of nasal cavity and paranasal sinuses
Maxillary division (CN V ²)				Foramen rotundum	Sensation from skin of face over maxilla including upper lip, maxillary teeth, mucosa of nose, maxillary sinuses, and palate	
Mandibular division (CN V ³)		Somatic (branchial) motor	Pons (motor nucleus of CN V)	Foramen ovale	Sensation from the skin over mandible, including lower lip and side of head, mandibular teeth, temporomandibular joint, and mucosa of mouth and anterior two thirds of tongue	
					Motor to muscles of mastication, mylohyoid, anterior belly of digastric, tensor veli palatini, and tensor tympani	
Abducent (CN VI)		Somatic motor	Pons (nucleus of CN VI)	Superior orbital fissure	Motor to lateral rectus to direct gaze laterally	
Facial (CN VII)		Somatic (branchial) motor	Pons (motor nucleus of CN VII)	Internal acoustic meatus, facial canal, and stylomastoid foramen	Motor to muscles of facial expression and scalp; also supplies stapedius of middle ear, stylohyoid, and posterior belly of digastric	
		Special sensory	Geniculate ganglion Synapse: nuclei of solitary tract			
		General sensory	Geniculate ganglion Synapse: sensory nucleus of CN V		Taste from anterior two thirds of tongue, and palate	
		Visceral motor	Presynaptic: pons (superior salivatory nucleus); Postsynaptic: pterygopalatine ganglion and submandibular ganglion		Sensation from skin of external acoustic meatus Parasympathetic innervation to submandibular and sublingual salivary glands, lacrimal gland, and glands of nose and palate	
Vestibulocochlear (CN VIII) Vestibular		Special sensory	Vestibular ganglion Synapse: vestibular nuclei	Internal acoustic meatus	Vestibular sensation from semicircular ducts, utricle, and saccule related to position and movement of head	
Cochlear		Special sensory	Spiral ganglion Synapse: cochlear nuclei		Hearing from spiral organ	
Glossopharyngeal (CN IX)		Somatic (branchial) motor	Medulla (nucleus ambiguus)	Jugular foramen	Motor to stylopharyngeus that assists with swallowing	
		Visceral motor	Presynaptic: medulla (inferior salivatory nucleus); Postsynaptic: otic ganglion		Parasympathetic innervation to parotid gland	
		Visceral sensory	Inferior ganglion		Visceral sensation from parotid gland, carotid body and sinus, pharynx, and middle ear	
		Special sensory	Inferior ganglion Synapse: nuclei of solitary tract		Taste from posterior third of tongue	
		General sensory	Superior ganglion Synapse: sensory nucleus of CN V		Cutaneous sensation from external ear	
Vagus (CN X)		Somatic (branchial) motor	Medulla (nucleus ambiguus)		Jugular foramen	Motor to constrictor muscles of pharynx, intrinsic muscles of larynx, muscles of palate (except tensor veli palatini), and striated muscle in superior two thirds of esophagus
		Visceral motor	Presynaptic: medulla; Postsynaptic: neurons in, on, or near viscera			Smooth muscle of trachea, bronchi, and digestive tract, cardiac muscle
		Visceral sensory	Inferior ganglion Synapse: nuclei of solitary tract			Visceral sensation from base of tongue, pharynx, larynx, trachea, bronchi, heart, esophagus, stomach, and intestine
		Special sensory	Inferior ganglion Synapse: nuclei of solitary tract			
		Somatic (general) sensory	Superior ganglion Synapse: sensory nucleus of trigeminal nerve			Taste from epiglottis and palate Sensation from auricle, external acoustic meatus, and dura mater of posterior cranial fossa
Spinal accessory nerve (CN XI)		Somatic motor	Cervical spinal cord		Motor to sternocleidomastoid and trapezius	
Hypoglossal (CN XII)		Somatic motor	Medulla (Nucleus of CN XII)	Hypoglossal canal	Motor to muscles of tongue (except palatoglossus)	

- **CRANIAL NERVE I: OLFACTORY:**
- Arise from **Olfactory receptor nerve cells** in the olfactory mucous membrane which is situated in the upper part of nasal cavity above the level of superior turbinate.
- Olfactory nerve bundles pass through **Cribriform plate** of Ethmoid bone.
- Enter the **Olfactory Bulb** in Cranial cavity.
- Olfactory Bulb is connected to olfactory area in cerebral cortex by **Olfactory Tract**.
- **Pure Sensory (SVA)**



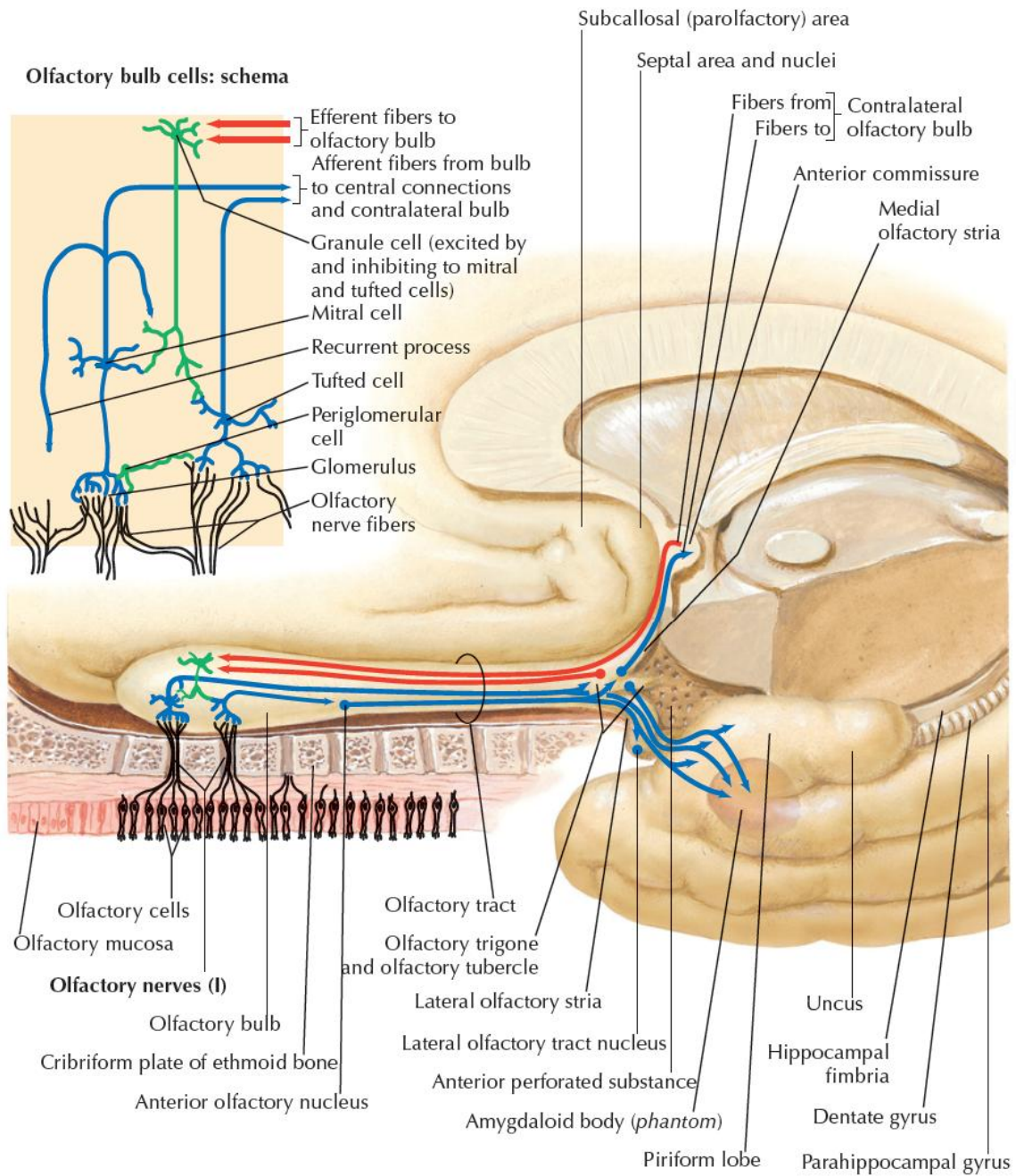
A. Medial View of Lateral Wall of Nasal Cavity



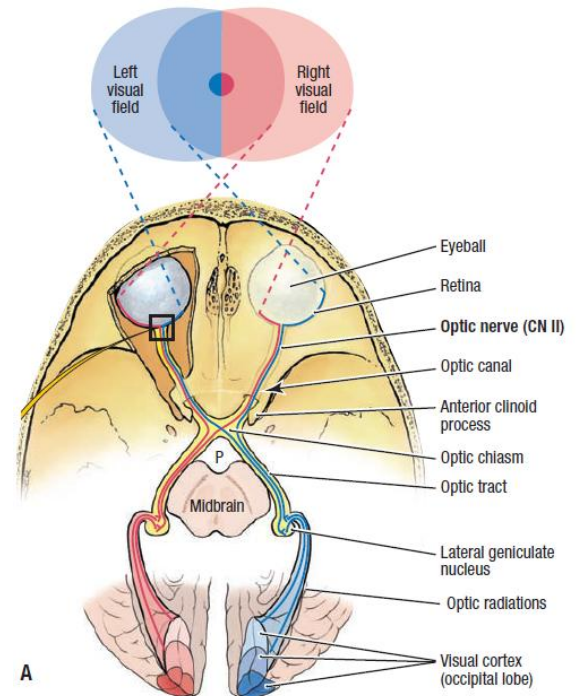
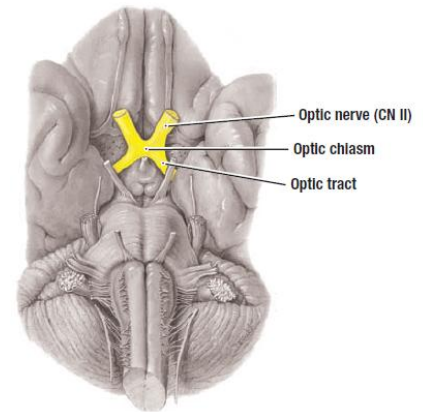
Anterior View

CRANIAL NERVE I: OLFACTORY NERVE

Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
SVA	Fibers originate in the neurosensory cells of the olfactory epithelium. The primary fibers travel through the cribriform plate to synapse on the secondary fibers within the olfactory bulb. These fibers continue posteriorly as the olfactory tract that carries the fibers to the olfactory areas.	The secondary fibers continue to synapse in the olfactory areas: • Lateral olfactory area • Anterior olfactory nucleus • Intermediate olfactory area • Medial olfactory area	The SVA fibers are responsible for the sense of smell.	Tumors of the olfactory lobe can affect the olfactory system.



- **CRANIAL NERVE II: OPTIC:**
- Composed of axons of the cells of **Ganglionic layer of Retina.**
- Emerges from the back of eyeball and leaves the orbital cavity through **Optic canal** to enter the cranial cavity.
- Optic nerve then unites with the optic nerve of opposite side to form **Optic Chiasma.**
- In optic Chiasma:
 - o Fibers from Medial half of each retina cross the midline and enter the **Optic tract** of opposite side.
 - o Fibers from Lateral half of each retina pass posteriorly in the optic tract of same side.
- Most fibers of Optic Tract terminate by synapsing with nerve cells in **Lateral Geniculate Body.**
- Axons of nerve cells of lateral geniculate body pass posteriorly as **Optic radiation** and terminate in the **Visual cortex** of the cerebral hemisphere.
- **Pure Sensory (SSA)**

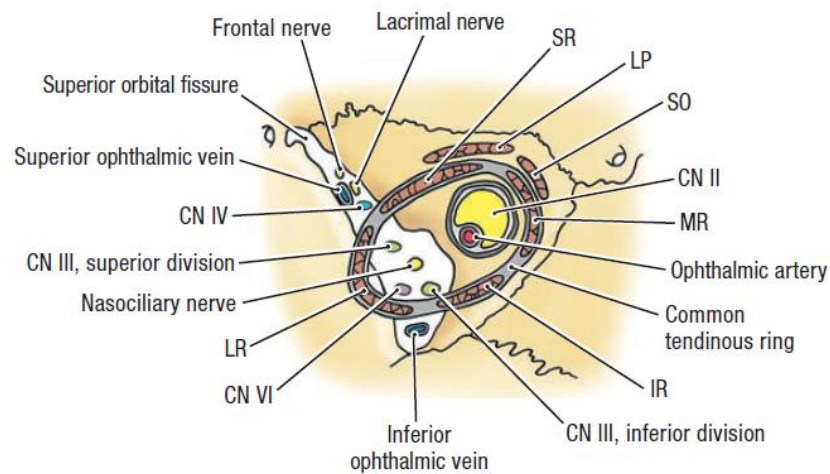
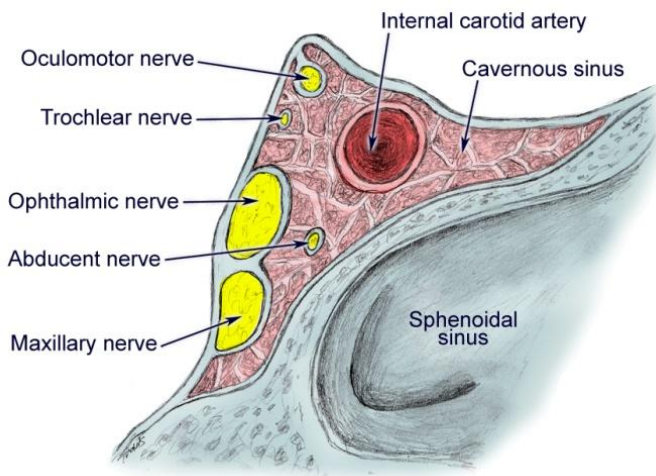
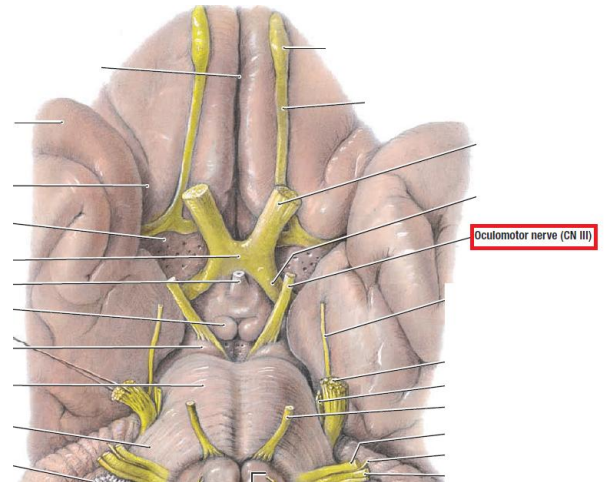


CRANIAL NERVE II: OPTIC NERVE

Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
SSA	Begins in the retina with the receptors of rods and cones that synapse on bipolar cells which synapse with ganglion cells	Ganglionic axons form the optic nerve that meets in an incomplete crossing at the optic chiasm where: • Nasal retinal fibers decussate to the opposite side • Temporal retinal fibers remain on the ipsilateral side These form an optic tract that terminates on the lateral geniculate nucleus Fibers from the lateral geniculate travel to synapse in the occipital lobe	The SSA fibers are responsible for vision	Lesions of the optic nerve lead to blindness Lesions of the optic chiasm lead to bitemporal hemianopsia Lesions of the optic tract lead to homonymous hemianopsia

- **CRANIAL NERVE III: OCULOMOTOR:**

- Begins in **Oculomotor Nucleus** in the Midbrain.
- Fibers emerges on Anterior surface of **Midbrain**.
- Continues into Middle Cranial Fossa in the **lateral wall of Cavernous Sinus** where it divides into Superior and Inferior ramus.
- Enter Orbital cavity through **Superior orbital fissure**.



- CN-III Nuclei:

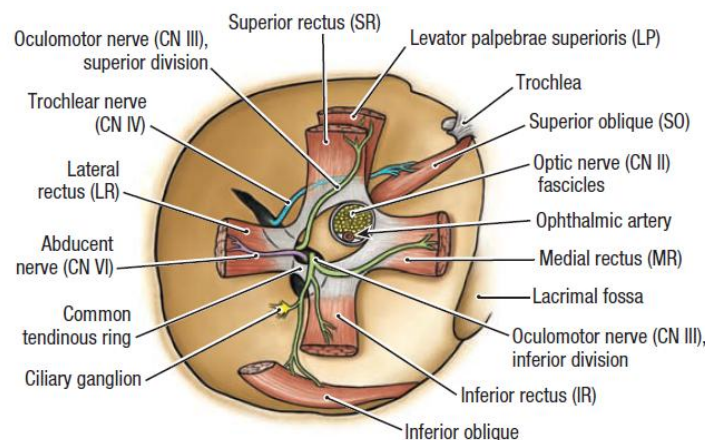
1. Oculomotor Nucleus:

- At Midbrain.
- GSE
- Supplies All Extrinsic eye muscles **Except** Superior oblique and Lateral Rectus.

2. Edinger-Westphal nucleus:

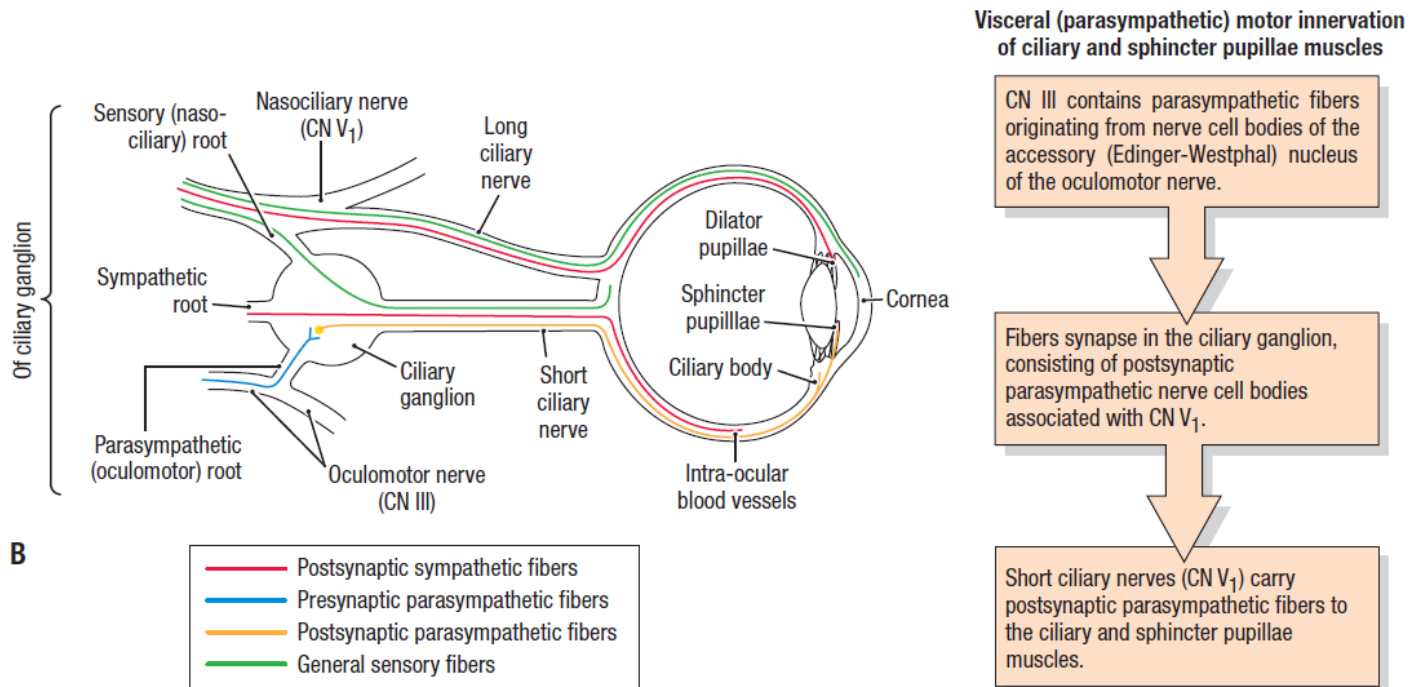
- GVE
- Supplies Constrictor Pupillae and Ciliary Muscle.
- Via ciliary ganglion.

- **Pure Motor.**
- GSE supply to the following Extrinsic muscles of eye:
 - 1. Levator Palpebrae Superioris:**
 - Superior division of CN-III
 - **Upper eyelid Elevation.**
 - 2. Superior Rectus:**
 - Superior division of CN-III
 - **Eye Elevation, Intorsion, and Adduction**
 - 3. Medial Rectus:**
 - Inferior division of CN-III
 - **Eye Adduction.**
 - 4. Inferior Rectus:**
 - Inferior division of CN-III
 - **Eye Depression, Extortion and Adduction.**
 - 5. Inferior Oblique.**
 - Inferior division of CN-III
 - **Eye Extorsion, Elevation and Abduction.**
- GVE supply to the following Intrinsic muscles of eye:
 - 1. Constrictor Pupillae of the iris:**
 - Pupil Constriction (Light reflex and Accommodation)
 - Controlled by Parasympathetic fibers origination from the **Edinger-Westphal nucleus**, travel along the oculomotor nerve (CN III), synapse in the ciliary ganglion, and then enter the eye via the short ciliary nerves.
 - 2. Ciliary Muscles.**
 - Accommodation and Regulation of trabecular meshwork pore size.
- **CN-III functions:**
 - Lifting the Upper eyelid.
 - Turning the Eye Upward, Downward and Medially.
 - Constricting the pupil.
 - Accommodation of the Eye.



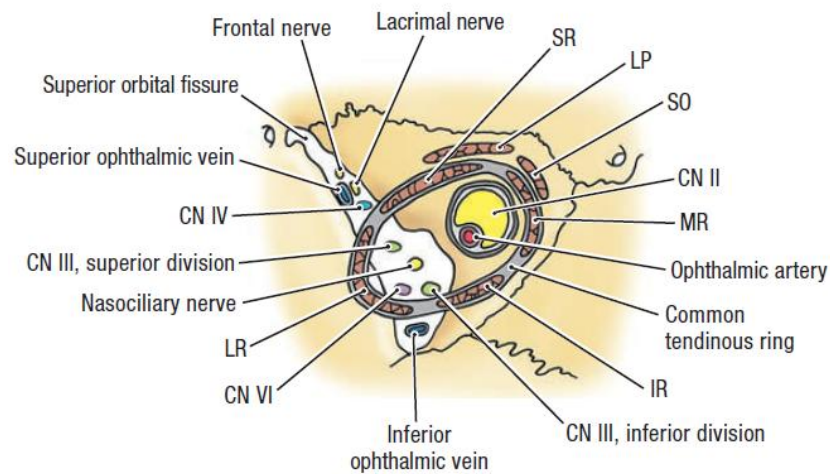
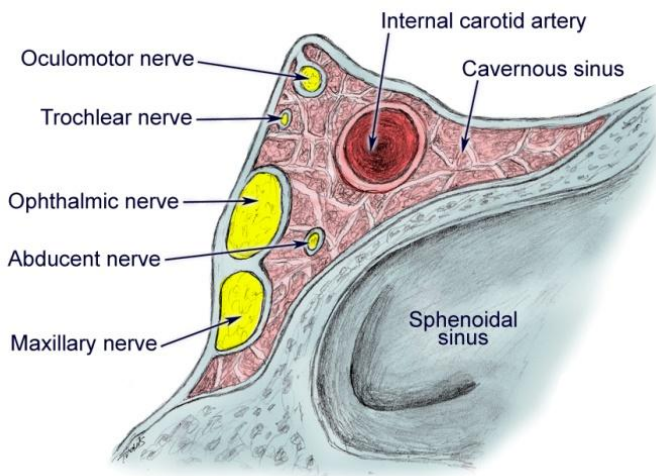
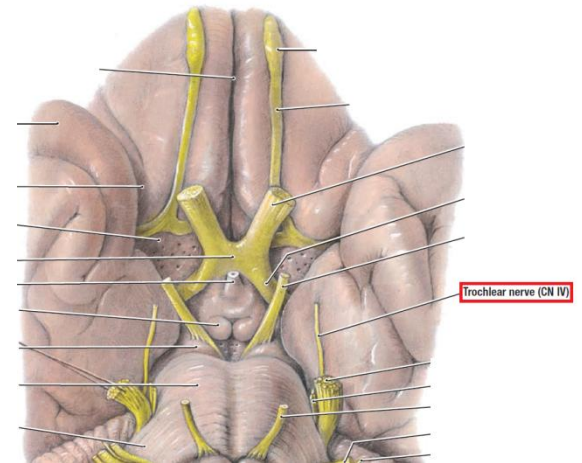
OCULOMOTOR NERVE				
Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSE	Begins in the oculomotor nucleus	Innervates the superior rectus, inferior rectus, medial rectus, inferior oblique, and levator palpebrae superioris mm.	GSE fibers are responsible for innervating the majority of the extraocular eye muscles	Lesions of the oculomotor nerve result in diplopia, lateral strabismus, ptosis, and mydriasis
GVE	Preganglionic parasympathetic fibers begin in the Edinger-Westphal nucleus	Innervates the sphincter pupillae and ciliary mm.	GVE fibers are responsible for providing the parasympathetic innervation to the intrinsic eye muscles	GVE fibers utilize 1 ganglion: • Ciliary

- Pupillary light reflex:
 - **Afferent:** Optic nerve **CN-II**
 - **Efferent:** Oculomotor nerve **CN-III** via Edinger-Westphal nucleus.
- Complete Oculomotor nerve palsy:
 1. Outward and Downward Eye displacement.
 2. Ptosis (drooping of the eyelid)
 3. Mydriasis (pupil dilation).



- **CRANIAL NERVE IV: TROCHLEAR:**

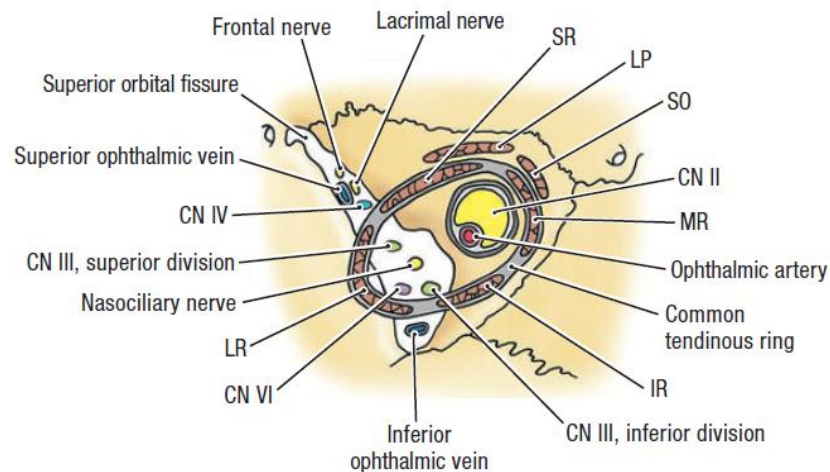
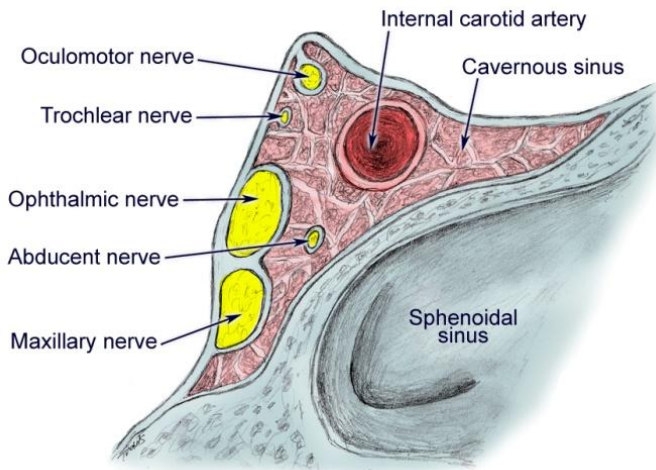
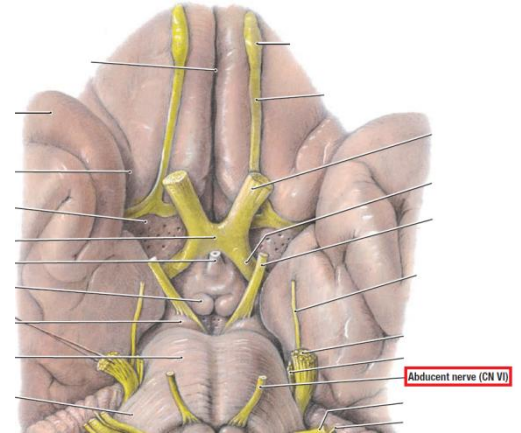
- Most slender CN.
- Begins in **Trochlear Nucleus** in the Midbrain.
- Fibers leave the Posterior surface of **Midbrain**.
- Immediately decussates with the nerve of the opposite side.
- Continues into Middle Cranial Fossa in the **Lateral wall of Cavernous Sinus**.
- Enter Orbital cavity through **Superior orbital fissure**.



- **Pure Motor.**
- GSE supply to one Extrinsic muscle of eye:
- 1. Superior Oblique Muscle:**
 - o **Eye Intorsion, Abduction and depression.**
 - o Turning the Eye Downward and Laterally.

TROCHLEAR NERVE				
Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSE	Begins in the trochlear nucleus	Innervates the superior oblique m.	GSE fibers are responsible for innervating 1 extraocular muscle of the eye: the superior oblique	The trochlear nerve exits the brainstem dorsally. Lesions of the trochlear n. result in diplopia. In trochlear n. lesions, the eye is adducted and elevated.

- **CRANIAL NERVE VI: ABDUCENT:**
- Begins in **Abducent Nucleus** which located beneath 4th ventricle in caudal portion of **Pons**.
- Emerges in Anterior surface **between Pons and Medulla oblongata**.
- Pass forward with Internal Carotid Artery through **Cavernous Sinus** in the middle cranial fossa.
- Enter Orbital cavity through **Superior orbital fissure**.

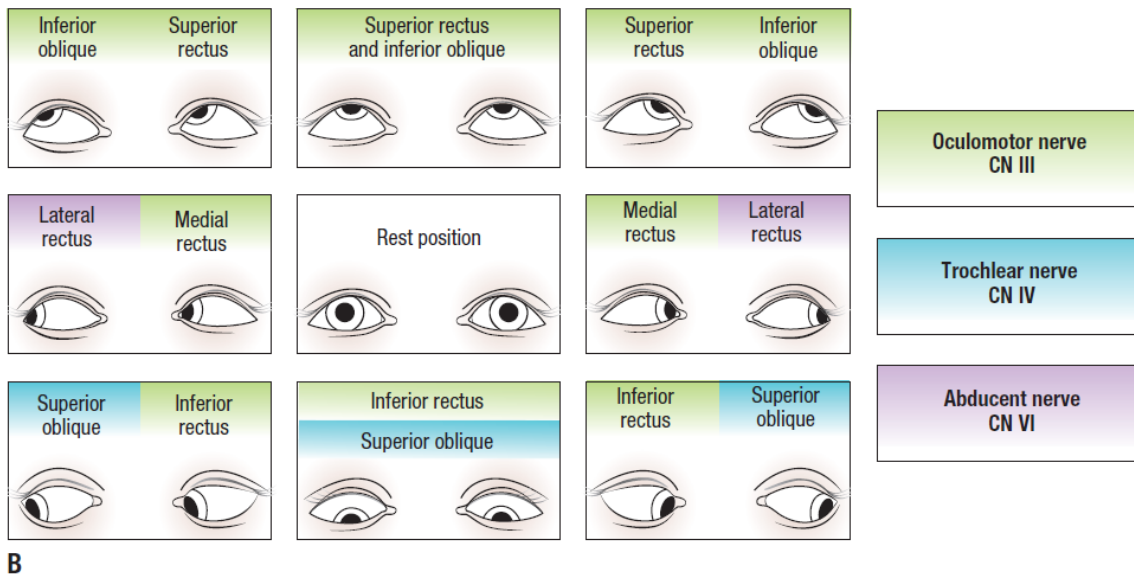
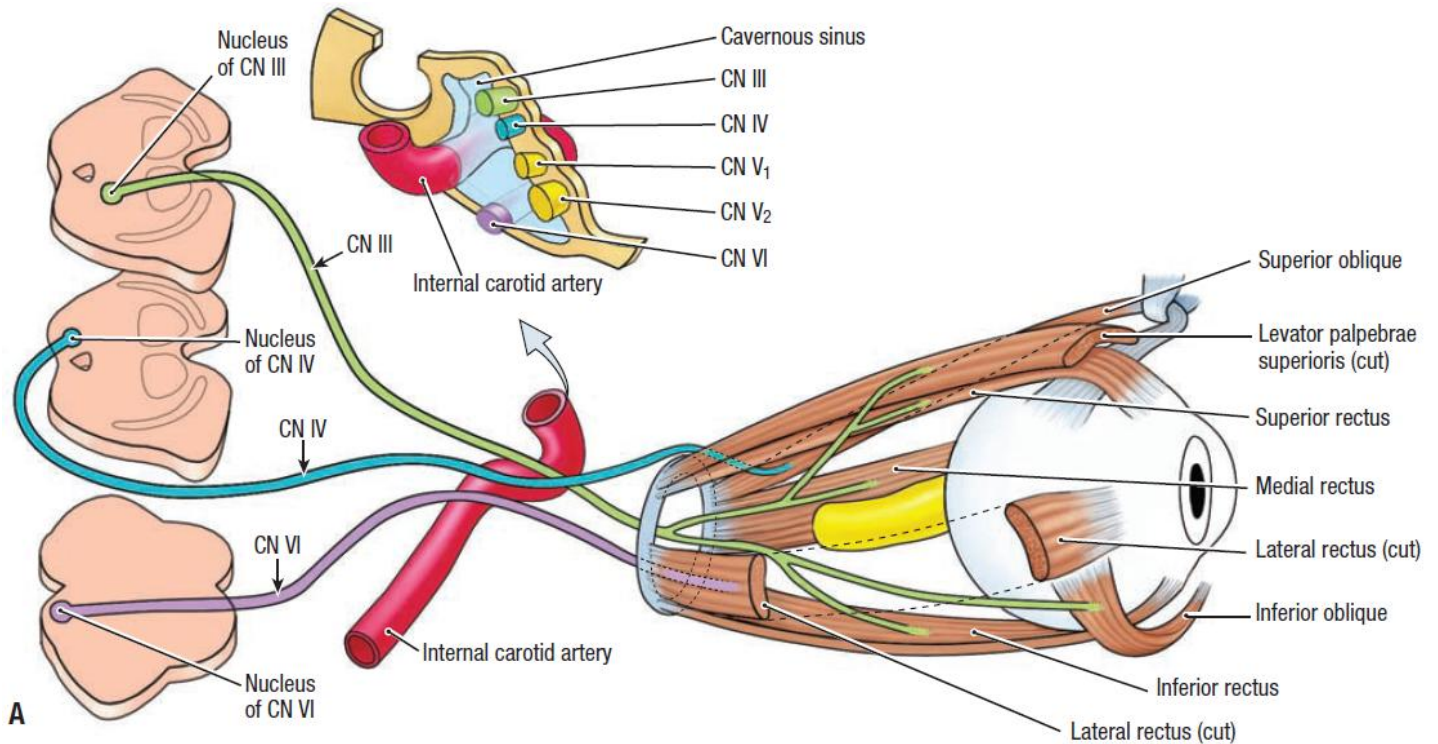


- **Pure Motor.**
- GSE supply to one Extrinsic muscle of eye:

1. Lateral Rectus:

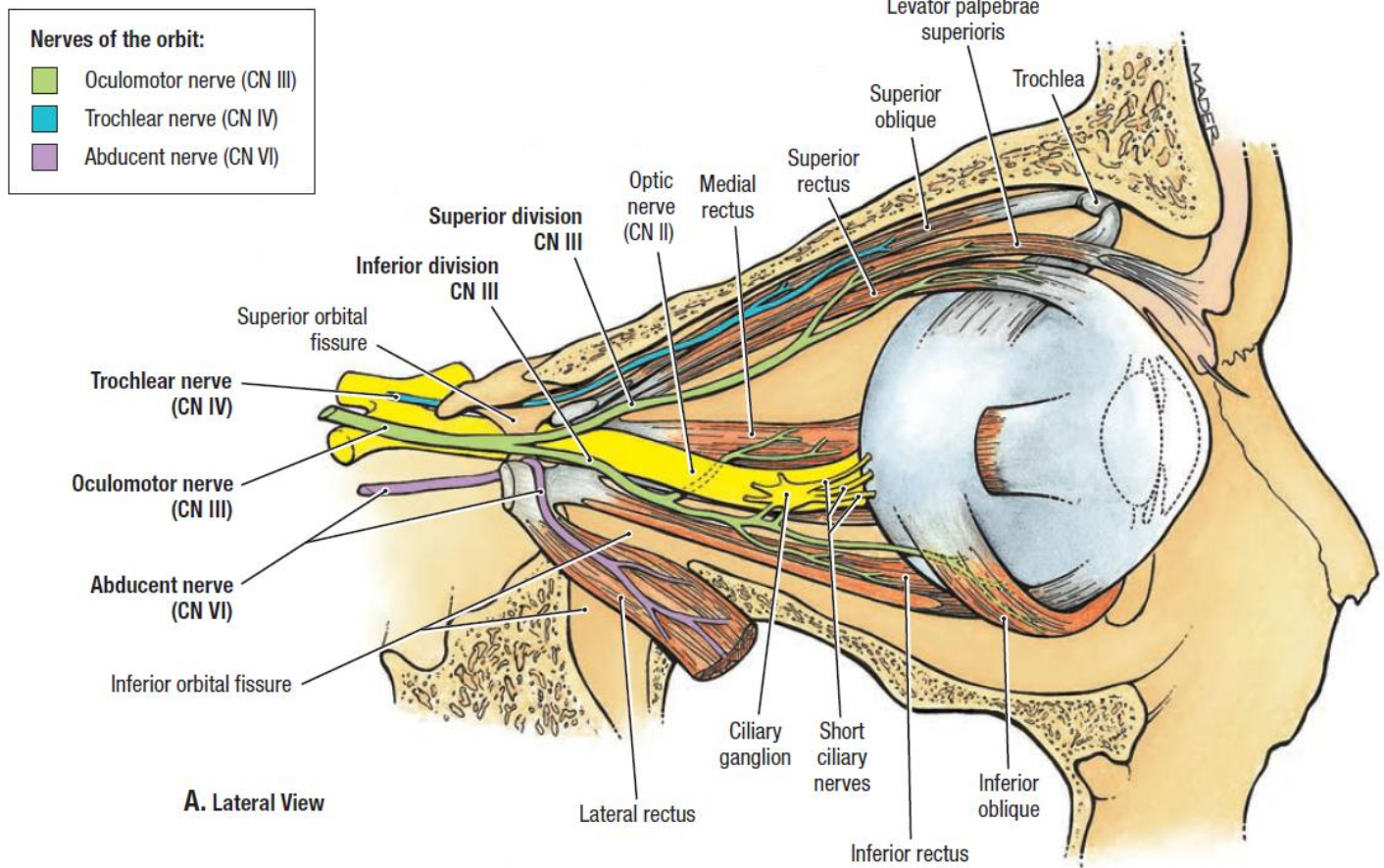
- **Eye Abduction**

ABDUCENS NERVE				
Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSE	Begins in the abducens nucleus	Innervates the lateral rectus m.	GSE fibers are responsible for innervating 1 extraocular muscle of the eye: the lateral rectus	Lesions of the abducens nerve result in diplopia and medial strabismus

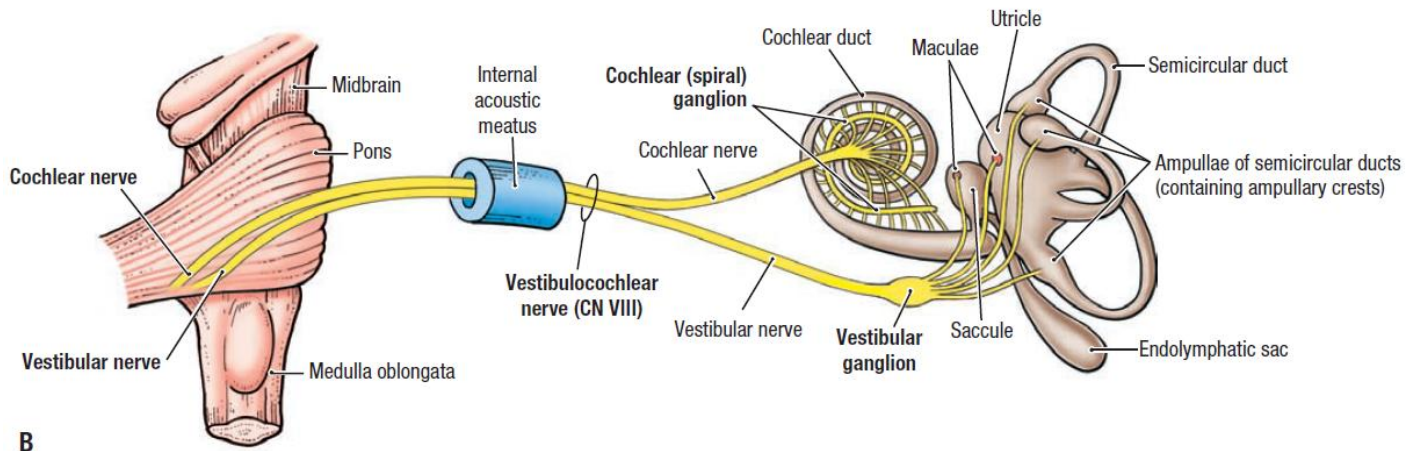
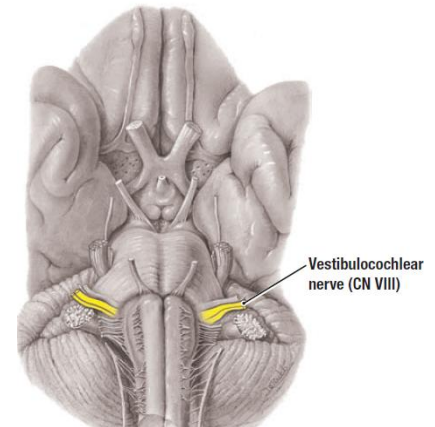


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Nerve	Functional Components	Cells of Origin/Termination	Cranial Exit	Distribution and Functions
Oculomotor	Somatic motor	Nucleus of CN III	Superior orbital fissure	Motor to superior, inferior, and medial recti, inferior oblique, and levator palpebrae superioris muscles; raises upper eyelid, directing gaze superiorly, inferiorly, and medially
	Visceral motor (parasympathetic)	Presynaptic: midbrain (Edinger-Westphal nucleus); Postsynaptic: ciliary ganglion		Motor to sphincter pupillae and ciliary muscle that constrict pupil and accommodate lens of eyeball
Trochlear	Somatic motor	Nucleus of CN IV		Motor to superior oblique that assists in directing gaze inferolaterally
Abducent	Somatic motor	Nucleus of CN VI		Motor to lateral rectus that directs gaze laterally



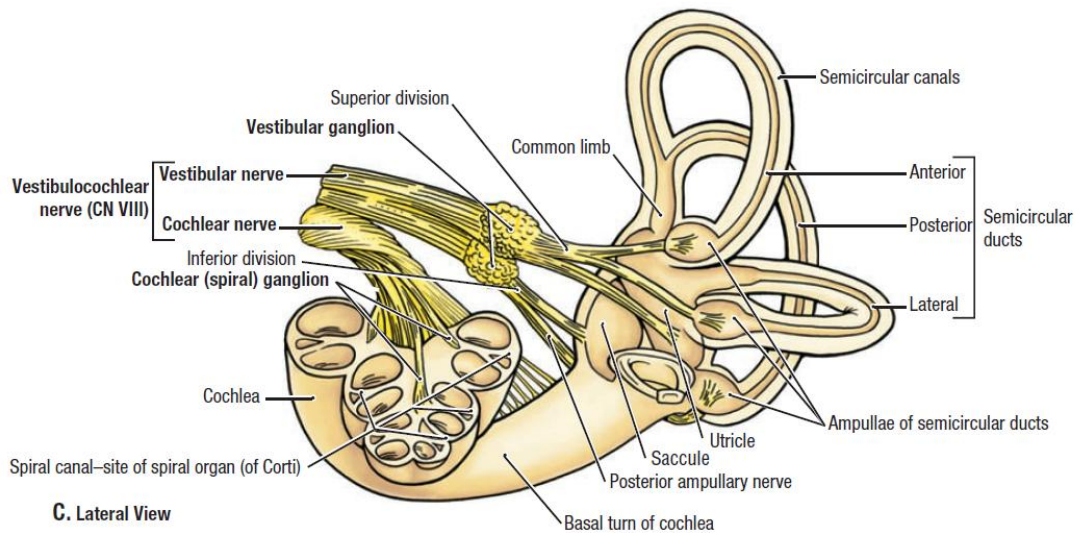
- **CRANIAL NERVE VIII: VESTIBULOCOCHLEAR:**
- Consist of 2 sets of Sensory Fibers:
 - o Vestibular Nerve
 - o Cochlear Nerve
- Leave the Anterior surface of brain **between Pons and Medulla oblongata**.
- Cross the Posterior cranial fossa and enter the **Internal Acoustic Meatus** with the Facial nerve.
- **Pure Sensory.**



B

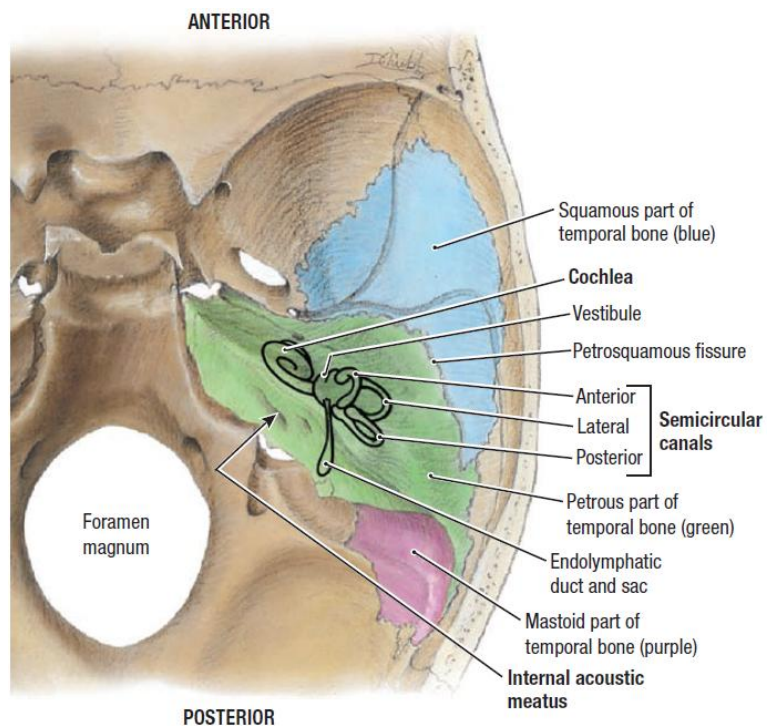
- **Vestibular Nerve:**
- Arising from neurons in the Vestibular (Scarpa's) ganglion in the outer part of IAM.
- **4 Vestibular nuclei:**
 1. Medial vestibular nucleus: (Medulla)
 2. Lateral vestibular nucleus: (Medulla)
 3. Inferior vestibular nucleus: (Medulla)
 4. Superior vestibular nucleus: (Pons)
- Divides into superior and inferior branches.
- **Superior Vestibular Nerve:**
 1. Superior SSC.
 2. Horizontal SSC.
 3. Utricle.
- **Inferior Vestibular Nerve:**
 1. Posterior SSC.
 2. Saccule.

Part of Vestibulocochlear Nerve	Functional Components	Cells of Origin/Termination	Cranial Exit	Distribution and Functions
Vestibular nerve	Special sensory	Vestibular ganglion/vestibular nuclei	Internal acoustic meatus	Vestibular sensation from semicircular ducts, utricle, and saccule related to position and movement of head
Cochlear nerve		Spiral ganglion/cochlear nuclei		Hearing from spiral organ



- **Cochlear Nerve:**
- Arising from neurons in the Spiral ganglion of Cochlea.
- 2 Cochlear nuclei in Inferior Cerebellar Peduncle:
 1. Dorsal cochlear nucleus (DCN)
 2. Ventral (Accessory) cochlear nucleus (VCN)

Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
SSA	Organ of Corti Cristae of semicircular canals Maculae of utricle and saccule	Cochlear and vestibular nuclei	SSA fibers travel from the various vestibulocochlear receptors to their respective nuclei in the brainstem	Vestibulocochlear and facial nn. both enter the internal acoustic meatus and can be affected by tumors in the region



- **Internal Acoustic Meatus (IAM):**
- A canal in the petrous part of the temporal bone.
- Carries nerves from inside the skull towards the middle and inner ear, namely cranial nerve VII and cranial nerve VIII. There are two internal auditory meati in most individuals, one on the left side of the skull and one on the right.
- The opening to the internal acoustic meatus is located inside the cranial cavity, near the center of the posterior surface of the petrous part of the temporal bone. The size varies considerably; its margins are smooth and rounded. The canal is short (about 1 cm) and runs laterally into the bone. At its end are the openings for three different canals, one of which is the facial canal.

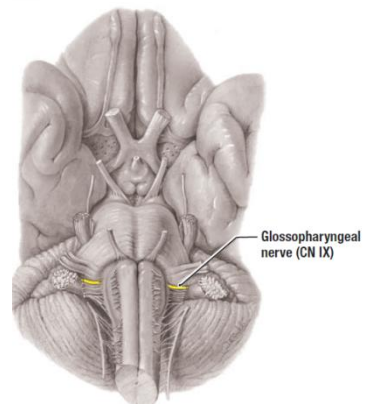
The internal acoustic meatus transmits the facial and vestibulocochlear nerves and the labyrinthine artery (an internal auditory branch of the basilar artery). The facial nerve travels through the facial canal, eventually exiting the skull at the stylomastoid foramen.

The opening of the meatus is called the porus acusticus internus, or its English translation, the internal acoustic opening.

The antero-superior part transmits the facial nerve and nervus intermedius and is separated from the postero-superior section, which transmits the superior vestibular nerve, by Bill's bar (named by William F. House). The falciform crest or transverse crest separates the superior part from the inferior part. The cochlear nerve runs antero-inferiorly and the inferior vestibular nerve runs postero-inferiorly.

- **CRANIAL NERVE IX: GLOSSOPHARYNGEAL:**

- Emerges from Lateral surface of **Medulla oblongata** between Olive and Inferior cerebellar peduncle.
- Passes in the posterior cranial fossa.
- Leaves the skull through **Jugular Foramen**.
- Descends through the upper part of the neck
- Passes between IJV and ICA.
- Passes together with styloid apparatus between Internal and External carotid arteries.
- Curves forward, forming an arch on Middle constrictor muscle.
- Passes deep to Hyoglossus.
- Pierces Middle constrictor to Palatine tonsil, Mucous membrane of the base of Tongue and Mucous glands of the mouth.
- **Mixed Motor and Sensory Nerve**.



- 4 Nuclei:

1. Nucleus Ambiguus: (Motor)

- At medulla oblongata.
- **StyloPharyngus muscle**.
- Branchial Efferent (SVE)

2. Inferior Salivatory Nucleus: (SecretoMotor)

- At medulla oblongata.
- Postsynaptic to Otic ganglion for **Parasympathetic innervation of Parotid gland**.
- GVE

3. Nucleus of Tractus Solitarius: (Sensory)

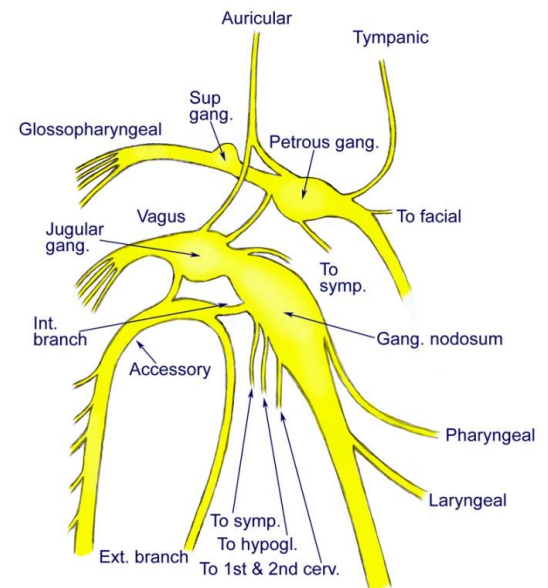
- SVA → **Taste fibers from Posterior 1/3 of Tongue**.
- GVA → **Mucus membrane of Nasopharynx, Oropharynx, Middle ear and Carotid body**.
- GVA is predominantly the sensory portion of pharyngeal plexus.

4. Spinal Trigeminal Nucleus: (Sensory)

- General Sensation from mucus membrane of Posterior 1/3 of Tongue and part of External ear.
- GSA

- Glossopharyngus Nerve has 2 Ganglionic swellings, which are the sensory ganglia of the nerve:

- Superior (Jugular) Ganglion.
- Inferior (Petrous) Ganglion.



- **Branches of Glossopharyngeal nerve:**

1. Tympanic Nerve: (Jacobson Nerve)

- Arises from Inferior ganglion.
- Ascends to Tympanic cavity.
- Divides in Tympanic cavity into branches to form Tympanic plexus upon surface of promontory.
- This plexus gives off:
 - 1. Lesser Superficial Petrosal Nerve**
 - 2. A branch to join Greater Superficial Petrosal Nerve**
 - 3. Branches to Tympanic cavity.**

2. Carotid Branches:

- Descend along Internal Carotid Artery
- Communicates with Pharyngeal branch of the vagus and with branches of Sympathetic.

3. Pharyngeal Branches:

- Unite with Pharyngeal branches of Vagus and Sympathetic.
- Form Pharyngeal plexus.

4. Nerve to Stylopharyngeus.

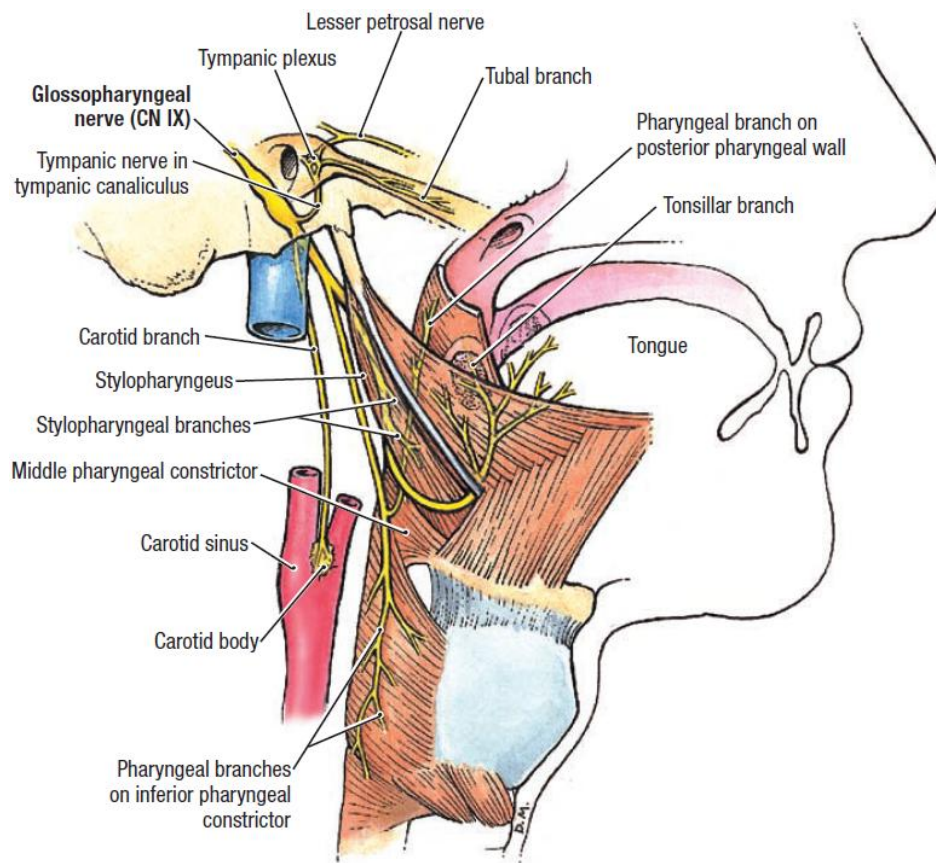
- Supply Stylopharyngeus Muscle.

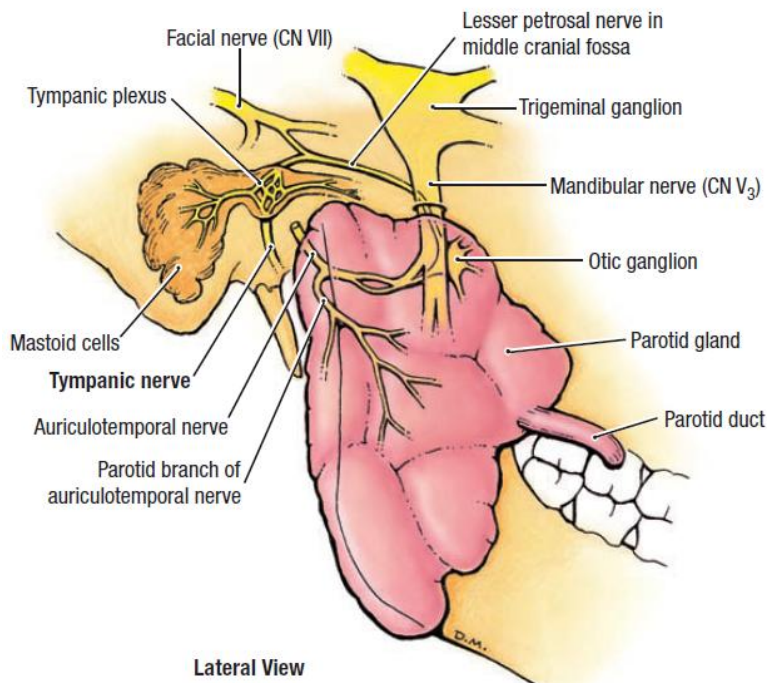
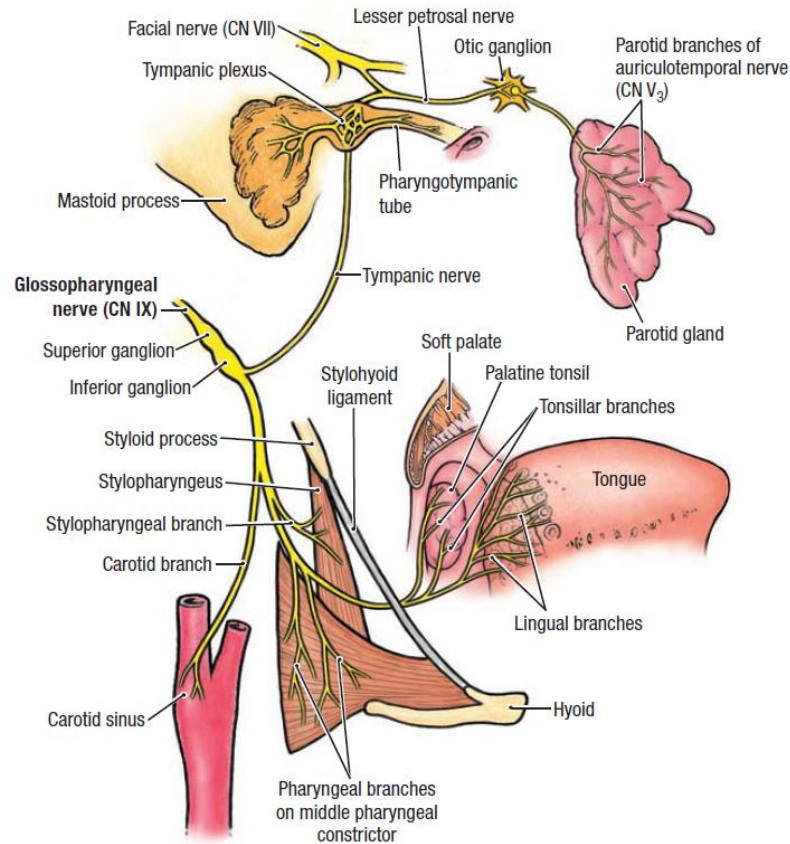
5. Tonsillar Branches:

- Supply Palatine tonsil, Soft palate.

6. Lingual Branches:

- Supply ucouc membrane and glands of the posterior part of the tongue.
- Communicates with Lingual nerve.





Visceral motor (parasympathetic) innervation of parotid gland

Tympanic nerve arises from CN IX and emerges with it from jugular foramen.

Tympanic nerve enters the middle ear via the tympanic canaliculus in petrous part of the temporal bone.

Tympanic nerve forms the tympanic plexus on the promontory of middle ear.

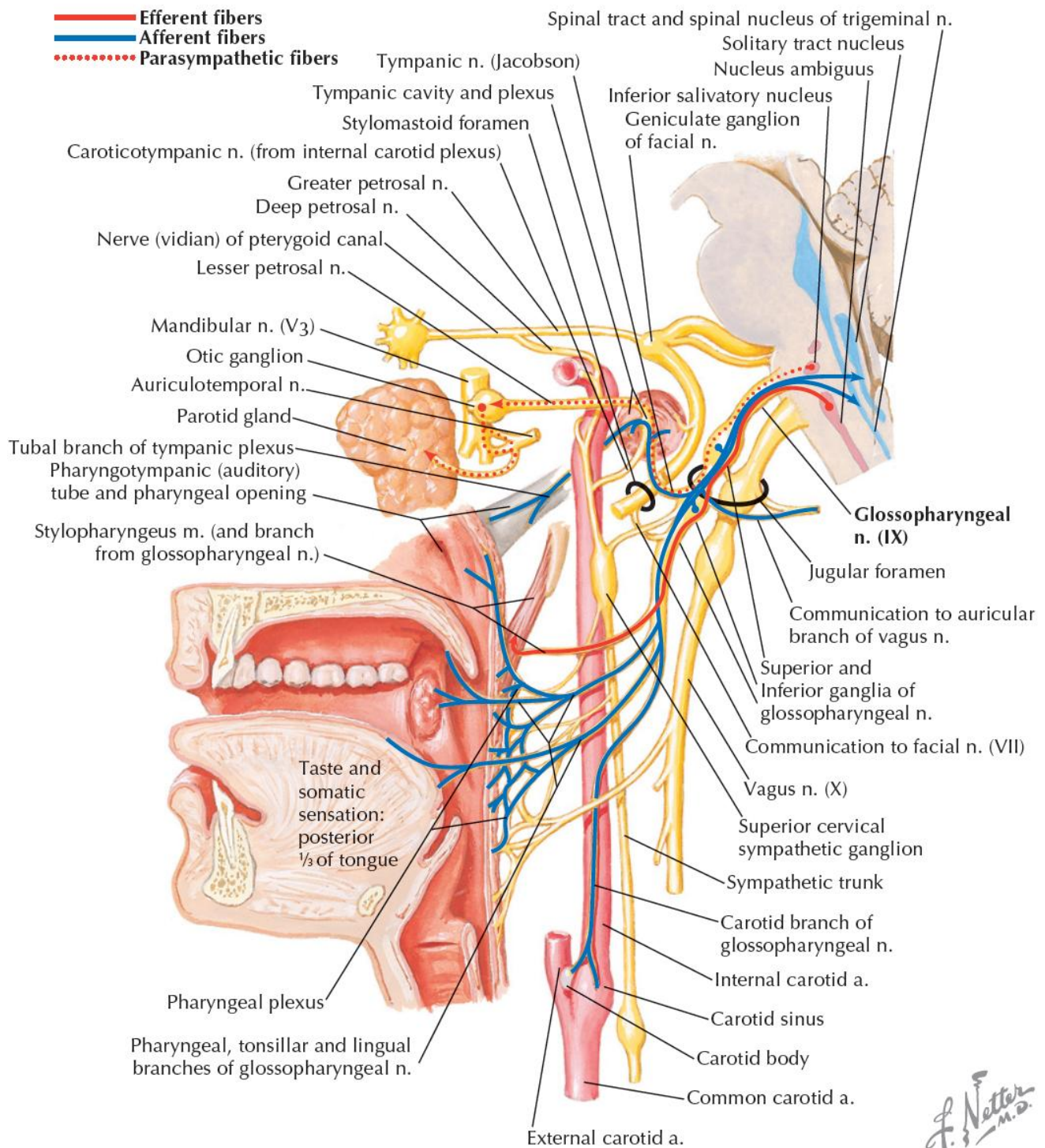
The lesser petrosal nerve arises as a branch of the tympanic plexus.

Lesser petrosal nerve penetrates roof of tympanic cavity (tegmen tympani) to enter middle cranial fossa.

Lesser petrosal nerve leaves the cranium through the foramen ovale.

Parasympathetic fibers synapse in the otic ganglion.

Postsynaptic fibers pass to parotid gland via branches of auriculotemporal nerve (CN V₃).



CRANIAL NERVE IX: GLOSSOPHARYNGEAL NERVE

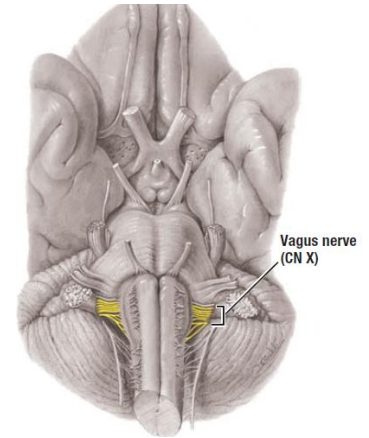
Riyadh et al. Notes

Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSA	Afferent fibers begin in the various receptors of the skin of the external ear and the posterior 1/3 of the tongue	Pain and temperature fibers terminate in the spinal nucleus of V	GSA fibers are responsible for providing sensory innervation to a small portion of the external ear and posterior 1/3 of the tongue GSA fibers of the glossopharyngeal n. utilize the trigeminothalamic lemniscus to carry their sensory impulses to consciousness	Nerve cell bodies for the primary fibers are located in the superior ganglion of IX
SVA	Afferent fibers begin in the taste receptors of the posterior 1/3 of the tongue	Primary afferent fibers travel in the tractus solitarius and terminate in the nucleus solitarius	SVA fibers are responsible for carrying the taste fibers from the circumvallate papillae and the taste buds on the posterior 1/3 of the tongue	Nerve cell bodies for the primary fibers are located in the inferior ganglion of IX
GVA	Afferent fibers begin in the various receptors of the mucous membranes of the nasopharynx, oropharynx, middle ear, carotid body, and carotid sinus	Primary afferent fibers travel in the tractus solitarius and terminate in the nucleus solitarius	GVA fibers utilize the same pathway as for the SVA fibers	The nerve cell bodies for the primary fibers are located in the inferior ganglion of IX GVA fibers are predominantly the sensory portion of the pharyngeal plexus
GVE	Preganglionic parasympathetic fibers begin in the inferior salivatory nucleus	Postganglionic parasympathetic fibers innervate parotid gland	The GVE fibers are responsible for providing the parasympathetic innervation to the parotid gland	GVE fibers utilize 1 ganglion: • Otic
SVE	Begins in the nucleus ambiguus	Innervates the stylopharyngeus m.	SVE fibers are responsible for innervating the muscles of the 3rd pharyngeal arch	Stylopharyngeus is the only muscle innervated by the glossopharyngeal n.

Nerve	Functional Components	Cells of Origin/Termination	Cranial Exit	Distribution and Functions
Glossopharyngeal	Somatic (branchial) motor	Nucleus ambiguus	Jugular foramen	Motor to stylopharyngeus that assists with swallowing
	Visceral motor	Presynaptic: inferior salivatory nucleus; postsynaptic: otic ganglion		Parasympathetic innervation to parotid gland
	Visceral sensory	Nuclei of solitary tract, spinal trigeminal nucleus/inferior ganglion		Visceral sensation from parotid gland, carotid body, carotid sinus, pharynx, and middle ear
	Special sensory	Nuclei of solitary tract /inferior ganglion		Taste from posterior third of tongue
	General sensory	Spinal trigeminal nucleus/superior ganglion		Cutaneous sensation from external ear

- **CRANIAL NERVE X: VAGUS:**

- Emerges from Anterior surface of **Medulla oblongata** between Olive and Inferior cerebellar peduncle.
- Passes in the posterior cranial fossa.
- Leaves the skull through **Jugular Foramen**.
- Descends through the neck Between the Carotid arteries and Internal Jugular vein within Carotid Sheath.
- It passes through the mediastinum of thorax, pierces the diaphragm with esophagus and terminates within Abdomen.
- Longest and most extensive distribution of all Cranial nerves.
- Mixed Motor and Sensory Nerve.



- 4 Nuclei:

1. Spinal Nucleus of Trigeminal: (Sensory)

- o GSA
- o General Sensation from Posterior portion of External ear, EAC and TM.

2. Nucleus of Tractus Solitarius: (Sensory)

- o SVA → Taste fibers from Epiglottis and Palate.
- o GVA → Mucus membrane of Hypopharynx and Larynx.

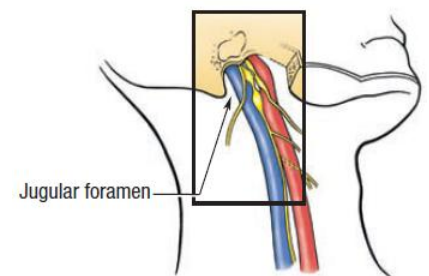
3. Spinal Trigeminal Nucleus: (Sensory)

- o General Sensation from mucus membrane of Posterior 1/3 of Tongue and part of External ear.

- o GSA

4. Nucleus Ambiguus: (Motor)

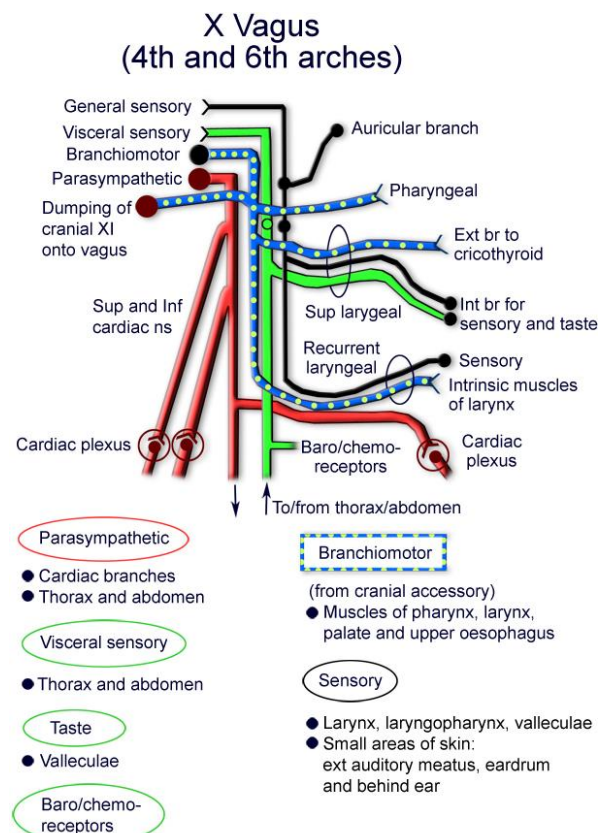
- o At medulla oblongata.
- o Innervates muscles of Pharynx and Larynx.
- o Branchial Efferent (SVE)
- o SVE is predominantly the Motor portion of pharyngeal plexus.

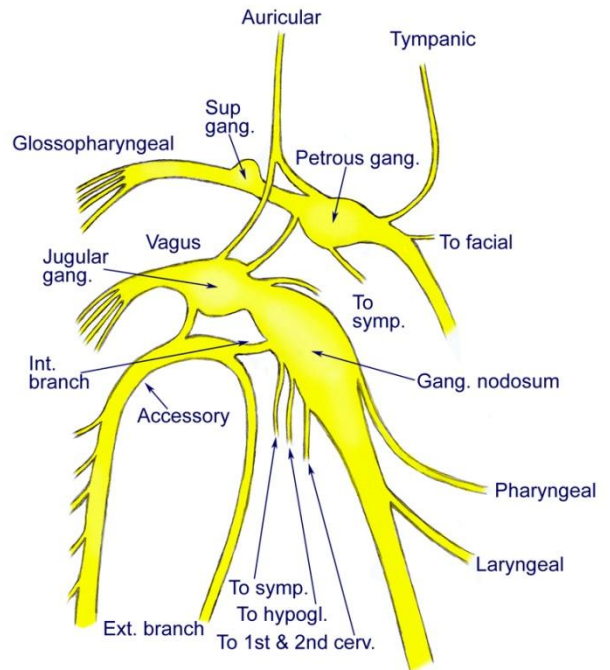
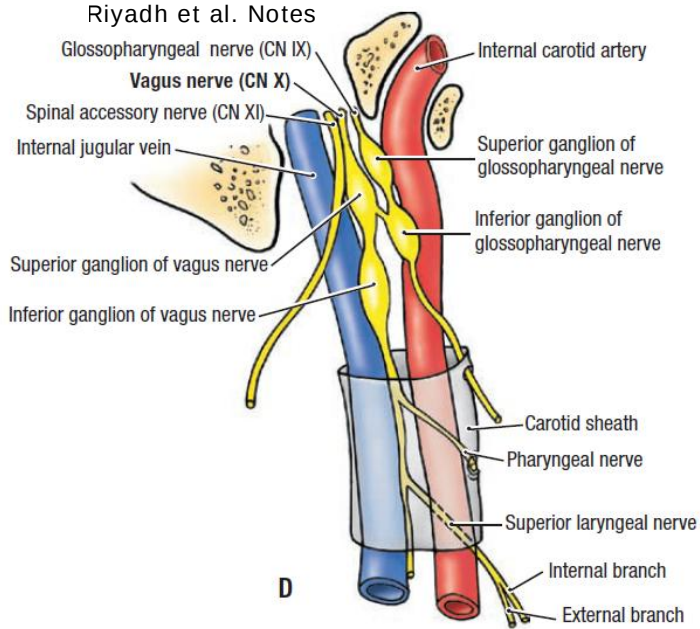


- Vagus Nerve has 2 Ganglionic swellings, which are the sensory ganglia of the nerve:

- o Superior (Jugular) Ganglion.
- o Inferior (Nodosum) Ganglion.

- Vagus nerve is joined by Cranial root of the Accessory nerve (CN-XI) just below the inferior ganglion.



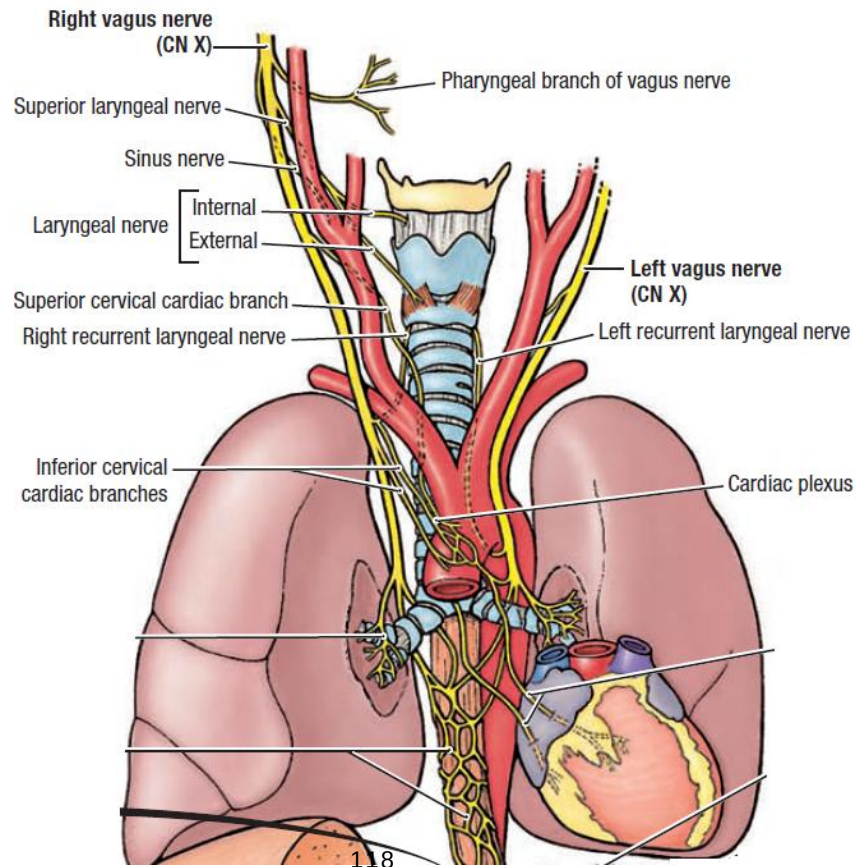


- **Right Vagus Nerve:**

- Crosses in front of 1st part of Subclavian Artery.
- Travels behind the innominate vessels.
- Reaches the thorax on the right side of the trachea.

- **Left Vagus Nerve:**

- Crosses in front of Left subclavian artery.
- Enter the thorax between the left common carotid and subclavian arteries.
- Descends on left side of the aortic arch.



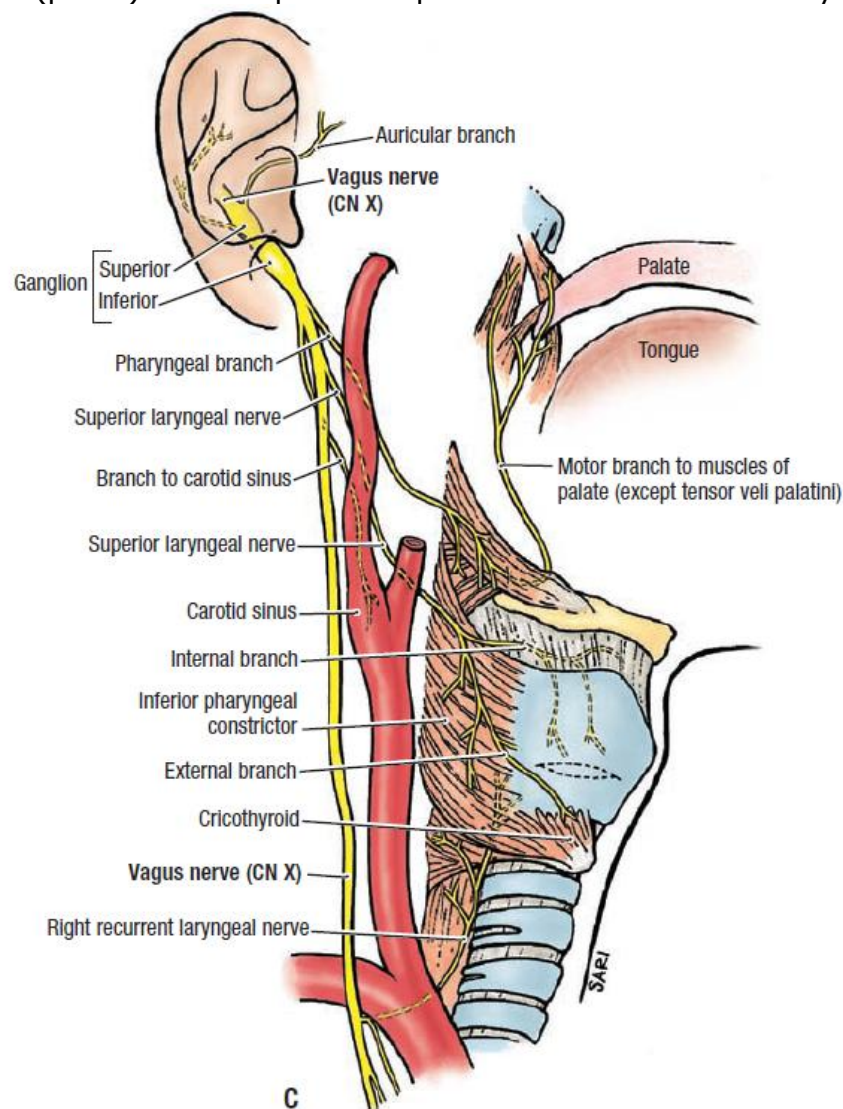
- **Branches of Vagus nerve in Jugular Foramen:**

1. **Meningeal branch:**

- Arises at Superior ganglion.
- Re-enters the cranium through the jugular foramen.
- Supply Dura of Posterior fossa.

2. **Auricular branch (Arnold's nerve):**

- Arises at Superior ganglion.
- Enters Mastoid canaliculus in lateral part of the jugular foramen.
- Exits again through Tympanomastoid suture of the temporal bone to reach the skin.
- Communicates with branches of Facial (CN-VII) and Glossopharyngeal (CN-IX) Nerves.
- Supplies sensations to Posterior aspect of the external ear (pinna) and the posterior part of the external auditory canal.



- **Branches of Vagus nerve in the Neck:**

1. Pharyngeal branches:

- Arise from Inferior Ganglion.
- Mixed sensory and motor fibers.
- Motor Fibers contributed by Accessory Nerve (CN-XI).
- Reach the middle constrictor muscle after crossing between the external and internal carotid arteries.
- Reach Pharyngeal plexus formed by Glossopharyngeal Nerve (CN-IX) and the Sympathetic chain.
- Supply Pharyngeal muscles and mucous membrane and palate **Except** Tensor palatini muscle.
- Vagal fibers from the pharyngeal plexus forms Inter-carotid plexus at the carotid bifurcation which mediates impulses set up by the chemoreceptors in the carotid body.

2. Superior laryngeal nerve

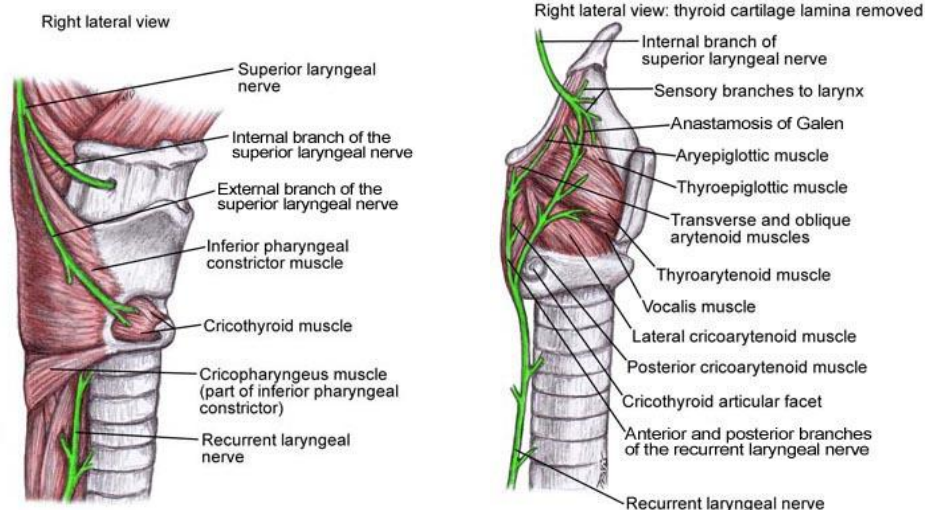
- Passes between External and Internal carotid arteries at the level of crossing of Hypoglossal Nerve (CN-XII).
- Divides into External and Internal branches at tip of the hyoid (2-3 cm superior to the superior pole of the thyroid).

1. Internal Laryngeal Nerve:

- Pierces Thyrohyoid membrane to enter the larynx.
- **Supplies General sensations to Most of mucosa ABOVE glottis:**
- Supplies mucosa of Laryngeal surface of Epiglottis, True and False vocal folds and Aryepiglottic fold, Arytenoid mucosa, Anterior wall of Hypopharynx, Upper esophageal sphincter, and part of the subglottis (Major part of Subglottis is innervated by ipsilateral recurrent nerve)

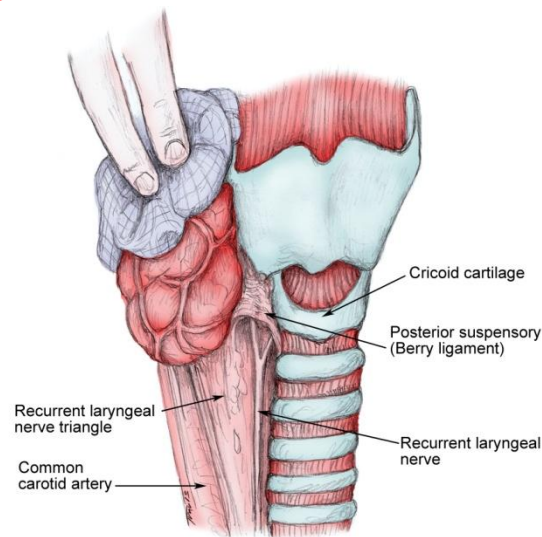
2. External Laryngeal Nerve:

- Passes inferiorly with Superior thyroid vessels to the inferior pharyngeal constrictor muscle.
- Supplies **Cricothyroid muscle and Inferior Pharyngeal constrictor.**
- Contributes innervations to the pharyngeal plexus.



3. Recurrent Laryngeal Nerve (RLN):

- Also known as Inferior laryngeal nerve.
- **Right (RLN)** branches from Right Vagus at root of neck (T1-T2)
- Loops around Right Subclavian artery.
- Travels inferior and posterior to Subclavian artery.
- Courses superiorly in Tracheoesophageal groove.
- Enter Larynx between Cricopharyngeus and Esophagus.
- **Left (RLN)** branches from Left Vagus in the Thorax.
- Loops around Aortic Arch distal to Ligamentum Arteriosus.
- Travels inferior and posterior to Aarch of Aorta.
- Ascend into the neck in between the trachea and esophagus.
- Main trunk RLN lies in a triangle bounded laterally by: Common carotid artery, IJV, and Vagus nerve and bounded medially by: Trachea and Esophagus.
- RLN passes under Posterior suspensory ligament of Berry before entering the larynx.
- Galen anastomosis (Ramus anastomoticus or Ansa of Galen) is a connection between RLN and Internal branch of SLN.
- **All Intrinsic laryngeal muscles are supplied by Ipsilateral RLN Except CricoThyroid muscle (by External branch of Superior Laryngeal Nerve).**
- **Interarytenoid muscle is the only one that receives a bilateral RLN supply.**
- **RLN gives Sensory and Secretomotor fibers Subglottis and Trachea.**



Nerve	Functional Components	Cells of Origin/Termination	Cranial Exit	Distribution and Functions
Vagus	Branchial motor	Nucleus ambiguus	Jugular foramen	Motor to constrictor muscles of pharynx, intrinsic muscles of larynx, muscles of palate (except tensor veli palatini), and striated muscle in superior two thirds of esophagus
	Visceral motor	Presynaptic: posterior (dorsal) nucleus of CN X; Postsynaptic: neurons in, on, or near viscera		Parasympathetic innervation to smooth muscle of trachea, bronchi, and digestive tract, cardiac muscle
	Visceral sensory	Nuclei of solitary tract, spinal trigeminal nucleus/ inferior ganglion		Visceral sensation from base of tongue, pharynx, larynx, trachea, bronchi, heart, esophagus, stomach, and intestine
	Special sensory	Nuclei of solitary tract/inferior ganglion		Taste from epiglottis and palate
	General sensory	Spinal trigeminal nucleus/superior ganglion		Sensation from auricle, external acoustic meatus, and dura mater of posterior cranial fossa

CRANIAL NERVE X: VAGUS NERVE

Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSA	Afferent fibers begin in the various receptors on a small part of the skin of the external ear	Pain and temperature fibers terminate in the spinal nucleus of V	The GSA fibers are responsible for providing sensory innervation to a very small portion of the external ear The GSA fibers of the glossopharyngeal n. utilize the trigeminothalamic lemniscus to carry their sensory impulses to consciousness	The nerve cell bodies for the primary fibers are located in the superior ganglion of X
SVA	Afferent fibers begin in the taste receptors of the epiglottic region and are scattered on the palate	Primary afferent fibers travel in the tractus solitarius and terminate in the nucleus solitarius	The SVA fibers are responsible for carrying the taste fibers from the epiglottic region and are scattered on the palate	The nerve cell bodies for the primary fibers are located in the inferior ganglion of X
GVA	Afferent fibers begin in the various receptors of the mucous membranes of the laryngopharynx, larynx, thorax, and abdomen	Primary afferent fibers travel in the tractus solitarius and terminate in the nucleus solitarius	The GVA fibers utilize the same pathway as for the SVA fibers	The nerve cell bodies for the primary fibers are located in the inferior ganglion of X
GVE	Preganglionic parasympathetic fibers begin in the dorsal motor nucleus of the vagus n.	Postganglionic parasympathetic fibers innervate thoracic and abdominal viscera	The GVE fibers are responsible for providing the parasympathetic innervation to the thoracic and abdominal viscera	The GVE fibers utilize: • Intramural ganglia
SVE	Begins in the nucleus ambiguus	Innervates the muscles of the pharynx (via the pharyngeal plexus) and the larynx	The SVE fibers are responsible for innervating the muscles of the 4th pharyngeal arch	The SVE fibers are the motor component to the pharyngeal plexus (muscles of pharynx) Lesions of the vagus paralyze the muscles of the larynx on the affected side

- **CRANIAL NERVE XI: ACCESSORY:**

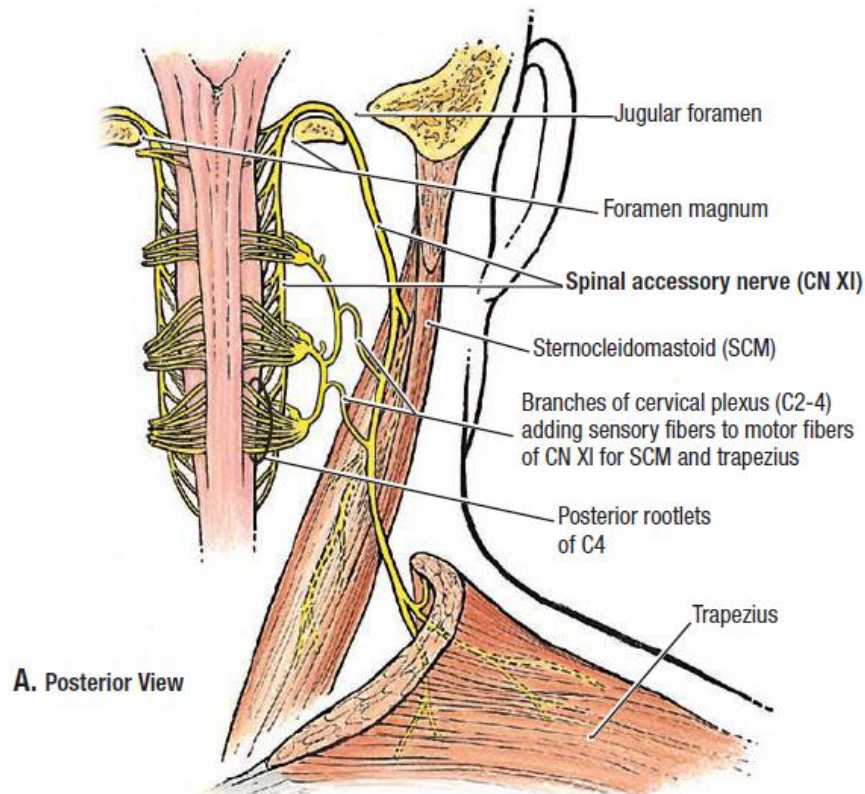
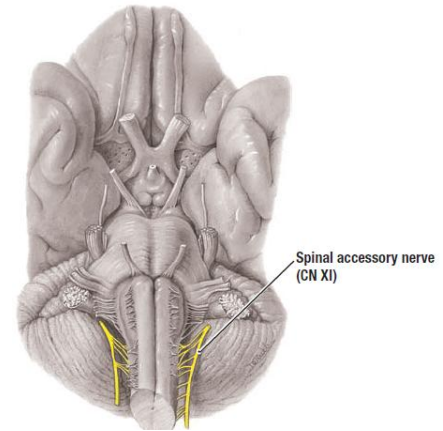
- Pure Motor nerve.
- Consist of Cranial Root and Spinal Root.

- **Cranial Root:**

- Arises from **Nucleus Ambiguus**.
- Emerge as four or five rootlets from side of **Medulla oblongata**, below Vagus Nerve.
- Runs to **Jugular foramen**, where it is connected with the Superior ganglion of the vagus.
- Form Nodosum ganglion distributed principally to Pharyngeal and Superior laryngeal nerve.
- **Considered part of Vagus Nerve (CN-X)**

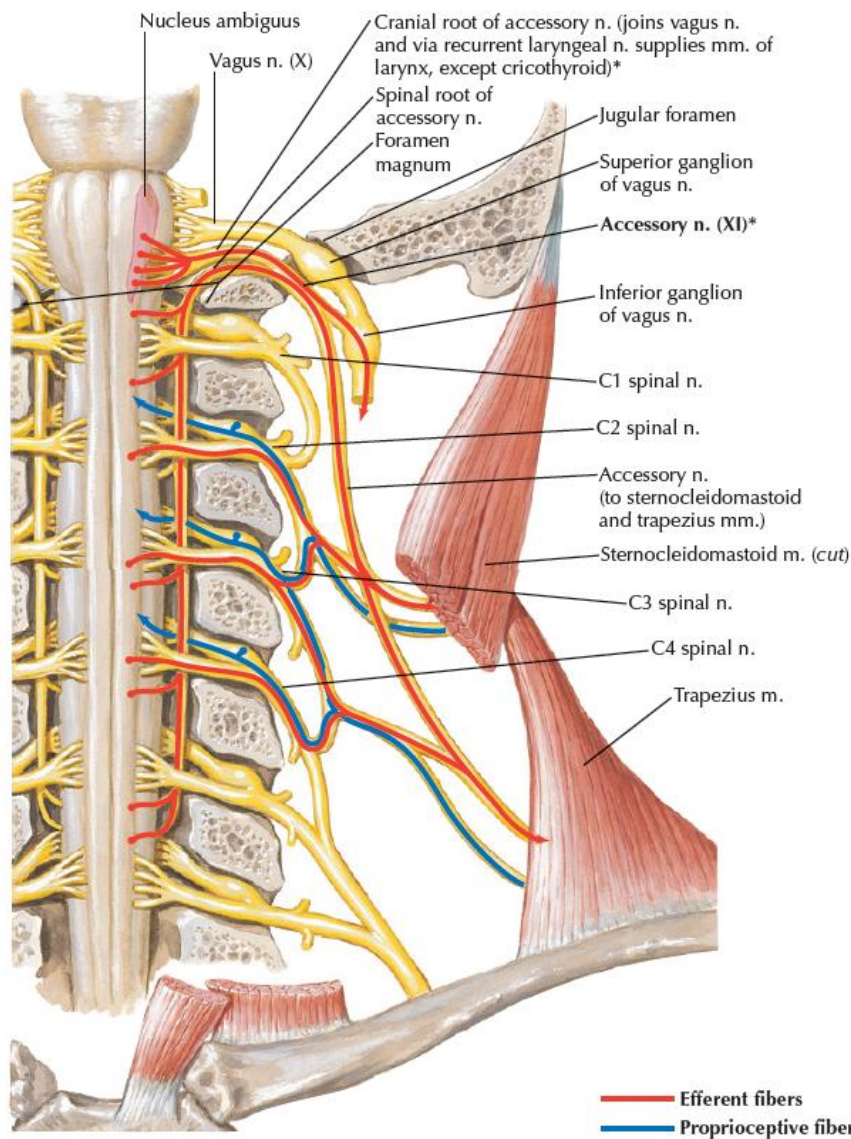
- **Spinal Root:**

- Arises from Nerve cells in the Anterior Gray horn of C1-C5.
- Ascends alongside the Spinal Cord and enters the skull through **Foramen Magnum**.
- Then Exits the skull by passing through **Jugular Foramen**.
- Descends obliquely to Upper part of SCM and pierces it.
- Courses obliquely across Posterior triangle of the neck.
- Ends in the deep surface of the Trapezius.
- **Motor Supply to SCM and Trapezius.**



CRANIAL NERVE XI: SPINAL ACCESSORY NERVE

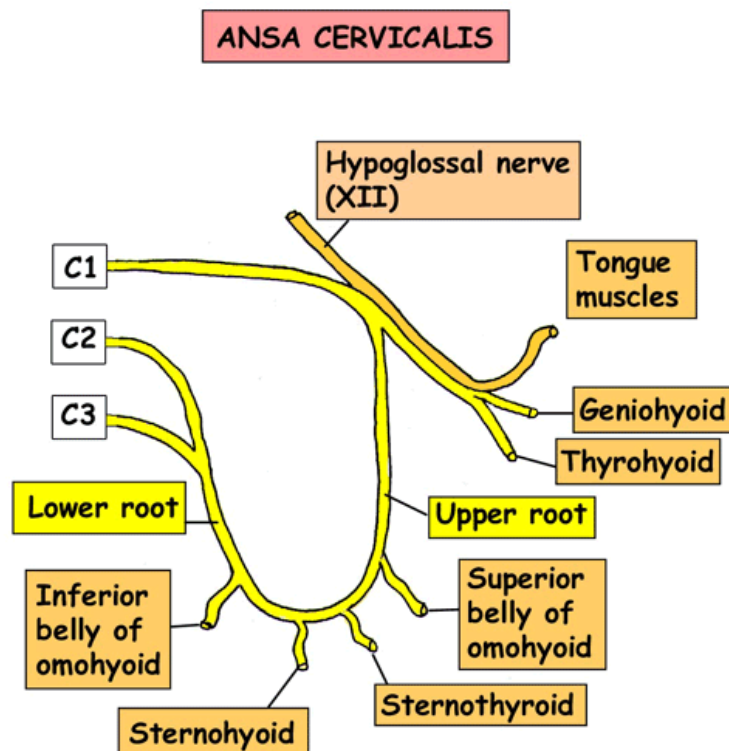
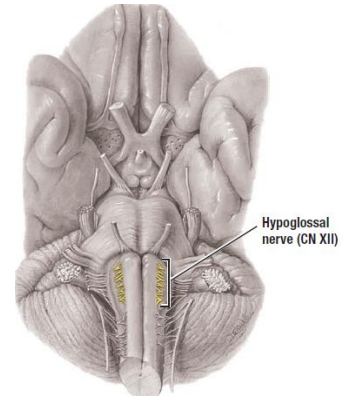
Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
SVE	Cranial part: Begins in the nucleus ambiguus Spinal part: Begins in the upper cervical levels of the spinal cord	Cranial part: Innervates the muscles of the pharynx (via the pharyngeal plexus) Spinal part: Innervates the trapezius and sternocleidomastoid mm.	These SVE fibers of the cranial part travel with the vagus n. and arise from the same nucleus (nucleus ambiguus) and often are considered to be the same	The cranial and spinal parts separate so the cranial part can join the pharyngeal plexus and the spinal part can innervate the sternocleidomastoid m. and pass through the posterior triangle until reaching the trapezius m.

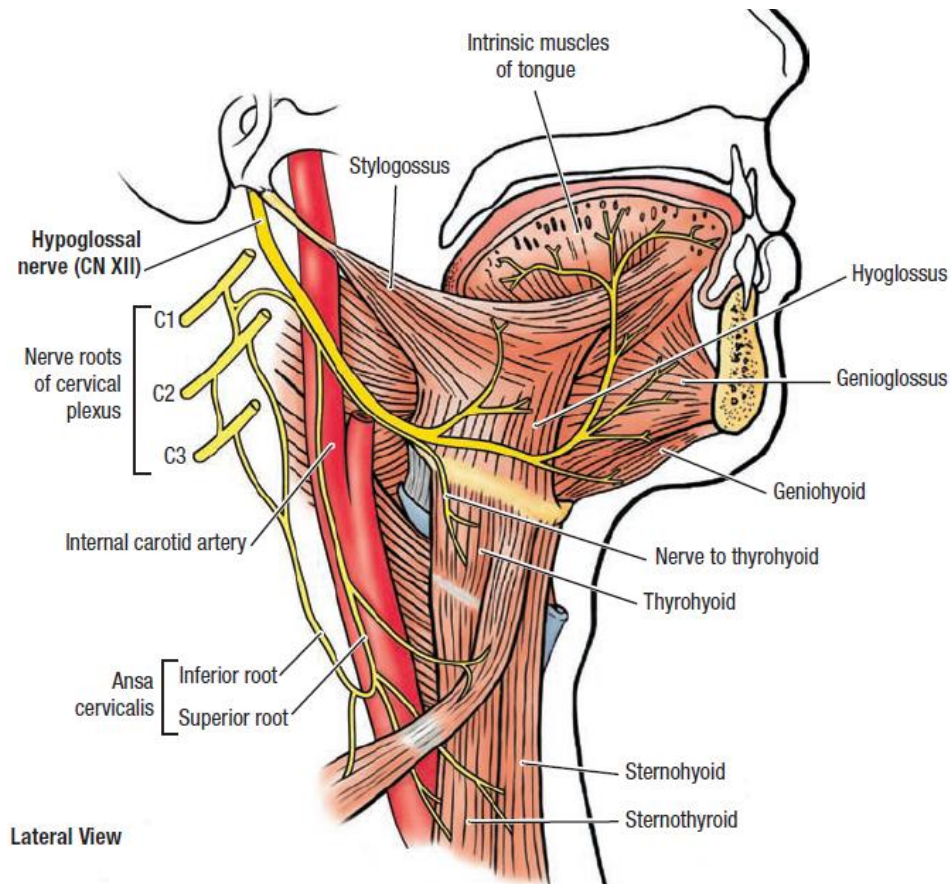


Recent evidence suggests that the accessory nerve lacks a cranial root and has no connection to the vagus nerve. Confirmation of this finding awaits further investigation.

- **CRANIAL NERVE XII: HYPOGLOSSAL:**

- Pure Motor Nerve.
- Arises from **Hypoglossal nucleus** in Medulla oblongata.
- Emerges in Antero-lateral sulcus between Pyramid and Olive.
- Crosses the Posterior Cranial Fossa.
- Leaves Skull through **Hypoglossal Canal**.
- Descends almost vertically to Neck.
- Passes forward between IJV & ICA (with CN-IX).
- Becomes superficial below Digastricus.
- Loops around Occipital artery.
- Crosses External carotid and Lingual arteries below the tendon of the Digastricus.
- Passes beneath Tendon of Digastric, Stylohyoid and Mylohyoid.
- Lying between Mylohyoid and Hyoglossus
- Communicates at anterior border of Hyoglossus with Lingual nerve.
- Continued forward to tip of the tongue
- **Innervates All muscles of Tongue Except Palatoglossus.**
- Controls Shape and movement of the Tongue.





CRANIAL NERVE XII: HYPOGLOSSAL NERVE

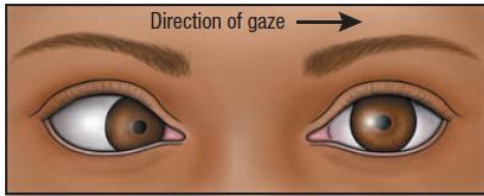
Functional Column	Origin of Fibers	Termination of Fibers	Summary	Comment
GSE	Begins in the hypoglossal nucleus	Innervates the genioglossus, hyoglossus, and styloglossus mm. and the intrinsic mm. of the tongue	The GSE fibers are responsible for innervating the major portion of the tongue musculature	Lesions of the hypoglossal n. cause the tongue to deviate to the side of the lesion on protrusion

SUMMARY OF CRANIAL NERVE LESIONS



Right eye: Downward and outward gaze, dilated pupil, eyelid manually elevated due to ptosis

A. Right oculomotor (CN III) nerve palsy

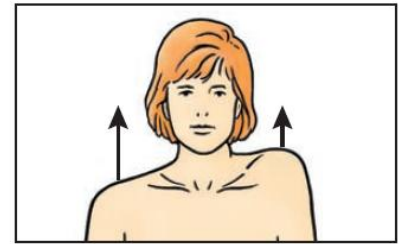


Right Left eye: Does not abduct

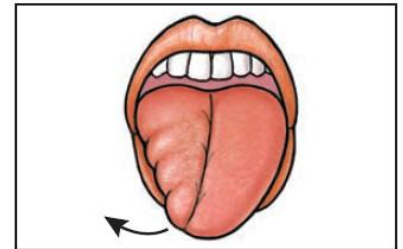
B. Left abducent (CN VI) nerve palsy



C. Right facial (CN VII) palsy (Bell palsy)



D. Right CN XI lesion



E. Right CN XII lesion

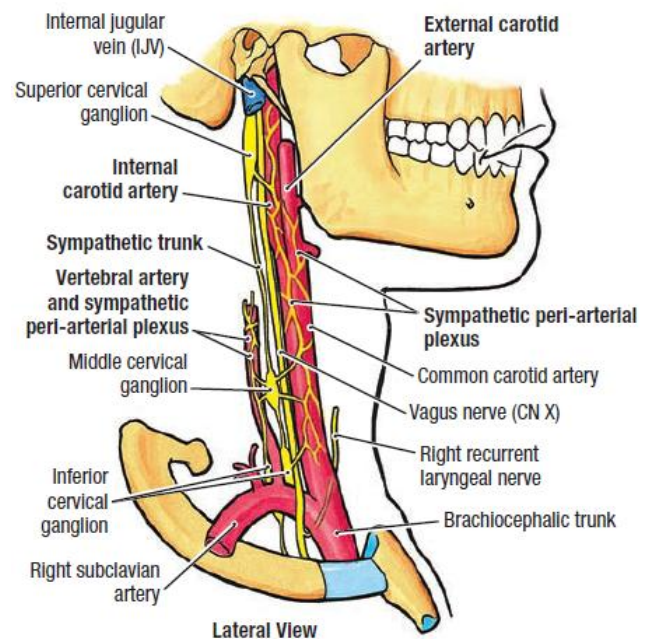
Nerve	Lesion Type and/or Site	Abnormal Findings
CN I	Fracture of cribriform plate	Anosmia (loss of smell); cerebrospinal fluid (CSF) rhinorrhea (leakage of CSF through nose)
CN II	Direct trauma to orbit or eyeball; fracture involving optic canal	Loss of pupillary constriction
	Pressure on optic pathway; laceration or intracerebral clot in temporal, parietal, or occipital lobes of brain	Visual field defects
	Increased CSF pressure	Swelling of optic disc (papilledema)
CN III	Pressure from herniating uncus on nerve; fracture involving cavernous sinus; aneurysms	Dilated pupil, ptosis, eye rotates inferiorly and laterally (down and out), pupillary reflex on the side of the lesion will be lost (A)
CN IV	Stretching of nerve during its course around brainstem; fracture of orbit	Inability to rotate adducted eye inferiorly
CN V	Injury to terminal branches (particularly CN V ₂) in roof of maxillary sinus; pathologic processes (tumors, aneurysms, infections) affecting trigeminal nerve	Loss of pain and touch sensations/paresthesia on face; loss of corneal reflex (blinking when cornea touched); paralysis of muscles of mastication; deviation of mandible to side of lesion when mouth is opened
CN VI	Base of brain or fracture involving cavernous sinus or orbit	Inability to rotate eye laterally; diplopia on lateral gaze (B)
CN VII	Laceration or contusion in parotid region	Paralysis of facial muscles; eye remains open; angle of mouth droops; forehead does not wrinkle (C)
	Fracture of temporal bone	As above, plus associated involvement of cochlear nerve and chorda tympani; dry cornea and loss of taste on anterior two thirds of tongue
	Intracranial hematoma ("stroke")	Weakness (paralysis) of lower facial muscles contralateral to the lesion, upper facial muscles are not affected because they are bilaterally innervated
CN VIII	Tumor of nerve	Progressive unilateral hearing loss; tinnitus (noises in ear); vertigo (loss of balance)
CN IX ^a	Brainstem lesion or deep laceration of neck	Loss of taste on posterior third of tongue; loss of sensation on affected side of soft palate; loss of gag reflex on affected side
CN X	Brainstem lesion or deep laceration of neck	Sagging of soft palate; deviation of uvula to unaffected side; hoarseness owing to paralysis of vocal fold; difficulty in swallowing and speaking
CN XI	Laceration of neck	Paralysis of sternocleidomastoid and superior fibers of trapezius; drooping of shoulder (D)
CN XII	Neck laceration; basal skull fractures	Protruded tongue deviates toward affected side; moderate dysarthria, disturbance of articulation (E)

- **Sympathetic Chain Ganglia:**

- Ganglia of the sympathetic nervous system.
- Contain approximately 20,000–30,000 nerve cell bodies.
- Bilaterally symmetric sympathetic chain ganglia.
- Located just ventral and lateral to the spinal cord.
- Extends from Upper neck down to the coccyx
- **Preganglionic nerves** from the spinal cord synapse end at one of the chain ganglia.
- **Postganglionic fiber** extends to a visceral organ.
- 21 or 23 pairs of Sympathetic ganglia
 - 3 Cervical
 - 12 Thoracic
 - 5 Lumbar
 - 5 Sacral
 - 1 Coccygeal.

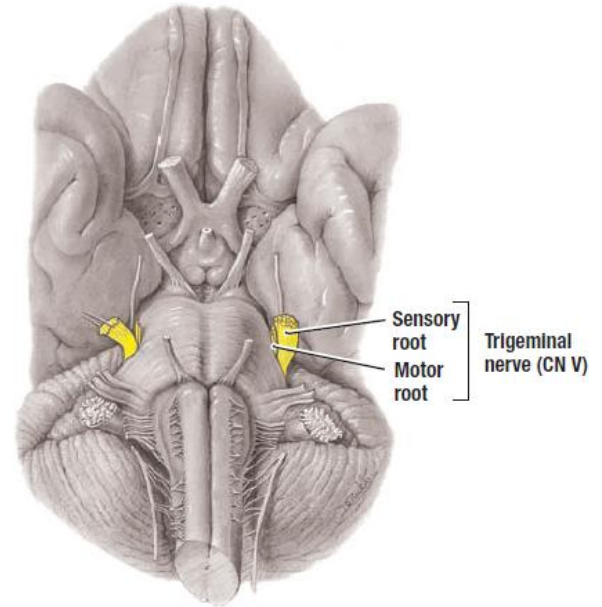
- **Cervical Sympathetic Chain Ganglia:**

- 3 Paravertebral ganglia located posterior to carotid sheath:
- 1. Superior Cervical Ganglion:**
 - Largest.
 - Adjacent to **C2-C3**
 - Target: (heart, head, neck)
- 2. Middle Cervical Ganglion:**
 - Smallest.
 - Adjacent to **C6**
 - Target: heart, neck
- 3. inferior Cervical Ganglion:**
 - Adjacent to C7.
 - May be fused with 1st Thoracic ganglion to form **Stellate ganglion.**
 - Target: heart, lower neck, arm, posterior cranial arteries



- **Trigeminal Nerve (CN-V)**

- Largest and most complex Cranial nerve.
- Origin:
 - o **Pons** at its junction with the Middle Cerebellar Peduncle.
- Mixed nerve supply:
 - o Large sensory root.
 - o Small Motor root.
- Passes from Posterior Cranial Fossa to reach Apex of Petrous part of Temporal bone in the Middle Cranial Fossa.
- Large Sensory root:
 - o Expands to form **Trigeminal Ganglion** (Gasserian or Semilunar ganglion).
 - o Sensory innervation to all areas of Face Except Angle of mandible by Great Auricular Nerve (C2-C3).
- Small Motor root:
 - o Situated below the Trigeminal Ganglion and completely separate from it.
- Main branches of Trigeminal Nerve:
 1. Ophthalmic Nerve (V1)
 2. Maxillary Nerve (V2)
 3. Mandibular Nerve (V3)



- **Trigeminal Nuclei:**

1. Main Sensory Nucleus:

- o Located in the **Pons**.
- o Ophthalmic division is in the lower part of the nucleus.
- o Mandibular division is in the upper part of the nucleus.
- o Receives Afferent general somatic sensations from the main 3 branches of the trigeminal Nerve.
- o Axons cross to the other side.
- o Ascends to Thalamic nuclei.
- o Relay in the Postcentral cerebral cortex.
- o Descending sensory fibers from Trigeminal (Gasserian) Ganglion course through the pons and medulla in the spinal tract of CN V.
- o End in the nuclei of this tract

2. Spinal Nucleus:

- o Located in the **Medulla**.
- o Receives information about deep/crude touch, pain, and temperature from the ipsilateral face.
- o Also receives Pain sensation from Facial (CN-VII), Glossopharyngeal (CN-IX) and Vagus Nerve (CN-X).

3. Motor Nucleus:

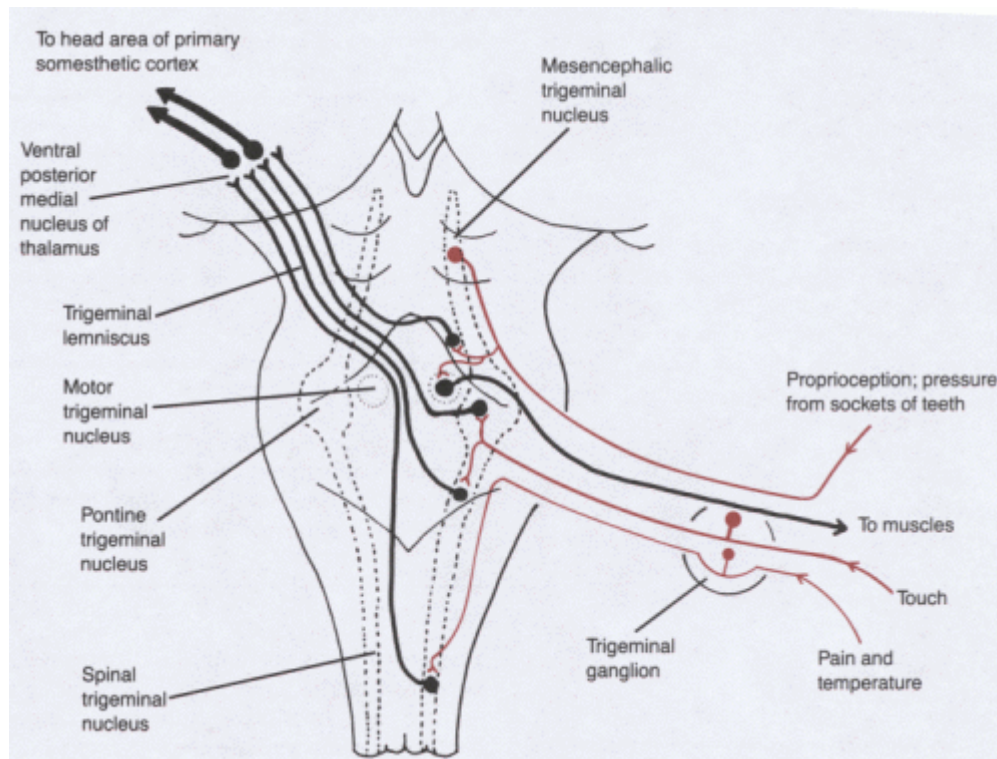
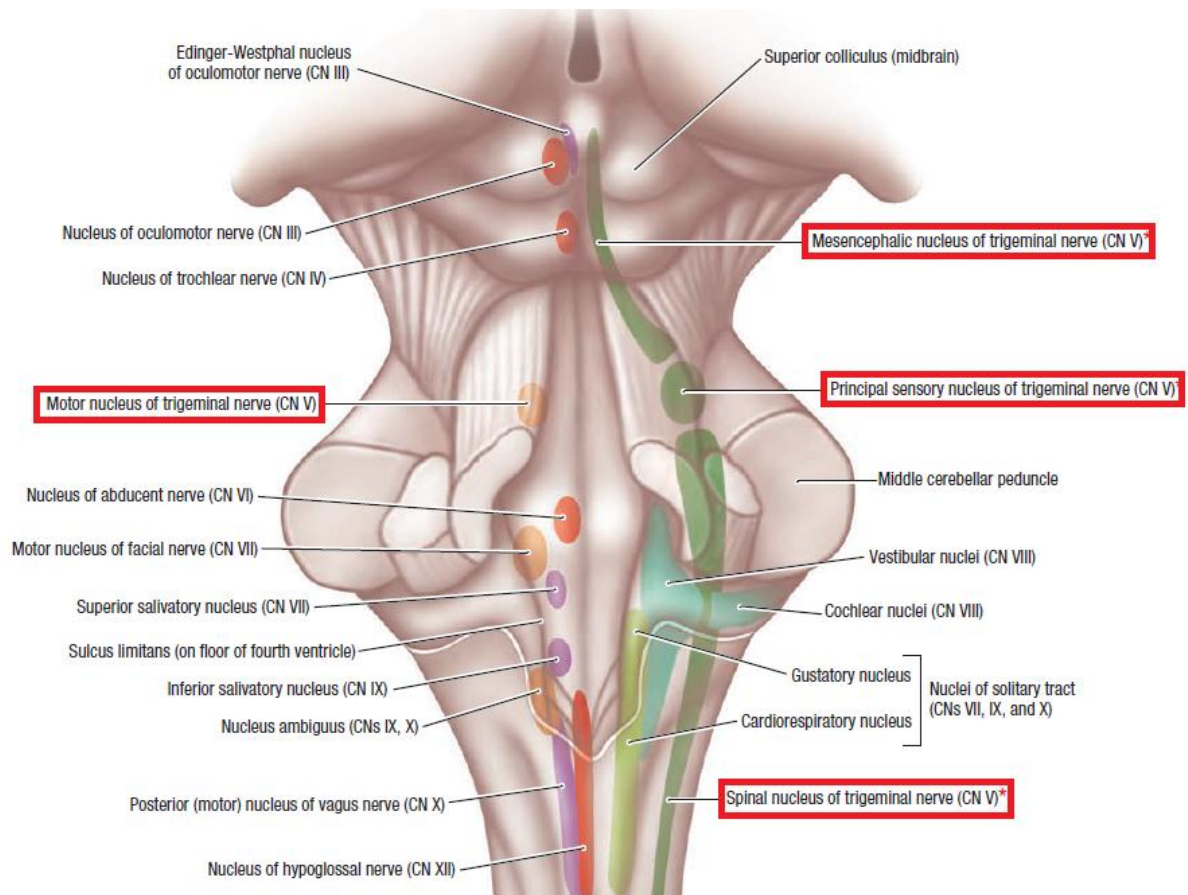
- Located in the **Pons**, ventromedial to the sensory nucleus.
- Lies near the lateral angle of the fourth ventricle in the rostral part of the pons.
- Receives cortical fibers for voluntary control of the muscles of mastication.
- Also receives input from the mesencephalic and sensory nuclei.
- Motor fibers are distributed in Mandibular division (V3).

4. Mesencephalic Nucleus:

- Located in the **Midbrain**.
- Receives proprioceptive fibers from all muscles of mastication and Extra-ocular muscles.
- Has connections to the motor nucleus of CN V.

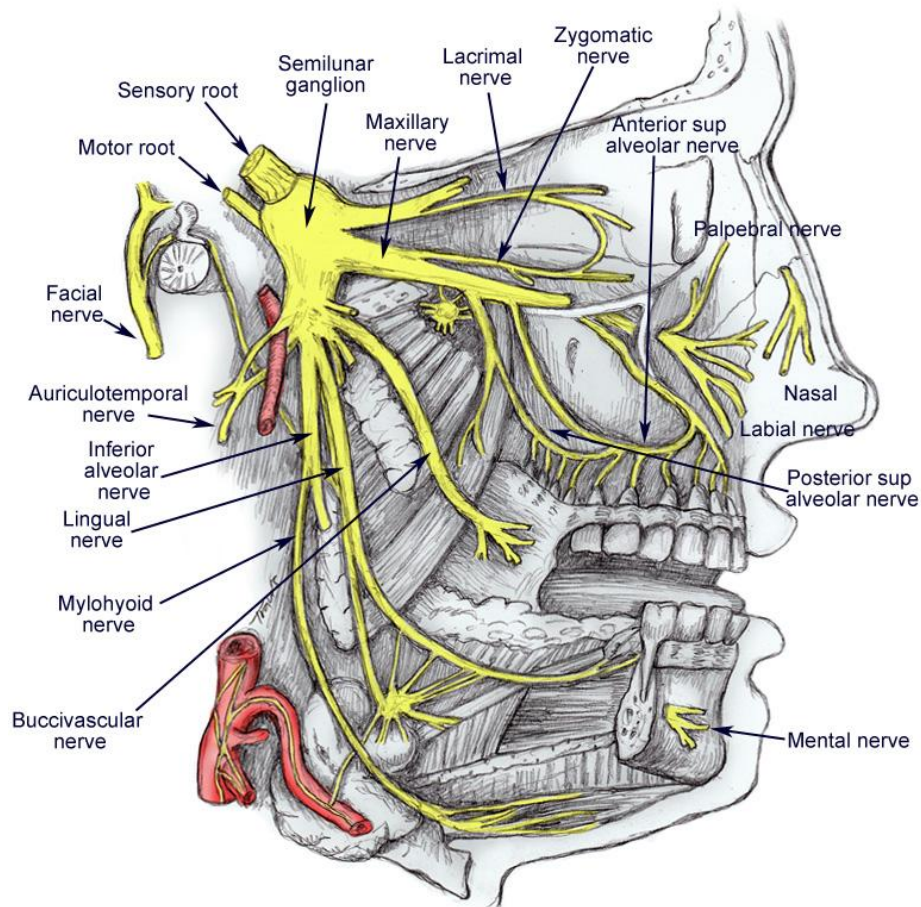
Components	Function	Central connection	Cell bodies	Peripheral distribution
Afferent General Somatic	General sensibility	Sensory nucleus V	Gasserian ganglion	Sensory branches of the ophthalmic, maxillary, and mandibular nerves to skin, mucous membranes of the face and head
Efferent Special Visceral	Mastication	Motor nucleus V	Motor nucleus V	Branches to Temporalis, Masseter, Pterygoids, Mylohyoid, Tensor tympani, and Tensor Palati
Afferent Proprioceptive	Muscular sensibility	Mesencephalic nucleus V	Mesencephalic nucleus V	Sensory endings in muscles of mastication

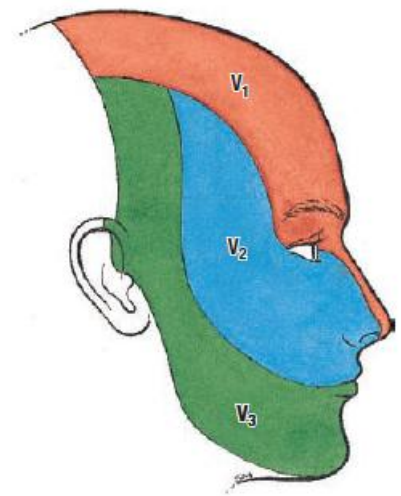
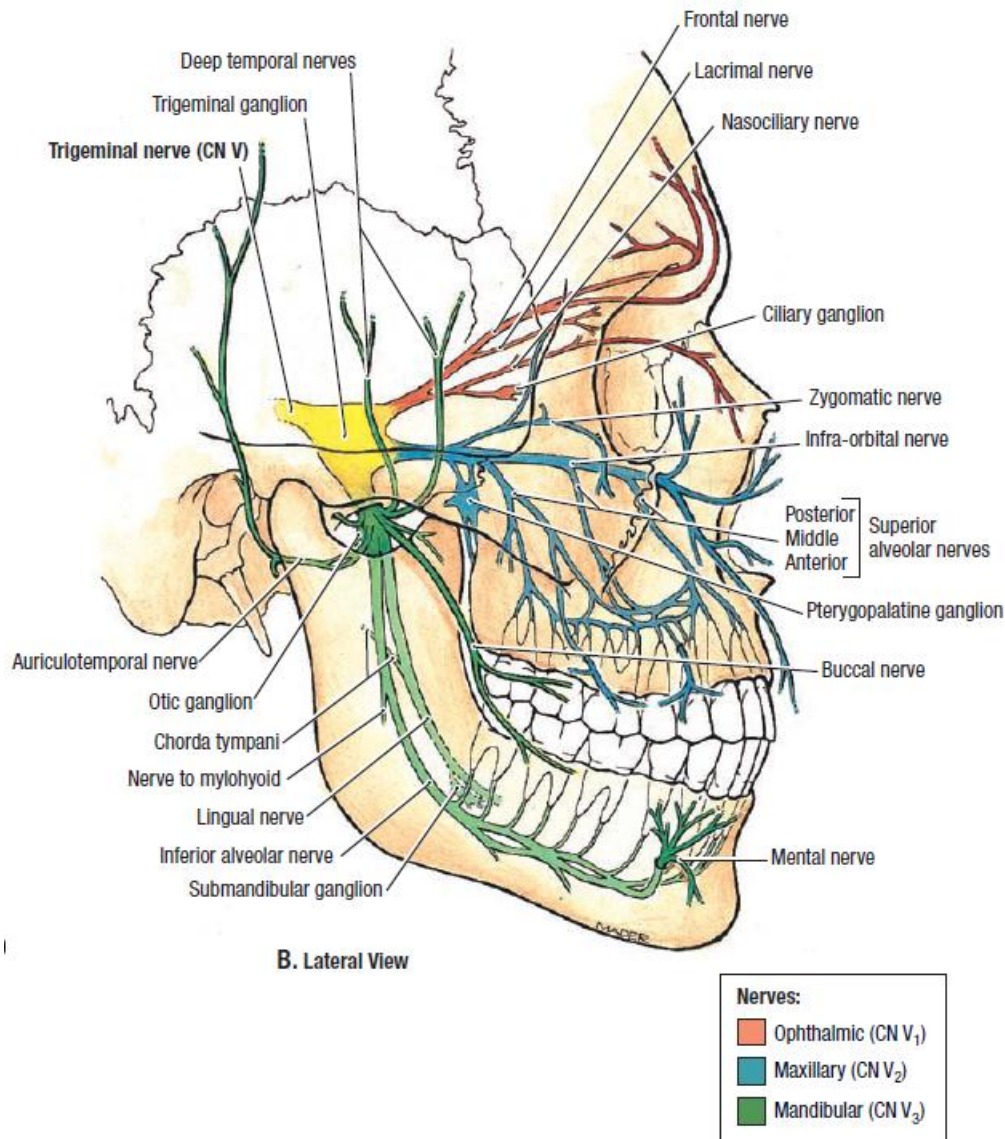
Type	Function	Pathway
Branchial Motor	Motor to muscles of mastication	CN V innervates Muscles of Mastication, Mylohyoid, Tensor tympani, Tensor veli palate, Anterior belly of digastric
General sensory	Sensory from surface of head and neck, sinuses, meninges and TM	Gasserian ganglion receives the Ophthalmic, Maxillary and Mandibular divisions and Sympathetic fibers from the Carotid plexus and sends branches to the dura. The four accessory ganglia are anatomically but not functionally associated with CN V



- **Trigeminal Ganglion:**

- Semilunar (Gasserian) Ganglion.
- Great sensory ganglion of CN-V.
- Lies in a depression on the petrous apex, within a dural fold called the **Meckel cave**.
- Contains the sensory cell bodies of the 3 branches of the trigeminal nerve (Ophthalmic, Maxillary and Mandibular).
- Ophthalmic and Maxillary nerves are Purely Sensory.
- Mandibular nerve has Mixed sensory and motor functions.
- Sensory roots of the 3 branches of CN V are received Anteriorly, then pass from the posterior aspect of the ganglion to the pons.
- Motor root passes under the ganglion to join the sensory division of the mandibular nerve and exits the skull through Foramen Ovale.
- The carotid plexus contributes sympathetic fibers to the gasserian ganglion.



**TABLE 9.5 TRIGEMINAL NERVE (CN V)**

Nerve	Functional Components	Cells of Origin/Termination	Cranial Exit	Distribution and Functions
Ophthalmic division (CN V ₁)	Somatic (general sensory)	Trigeminal ganglion/spinal, principal and mesencephalic nucleus of CN V	Superior orbital fissure	Sensation from cornea, skin of forehead, scalp, eyelids, nose, and mucosa of nasal cavity and paranasal sinuses
Maxillary division (CN V ₂)			Foramen rotundum	Sensation from skin of face over maxilla including upper lip, maxillary teeth, mucosa of nose, maxillary sinuses, and palate
Mandibular division (CN V ₃)			Foramen ovale	Sensation from the skin over mandible, including lower lip and side of head, mandibular teeth, temporomandibular joint, and mucosa of mouth and anterior two thirds of tongue
	Somatic (branchial) motor	Motor nucleus of CN V		Motor to muscles of mastication, mylohyoid, anterior belly of digastric, tensor veli palatini, and tensor tympani

- **Branches of Trigeminal Nerve:**

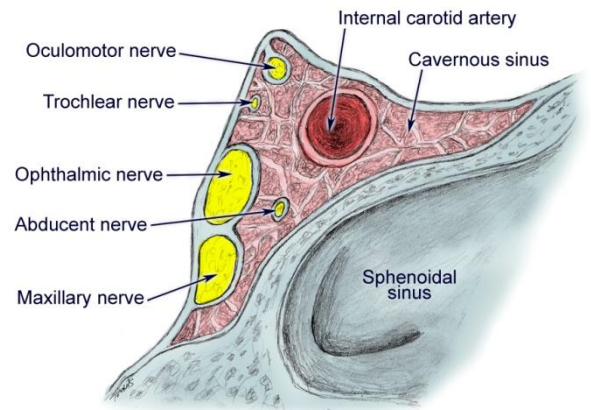
1. Ophthalmic Nerve (V1):
 - Superior Orbital Fissure.
 - Pure Sensory.
2. Maxillary Nerve (V2):
 - Foramen Rotundum
 - Pure Sensory.
3. Mandibular Nerve (V3):
 - Foramen Ovale.
 - Mixed Sensory and Motor

- **Ophthalmic Nerve (V1):**

- First branch of Trigeminal nerve (CN-V).
- Arises from the convex surface of the gasserian ganglion.
- Passes forward along the lateral wall of Cavernous sinus in Middle Cranial Fossa under Trochlear Nerve (CN-IV) and above Maxillary Nerve (V2).
- Exits the Skull and enters the Orbit through **Superior Orbital Fissure.**
- **Pure sensory nerve.**

- Carries Sensory information from:

- Scalp
- Forehead
- Upper eyelid
- Conjunctiva
- Cornea
- Nose (including the tip of the nose, EXCEPT Alae nasi).
- Nasal mucosa
- Frontal sinuses
- Parts of the meninges (Dura and blood vessels).



- Receives sympathetic filaments from:

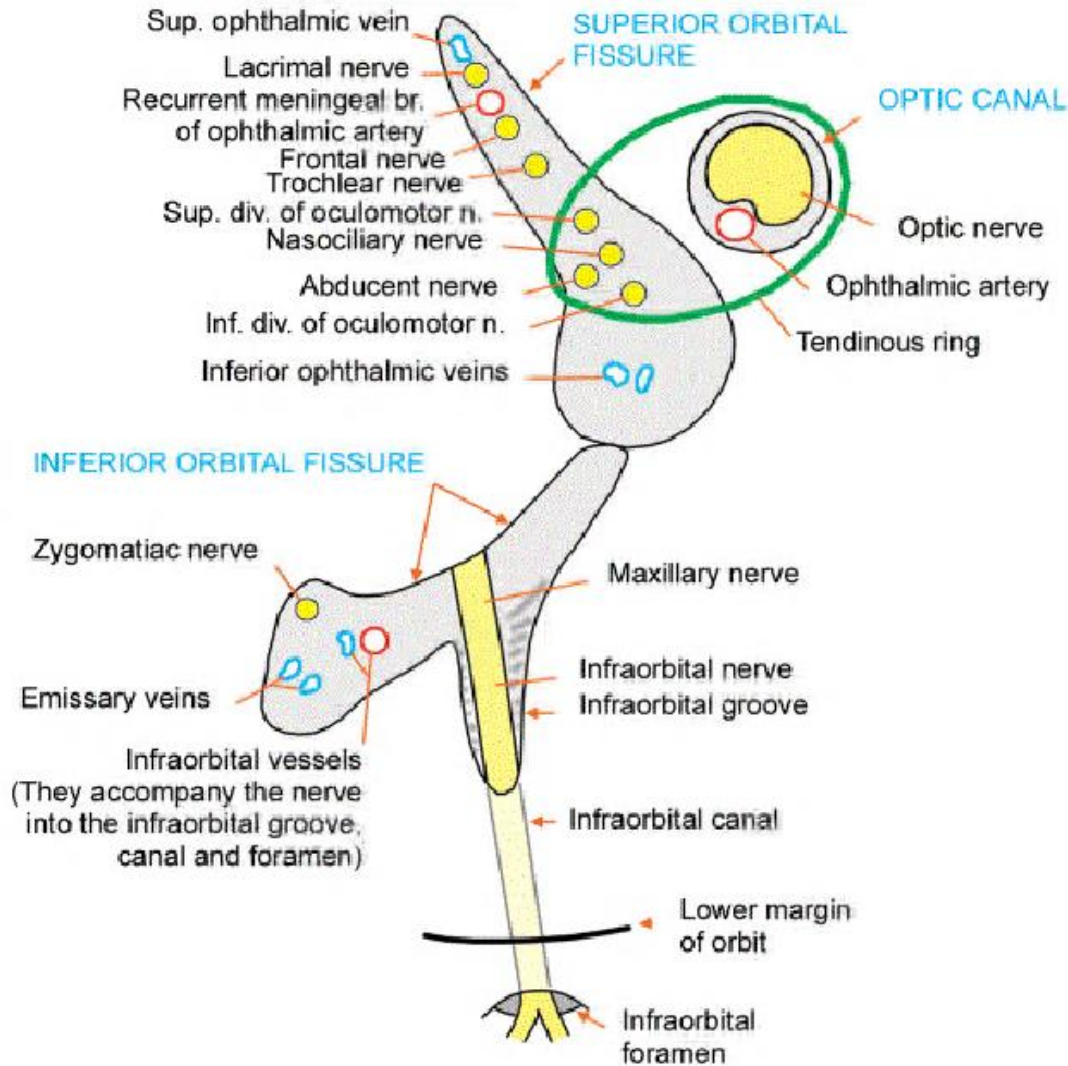
- Cavernous sinus
- Communicating branches from Oculomotor Nerve (CN-III) and Trochlear Nerve (CN-IV).

- **Ophthalmic Nerve Branches:**

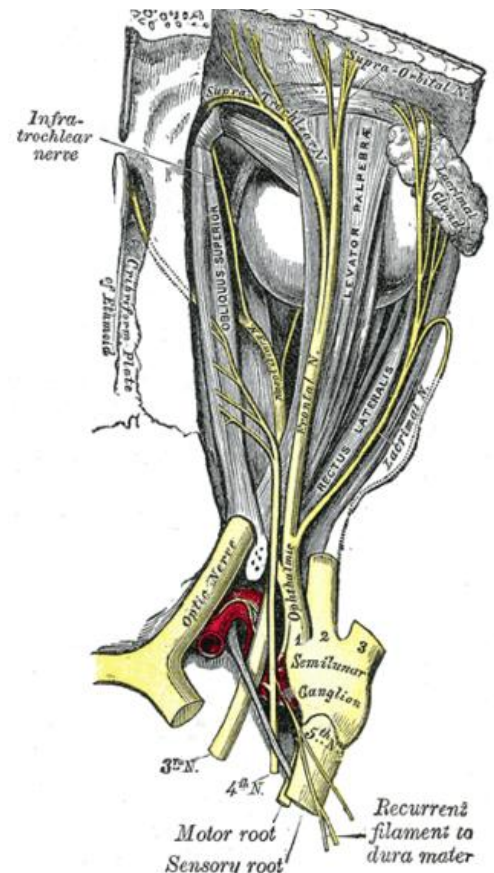
- One branch before it exits the skull:
 1. Dural branch.

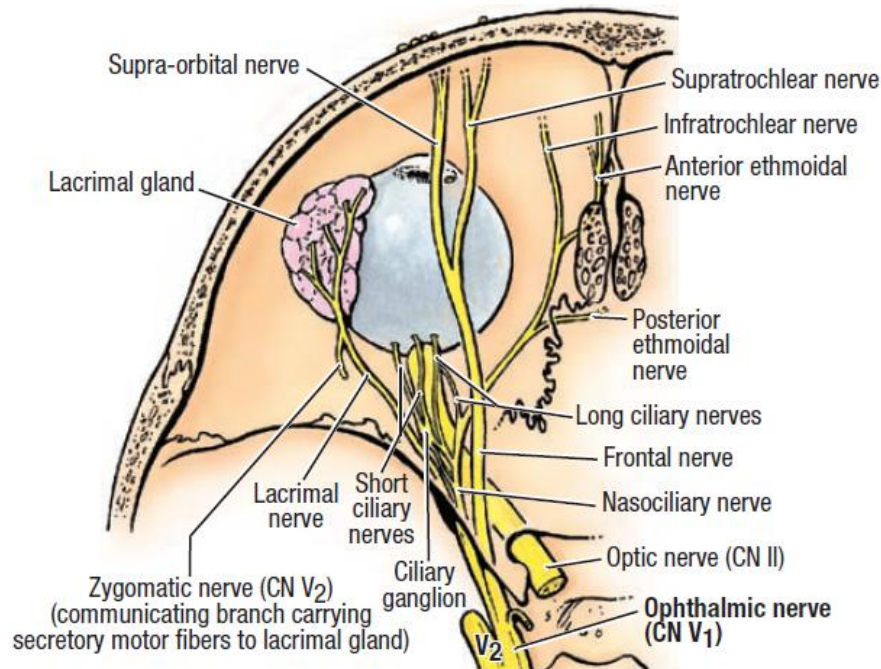
- Three branches after it enters the orbit through Superior Orbital Fissure.:

1. Frontal Nerve.
2. Lacrimal Nerve.
3. Nasociliary Nerve.

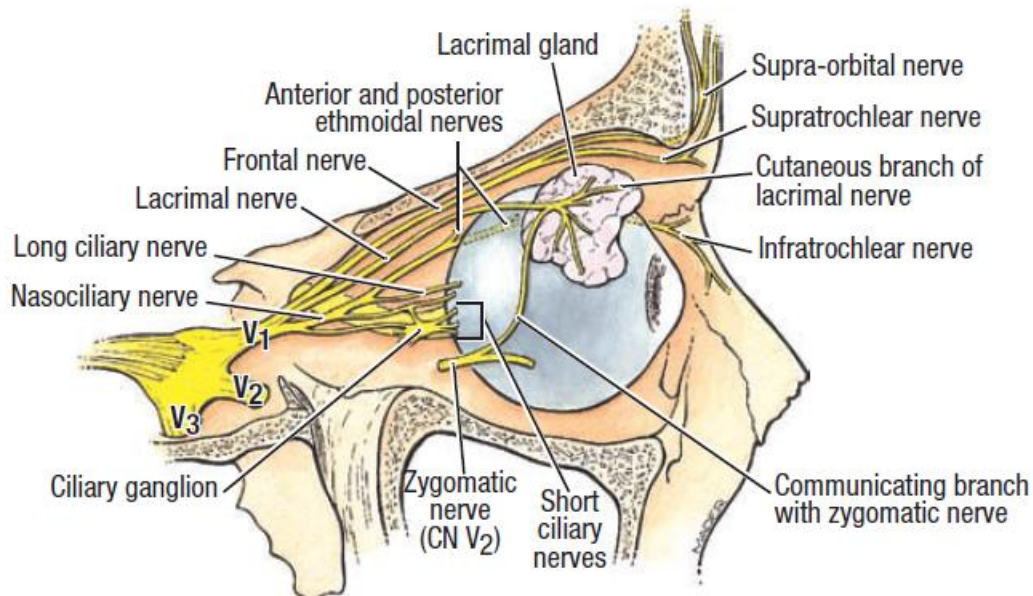


- **Frontal Nerve:**
- Largest branch of Ophthalmic nerve (V1).
- Passes in the lateral part of Superior Orbital Fissure:
 - o Below Lacrimal nerve and above Trochlear nerve (CN-IV).
- **Divides in the orbit into:**
- 1. **Supraorbital Nerve:**
 - o Terminal branch of Frontal nerve.
 - o Exits the skull through **Supraorbital Foramen.**
 - o Supplies Upper Lid and Scalp.
- 2. **Supratrochlear Nerve:**
 - o Exits the medial orbit.
 - o Supplies Conjunctiva and skin of upper lid, Lower and medial parts of the forehead and frontal sinus mucosa.





- **Lacrimal Nerve:**
- Second branch of Ophthalmic nerve (V1).
- Arises in the narrow, lateral part of the superior orbital fissure.
- Passes along upper border of lateral rectus, with Lacrimal artery.
- Enters Lacrimal gland.
- Pierces the orbital septum, and ends in the skin of the upper eyelid.
- Supplies:
 - Lacrimal gland
 - Conjunctiva
 - Upper lid.
- Receives a communication from Zygomatic branch of Maxillary Nerve (V2):
 - Represents Postganglionic Parasympathetic secretory fibers from Sphenopalatine Ganglion to Lacrimal gland.
 - The preganglionic fibers reach the ganglion via the Greater Petrosal and Vidian nerves from Facial Nerve (CN-VII).



- **Nasociliary Nerve:**

- Third branch of Ophthalmic nerve (V₁).

- Branches:

1. **Anterior Ethmoid Nerve:**

- Passes to Anterior Ethmoid Foramen lateral to the crista galli.
- Supply Anterior Ethmoid and Frontal sinuses.
- Supply Anterior part of the septum and lateral nasal wall.

2. **External Nasal Nerve:**

- Supply skin of the nasal tip.

3. **Posterior Ethmoid Nerve:**

- Supply Posterior Ethmoid and Sphenoid sinuses.

4. **Infratrochlear Nerve:**

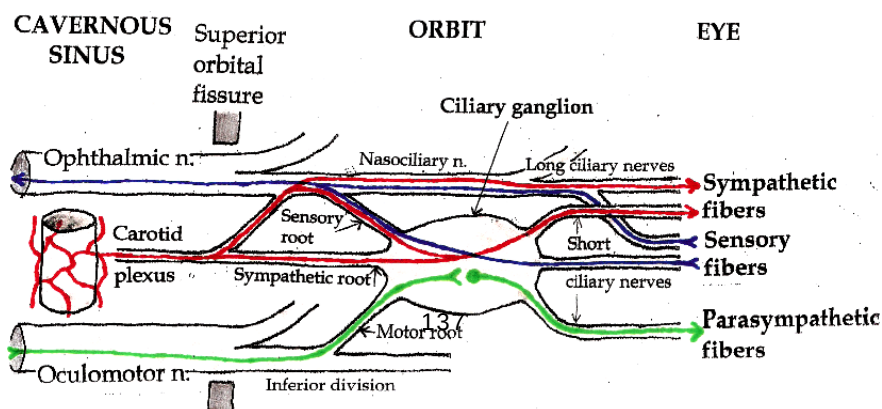
- Supply skin of the eyelids and side of nose, conjunctiva, lacrimal sac and caruncle.

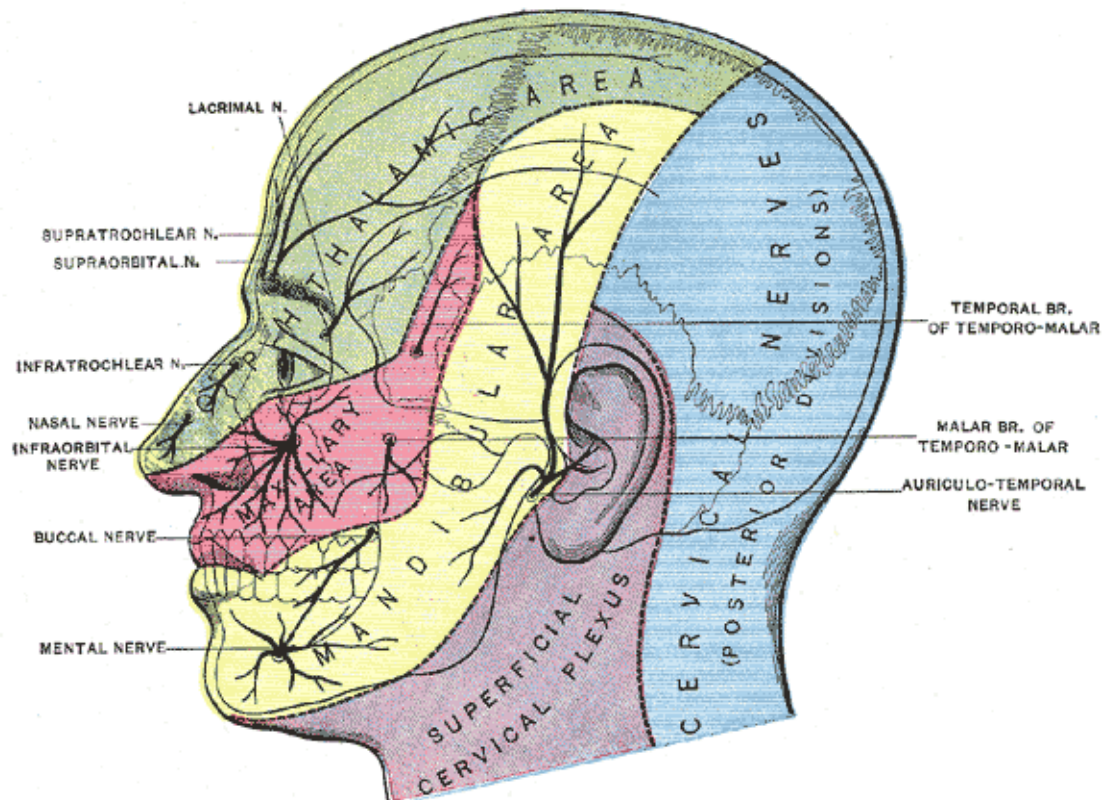
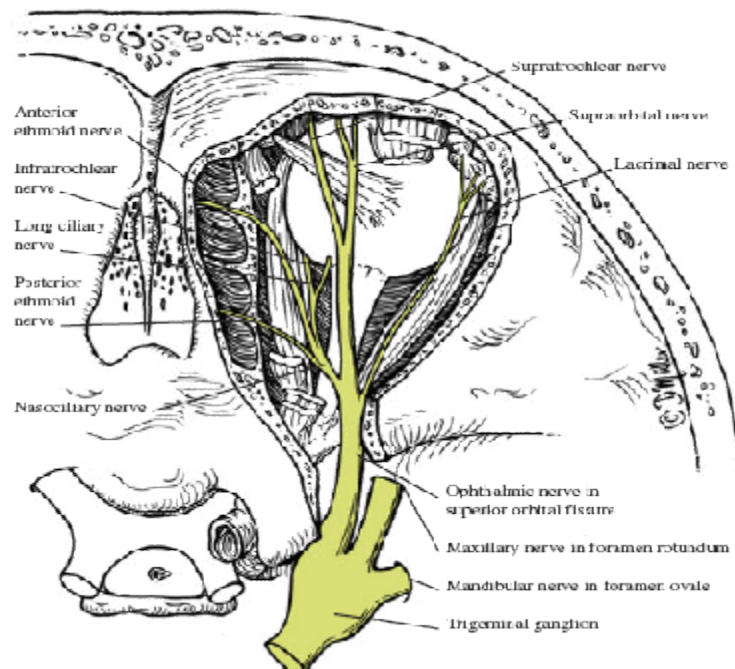
5. **Ciliary Ganglion:**

- Supply cornea, iris, and ciliary body.

6. **Long ciliary nerves:**

- Two or Three in number.
- Sensory innervation to the eyeball, including the cornea.
- Contain sympathetic fibers from the superior cervical ganglion to the dilator pupillae muscle.

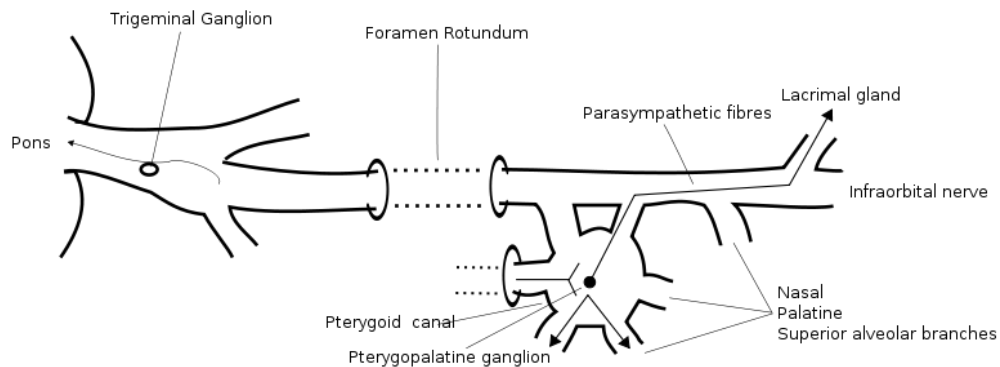
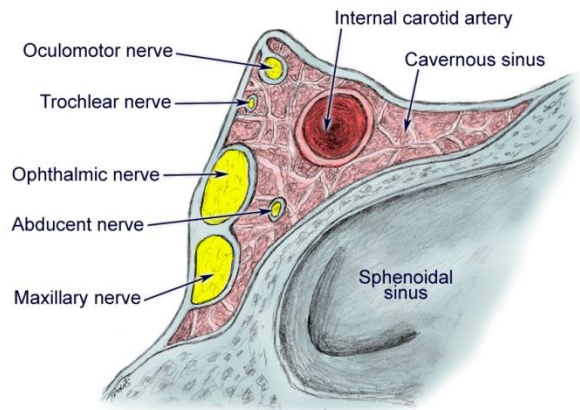


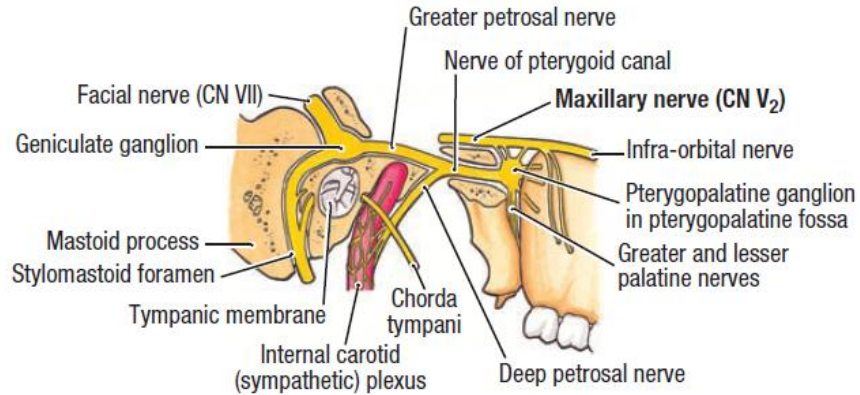


Ophthalmic Nerve Branches and Distribution

Nerve	Branches	Distribution
Frontal nerve	• Supraorbital nerve • Supratrochlear nerve	• Upper lid, frontalis muscle, scalp • Conjunctiva, upper lid, forehead
Lacrimal nerve	Receives branch from the zygomatic nerve of the maxillary	Lacrimal gland, conjunctiva, upper lid
Nasociliary nerve	• Anterior ethmoid nerve • Branches to ciliary ganglion • Posterior ethmoid nerve • 2-3 long ciliary nerves	• Frontal, anterior, ethmoid sinuses • Anterior septum, nasal wall • Cornea, iris, ciliary body • Posterior ethmoid sphenoid sinuses • Eye

- **Maxillary Nerve (V2):**
- Second branch of Trigeminal nerve (CN-V).
- Passes forward along the lateral wall of Cavernous sinus in Middle Cranial Fossa under Ophthalmic Nerve (V1).
- Exits the Skull through **Foramen Rotundum**.
- Crosses Pterygopalatine Fossa.
- Enter the orbit through **Inferior Orbital Fissure**.
- Appears upon the face at **Infraorbital Foramen**.
- **Pure sensory nerve.**
- Carries Sensory information from:
 - Lower eyelid
 - Cheek
 - Nares
 - Upper lip
 - Upper teeth and gums
 - Nasal mucosa
 - Palate
 - Roof of the pharynx
 - Maxillary, Ethmoid and sphenoid sinuses
 - Parts of the meninges.





- **Maxillary Nerve Branches:**

- **In the Cranium:**

1. Middle Meningeal Nerve.

- **In the Pterygopalatine Fossa:**

1. Zygomatic Nerve:

- Zygomatico-Temporal Nerve:

- In the lateral wall of the orbit.
- Gives a branch to Lacrimal nerve of (V1).
- Carries postganglionic fibers from the
- sphenopalatine ganglion for lacrimation.

- Zygomatico-Facial Nerve:

- Inferiorly situated
- Supplies the skin of the cheek

2. Sphenopalatine Nerve:

- Two nerves.
- Unite Sphenopalatine ganglion to the maxillary nerve (V2).
- Transmit Afferent sensations from the nose, palate, and pharynx.
- Carry Preganglionic Parasympathetic fibers (which derived from Facial nerve via the greater petrosal and vidian nerves) to Lacrimal nerve that go to the lacrimal gland.

3. Posterior Superior Alveolar Nerve:

- Two nerves.
- Supply mucosa of the posterior cheek and gingiva.

4. Nasopalatine Nerve:

- Enters nasal cavity through the sphenopalatine foramen.
- Passes across roof of nasal cavity below the orifice of the sphenoidal sinus to reach the septum.

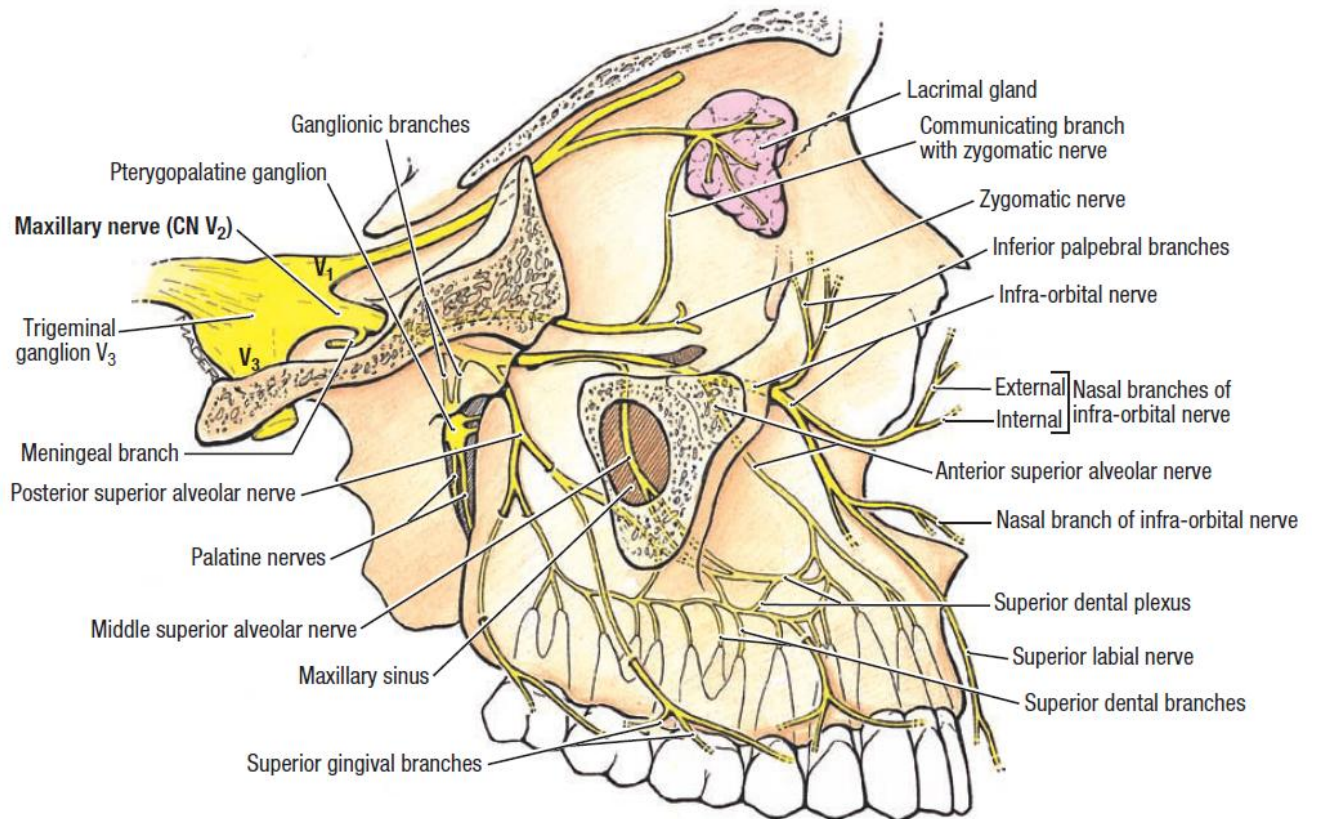
- Descends to roof of the mouth through Incisive canal and communicates with the corresponding nerve of the opposite side and with the greater palatine nerve.
- Supplies the palatal structures around the upper central and lateral incisors and the canines (the upper front six teeth).
- It also furnishes a few filaments to the mucous membrane of the nasal septum.

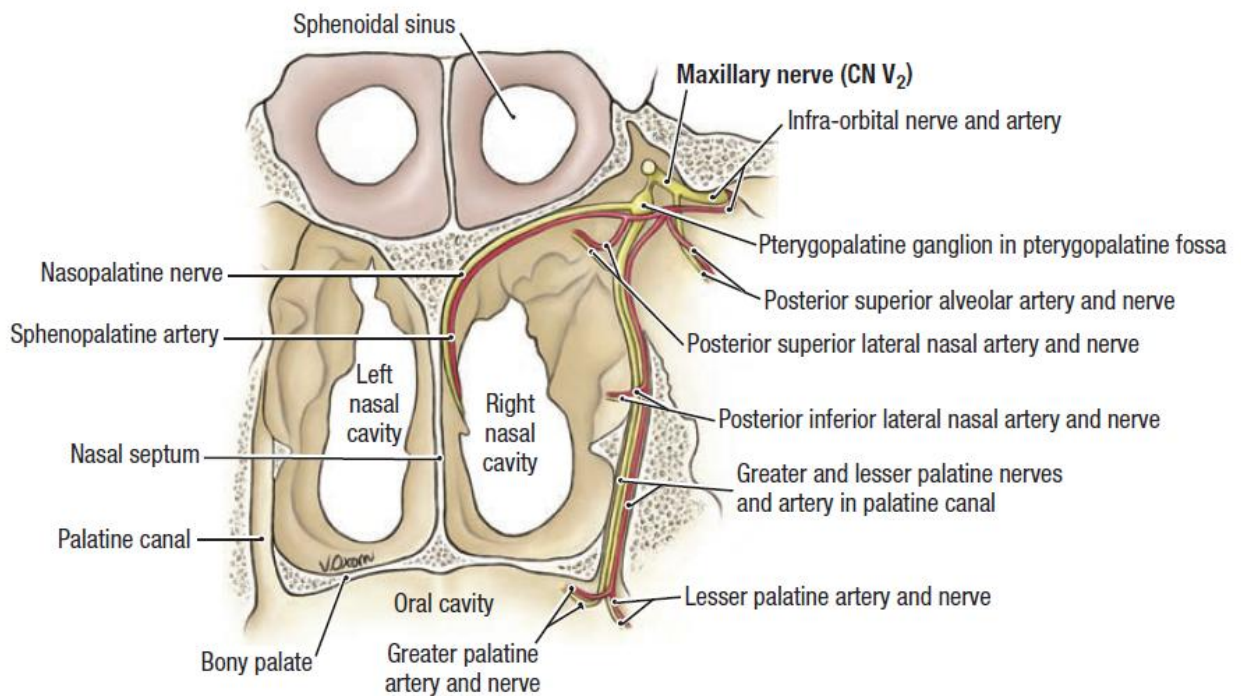
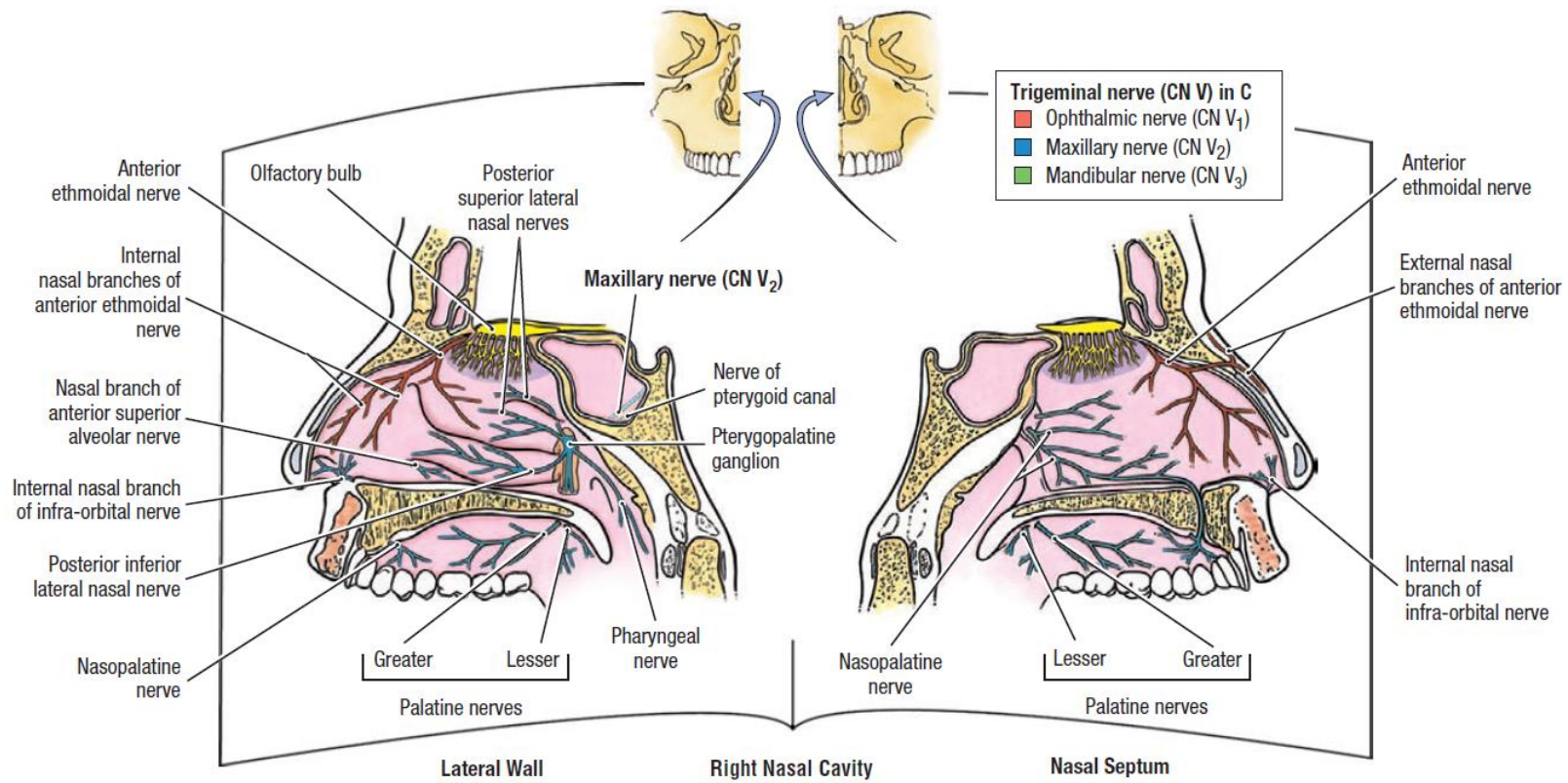
- In the Infraorbital canal:

1. Anterior Superior Alveolar nerve
2. Middle Superior Alveolar nerve

- On the face:

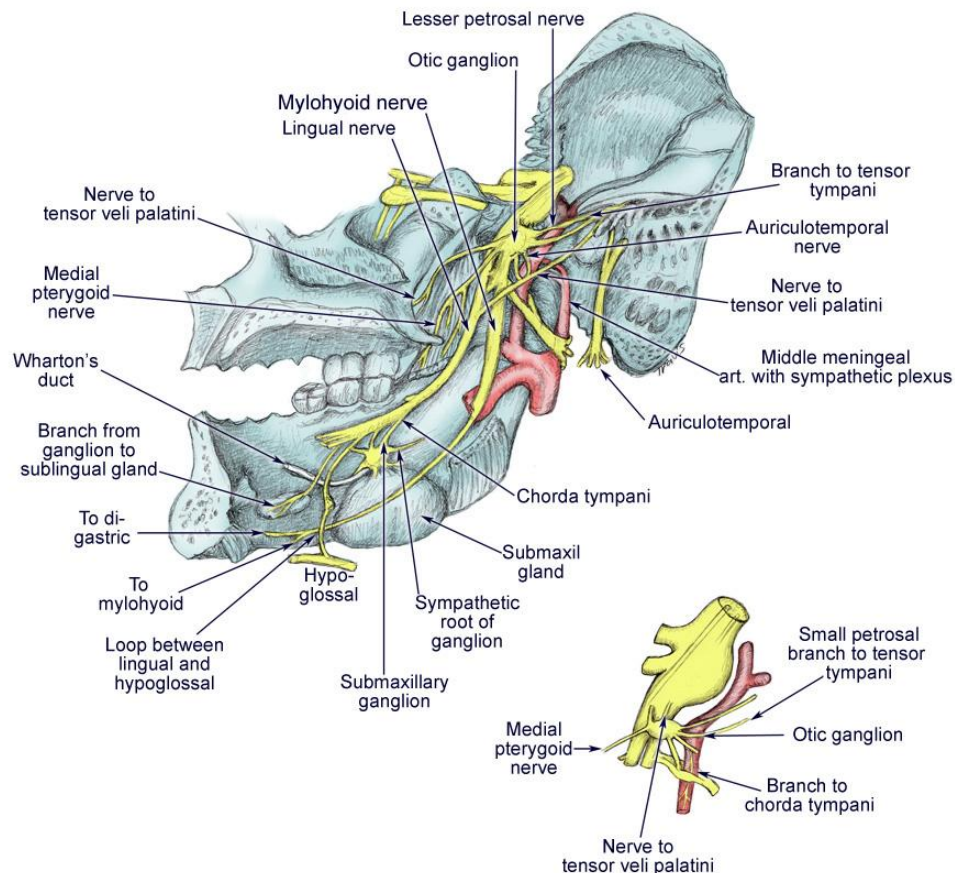
1. Infraorbital Nerve:
 - Inferior palpebral Nerve:
 - Supply Lower eyelid.
 - Nasal branch:
 - Supply part of the nasal vestibule.
 - Superior labial Nerve:
 - Supply Upper lip.





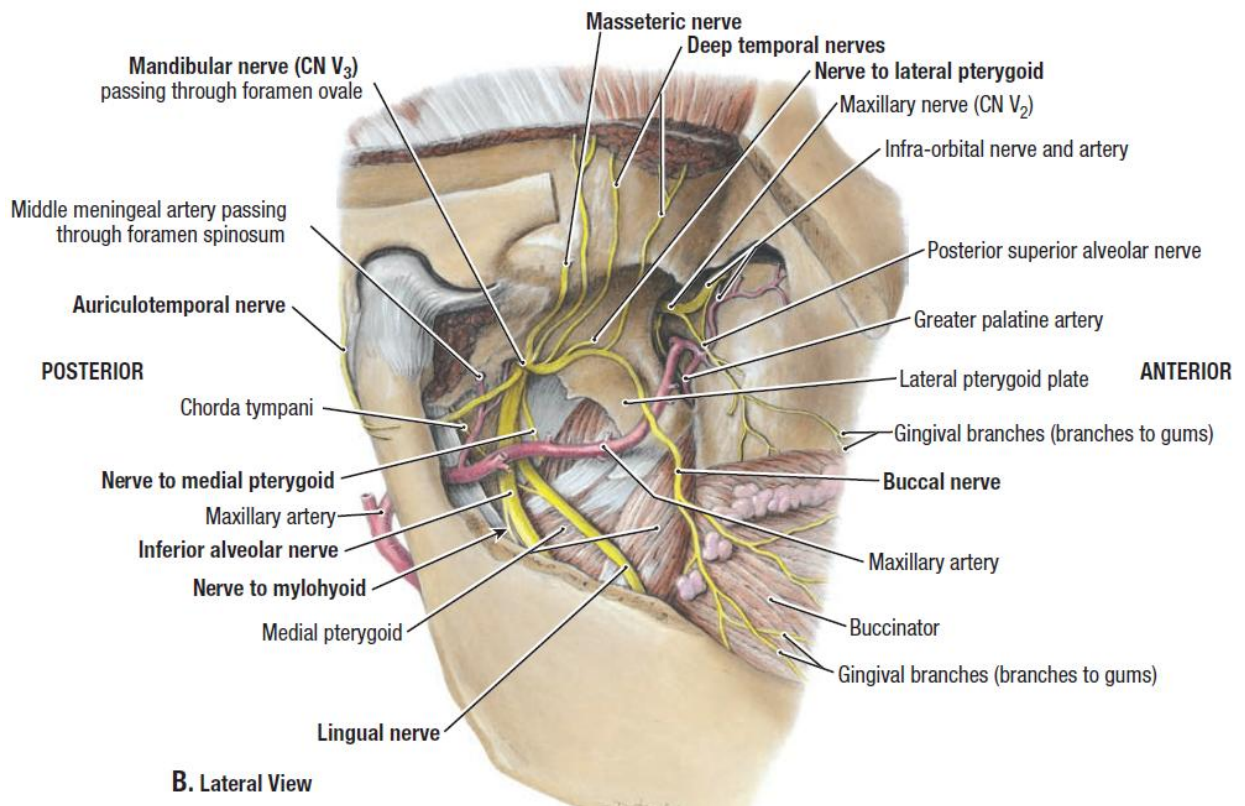
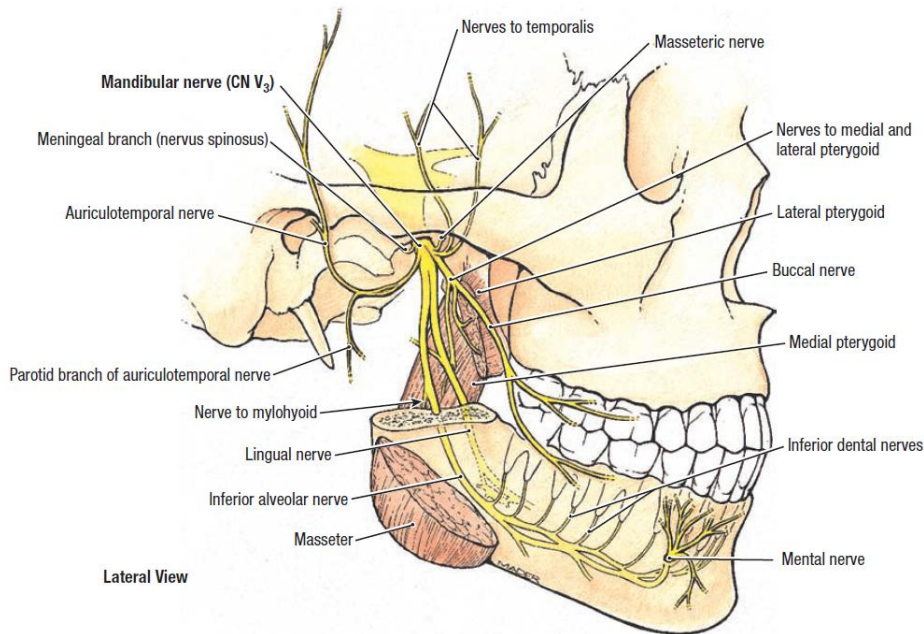
Nerve	Branches	Distribution
Middle meningeal nerve		Dura
Zygomatic nerve	• Zygomatico-temporal • Zygomatico-facial	• Lacrimal gland • Forehead • Cheek
Pterygopalatine nerve	• 2 branches unite sphenopalatine ganglion and maxillary nerve • Greater palatine nerve • Posterior superior nasal nerve • Pharyngeal	• Nasal cavity, pharynx, palate • Soft and hard palate • Superior, middle turbinate, septum • Nasopharynx
Posterior superior alveolar nerve	• Middle, anterior, superior alveolar, and nasal nerves	• Gums, posterior cheek, teeth (canine, incisors, premolar), nasal floor

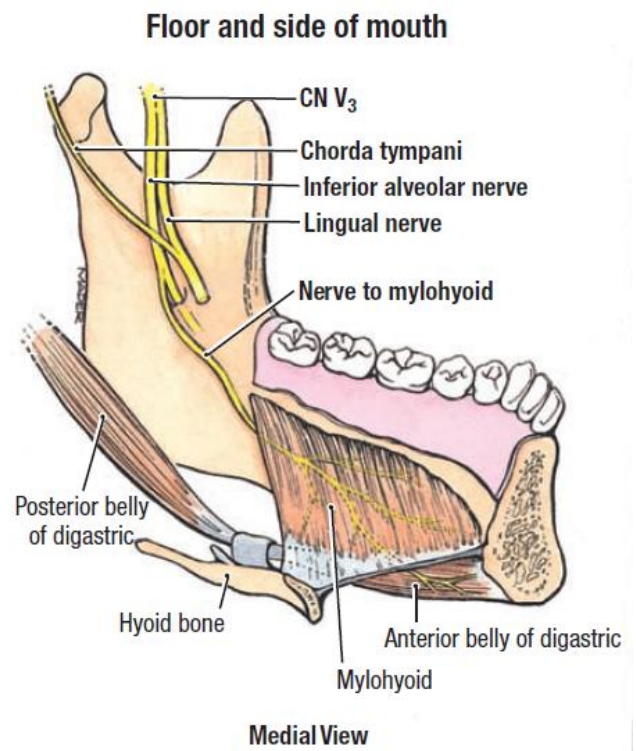
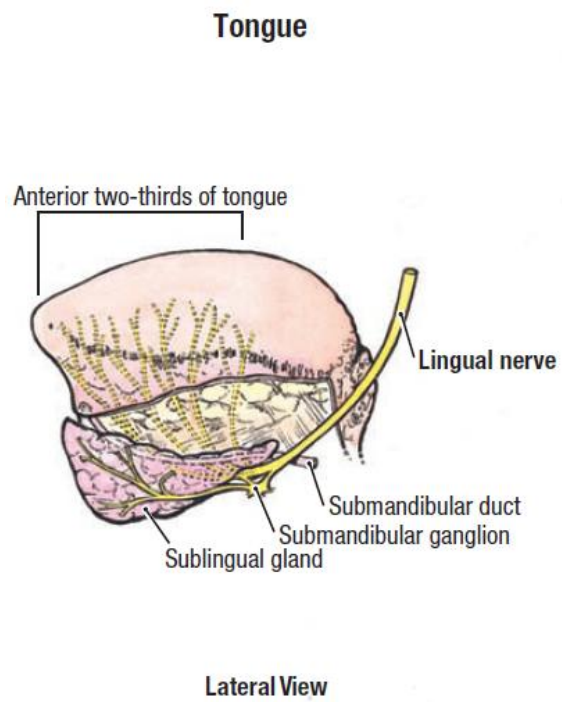
- **Mandibular Nerve (V3):**
- Third and Largest branch of Trigeminal nerve (CN-V).
- Exits the Skull through **Foramen Ovale**.
- **Mixed sensory and motor fibers.**
- Carries Sensory information from:
 - o Lower lip
 - o Lower teeth, gums, the chin and jaw (except the angle of the mandible, which is supplied by C2-C3).
 - o Parts of External ear
 - o Parts of the meninges.
- Carries touch/position and pain/temperature sensations from the mouth.
- Does not carry taste sensation (the chorda tympani is responsible for taste), but one of its branches, the lingual nerve, carries multiple types of nerve fibers that do not originate in the mandibular nerve.
- Motor branches of the trigeminal nerve are distributed in the mandibular nerve and originate in the motor nucleus of the fifth nerve, which is located near the main trigeminal nucleus in the pons.



- **Mandibular Nerve Branches:**

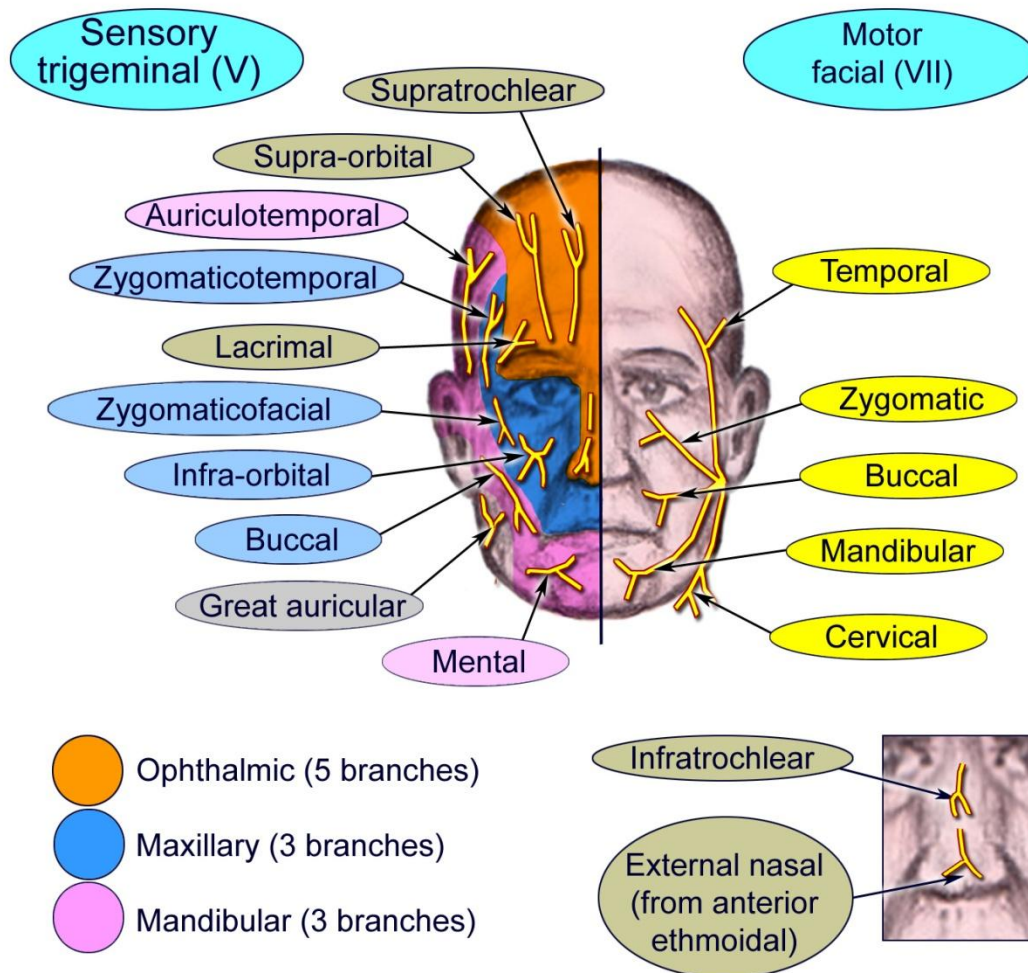
1. **Recurrent Meningeal nerve:**
 - Enters the skull via **Foramen spinosum** with Meningeal artery.
2. **Medial Pterygoid nerve:**
 - After passing through Otic ganglion without synapsing, this nerve supplies:
 1. **Medial pterygoid**
 2. **Tensor veli palatini**
 3. **Tensor tympani muscles**
3. **Masseteric nerve:**
 - Passes through Mandibular notch to innervate:
 1. **Masseter muscle**
 2. **Temporomandibular joint (TMJ)**
4. **Deep temporal nerves:**
 - Supply: **Temporal muscle**
5. **Lateral pterygoid nerve:**
 - Supply: **Lateral pterygoid.**
6. **Buccal nerve:**
 - Divides into:
 1. **Temporal branch**
 2. **Buccinator branche**
7. **Auriculotemporal nerve:**
 - Begins as 2 roots that encircle the middle meningeal artery.
 - Then forms a single trunk medial to the neck of the mandible
 - It emerges superficially between the ear and the mandibular condyle deep to the parotid gland.
 - Ends in 2 superficial temporal branches.
 - **Autonomic supply to the parotid gland.**
8. **Lingual nerve:**
 - Supplies mucous membrane of the anterior two-thirds of tongue.
 - Runs parallel to Inferior alveolar nerve.
 - Joined by Chorda tympani nerve of Facial nerve (CN-VII) near Internal maxillary artery.
 - Courses forward between Hyoglossus muscle and Deep part of Submandibular gland.
 - As it passes forward, crosses the submandibular (Wharton) duct along the tongue to its tip.
 - Supplies mucous membrane of the anterior two-thirds of tongue.
 - **Lingual nerve could be injured in this location during surgery on the floor of mouth or during excision of the submandibular gland.**
9. **Inferior alveolar nerve:**
 - Accompanies the Inferior alveolar artery in the mandibular foramen and courses into the mandibular canal.
 - Exit through the **Mental foramen.**
 - Divides into two terminal branches, incisive and mental.





Nerve	Branches	Distribution
Recurrent meningeal		Dura
Medial pterygoid		Medial pterygoid, Tensor veli palatini, Tensor tympani muscles
Masseteric		Masseter muscle, Temporomandibular joint
Deep temporal (x2)		Temporalis muscle
Lateral pterygoid		Lateral pterygoid muscle
Buccal	• Temporal nerve (upper) • Buccinator nerve (lower)	Skin of cheek, mucous membrane of mouth, and gingiva
Auriculotemporal	• Communication with facial nerve, and otic ganglion, • Articular nerve • Parotid gland	Parasympathetic and sympathetic supply to the parotid gland, after relay in the otic ganglion
8) Lingual	Communicates with CN VII via chorda tympani	Taste sensations to the anterior third of tongue
9) Inferior alveolar	• Mylohyoid • Dental • Incisive • Mental	Mylohyoid, Anterior belly of digastric, Molars, premolars, canine, incisors Lower lip, and chin

Face: motor and sensory supply



Facial nerve branches

Temporal: frontalis and procerus

Zygomatic 1: eye and around orbit

Zygomatic 2: mid face and smile

Buccal: buccinator and upper lip

Mandibular: lower lip and orbicularis oris

Cervical: platysma

(Note: proprioception is supplied by trigeminal)

TABLE 9.6 BRANCHES OF OPHTHALMIC NERVE (CN V₁)

Function	Riyadh et al. Notes	Branches
Ophthalmic nerve (CN V₁) Somatic sensory only at origin from trigeminal ganglion Visceral motor: extracranially, conveys (1) postsynaptic parasympathetic fibers from ciliary ganglion to ciliary body and sphincter of pupillae; (2) postsynaptic parasympathetic fibers from communicating branch of zygomatic nerve (CN V ₂) to lacrimal gland; and (3) postsynaptic sympathetic fibers from internal carotid plexus to dilator pupillae and intra-ocular blood vessels. Passes through superior orbital fissure to enter orbit Supplies general sensory innervation to cornea, superior bulbar and palpebral conjunctiva, mucosa of anterosuperior nasal cavity, frontal, ethmoidal, and sphenoidal sinuses, anterior and supratentorial dura mater, skin of dorsum of external nose, superior eyelid, forehead and anterior scalp.		Somatic sensory CN V₁
		Somatic sensory branches: Tentorial nerve (an intracranial meningeal branch) Lacrimal nerve [terminal portion also receives postsynaptic parasympathetic fibers from zygomatic nerve (CN V ₂) and conveys them to lacrimal gland] Frontal nerve Supra-orbital nerve Supratrochlear nerve Nasociliary nerve Sensory root of ciliary ganglion Long and short ciliary nerves [also convey postsynaptic sympathetic fibers (from internal carotid plexus to eyeball additionally, short ciliary nerves convey postsynaptic parasympathetic fibers from ciliary ganglion to eyeball)] Anterior and posterior ethmoidal nerves Anterior meningeal nerves Internal and external nasal branches Infratrochlear nerve

TABLE 9.7 BRANCHES OF MAXILLARY NERVE (CN V₂)

Function		Branches
Maxillary nerve (CN V₂) Somatic sensory only (proximally, at origin from trigeminal ganglion) Visceral motor: distally, conveys (1) postsynaptic parasympathetic fibers from pterygopalatine ganglion (presynaptic fibers are from CN VII via greater petrosal nerve and nerve of pterygoid canal); and (2) postsynaptic sympathetic fibers from superior cervical ganglion via internal carotid plexus (presynaptic fibers are from intermediolateral column of gray matter, spinal cord segments T1–T3). Passes through foramen rotundum to enter pterygopalatine fossa Supplies dura mater of anterior aspect of lateral part of middle cranial fossa; conjunctiva of inferior eyelid; mucosa of postero-inferior nasal cavity, maxillary sinus, palate, and anterior part of superior oral vestibule; maxillary teeth; and skin of lateral external nose, inferior eyelid, anterior cheek, and upper lip.		Somatic sensory CN V₂
		Somatic sensory branches: Meningeal branch Zygomatic branch Zygomaticofacial branch Zygomaticotemporal branch Communicating branch to lacrimal nerve Ganglionic branches to (sensory root of) pterygopalatine ganglion Infra-orbital nerve Posterior, middle, and anterior superior alveolar branches Superior dental plexus and branches Superior gingival branches Inferior palpebral branches External and internal nasal branches Superior labial branches Greater palatine nerve Posterior inferior lateral nasal nerves Lesser palatine nerves Posterior superior lateral and medial nasal branches Nasopalatine nerve Pharyngeal nerve

TABLE 9.8 BRANCHES OF MANDIBULAR NERVE (CN V₃)

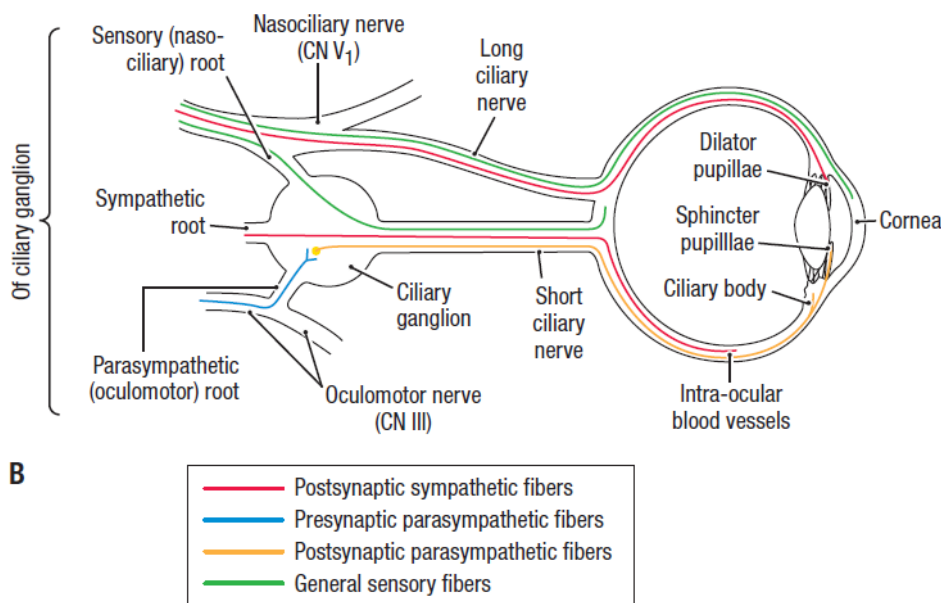
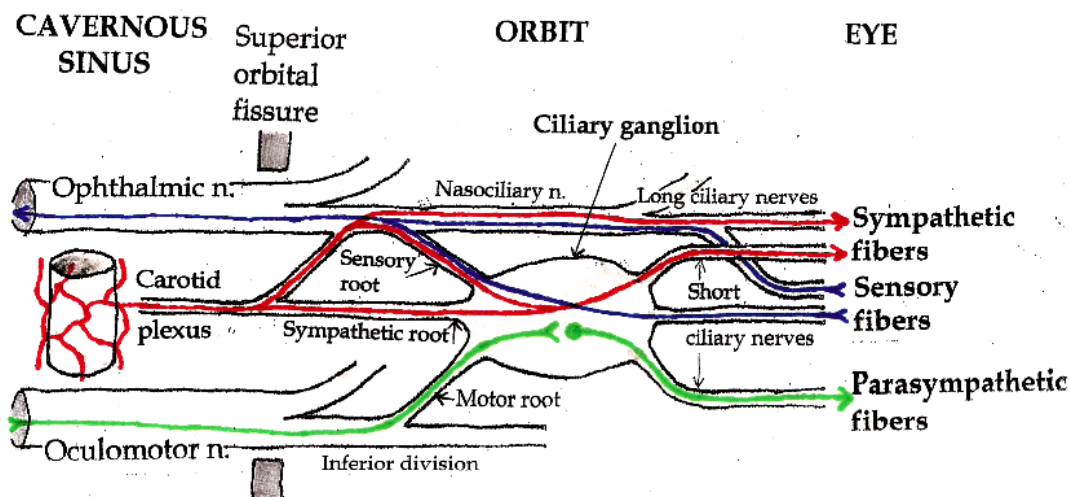
Function		Branches
Mandibular nerve (CN V₃) Somatic sensory and somatic (branchial) motor Special sensory: extracranially, conveys taste fibers (from CN VII via chorda tympani nerve) to anterior 2/3 of tongue Visceral motor: extracranially, conveys (1) presynaptic parasympathetic fibers to submandibular ganglion (presynaptic fibers are from CN VII via chorda tympani nerve); (2) postsynaptic parasympathetic fibers from submandibular ganglion to submandibular and sublingual glands; and (3) postsynaptic parasympathetic fibers from otic ganglion to parotid gland. Passes through foramen ovale to enter infratemporal fossa Supplies general sensory innervation to mucosa of anterior 2/3 of tongue, floor of mouth, and posterior and anterior inferior oral vestibule; mandibular teeth; and skin of lower lip, buccal and temporal regions of face, and external ear (anterior superior auricle, upper external auditory meatus, and tympanic membrane). Supplies motor innervation to all 4 muscles of mastication, mylohyoid, anterior belly of digastric, tensor tympani and tensor veli palatini		Somatic sensory CN V₃
		Somatic sensory branches: Meningeal branch (nervus spinosum) Buccal nerve Auriculotemporal nerve (also conveys visceral motor fibers) Superficial temporal branches Parotid branches Lingual nerve (also conveys visceral motor and special sensory fibers) Inferior alveolar nerve Nerve to mylohyoid Inferior dental plexus Inferior dental branches Inferior gingival branches Mental nerve Somatic (branchial) motor branches: Masseteric nerve Medial and lateral pterygoid branches Deep temporal nerves Nerve to mylohyoid Nerve to tensor tympani Nerve to tensor veli palatini
		Somatic motor CN V₃

- **Parasympathetic Ganglia:**

- Four small parasympathetic ganglia are associated anatomically (but not functionally) with the branches of the trigeminal nerve.

1. Ciliary ganglion:

- Associated with the ophthalmic nerve (V1).
- Has 3 roots:
 - 1. Sympathetic root** from Nasociliary nerve to dilator pupillae muscle of the eye.
 - 2. Sensory root** from the Nasociliary nerve to the cornea
 - 3. Parasympathetic root** from Nerve to inferior oblique (CN-III) from Edinger Westphal nucleus and caudal central nucleus to supply the sphincter pupillae and ciliary muscles.



Visceral (parasympathetic) motor innervation of ciliary and sphincter pupillae muscles

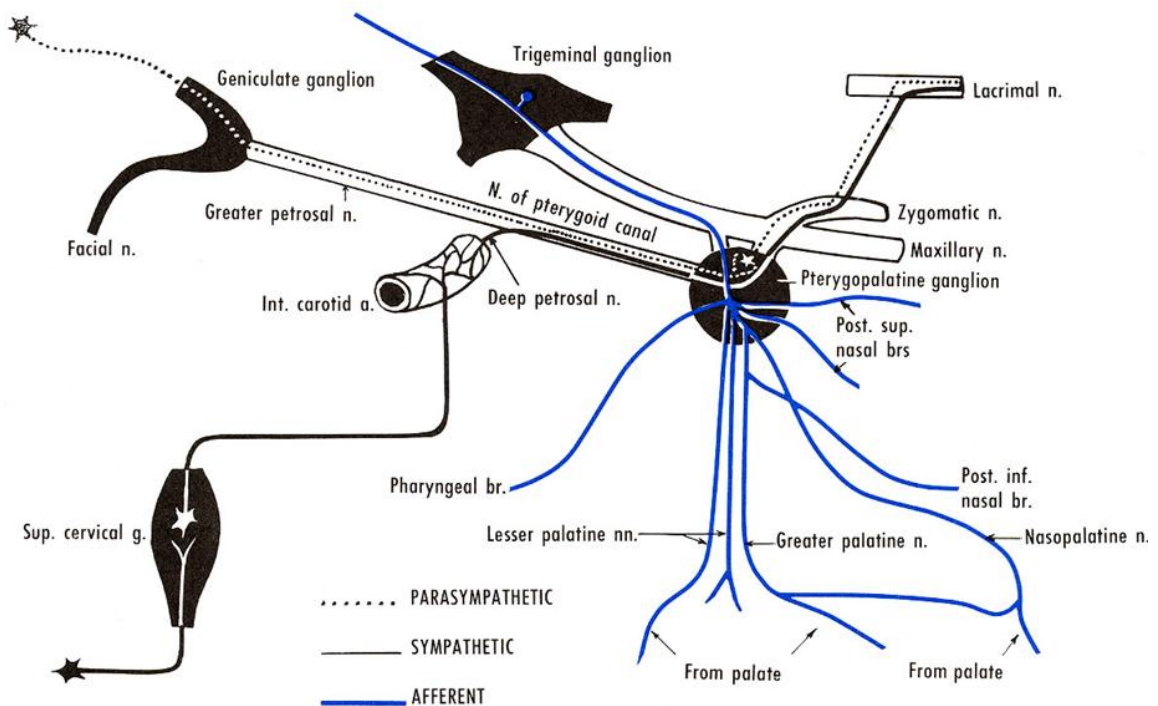
CN III contains parasympathetic fibers originating from nerve cell bodies of the accessory (Edinger-Westphal) nucleus of the oculomotor nerve.

Fibers synapse in the ciliary ganglion, consisting of postsynaptic parasympathetic nerve cell bodies associated with CN V₁.

Short ciliary nerves (CN V₁) carry postsynaptic parasympathetic fibers to the ciliary and sphincter pupillae muscles.

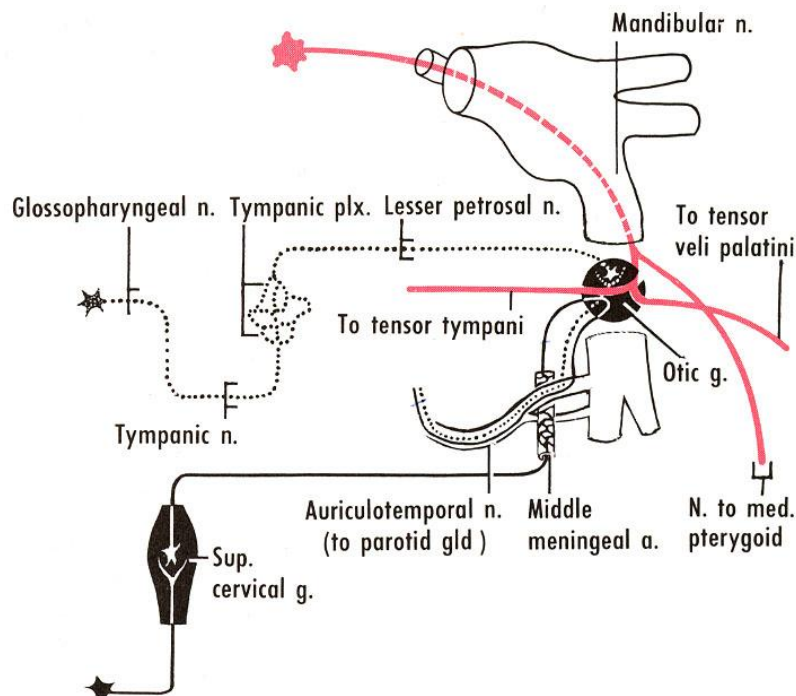
2. Pterygopalatine ganglion:

- Placed in the pterygopalatine fossa, just below the maxillary nerve as it crosses the fossa.
- Associated with the Maxillary nerve (V2).
- Receives a sensory, a motor, and a sympathetic root.
- **Sensory root:**
 - o From Sphenopalatine branch of the maxillary nerve (V2).
- **Secreto-motor root:**
 - o From **Greater superficial petrosal nerve** from the genicular ganglion of the facial nerve (CN-VII).
 - o Passes through the hiatus of the facial canal, enters the cranial cavity, and runs forward below the dura mater in a groove on the anterior surface of the petrous portion of the temporal bone.
 - o Then enters the vidian canal, just anterior to foramen lacerum.
 - o Joins the **Deep petrosal nerve** (given off from the carotid plexus, and runs through the carotid canal lateral to the internal carotid artery) to form the **Nerve of the pterygoid canal (Vidian nerve)** to enter pterygopalatine fossa from behind.
- **Branches of Pterygopalatine ganglion:**
 1. Lacrimal branches:
 - o Via zygomatic branch of Maxillary Nerve (V2) to Lacrimal gland.
 2. Palatine nerves:
 - o Descends through Pterygopalatine canal.
 - o Emerges upon the hard palate through the Greater palatine foramen.
 3. Nasal branches:
 - o Distributed to the septum and lateral nasal wall.
 4. Pharyngeal nerve:
 - o Distributed to the mucous membrane of nasopharynx.



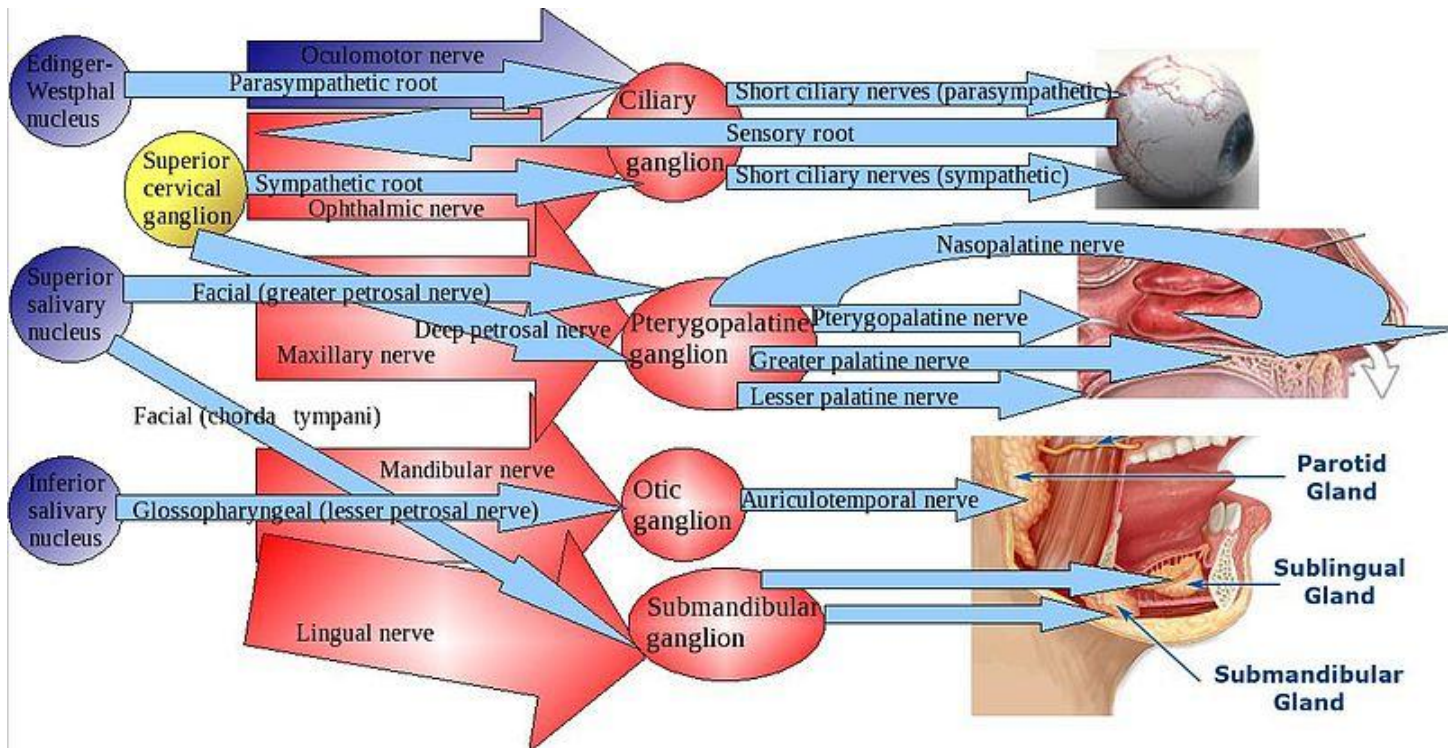
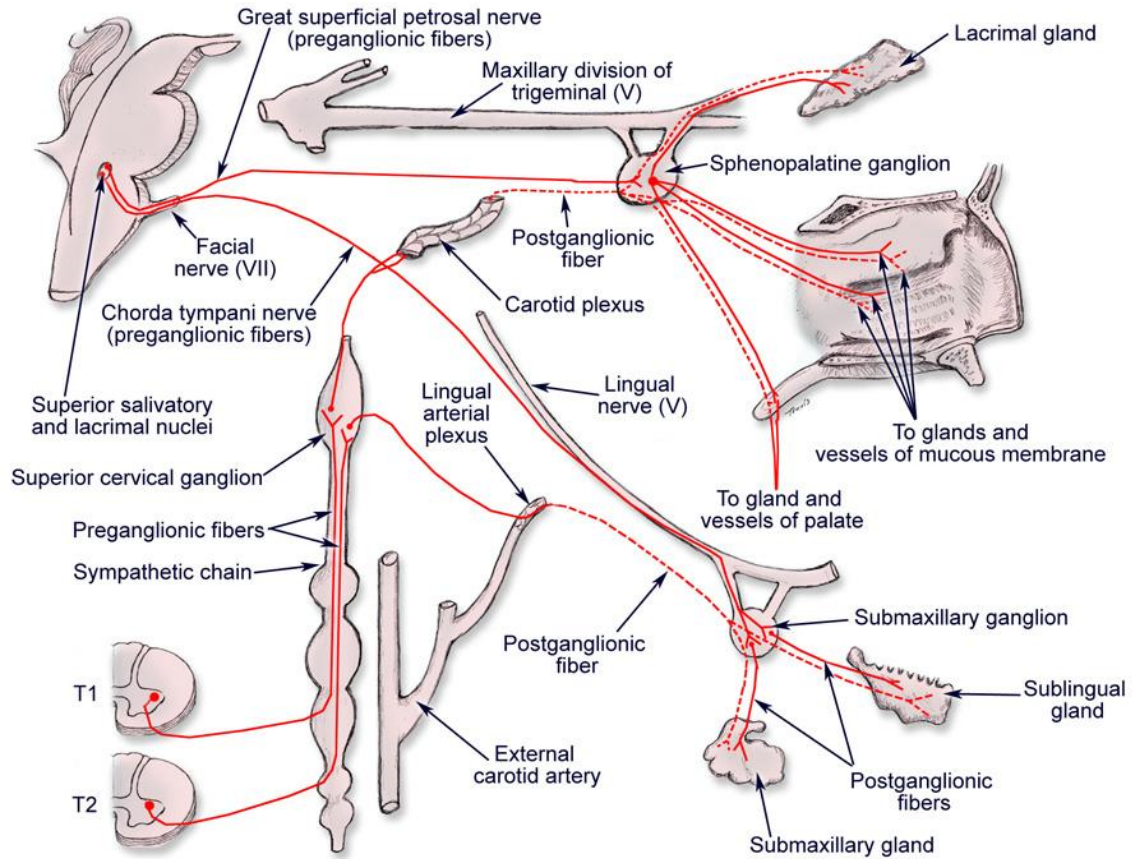
3. Otic ganglion:

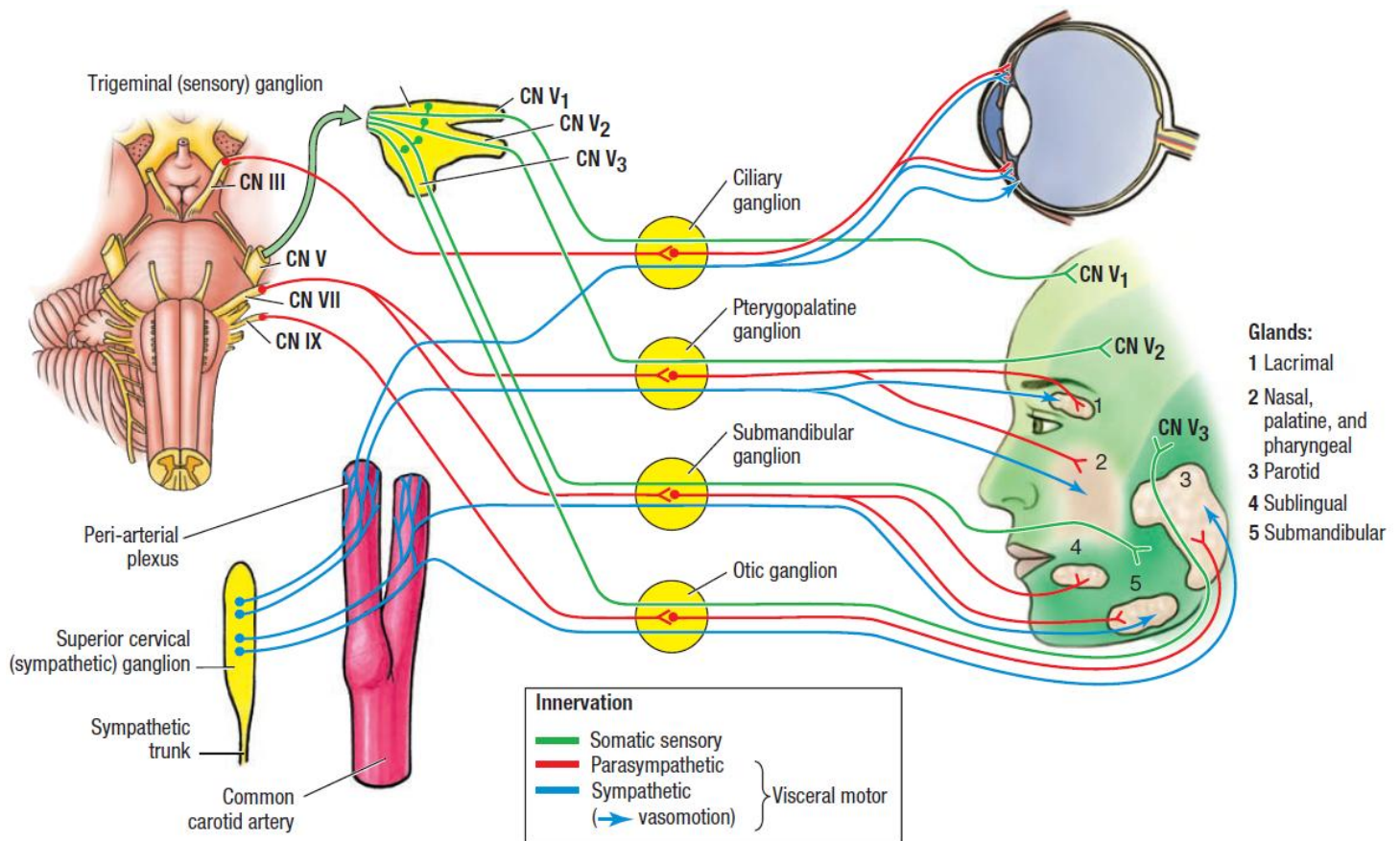
- Situated immediately below the foramen ovale.
- On the medial surface of the mandibular nerve.
- Medially, it is related to cartilaginous part of ET.
- Associated with the Mandibular nerve (V3).
- Preganglionic parasympathetic fibers:
 - From Inferior Salivatory Nucleus of the medulla via Glossopharyngeal nerve (CN-IX).
- Postganglionic parasympathetic fibers:
 - Pass with the Auriculotemporal nerve to Parotid gland.



4. Submandibular ganglion:

- Situated above the deep portion of the submandibular gland, on the hyoglossus.
- Suspended from the lingual nerve by two filaments. Through the posterior of these it receives a branch from the chorda tympani nerve which runs in the sheath of the lingual.
- Associated with the Mandibular nerve (V3).
- Preganglionic parasympathetic fibers:
 - From Superior Salivatory Nucleus of the medulla via chorda tympani nerve (CN-VII).

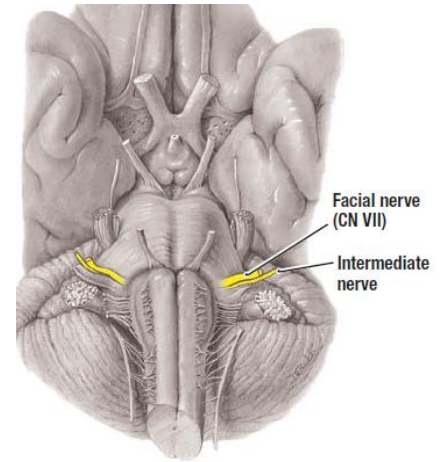




Ganglion	Location	Parasympathetic Root (Nucleus of Origin) ^a	Sympathetic Root	Main Distribution
Ciliary	Between optic nerve and lateral rectus, close to apex of orbit	Inferior branch of oculomotor nerve (CN III) (Edinger-Westphal nucleus)	Branch from internal carotid plexus in cavernous sinus	Parasympathetic postsynaptic fibers from ciliary ganglion pass to ciliary muscle and sphincter, pupillae of iris; sympathetic postsynaptic fibers from superior cervical ganglion pass to dilator pupillae and blood vessels of eye
Pterygopalatine	In pterygopalatine fossa, where it is attached by pterygopalatine branches of maxillary nerve; located just anterior to opening of pterygoid canal and inferior to CN V ₂	Greater petrosal nerve from facial nerve (CN VII) (superior salivatory nucleus)	Deep petrosal nerve, a branch of internal carotid plexus that is continuation of postsynaptic fibers of cervical sympathetic trunk; fibers from superior cervical ganglion pass through pterygopalatine ganglion and enter branches of CN V ₂	Parasympathetic postsynaptic fibers from pterygopalatine ganglion innervate lacrimal gland through zygomatic branch of CN V ₂ ; sympathetic postsynaptic fibers from superior cervical ganglion accompany branches of pterygopalatine nerve that are distributed to the nasal cavity, palate, and superior parts of the pharynx
Otic	Between tensor veli palatini and mandibular nerve; lies inferior to foramen ovale	Tympanic nerve from glossopharyngeal nerve (CN IX); tympanic nerve continues from tympanic plexus as lesser petrosal nerve (inferior salivatory nucleus)	Fibers from superior cervical ganglion travel via plexus on middle meningeal artery	Parasympathetic postsynaptic fibers from otic ganglion are distributed to parotid gland through auriculotemporal nerve (branch of CN V ₃); sympathetic postsynaptic fibers from superior cervical ganglion pass to parotid gland and supply its blood vessels
Submandibular	Suspended from lingual nerve by two short roots; lies on surface of hyoglossus muscle inferior to submandibular duct	Parasympathetic fibers join facial nerve (CN VII) and leave it in its chorda tympani branch, which unites with lingual nerve (superior salivatory nucleus)	Sympathetic fibers from superior cervical ganglion travel via the plexus on facial artery	Postsynaptic parasympathetic fibers from submandibular ganglion are distributed to the sublingual and submandibular glands; sympathetic fibers supply sublingual and submandibular glands and appear to be secretomotor

- **CRANIAL NERVE VII: FACIAL:**

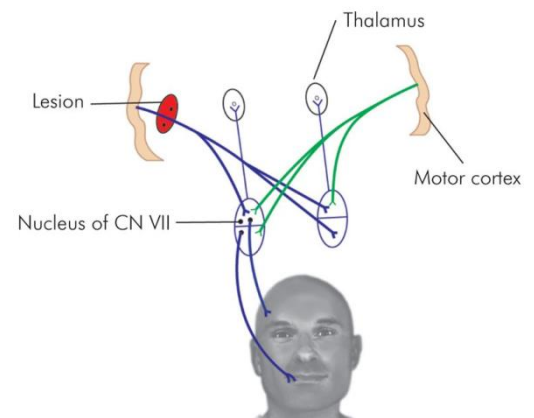
- Nerve of Facial expression.
- Facial Motor Nucleus is located in **Pons**, beneath the Fourth ventricle.
- Composed of 10,000 Neurons.
 - o 7,000 Neurons are Myelinated and innervate muscles of Facial expression (Motor).
 - o 3,000 Neurons are Somatosensory and Secretomotor and make up **Nervus Intermedius** (Nerve of Wrisberg)
- Mixed Motor and Sensory Nerve.



- 3 Nuclei:

1. Motor Nucleus:

- Located in Pons beneath the Fourth ventricle.
- Facial neurons pass around Abducens nucleus as they emerge from Brainstem.
 - o Involvement of Facial Nerve Nucleus and Abducens Nerve nucleus are suggestive of a lesion near Fourth ventricle.
- Receives fibers from Precentral gyrus.
- Upper part of Motor Nucleus:
 - o Innervates Forehead muscles.
 - o Receives fibers from both Cerebral hemispheres.
 - o Function of Forehead is preserved in Supranuclear lesions (UMNL) because of Bilateral innervation.
- Lower part of Motor Nucleus:
 - o Innervates Lower Face.
 - o Receives only Crossed fibers from one hemisphere.
 - o Function of Forehead is Lost in (LMNL) because of Unilateral innervation.
- Motor Nucleus also receives fibers from Thalamus:
 - Provides involuntary control to facial muscles.
 - Emotional movements (Smiling and crying) are Preserved in Supranuclear lesions (UMNL) because of these fibers from Thalamus.
 - With Nuclear and Infranuclear lesions, Loss of involuntary and voluntary facial movement occurs.
- SVE:
 - o Responsible for innervating muscles of 2nd Pharyngeal Arch:
 1. Muscles of Facial Expression.
 2. Stapedius.
 3. Stylohyoid.
 4. Posterior belly of Digastric.



2. Superior Salivatory Nucleus:

- Located in Pons.
- **GVE (Parasympathetic):**
 1. Seromucinous Nasal Glands
 2. Seromucinous Palatine Glands
 3. Lacrimal Gland
 4. Sublingual Salivary Gland
 5. Submandibular Salivary Gland

3. Nucleus Solitarius:

- Located in Medulla.
- **SVA :**
 - o Taste from Anterior 2/3 of Tongue and Skin.

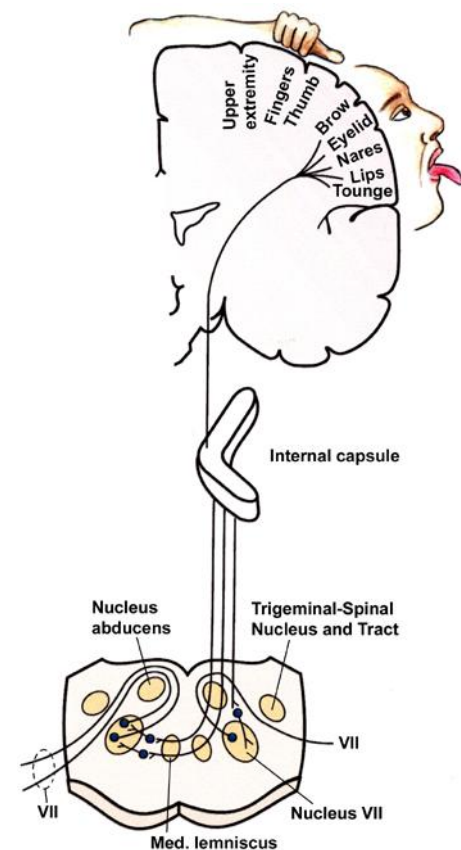
- **Components of CN-VII:**

1. Special Visceral Efferent (SVE): (Motor)

- Begins in Motor Nucleus of Facial nerve in Pons.
- Carried in Motor root of Facial nerve.
- **Innervation Pathways:**
 - o Premotor cortex
 - o → Corticobulbar Tract
 - o → Bilateral Facial Motor Nuclei
 - o → 2nd Pharyngeal Arch muscles
 1. Muscles of Facial Expression.
 2. Stapedius.
 3. Stylohyoid.
 4. Posterior belly of Digastric.
- In Bell's palsy, Muscles innervated by SVE fibers are paralyzed.

2. General Visceral Efferent (GVE): (Parasympathetic)

- Begins in Superior Salivatory Nucleus in Pons.
- Carried in Nervus Intermedius.
- **1st Innervation Pathways:**
 - o Superior Salivatory Nucleus (**Pons**)
 - o → Nervus Intermedius (**CN-VII**)
 - o → Greater Superficial Petrosal Nerve
 - o Joins Deep Petrosal Nerve (Sympathetic fibers from Cervical Plexus).
 - o → Vidian Nerve (Nerve of Pterygoid canal).
 - o → Spheno/Pterygo-palatine Ganglion.
 - o → Postganglionic Parasympathetic Fibers
 - o → Joins ZygomaticoTemporal Nerve (V2)
 - o →
 1. Seromucinous Nasal Glands.
 2. Seromucinous Palatine Glands.
 3. Lacrimal Gland.



- 2nd Innervation Pathways:

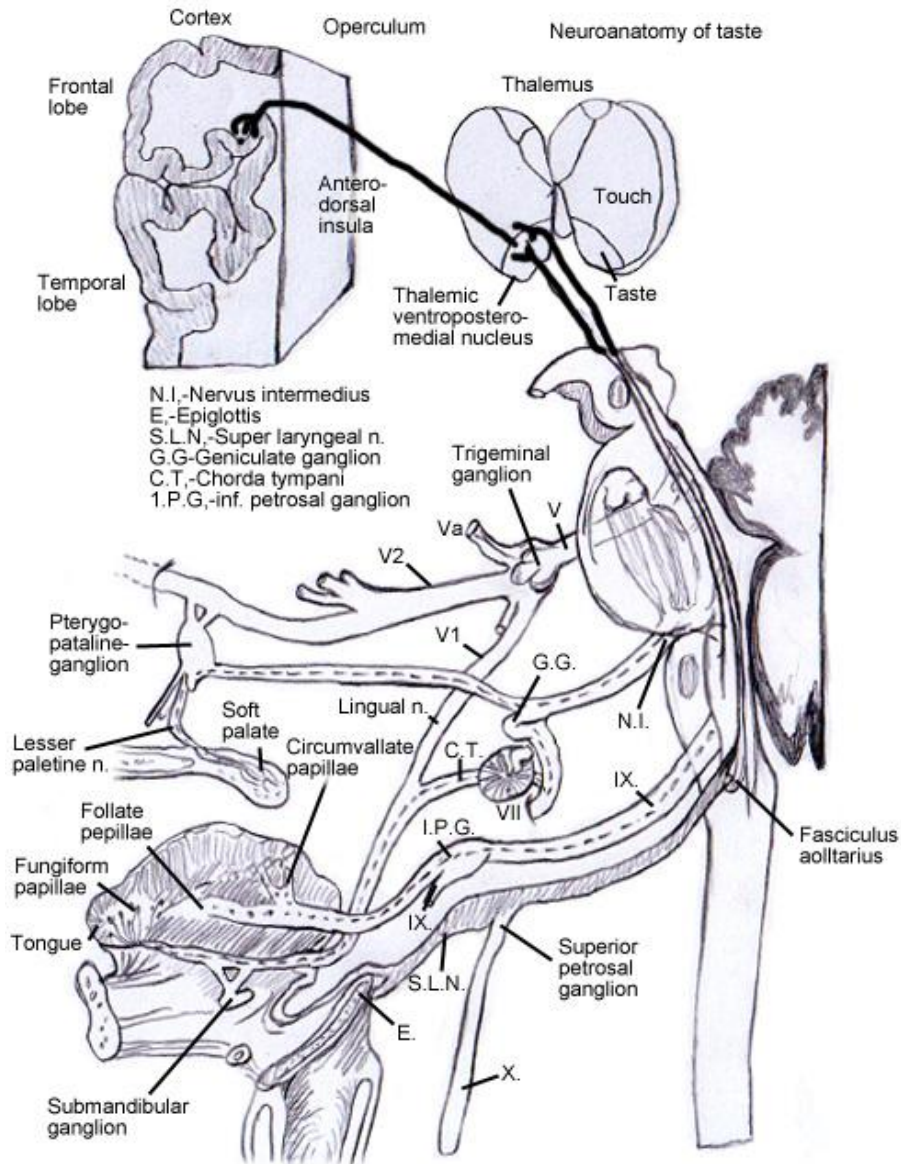
- Superior Salivatory Nucleus (**Pons**)
- → Nervus Intermedius (**CN-VII**)
- → Chorda Tympani.
- → Lingual Nerve.
- → Submandibular Ganglion.
- → Postganglionic Parasympathetic Fibers
- →
 1. Submandibular Salivary Gland
 2. Sublingual Salivary Gland

3. General Sensory Afferent (GSA): (Sensory)

- Carried in Nervus Intermedius.
- Supplies Sensation to Auricular concha, Postauricular skin, Posterosuperior part of EAC and TM.
- Cell bodies housed in Genuate ganglion.

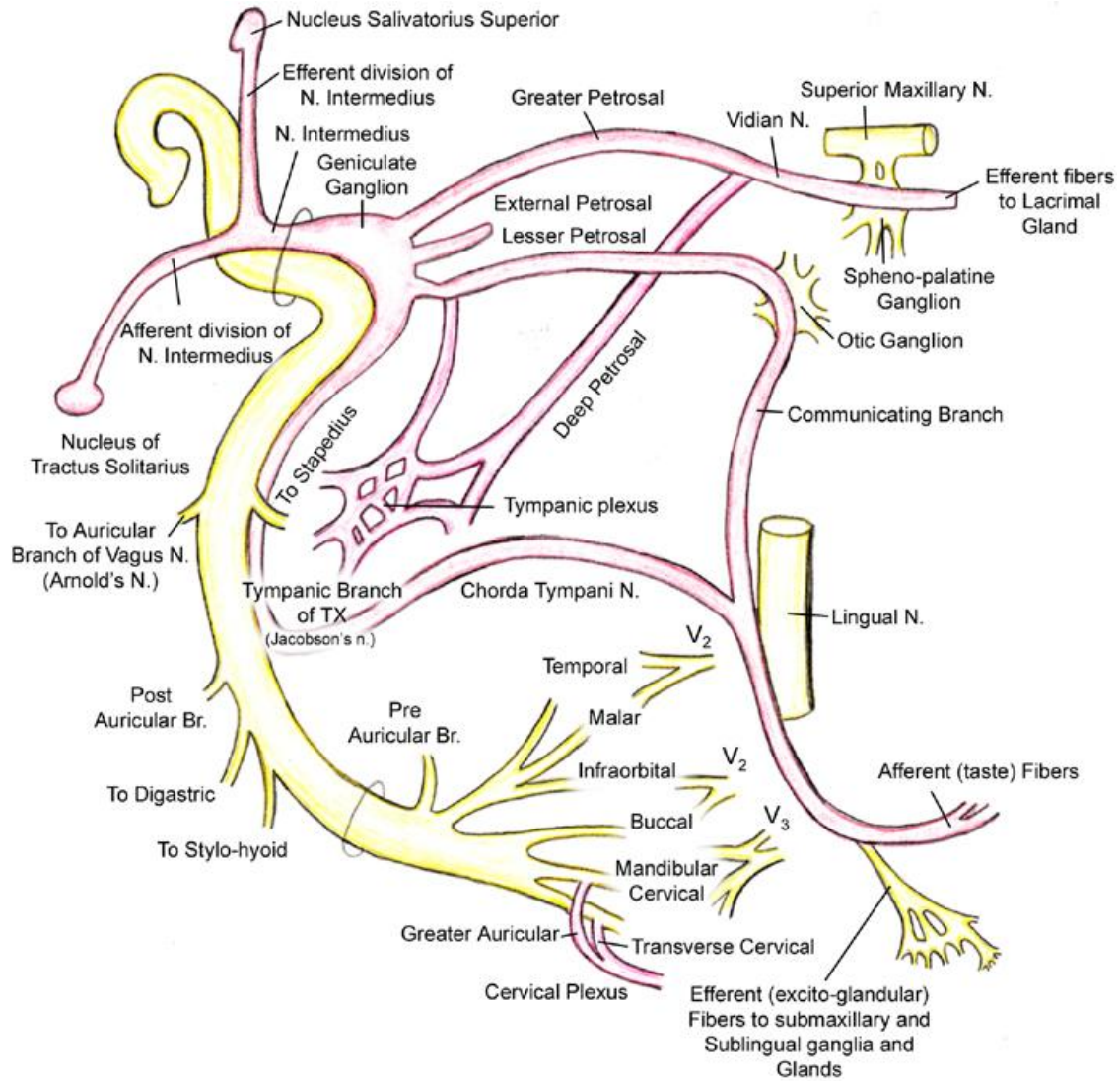
4. Special Visceral Afferent (SVA): (Taste)

- Carried in Nervus Intermedius.
- Begins in Nucleus Solitarius in Medulla.
 - → Tractus Solitarius.
 - → Nervus Intermedius (**CN-VII**)
 - → Genuate Ganglion
 - → Chorda Tympani.
 - → Lingual Nerve.
 - → Taste from Anterior 2/3 of Tongue
- Nucleus Solitarius in Medulla.
 - → Tractus Solitarius.
 - → Nervus Intermedius (**CN-VII**)
 - → Genuate Ganglion
 - → Greater Superficial Petrosal Nerve
 - Joins Deep Petrosal Nerve (Sympathetic fibers from Cervical Plexus).
 - → Vidian Nerve (Nerve of Pterygoid canal).
 - → Spheno/Pterygo-palatine Ganglion.
 - → Lesser Palatine Nerve (V2)
 - → Taste from Soft and Hard Palate.



- Nervus Intermedius carries:

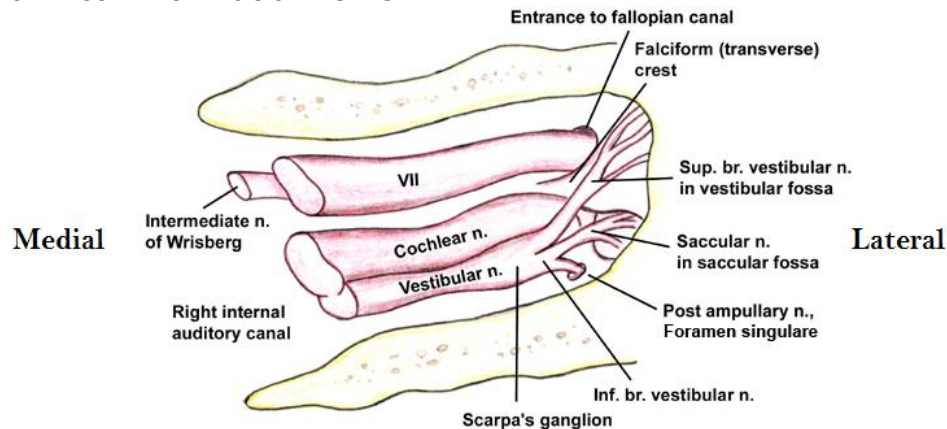
1. Taste fibers from:
 - Anterior 2/3 Tongue via Chorda tympani nerve.
 - Palate via Palatine and Greater Petrosal Nerves.
2. Preganglionic Parasympathetic Innervation to:
 - Submandibular Salivary Gland
 - Sublingual Salivary Gland
 - Lacrimal Gland.
3. Cutaneous sensory from:
 - Auricular concha
 - Postauricular skin
 - Posterosuperior part of EAC and TM.



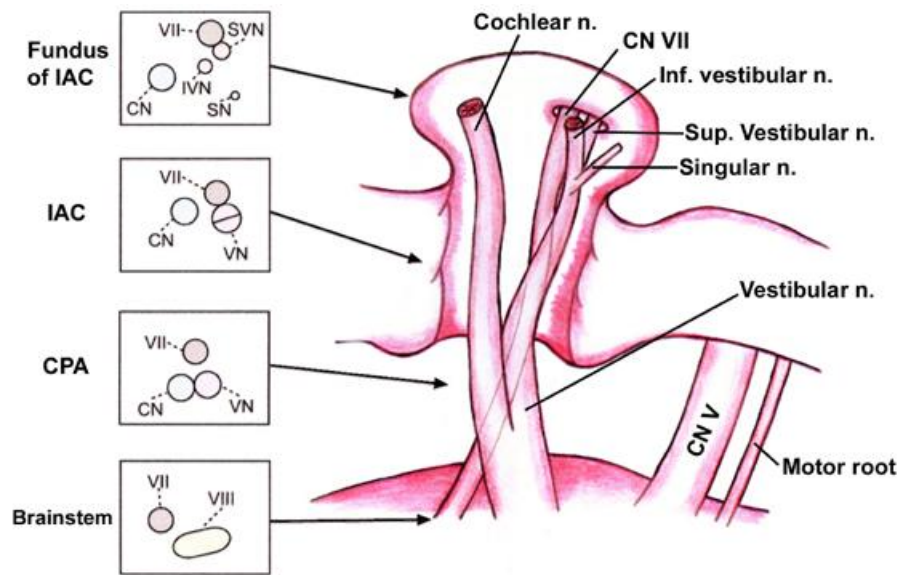
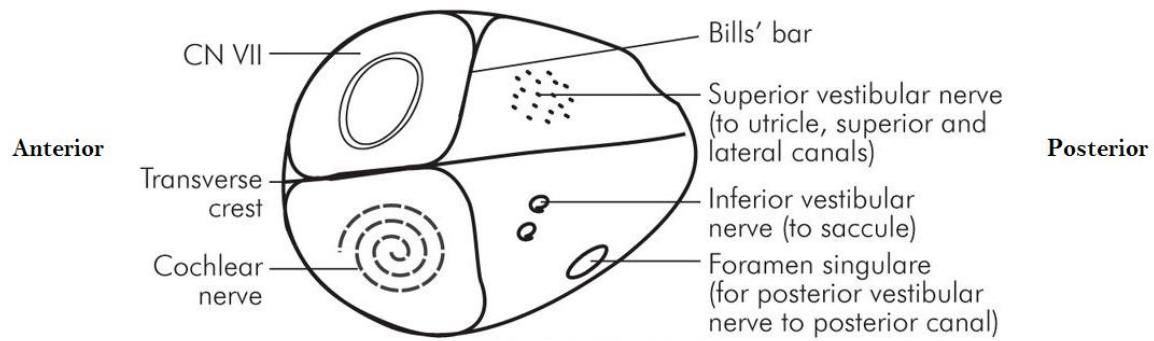
- Parts of Facial Nerve (CN-VII):

1. Intra-Cranial Part.
2. Intra-Temporal Part:
 1. Meatal Segment.
 2. Labirithine Segment.
 3. Tympanic (Horizontal) Segment.
 4. Mastoid (Vertical) Segment.
3. Extra-Temporal Parts.

- **Intra-Cranial Part of CN-VII:**
- **Pons → Porus Acousticus (Medial opening of IAM)**
- 15-17 mm in length.
- Facial nerve emerges from Brainstem with Nervus intermedius.
- Nervus intermedius gained its name from its position as it courses across Cerebellopontine angle (CPA) between Facial Nerve (CN-VII) and Vestibulocochlear Nerve (CN-VIII)
- Facial nerve and Nervus intermedius lie above and slightly anterior to CN-VIII.
- Motor Root of Facial Nerve joins Nervus intermedius in CPA/IAM to form common Facial Nerve.



- Lesions at level of CPA/IAM result in disturbances in:
 - Tearing
 - Taste
 - Salivary gland flow
 - Hearing
 - Balance
 - Facial function
- **At Fundus of IAM (Lateral Opening):**
 - Facial nerve and Nervus intermedius enter IAM with Vestibulocochlear Nerve.
 - Falciform Crest (Transverse Crest) divides IAM into Superior and Inferior compartments.
 - Bill's bar divides IAM into Anterior and Posterior compartments
- **Seven-up Coke-down:**
 - Facial Nerve runs into Superior-Anterior Compartment.
 - Vestibulocochlear Nerve enters IAM Inferiorly and Divides into:
 1. Superior Vestibular Nerve:
 - Runs into Superior-Posterior Compartment.
 2. Inferior Vestibular Nerve:
 - Runs into Inferior-Posterior Compartment.
 3. Cochlear Nerve:
 - Runs into Inferior-Anterior Compartment.



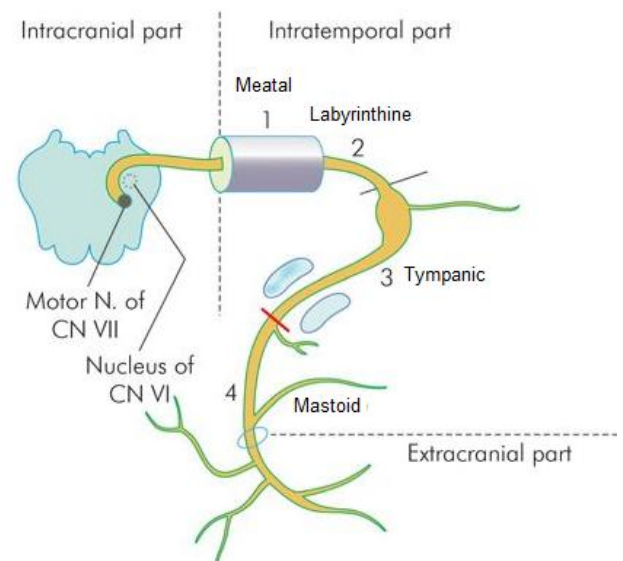
- **Intra-Temporal Part of CN-VII:**

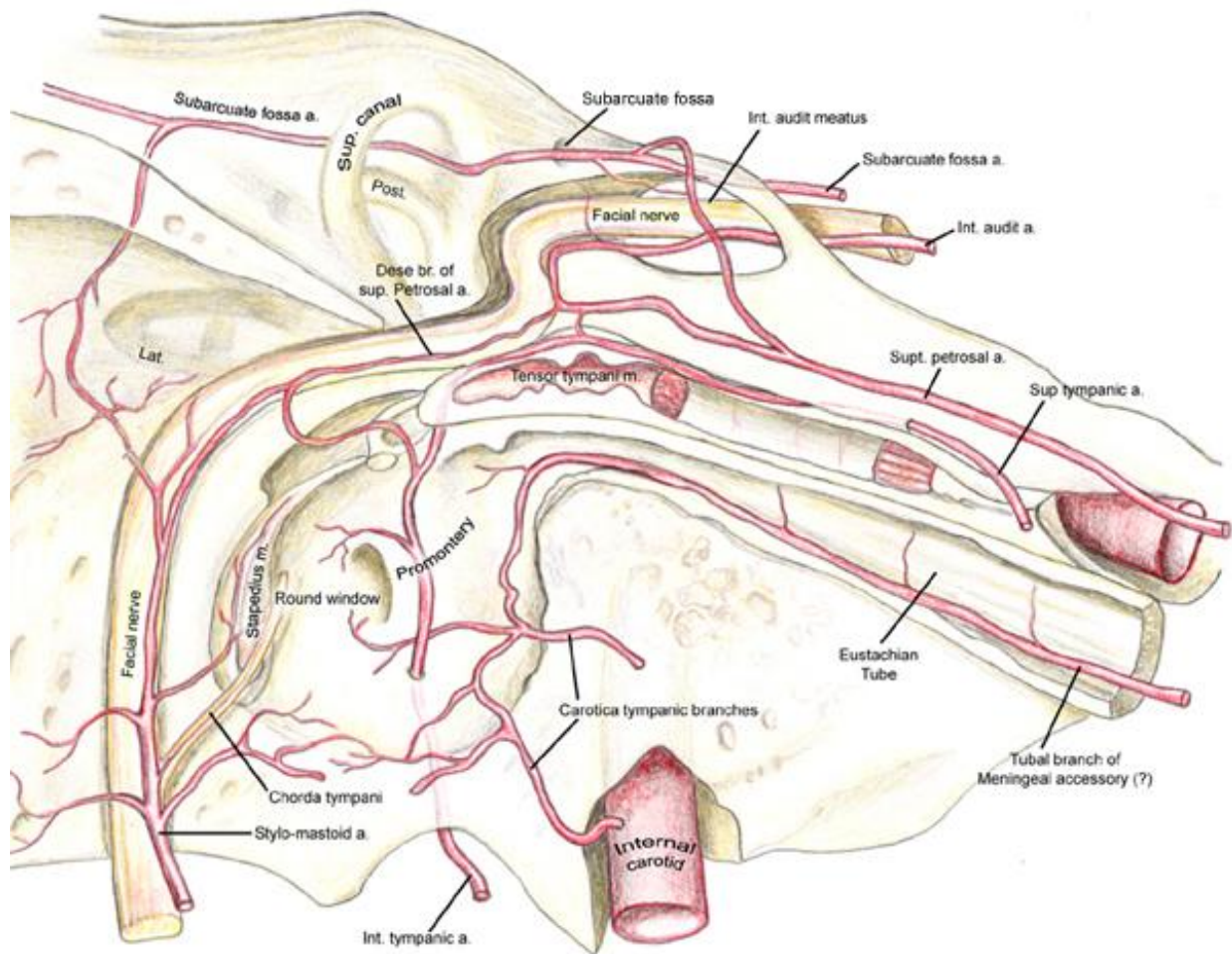
- Facial nerve travels through Petrous part of Temporal bone in a bony canal called Fallopian canal.

- o Porus Acousticus (Medial opening of IAM) → Stylomastoid Foramen.
- o Inflammatory processes involving the central nervous system (CNS) and Facial nerve or traumatic injuries to Temporal bone can produce unique complications.

- **Divides into 4 segments:**

1. Meatal Segment.
2. Labrinthine Segment.
3. Tympanic (Horizontal) Segment.
4. Mastoid (Vertical) Segment.





1. Meatal Segment:

- Porus Acusticus → IAM Fundus
- Within IAM.
- 8-10 mm in length.
- Facial Nerve runs into Superior-Anterior Compartment.
- Separated by Falciform crest inferiorly and Bill's bar Posteriorly.
- Meatal segment is ensheathed within an extension of Meninges.

2. Labyrinthine Segment:

- **IAM Fundus → Geniculate Ganglion**
- 4 mm in Length.
- 0.61-0.68 mm in Diameter.
- Narrowest and Shortest segment of Fallopian canal.
 - Susceptible to compression by Edema.
- Only segment that lacks Anastomosing Arterial cascades.
 - Susceptible to embolic phenomena, low-flow states, and vascular compression.
- Location:
 - Directed obliquely forward.
 - Perpendicular to axis of Temporal bone.
 - Immediately posterior to Cochlea.
 - Posterolateral to ampullated ends of Horizontal and Superior SCC.
 - Rests on Anterior part of Vestibule.
- Facial nerve takes a turn Posteriorly forming a Genu (1st Genu).

- Geniculate Ganglion:

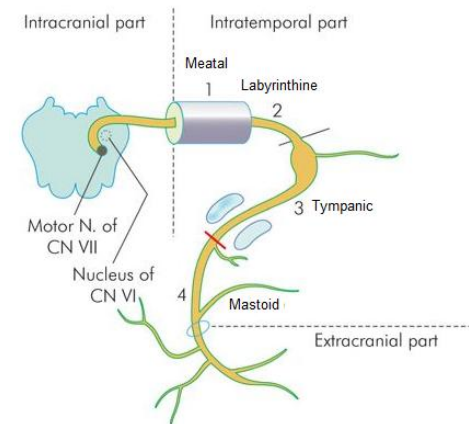
- Located at 1st Genu.
- Formed by juncture of Facial nerve and Nervus intermedius into a common trunk.
- Houses cell bodies of Sensory and Taste cells from Anterior 2/3 of tongue and palate.
- Gives Three branches:

1. Greater Superficial Petrosal Nerve:

- 1st branch of Facial nerve.
- Exits Petrous temporal bone via Greater Petrosal Foramen.
- Enters Middle cranial fossa.
- Passes deep to Trigeminal ganglion.
- Travels through Foramen lacerum to Pterygoid Canal.
- Joins Deep petrosal nerve.
- Become Nerve of Pterygoid canal (Vidian nerve).
- Synapse in Pterygopalatine ganglion.
- Postganglionic parasympathetic fibers Carried via branches of Maxillary (V2) divisions of Trigeminal nerve (CN V).
- Innervate Lacrimal gland and Mucus glands of Nasal and Oral cavities.

2. External Petrosal Nerve:

- Inconstant branch.
- Carries sympathetic fibers to Middle meningeal Artery.



3. Lesser Petrosal Nerve:

- Carries secretory fibers to Parotid gland.
- Carries Parasympathetic contributions from the Tympanic plexus (from CN-IX) and Nervus intermedius.

3. Tympanic (Horizontal) Segment:

- **Geniculate Ganglion → Second Genu.**
- 11.0 mm in length.
- Most common site of Dehiscence in Fallopian canal (**25-55%**)
- Located in Middle ear:
 - Inferior to Horizontal SCC.
 - Posterior to Cochleariform process.
 - Above and Posterior to Oval window and Stapes.
 - Passes distal to Pyramidal eminence, where Facial nerve makes a Second turn (2nd Genu).

4. Mastoid (Vertical) Segment:

- **Second Genu → Stylomastoid Foramen.**
- 13.0 mm in length.
- Longest part of Intra-Temporal course of Facial nerve.
- Most common Facial nerve segment injured in Middle ear surgery is at Pyramidal turn.
- Pass vertically from 2nd genu (Lateral and posterior to Pyramidal) down anterior wall of Mastoid process to Stylomastoid foramen. During middle ear surgery, the facial nerve is most commonly injured at the pyramidal turn.
- Gives 3 branches:

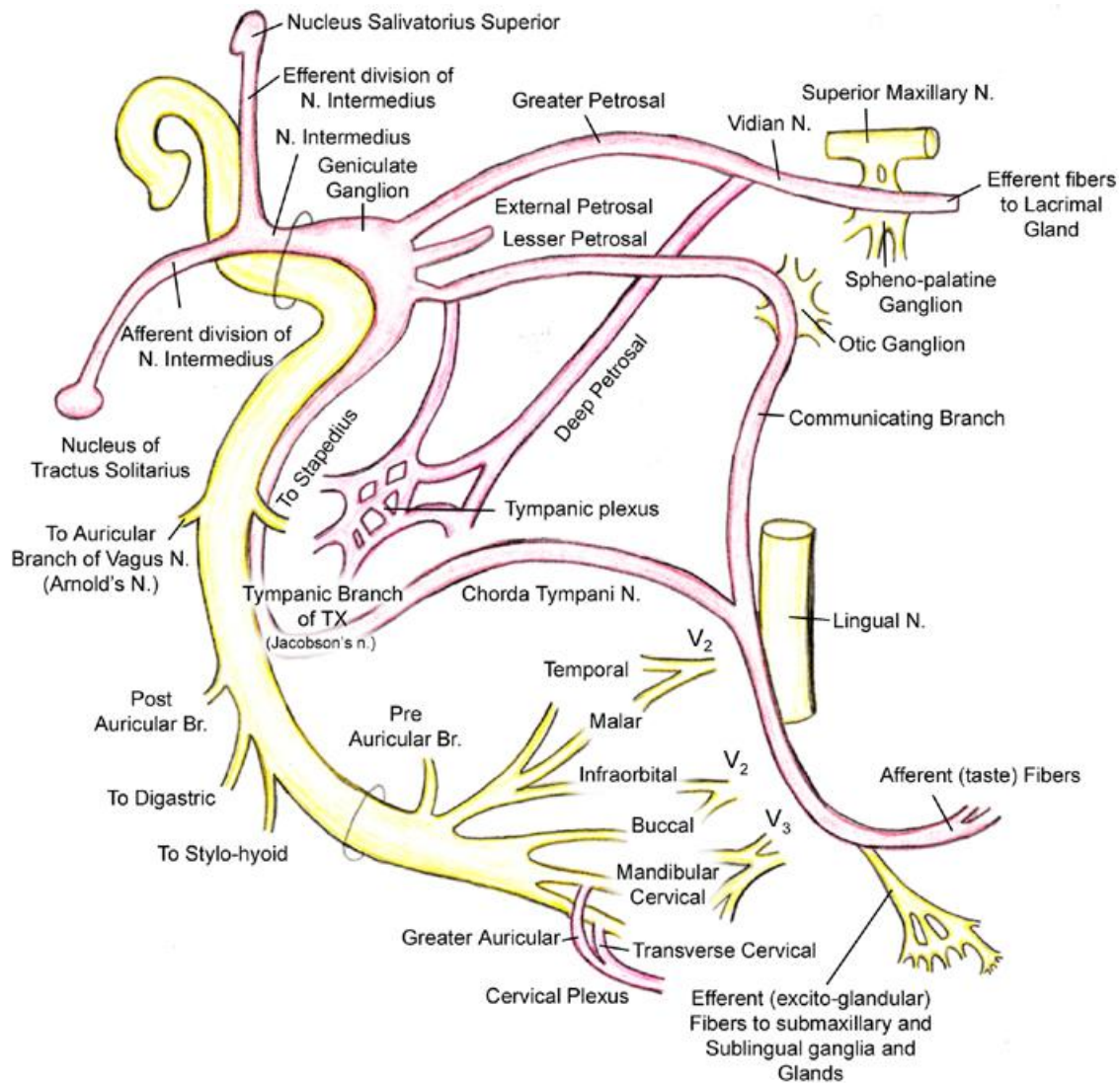
1. Nerve to Stapedius muscle.

2. Communicating Branch:

- Joins Auricular branch of Vagus nerve (Arnold's nerve) just distal to Nerve to Stapedius muscle.
- Carries Sensory fibers from Concha, Retroauricular groove and Posterior wall of EAC.

3. Chorda tympani nerve:

- Terminal branch of Nervus intermedius.
- Runs laterally in the middle ear.
- Between Incus and Handle of Malleus.
- Crosses Middle ear cavity.
- Exits through Petrotympanic Fissure (Canal of Huguier).
- Joins Lingual nerve.
- Carries:
 - Preganglionic secretomotor fibers to Submandibular and Sublingual glands.
 - Taste fibers from Anterior 2/3 of Tongue.



- **Extra-Temporal Part of CN-VII:**

- Facial nerve exits Fallopian canal via Stylomastoid foramen.
- Travels between Digastric and Stylohyoid muscles.
- Lateral to Styloid process.

- Branches just below Stylomastoid foramen:

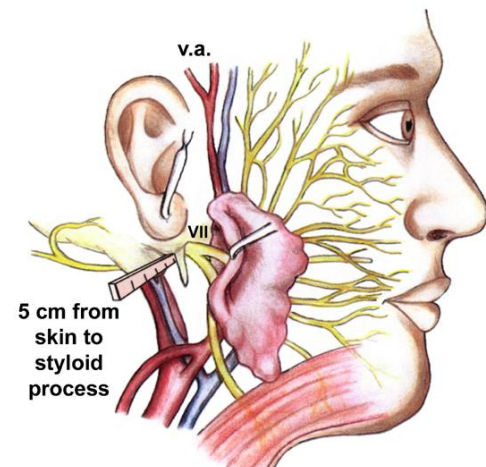
1. Posterior Auricular Nerve:

- Posterior Auricular Muscle.
- Occipital belly of Occipitofrontalis Muscle.

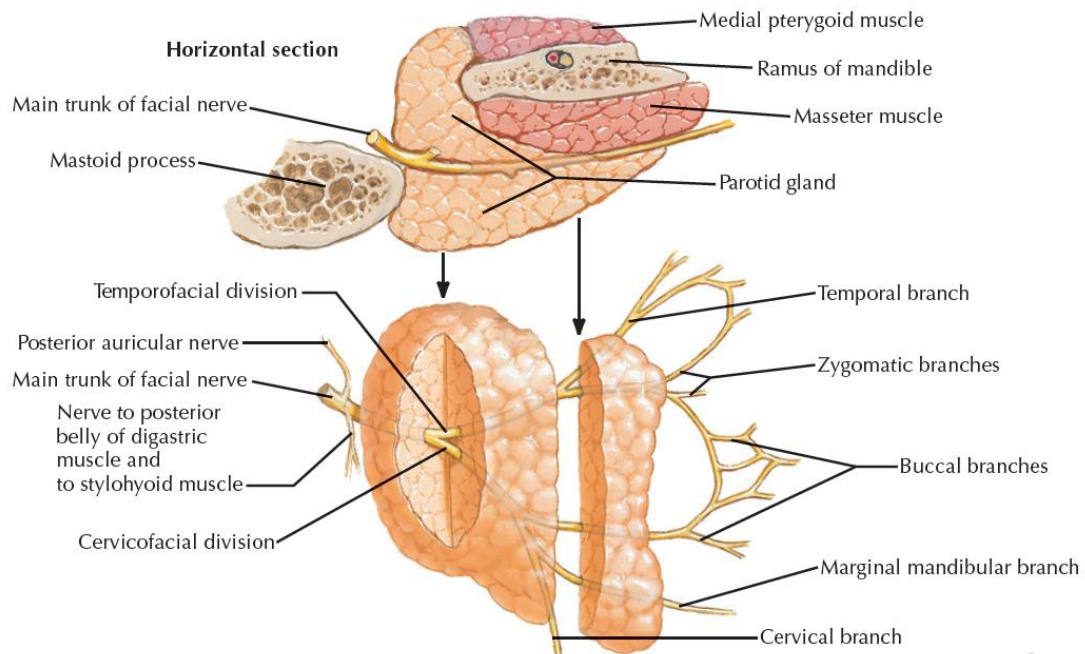
2. Nerve to Stylohyoid.

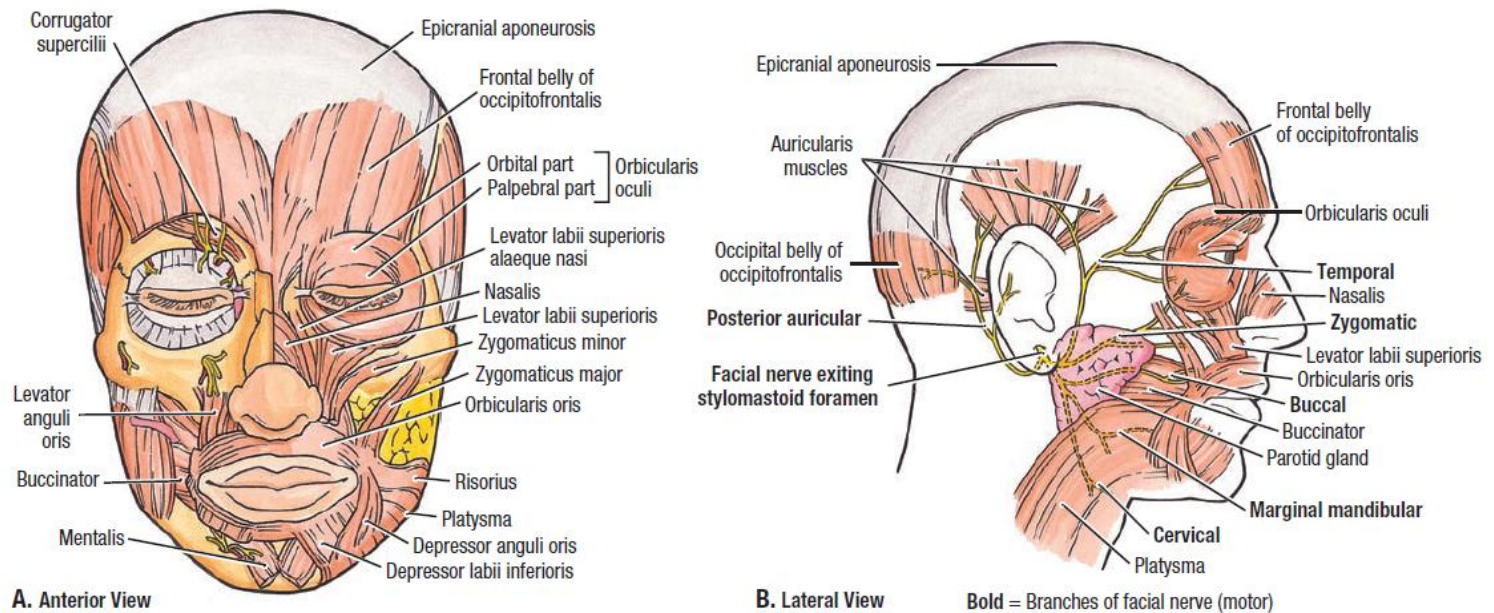
3. Nerve to Posterior Digastric.

- Facial nerve then penetrates Parotid gland.
- Lies in a fibrous plane that separates Deep and Superficial lobes of Parotid gland.



- Divides at Pes Anserinus in Parotid gland into:
 - 1. Temporal-Facial Division.**
 - 2. Cervico-Facial Division.**
- Followed by 5 Terminal branches:
 - 1. Temporal Branch:**
 - Frontalis.
 - Corrugator supercillii.
 - Procerus.
 - Upper orbicularis oculi.
 - 2. Zygomatic Branch:**
 - Lower orbicularis oculi.
 - Abundant Anastomotic supply Buccal branch.
 - 3. Buccal Branch:**
 - Zygomaticus Major and Minor muscles.
 - Levator Anguli oris.
 - Buccinator.
 - Upper orbicularis oris.
 - Abundant Anastomotic supply with Zygomatic branch.
 - 4. Marginal Mandibular Branch:**
 - Lower orbicularis oris.
 - Depressor Anguli oris
 - Depressor labii inferioris
 - Mentalis.
 - 5. Cervical Branch:**
 - Platysma.



**TABLE 7.2 MAIN MUSCLES OF FACIAL EXPRESSION^a**

Muscle ^a	Origin	Insertion	Action
Occipitofrontalis, frontal belly	Epicranial aponeurosis	Skin of and subcutaneous tissue of eyebrows and forehead	Elevates eyebrows and wrinkles skin of forehead; protracts scalp (indicating surprise or curiosity)
Occipitofrontalis, occipital belly	Lateral two-thirds of superior nuchal line	Epicranial aponeurosis	Retracts scalp; increasing effectiveness of frontal belly
Orbicularis oculi	Medial orbital margin, medial palpebral ligament; lacrimal bone	Skin around margin of orbit; superior and inferior tarsal plates	Closes eyelids; palpebral part does so gently; orbital part tightly (winking)
Orbicularis oris	Medial maxilla and mandible; deep surface of perioral skin; angle of mouth (modiolus)	Mucous membrane of lips	Tonus closes oral fissure; phasic contraction compresses and protrudes lips (kissing) or resists distension (when blowing)
Levator labii superioris	Infra-orbital margin (maxilla)	Skin of upper lip	Part of dilators of mouth; retract (elevate) and/or evert upper lip; deepen nasolabial sulcus (showing sadness)
Zygomaticus minor	Anterior aspect, zygomatic bone		
Buccinator	Mandible, alveolar processes of maxilla and mandible; pterygomandibular raphe	Angle of mouth (modiolus); orbicularis oris	Presses cheek against molar teeth; works with tongue to keep food between occlusal surfaces and out of oral vestibule; resists distension (when blowing)
Zygomaticus major	Lateral aspect of zygomatic bone	Angle of mouth (modiolus)	Part of dilators of mouth; elevate labial commissure—bilaterally to smile (happiness); unilaterally to sneer (disdain)
Risorius	Parotid fascia and buccal skin (highly variable)		Part of dilators of mouth; widens oral fissure
Platysma	Subcutaneous tissue of infraclavicular and supraclavicular regions	Base of mandible; skin of cheek and lower lip; angle of mouth (modiolus); orbicularis oris	Depresses mandible (against resistance); tenses skin of inferior face and neck (conveying tension and stress)

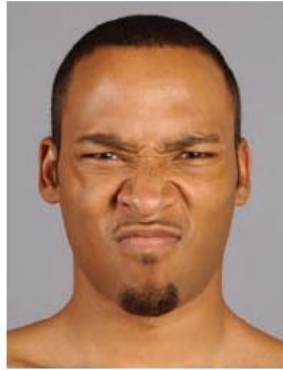
^aAll of these muscles are supplied by the facial nerve (CN VII).



Occipitofrontalis



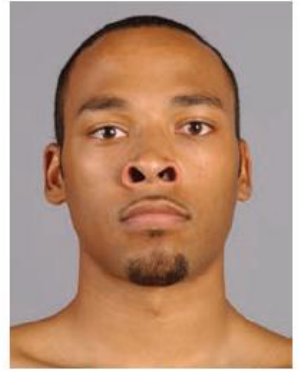
Corrugator supercilii



Procerus + transverse part of nasalis



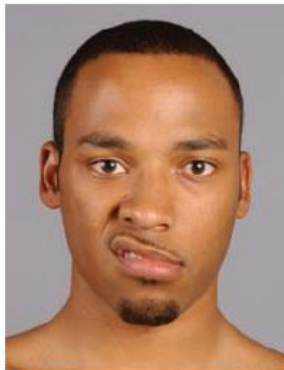
Orbicularis oculi



Lev. labii sup. alaeque nasi +
alar part of nasalis



Buccinator + orbicularis oris



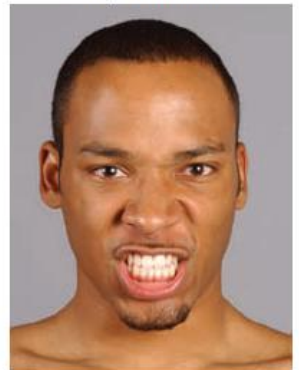
Zygomaticus major + minor



Risorius



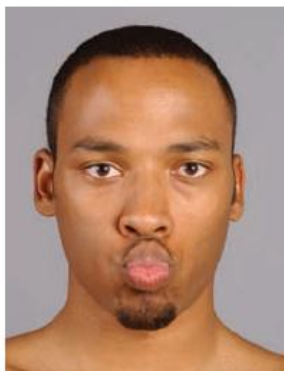
Risorius + depressor labii inferioris



Levator labii sup. + depressor labii



Dilators of mouth:
Risorius plus levator labii superioris
+ depressor labii inferioris



Orbicularis oris



Depressor anguli oris

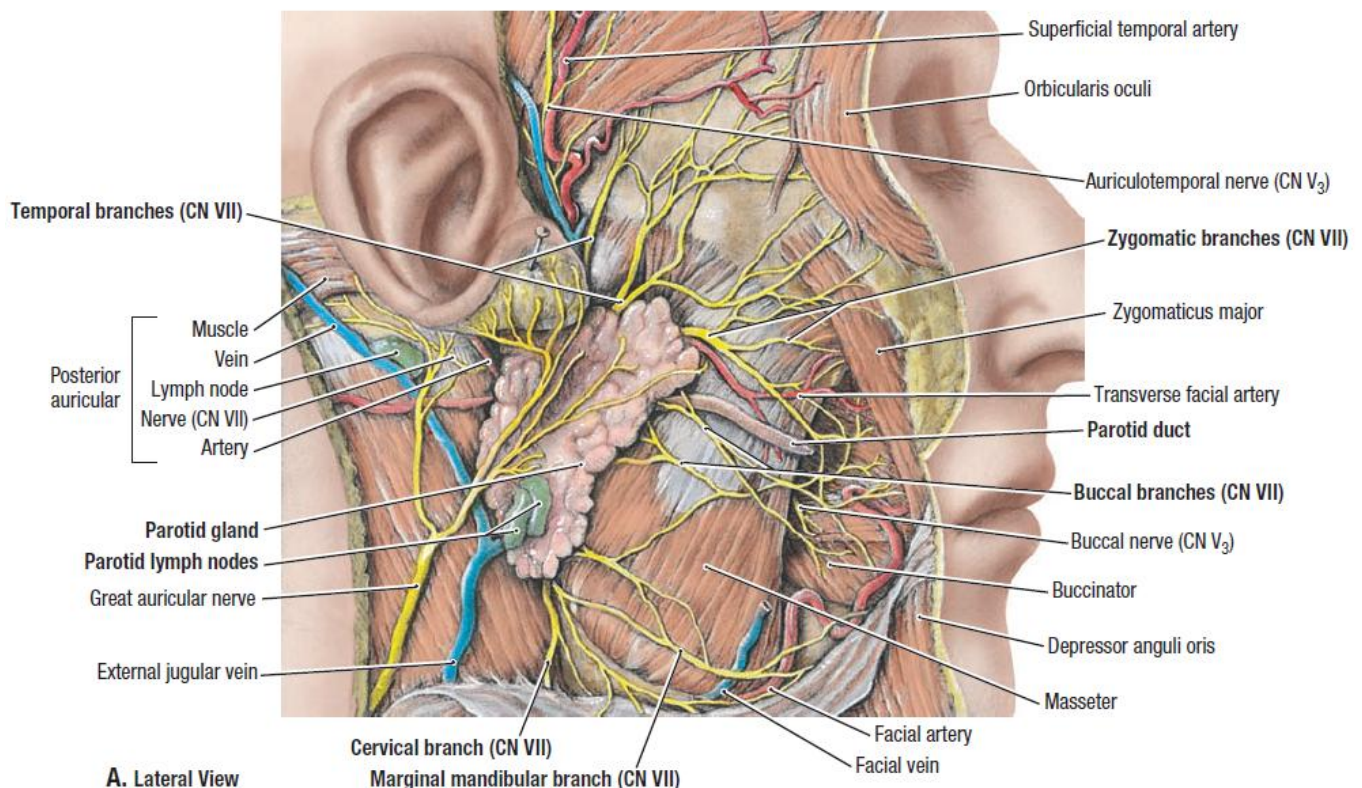


Mentalis



Platysma

- Temporal and Marginal Mandibular branches are at highest risk during surgical procedures and are usually terminal connections without anastomotic connections.
- **Temporal branch:**
 - Exits the parotid gland and runs within the SMAS over Zygomatic arch into Temple region.
 - Enters undersurface of Frontalis muscle and lies superficial to Deep Temporalis fascia.
 - Can be roughly located along a line extending from attachment of Lobule (5 mm below Tragus), Anterior and Superior to a point 1.5 cm above lateral aspect of ipsilateral eyebrow
 - To avoid injury to Temporal branch during elevation of Facial flaps, Surgeon should elevate either in a subcutaneous plane or deep to SMAS.
- **Marginal Mandibular Branch:**
 - Lies along Body of the mandible (80%) or within 1-2cm below (20%).
 - Lies deep to Platysma throughout much of its course.
 - More superficial approximately 2 cm lateral to Corner of the mouth and ends on undersurface of the muscles.
 - Injury to Marginal branch results in paralysis of Muscles that depress Corner of the mouth.



- **Superficial MusculoAponeurotic System (SMAS):**
- Superficial Fascial layer extends throughout Cervical facial region.
- In Scalp:
 - The equivalent of SMAS is Galea Aponeurotica.
 - Splits to ensheath:
 - Frontalis
 - Occipitalis
 - Procerus
 - Postauricular muscle
- in Upper Face:
 - SMAS ends at level of Zygoma because of attachments of fascial layers to Zygomatic arch.
 - Neurovascular structures exit their bony foramina and penetrate SMAS to run within its superficial aspects or on its surface.
- In Middle Face:
 - Facial nerve (**CN-VII**) runs on top of Masseter muscle just below the SMAS.
- In Lower Face:
 - SMAS invests Facial muscles.
 - Continuous with Platysma muscle.
 - Facial nerve (**CN-VII**) runs on top of Masseter, deep to Platysma and SMAS and innervates the muscles on their undersurfaces.
 - Except for:
 1. Buccinator
 2. Levator Anguli Oris
 3. Mentalis
- Temporoparietal Fascia:
 - Not continuous with SMAS.
 - Extends from Zygomatic arch as extension of Deep Temporal Fascia.
 - In Temporal region:
 - Temporal branch of Facial nerve crosses Zygomatic arch and courses within Superficial layer of Deep temporal fascia (Temporoparietal Fascia).

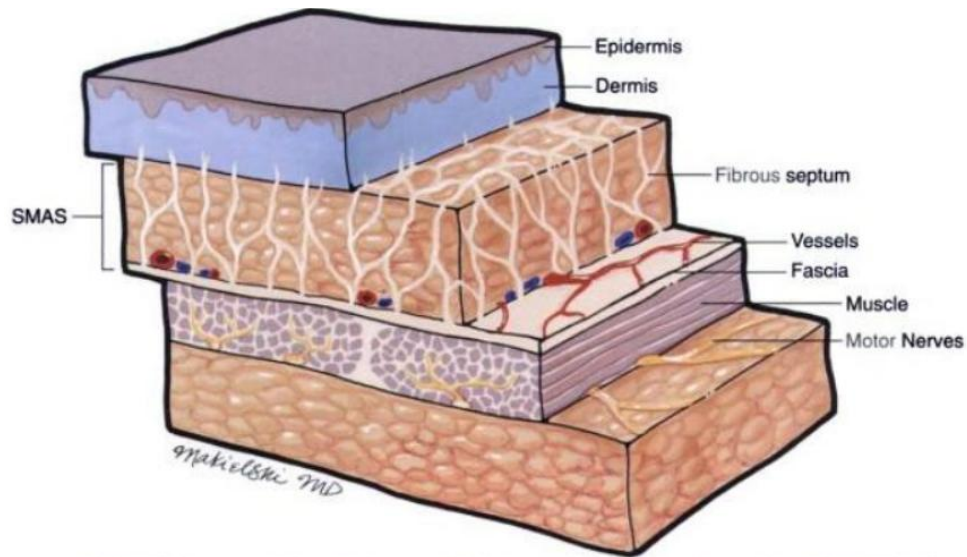


FIG. 6.1. Cross section of the superficial musculoaponeurotic system (SMAS) in the lower face. The SMAS envelops the facial musculature in a fibrous sheath of varying degrees of thickness and thus connects the muscles for coordination of facial expression. Fibrous septae to the dermis create a meshwork connecting the skin to the underlying muscles of expression. Motor nerves enter the deep surface of the muscles. Sensory nerves and vessels originate deep to the fascia, but their terminal branches run within or superficial to it.

- **Blood Supply of Facial Nerve (CN-VII):**
- In Pons:
 - Anterior Inferior Cerebellar Artery (**AICA**).
- In IAM:
 - Labyrinthine Artery.
- Geniculate ganglion:
 - Superficial petrosal Artery.
- Mastoid Segment:
 - Stylomastoid Artery.
- Extra-Temporal part:
 - Posterior Auricular Artery.
- Venous drainage parallels the arterial blood supply.

- Surgical Landmarks of Facial Nerve in Ear and Mastoid Surgery:

1. Processus Cochleariformis:

- Geniculate ganglion (1st Genu) lies Anterior to it.
- Tympanic segment of CN-VII starts at this level.

2. Horizontal SCC:

- Tympanic segment of CN-VII runs below Horizontal SCC.
- 2nd Genu of Facial nerve runs Infero-lateral to Horizontal SCC.

3. Oval Window:

- Tympanic segment of CN-VII runs above Oval window (Stapes).

4. Short Process of Incus:

- Tympanic segment of CN-VII lies medial to Short process of Incus at level of Aditus.

5. Pyramid:

- Mastoid segment of CN-VII runs behind Pyramid and Posterior Tympanic sulcus.

6. Facial Recess:

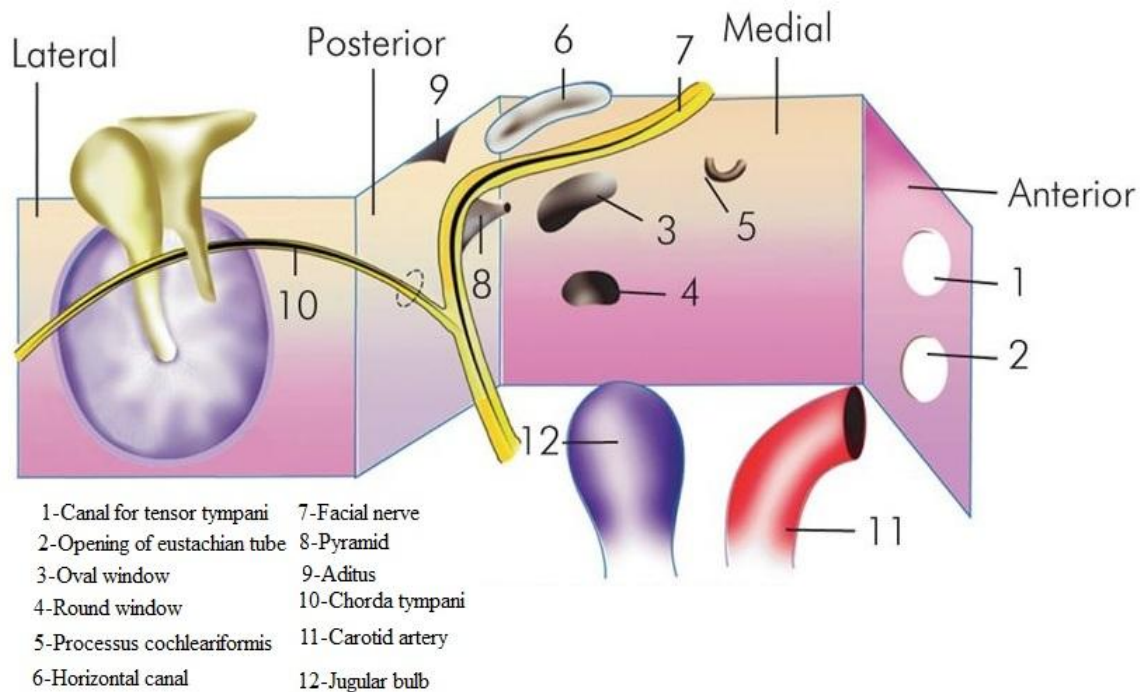
- Long process of Incus points toward Facial recess.
- Chorda tympani nerve serves at Lateral margin of Triangular facial recess.
- Chorda tympani nerve can be exposed along its length and can be followed inferiorly and medially to its takeoff from the main trunk of Facial nerve.

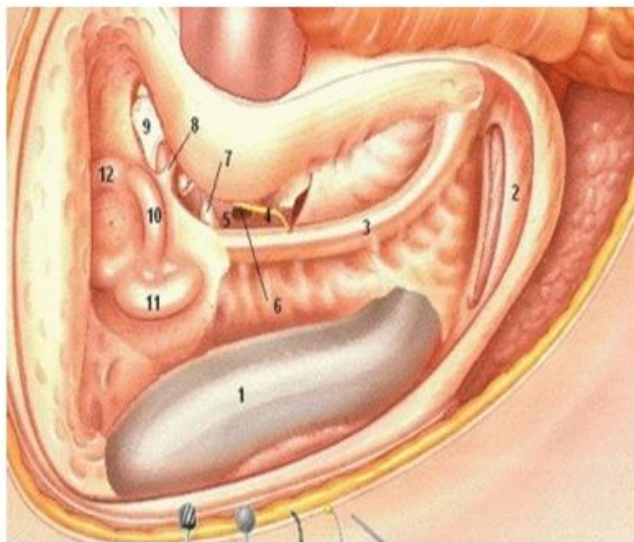
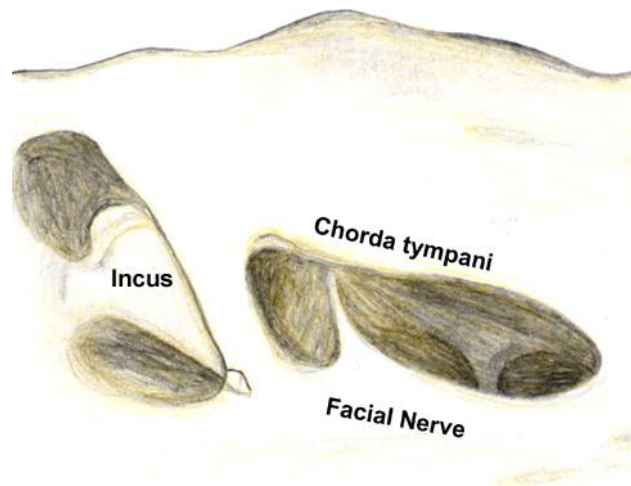
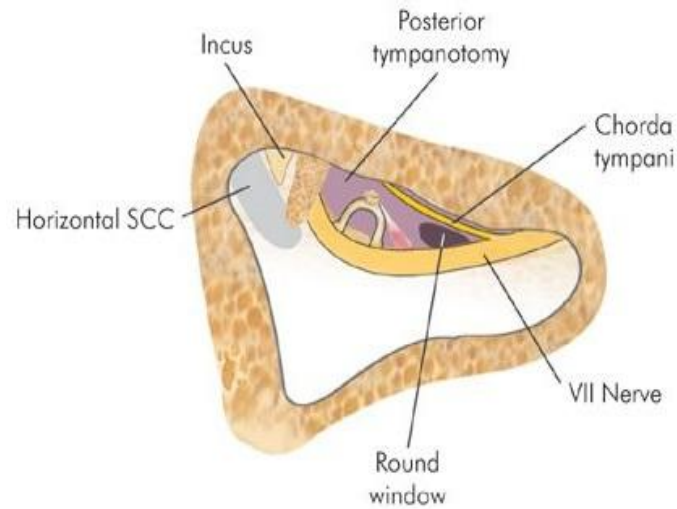
7. TympanoMastoid suture:

- Mastoid segment of CN-VII runs behind this suture.

8. Digastric Ridge:

- Mastoid segment of CN-VII leaves Mastoid at Anterior end of Digastric Ridge.





1. Sigmoid sinus
2. Digastric ridge and posterior belly of digastric muscle
3. Facial nerve (skeletonized mastoid segment)
4. Chorda tympani
5. Facial recess
 - ◊ triangle bound by: chorda tympani, facial nerve and incus buttress
6. Round window
 - ◊ opens into cochlear scala tympani
7. Pyramidal eminence containing stapedius muscle
8. Incus buttress
9. Incus body
10. Horizontal semicircular canal
11. Posterior semicircular canal
12. Superior semicircular canal

- **Surgical Landmarks of Facial Nerve in Parotid Surgery:**

1. Cartilaginous Pointer:

- Sharp triangular piece of cartilage of Pinna points to Facial Nerve.
- Extra-Temporal part of Facial Nerve lies 1 cm inferior and medial to the pointer.

2. Tympanomastoid Suture:

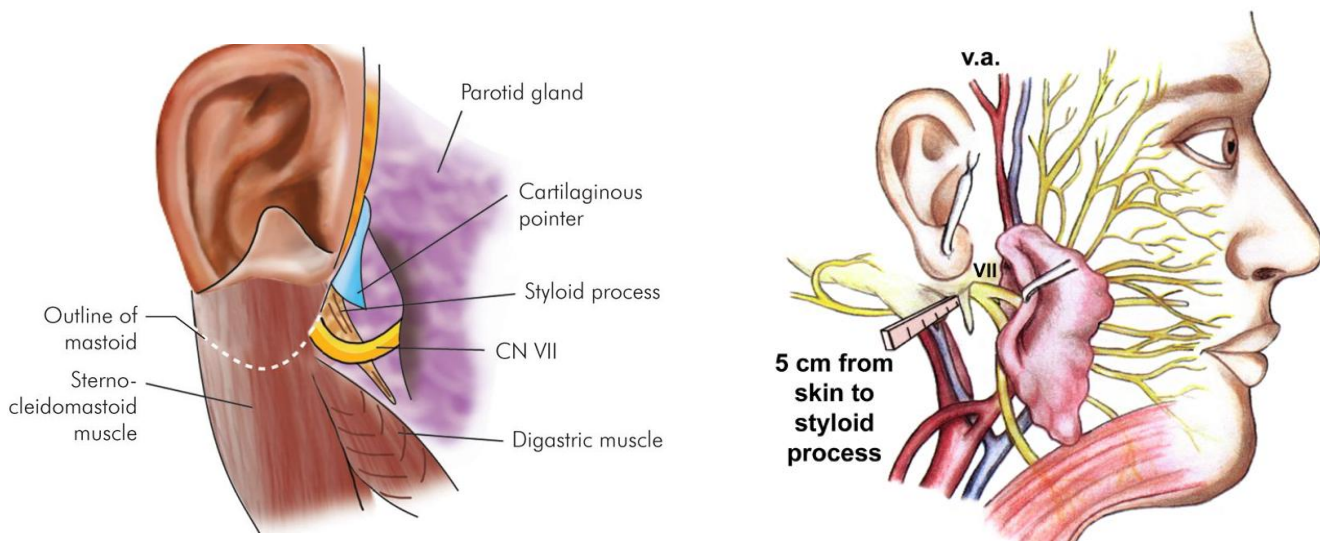
- Between Mastoid and Tympanic part of Temporal bone.
- Facial Nerve lies 6-8 mm deep to this suture in Stylomastoid foramen.

3. Styloid Process.

- Facial Nerve crosses lateral to Styloid process.

4. Posterior belly of Digastric.

- Facial Nerve lies between attachment of Posterior belly of digastric muscle to Digastric groove and Styloid process.



- In Adults, Line drawn between Mastoid tip and Angle of Mandible:

- Superior limits of a Neck dissection.
- Removal of Parotid inferior to this line is safe.

- **In infants and young children:**

- These landmarks are not applicable.
- Differences in rate of anatomic development of Parotid and Mastoid.
- Modified Blair incision most commonly used in adults is avoided in children.
- Facial nerve is located more superficially.
- Risk of Facial nerve injury is increased with elevation of the skin flaps.

Anatomy of Pharynx:

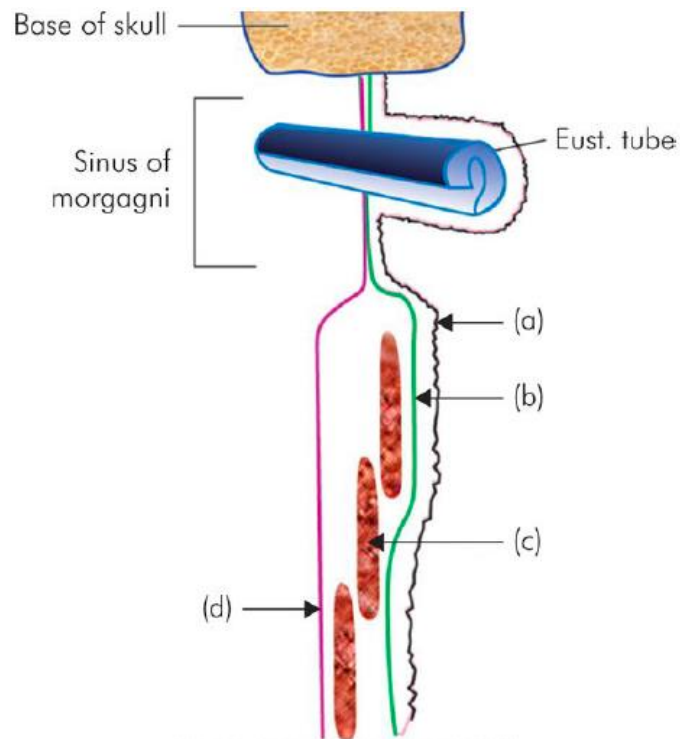
- A conical fibromuscular tube forming upper part of the air and food passages.
- It is 12-14 cm (5in) long, extending from base of the skull (basiocciput and basisphenoid) to the lower border of cricoid cartilage where it becomes continuous with the oesophagus (6th cervical vertebra).
- The width of pharynx is 3.5 cm at its base and this narrows to 1.5 cm at pharyngo-oesophageal junction, which is the narrowest part of digestive tract apart from the appendix.



Structure of Pharyngeal Wall:

1. Mucous membrane (A)
2. Pharyngeal aponeurosis/Pharyngobasilar fascia (B)
3. Muscular coat (C)
4. Buccopharyngeal fascia (D)

(Some will add submucosa layer between mucosa and pharyngeal aponeurosis)



1. **Mucous membrane:**

- Continuous with mucous membrane of eustachian tubes, nasal cavities, mouth, larynx and oesophagus.
- The epithelium is ciliated columnar in the nasopharynx and stratified squamous elsewhere.
- There are numerous mucous glands

2. **Pharyngeal aponeurosis (Pharyngobasilar fascia):**

- A fibrous layer which lines the muscular coat
- Thick near the base of skull but is thin and indistinct inferiorly.
- It fills up the gap left in the muscular coat near the base of skull.

3. Muscular coat

Two layers of muscles with three muscles in each layer.

(A) External layer:

These muscles constrict the pharynx during swallowing.

Contains:

- Superior constrictor (Inner most).
- Middle constrictor.
- Inferior constrictor (Outer most).

NAME	ORIGIN	INSERTION	ACTION	NERVE
Superior constrictor	Medial pterygoid plate, pterygoid hamulus, pterygoamandibular raphe, mylohyoid line	Pharyngeal tubercle, pharyngeal raphe	Constricts the pharynx	Vagus nerve through the pharyngeal plexus
Middle constrictor	Greater & lesser horns of hyoid bone & Stylohyoid ligament	pharyngeal raphe	Constricts the pharynx	Vagus nerve through the pharyngeal plexus
Inferior constrictor	Arch of cricoid cartilage & oblique line of thyroid cartilage	pharyngeal raphe	Constricts the pharynx	Vagus nerve through the pharyngeal plexus

(B) Internal layer (Longitudinal muscles):

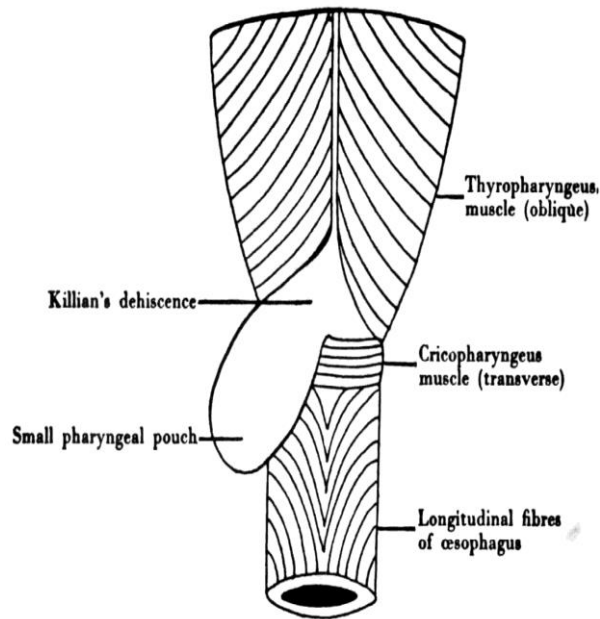
Contains:

- Stylopharyngeus (CN IX).
- Salpingopharyngeus (CN X).
- Palatopharyngeus (CN X).

NAME	ORIGIN	INSERTION	ACTION	NERVE
Salpingopharyngeus	Cartilage of auditory tube	Lateral wall of pharynx	Elevate the larynx in swallowing And the pharynx	Vagus nerve through the pharyngeal plexus
Palatopharyngeus	Posterior border of the hard palate & palatine aponeurosis	Thyroid cartilage, lateral wall of pharynx	Elevate the pharynx in swallowing Pulls the palatopharyngeal fold medially	Vagus nerve through the pharyngeal plexus
Stylopharyngeus	Styloid process	Thyroid cartilage	Elevate the pharynx in swallowing	Glossopharyngeal nerve

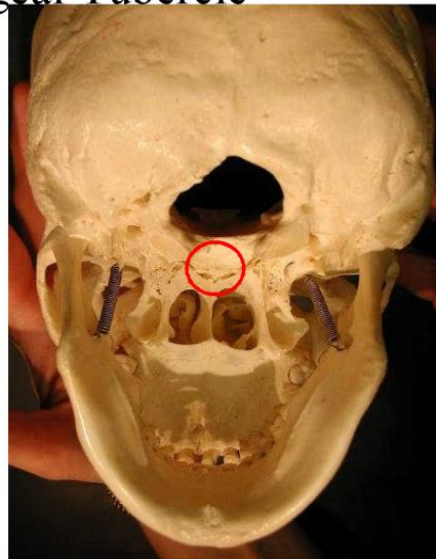
Inferior Constrictor is divided into 2 parts:

- Thyro-pharyngeus (Oblique)
- Crico-pharyngeus (Transverse)
- Between them is KILLIAN DEHISCENCE (Site for Pharyngeal pouch).



The Pharyngeal Tubercle

- Located on cranial base anterior to foramen magnum
- On occipital bone
- The **Pharyngeal Raphe** descends from the pharyngeal tubercle



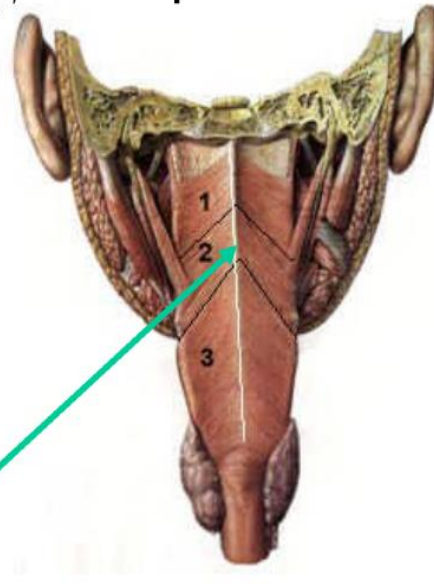
The Pharynx

- Runs through C6
- Located anterior to bodies of cervical vertebrae C1-C6

http://biology.clc.uc.edu/fankhauser/Labs/Anatomy/_&_Physiology/A&P201/Skeletal/skull_from_belowPB012152up.JPG

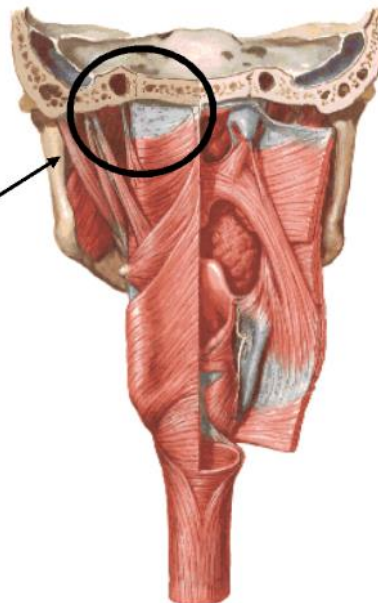
The Pharyngeal Raphe

- Descends from the pharyngeal tubercle
- Serves as the continuous attachment of the external group of pharyngeal muscles.



There are gaps between the constrictors through which structures enter/exit the pharynx

The Pharyngobasilar Fascia – Superior to the superior constrictor closes gap between superior constrictor and base of skull

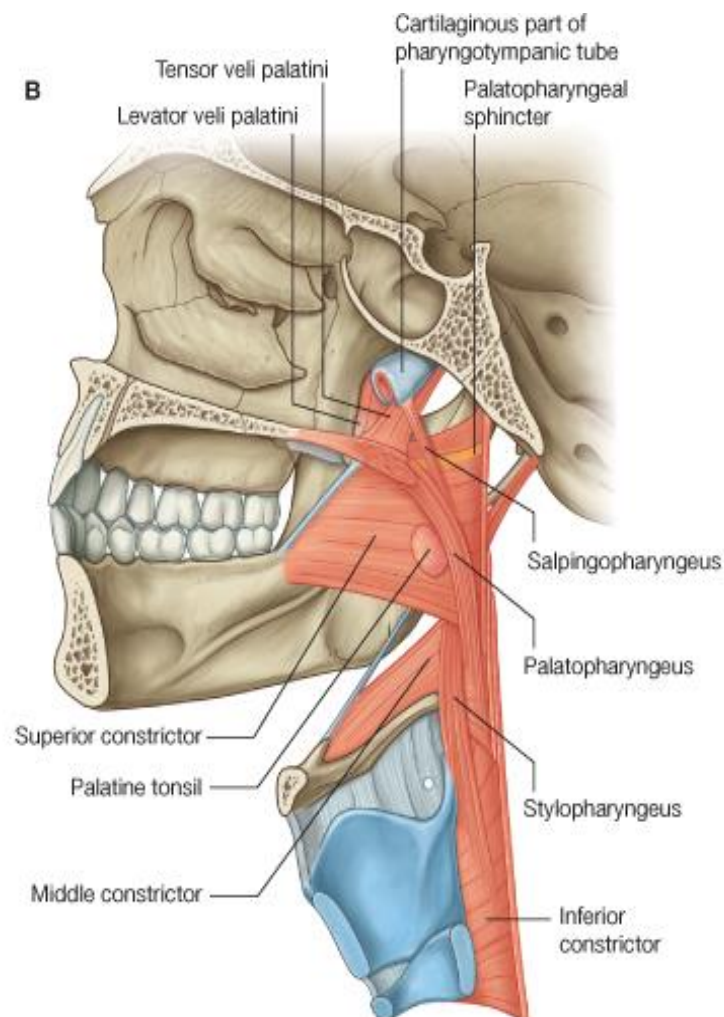


Netter, Frank H., *Atlas of Human Anatomy*. Ciba-Geigy Corporation, Summit, N.J. 1993. Plate 61.

Gap #1: (Sinus of Morgagni):

Gap between the superior constrictor and base of the skull:

- Eustachian tube
- Tensor veli palatini m.
- Levator veli palatini m.
- Ascending palatine artery (branch from facial a.)
- ? Ascending pharyngeal a.



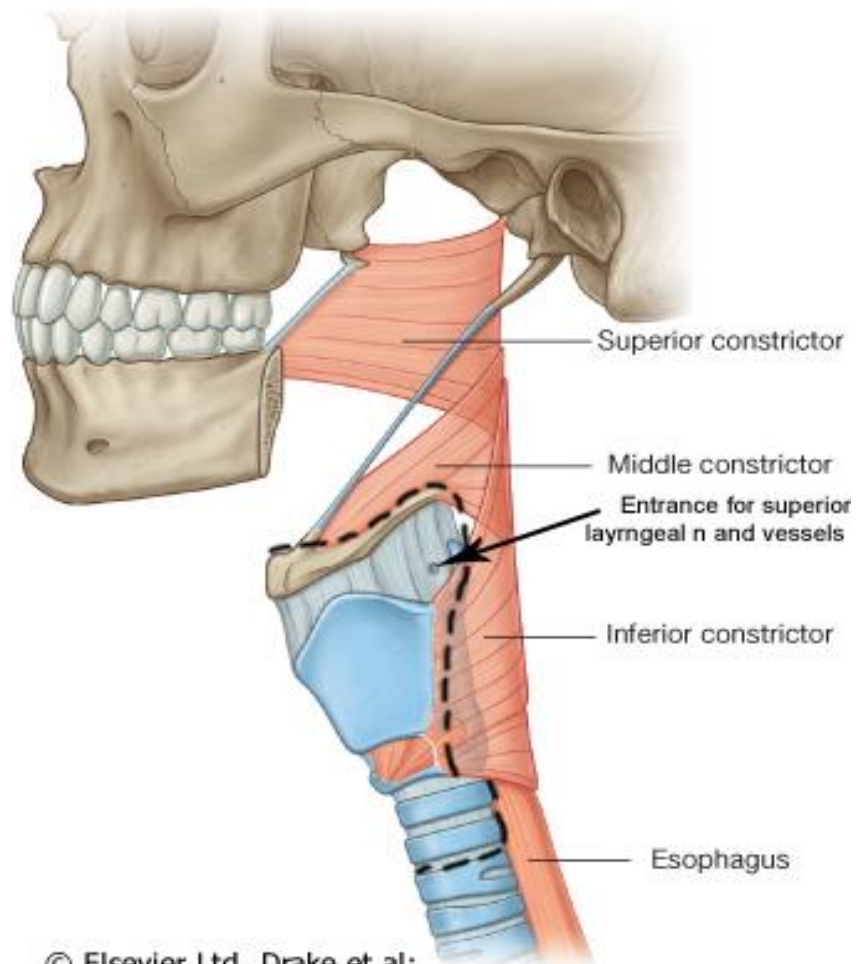
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Gap # 2:

Gap between superior and middle constrictor muscle:

- Stylopharyngeous muscle.
- Glossopharyngeal nerve
- Styloglossus muscle.
- ? Stylohyoid ligament
- ? Tonsillar branch of ascending palatine

Suprahyoid and Piriformis Recess Incisions



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Gap # 3:

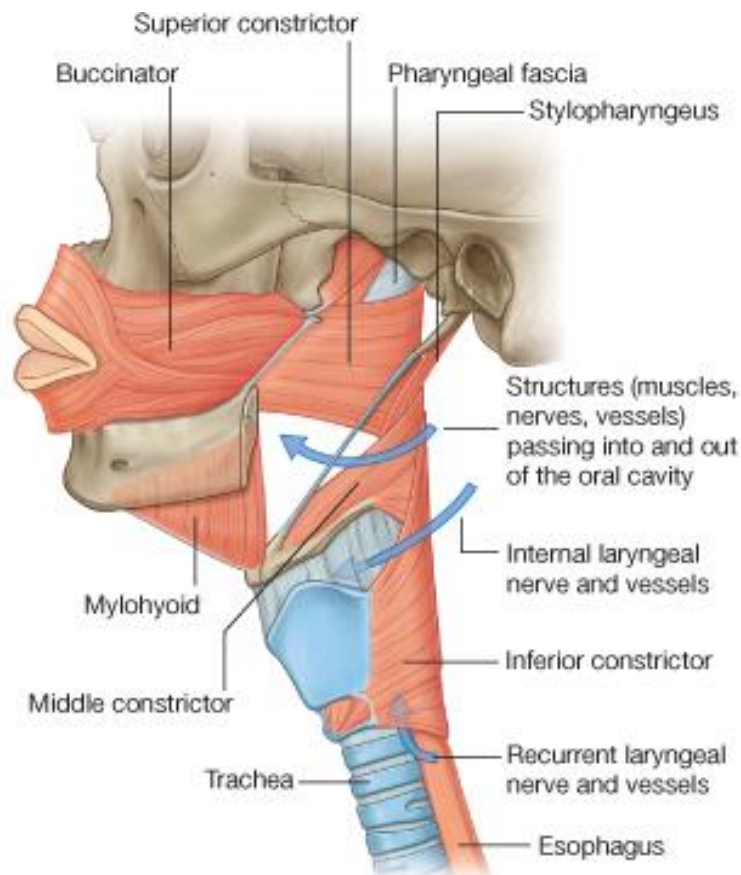
Gap between middle and inferior (thyrohyoid membrane):

- Superior laryngeal vessels
- Internal laryngeal nerve.

Gap # 4:

Gap inferior to inferior constrictor:

- Recurrent laryngeal N
- Inferior laryngeal vessels

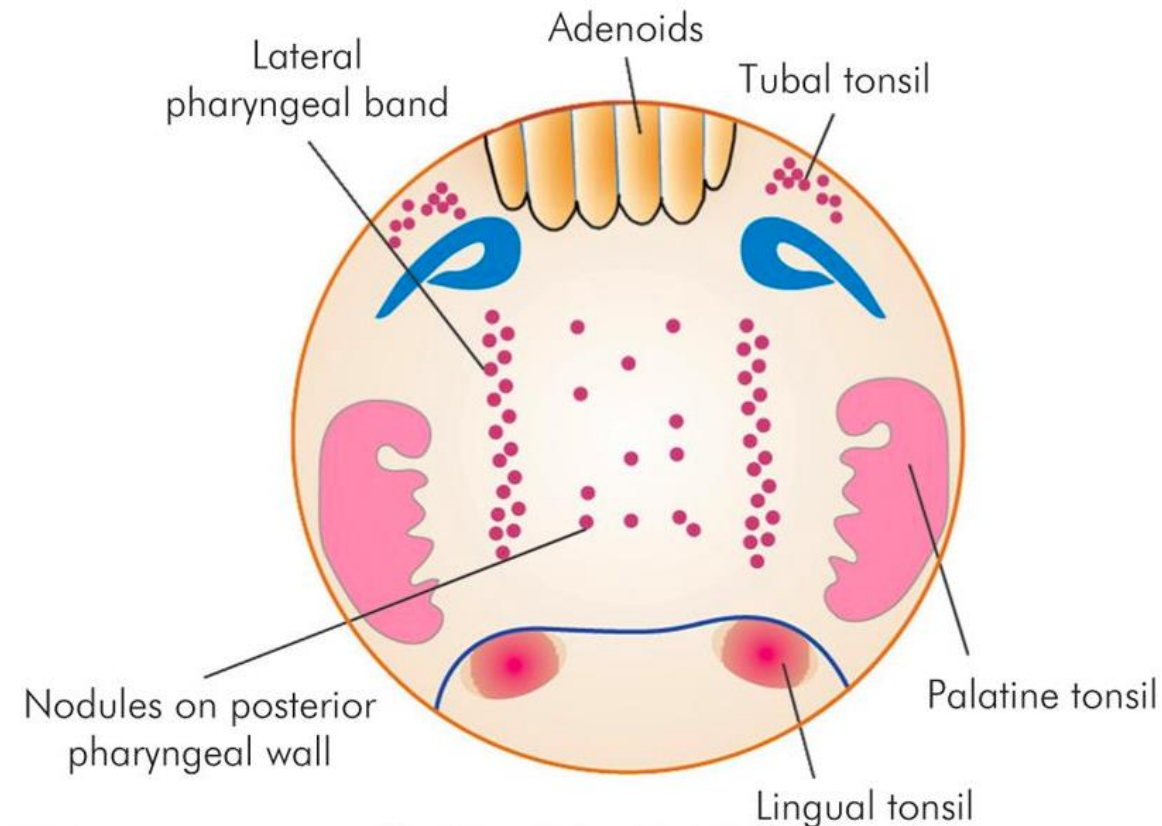


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Waldeyer's Ring:

- Scattered throughout the pharynx in its subepithelial layer
- Lymphoid tissue which is aggregated at places to form masses
- (Differ from other body lymph nodes that they have only efferent pathway rather than efferent and afferent, and they are B cell predominant rather than T cell predominant, localized in around pharynx area rather than scattered all over the body)

1. Nasopharyngeal tonsil or the adenoids
2. Palatine tonsils or simply the tonsils
3. Lingual tonsil
4. Tubal tonsils (in fossa of Rosenmuller)
5. Lateral pharyngeal bands
6. Nodules (in posterior pharyngeal wall).

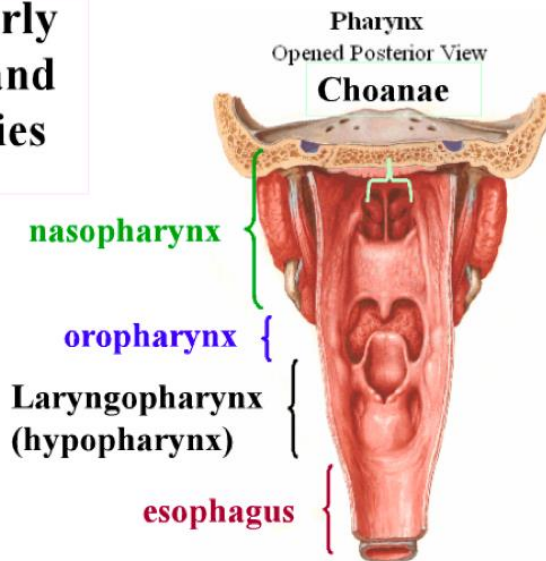


Pharyngeal Spaces: (See Neck spaces for more details):

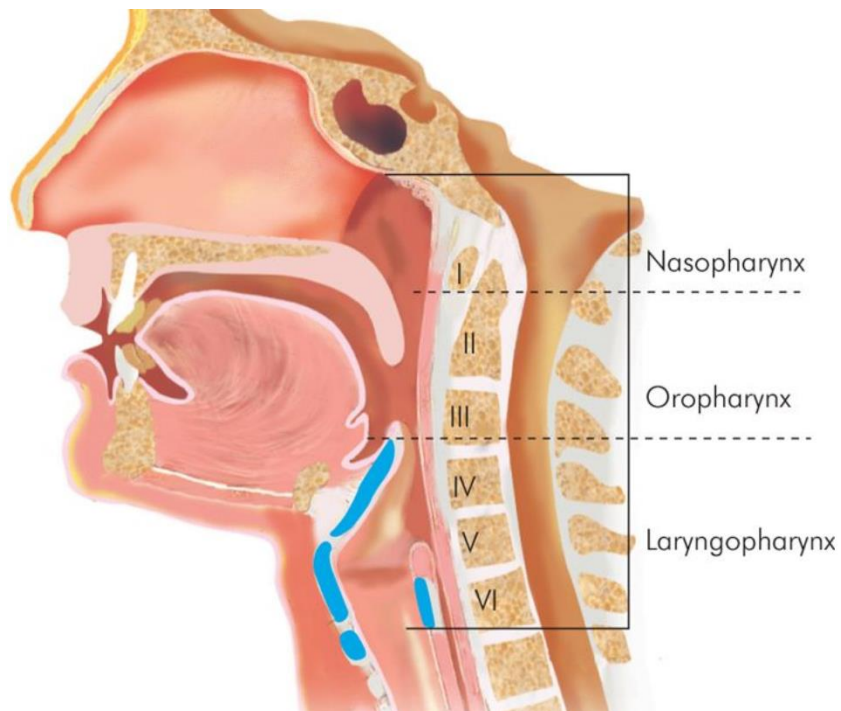
1. Retropharyngeal space: situated behind the pharynx
2. Parapharyngeal space: situated on the side of pharynx

Openings of the pharynx:

**It is open anteriorly
to nasal, oral, and
laryngeal cavities**



Divisions of Pharynx:

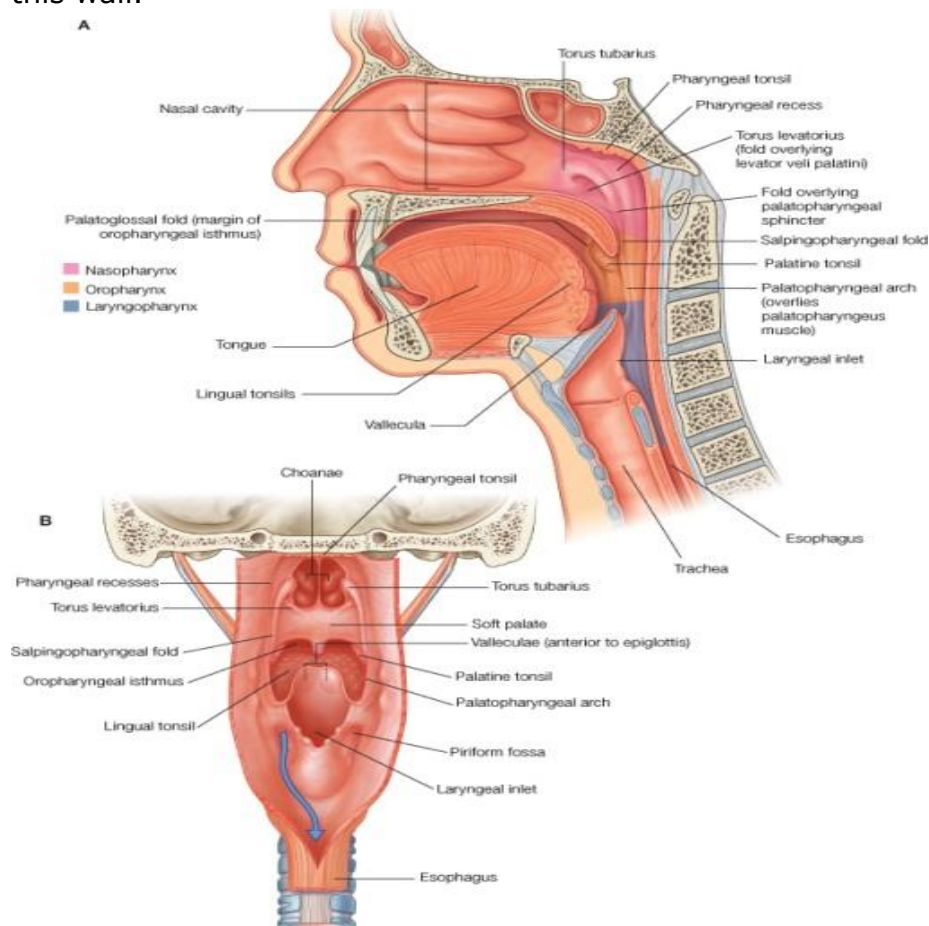


Nasopharynx (Epipharynx):

Uppermost part of the pharynx

It lies behind the nasal cavities and extends from the base of skull to the soft palate or the level of the horizontal plane passing through the hard palate

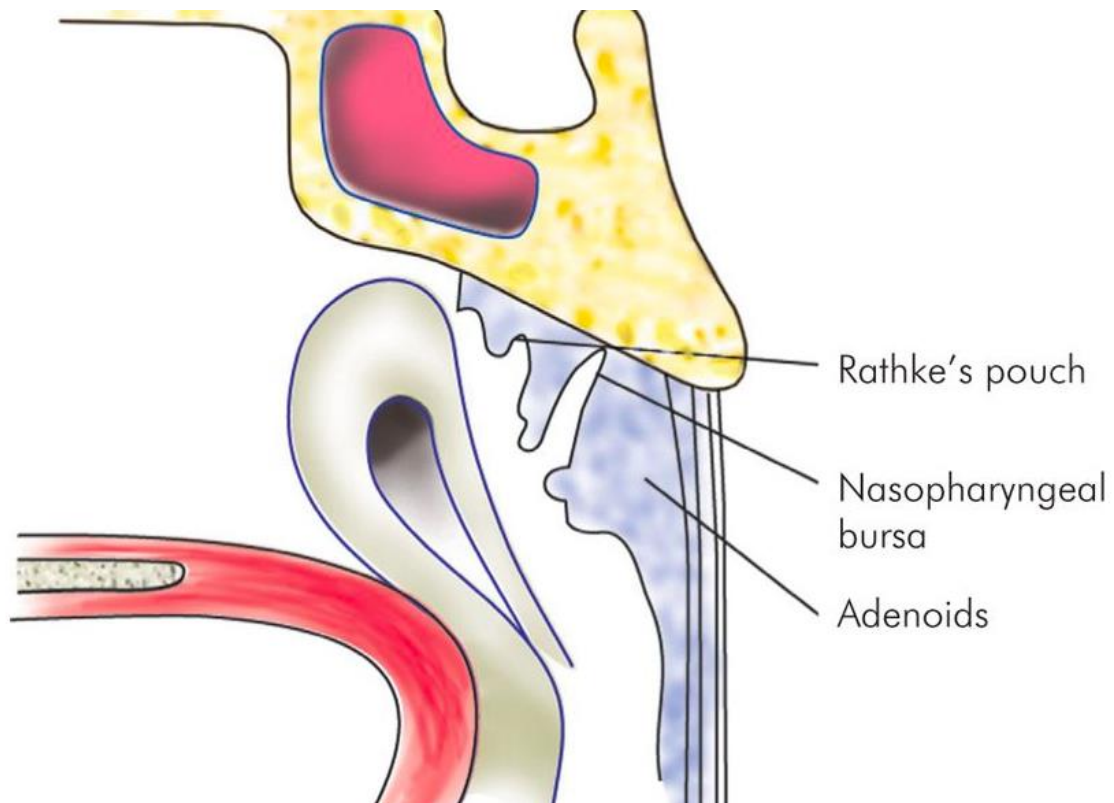
- **Roof** of the nasopharynx is formed by basisphenoid and basiocciput (fornix).
- **Posterior wall** is formed by arch of the atlas vertebra covered by prevertebral muscles and fascia.
- Both the roof and the posterior wall imperceptibly merge with each other.
- **Floor** is formed by the soft palate anteriorly but is deficient posteriorly. It is through this space-the *nasopharyngeal isthmus* that the nasopharynx communicates with the oropharynx.
 - Sealed off by elevation of the soft palate and contraction of portions of the superior pharyngeal constrictor (palatopharyngeal sphincter) (passavant's ridge) during swallowing
- **Anterior wall** is formed by posterior nasal apertures or *choanae*, separated from each other by the posterior border of the nasal septum. Posterior ends of nasal turbinates and meatuses are seen in this wall.



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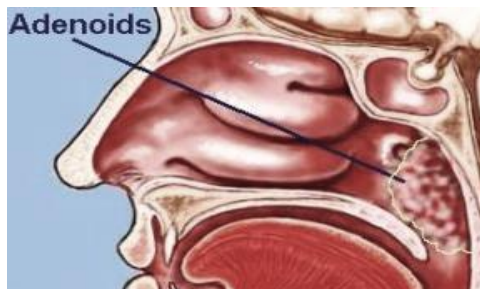
Lateral wall:

- Opening of eustachian tube situated 1.25 cm behind the posterior end of inferior turbinate.
- It is bounded above and behind by an elevation called torus tubarius raised by the cartilage of the tube.
- Above and behind the tubal elevation is a recess called fossa of Rosenmuller, which is the commonest site for origin of carcinoma.
- A ridge extends from the lower end of torus tubarius to the lateral pharyngeal wall and is called the salpingopharyngeal fold (raised by the corresponding muscle).



Nasopharyngeal Tonsil (Adenoids)

- See Adenoid lecture for more details.



Nasopharyngeal Bursa:

- An epithelial-lined median recess within the adenoid mass
- Extends from pharyngeal mucosa to the periosteum of the basiocciput.
- It represents the attachment of notochord to the pharyngeal endoderm during embryonic life.
- When infected, it may be the cause of persistent postnasal discharge or crusting. Sometimes an abscess can form in the bursa (Thornwaldt's disease).

Rathke's Pouch:

- A dimple above the adenoids and is resemble the buccal mucosal invagination, to form the anterior lobe of pituitary.
- A craniopharyngioma may arise from it.

Tubal Tonsil (tonsils of Garlach):

- Collection of subepithelial lymphoid tissue situated at tubal elevation.
- Continuous with adenoid tissue and forms a part of Waldeyer's ring.
- When enlarged due to infection, it causes eustachian tube occlusion.

Passavant's Ridge:

- It is a mucosal ridge raised by fibres of palatopharyngeus.
- It encircles the posterior and lateral walls of nasopharyngeal isthmus.
- Soft palate, during its contraction, makes firm contact with this ridge to cut off nasopharynx from the oropharynx during the deglutition or speech.

Velopharyngeal sphincter.

- Closure by:
- Postero-superior movement of the soft palate.
- Medial movement of the lateral pharyngeal wall.
- Slight anterior movement of the post .ph.wall (passavent ridge).

Epithelial Lining of Nasopharynx:

- Posterior extension of nasal cavity.
- It is lined by pseudostratified ciliated columnar epithelium.

Lymphatic Drainage:

- Upper deep cervical nodes either directly or indirectly through retropharyngeal and parapharyngeal lymph nodes.
- They also drain into spinal accessory chain of nodes in the posterior triangle of the neck.
- Lymphatics of the nasopharynx may also cross midline to drain into contralateral lymph nodes

Oropharynx:

- From the plane of hard palate above to the plane of hyoid bone below.
- It lies opposite the oral cavity with which it communicates through *oropharyngeal isthmus*.
- The latter is bounded above, by the soft palate; below, by the upper surface of tongue, and on either side, by palatoglossal arch (anterior pillar).
- Covered by non-keratinising stratified squamous epithelium

Boundaries of Oropharynx:

Posterior wall:

- Retropharyngeal space
- Second and upper part of the third cervical vertebrae

Anterior wall (deficient above, where oropharynx communicates with the oral cavity) but below:

(A) Base of tongue, posterior to circumvallate papillae.

(B) Lingual tonsils

- One on either side, situated in the base of tongue.
- They may show compensatory enlargement following tonsillectomy or may be the seat of infection.

(C) Valleculae.

- They are cup-shaped depressions lying between the base of tongue and anterior surface of epiglottis
- Each is bounded medially by the median glossoepiglottic fold and laterally by pharyngoepiglottic fold (Seat of retention cysts)

Lateral wall:

(A) Palatine (faucial) tonsil

(B) Anterior pillar: formed by Palatoglossus muscle.

(C) Posterior pillar: formed by Palatopharyngeus muscle.

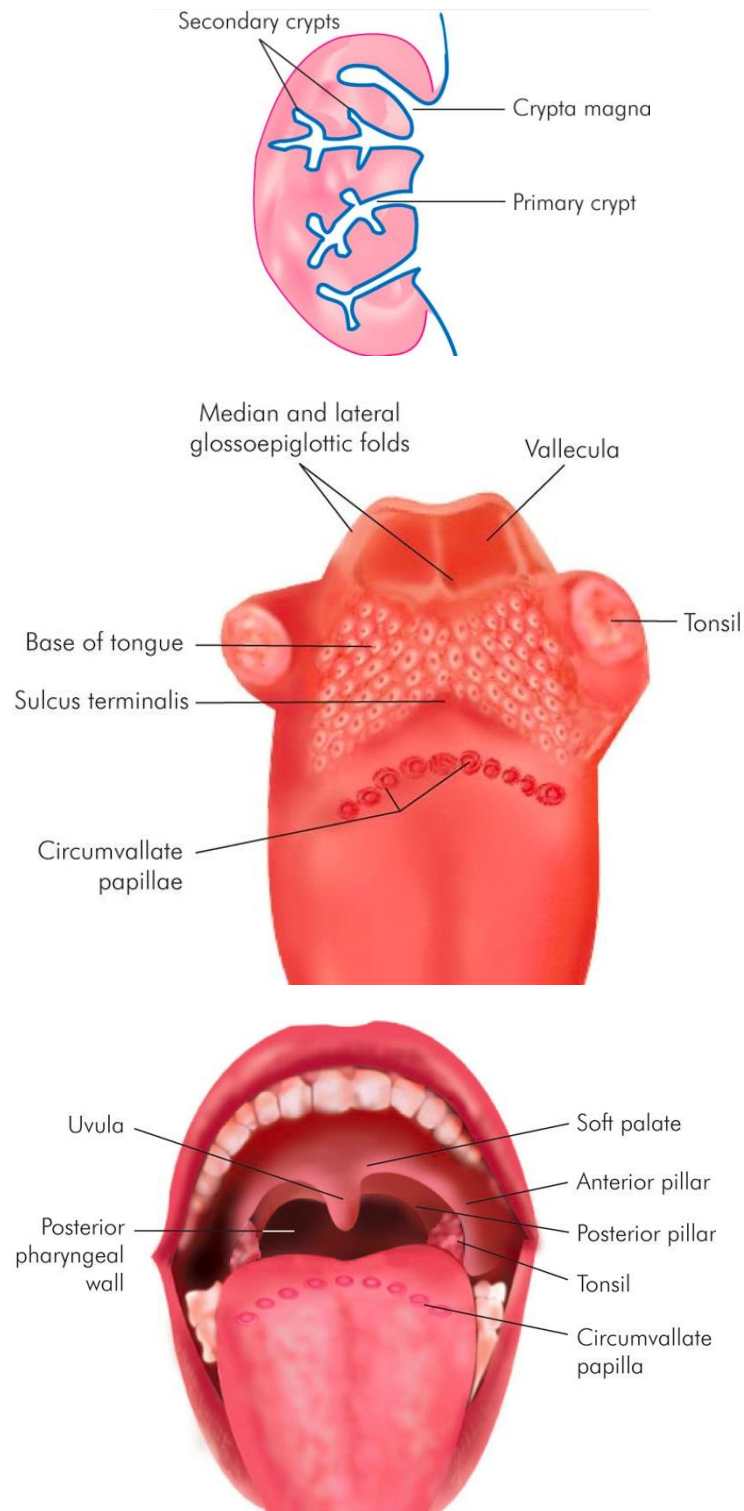
- Anterior and posterior pillars diverge from the soft palate and enclose a triangular depression called *tonsillar fossa* in which is situated the palatine tonsil)
- Boundary between oropharynx above and the hypopharynx below is formed by upper border of epiglottis and the pharyngoepiglottic folds.

Lymphatic Drainage:

- Drain into upper jugular chain particularly the jugulodigastric (tonsillar) node.
- The soft palate, lateral and posterior pharyngeal walls and the base of tongue also drain into retropharyngeal and parapharyngeal nodes and from there to the jugulodigastric and posterior cervical group.
- The base of tongue may drain bilaterally.

Palatine tonsils:

- See Tonsils lecture for more details.



Hypopharynx (Laryngopharynx):

- Lowest part of the pharynx
- Behind and partly on the sides of the larynx.
- Superior limit is the plane passing from the body of hyoid bone to the posterior pharyngeal wall
- Inferior limit is lower border of cricoid cartilage where hypopharynx becomes continuous with oesophagus.
- Hypopharynx lies opposite the 3rd, 4th, 5th, 6th cervical vertebrae.
- Clinically, it is subdivided into three regions-the
 - 1-** Pyriform sinus
 - 2-** Post-cricoid region
 - 3-** Posterior pharyngeal wall.

1. Pyriform sinus (fossa).

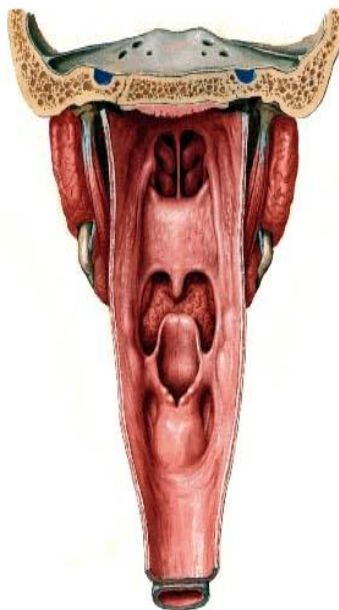
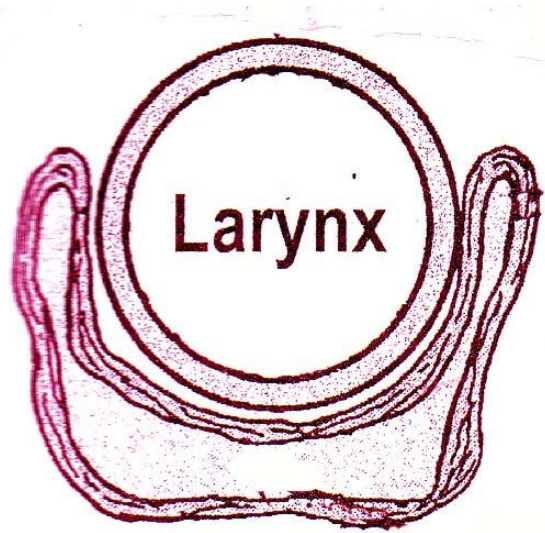
- Lies on either side of the larynx
- Extends from pharyngoepiglottic fold to upper end of esophagus.
- Bounded laterally by the thyrohyoid membrane and the thyroid cartilage
- Medially by the aryepiglottic fold, posterolateral surfaces of arytenoid and cricoid cartilages.
- It forms the lateral channel for food.
- Foreign bodies may lodge in the pyriform fossa.
- Internal laryngeal nerve runs submucosally in the lateral wall of the sinus and thus is easily accessible for local anaesthesia. It is also through this nerve that pain is referred to the ear in carcinoma of the pyriform sinus.

2. Post-cricoid region:

- It is the part of the anterior wall of laryngopharynx between the upper and lower borders of cricoid lamina.
- It is a common site for carcinoma in females suffering from Plummer-Vinson syndrome.

3. Posterior pharyngeal wall:

- It extends from the level of hyoid bone to the level of cricoarytenoid joint

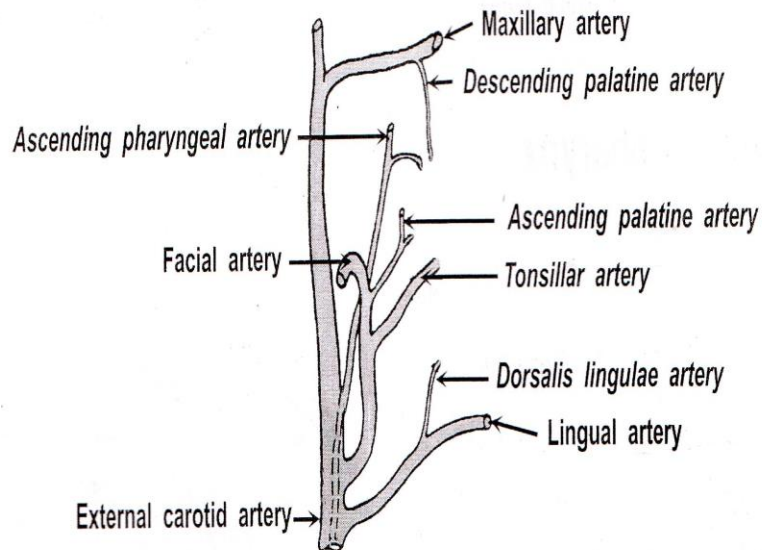


Lymphatic Drainage:

- Pyriform sinus is richly supplied by lymphatics which exit through the thyrohyoid membrane and drain into the upper jugular chain
- Lymphatics of the posterior wall terminate in the lateral pharyngeal or parapharyngeal nodes and thence to the deep cervical lymph nodes
- Lymphatics of post-cricoid region also drain into the parapharyngeal nodes but may also drain into nodes of supraclavicular and paratracheal chain.
- Rich lymphatic network of pyriform fossae explains the high frequency with which nodal metastases are seen in carcinoma of this region.

Blood Supply of the Pharynx:

1. Ascending pharyngeal artery
 2. Ascending palatine and tonsillar branch of the facial artery
 3. Descending palatine artery or pharyngeal branches of maxillary artery.
 4. Lingual artery.
- Sometimes branches from superior and inferior thyroid arteries.



Venous drainage of the Pharynx

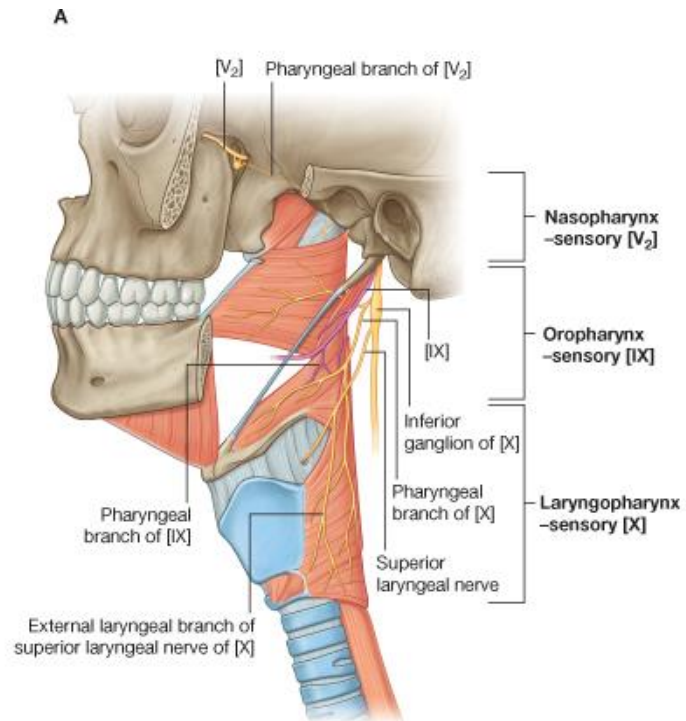
Vein	Course
Pharyngeal plexus	Located on the outer surface of the pharynx in the buccopharyngeal fascia Gives rise to pharyngeal vv., which drain into the internal jugular v. and also into the pterygoid plexus of veins along the lateral pterygoid m. The pharyngeal vv. also may drain into the facial, lingual, or superior thyroid v.

Sensory innervations of pharynx:

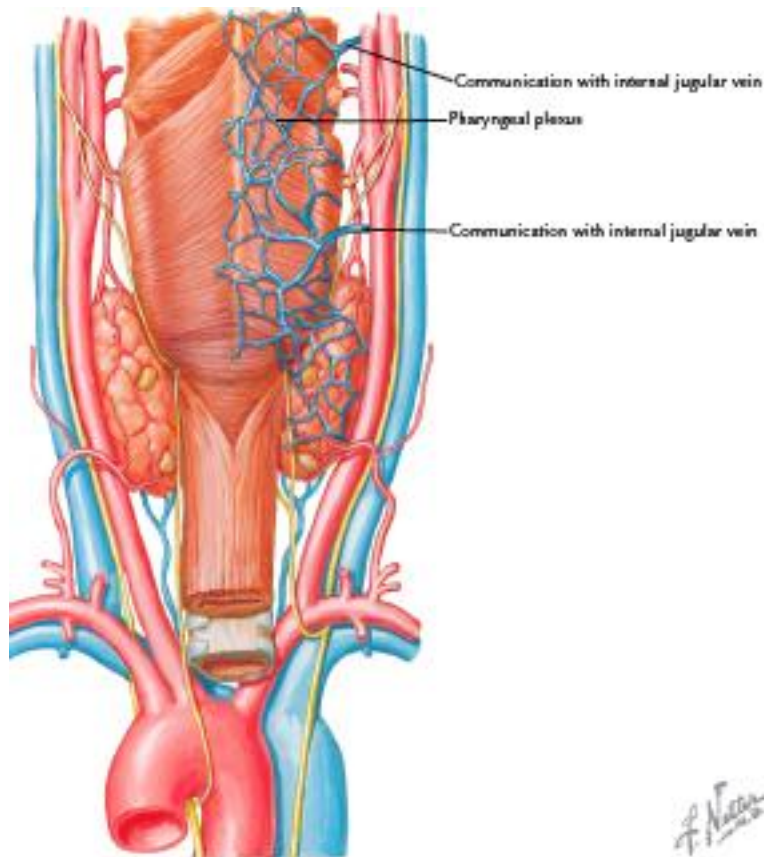
- Nasopharynx: Maxillary (V2) of CN-V.
- Oropharynx: CN-IX.
- Hypopharynx: CN-X.

Motor innervations of pharynx:

- All muscles of the pharynx are supplied by pharyngeal plexus (CN-X)
except Stylopharyngeous supplied by Glossopharyngeous nerve



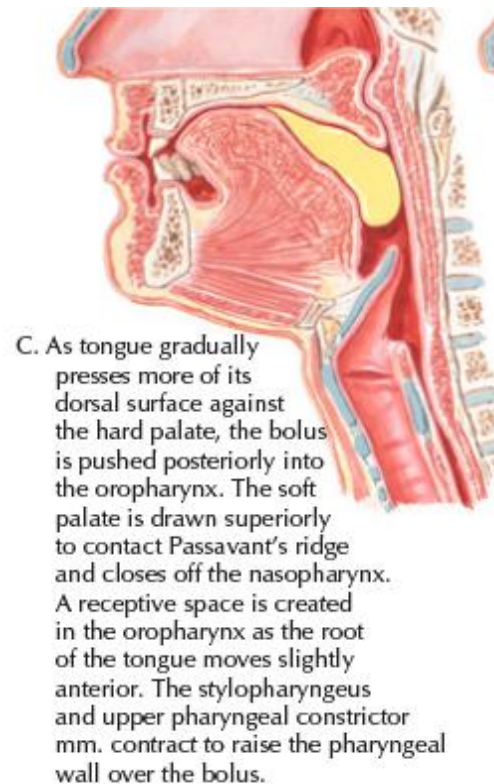
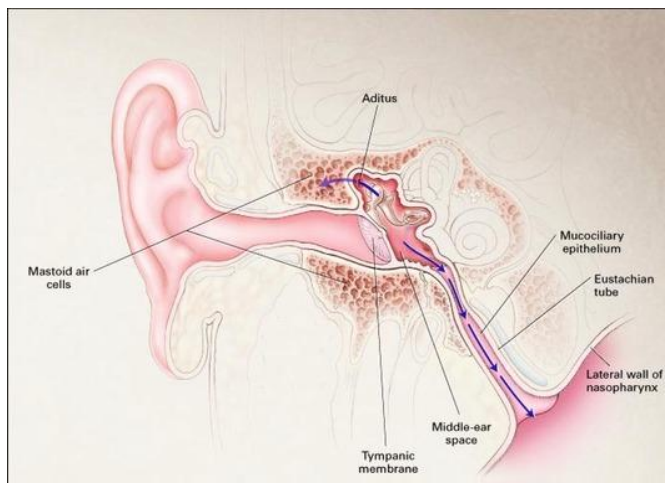
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Physiology of the pharynx:

Functions of Nasopharynx:

1. Acts as a conduit for air, which has been warmed and humidified in the nose, in its passage to the larynx and trachea.
2. Through the eustachian tube, it ventilates the middle ear and equalizes air pressure on both sides of tympanic membrane.
(This function is important for hearing)
3. Elevation of the soft palate against posterior pharyngeal wall and the Passavant's ridge helps to cut off nasopharynx from oropharynx.
(This function is important during swallowing, vomiting, gagging and speech.)
4. Acts as a resonating chamber during voice production.
(Voice disorders are seen in nasopharyngeal obstruction and velopharyngeal incompetence)
5. Acts as a drainage channel for the mucus secreted by nasal and nasopharyngeal glands.



Functions of Oropharynx:

1. As a conduit for passage of air and food.
2. Helps in the pharyngeal phase of deglutition.
Base of tongue against the hard palate close oral cavity (oropharyngeal isthmus),
Velopharyngeal sphincter close nasopharynx (nasopharyngeal isthmus)
3. Forms part of vocal tract for certain speech sounds.
4. Helps in appreciation of the taste. Taste buds are present in the base of tongue, soft palate, anterior pillars and posterior pharyngeal wall.
5. Provides local defence and immunity against harmful intruders into the air and food passages.

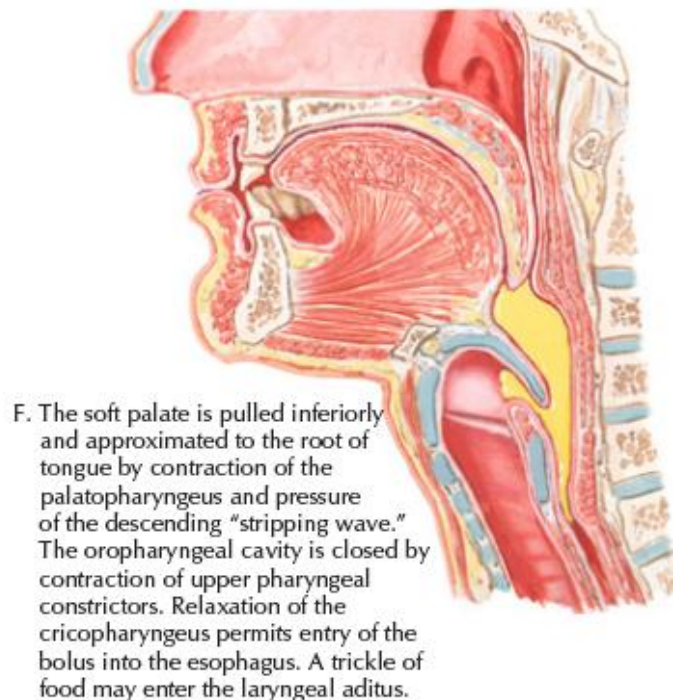
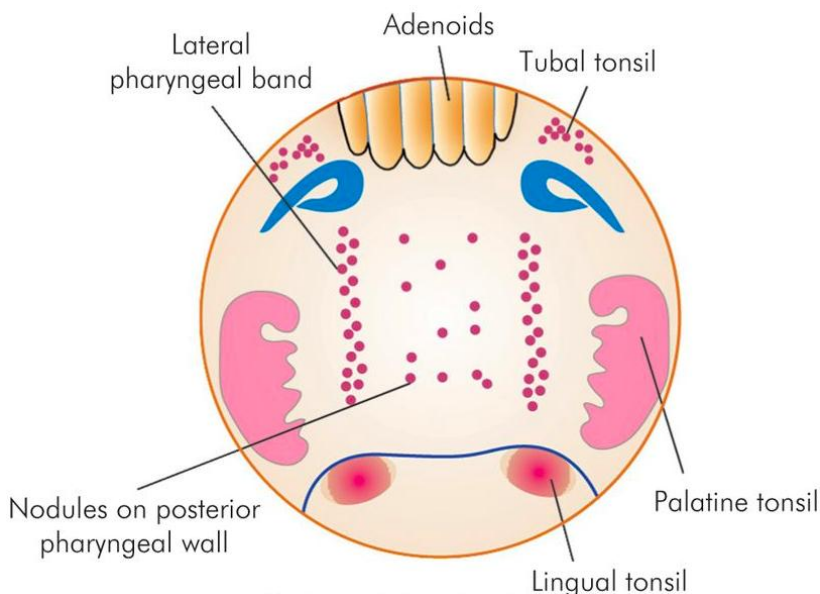
This function is subserved by subepithelial masses of lymphoid tissues scattered as Waldeyer's ring.

They are strategically placed at the portals of air and food entry and act as protective sentinels.

B-lymphocytes in the germinal centers of the follicles produce secretory antibodies of IgA class

Whereas T-lymphocytes in parafollicular region produce cell-mediated immunity against various viruses, bacteria and fungi.

Pathogens which happen to enter into these lymphoid masses are dealt by IgM and IgG antibodies secreted by plasma cells.



Functions of Hypopharynx:

- Like oropharynx, is a common pathway for air and food, provides a vocal tract for resonance of certain speech sounds and helps in deglutition.
- There is coordination between contraction of pharyngeal muscles and relaxation of cricopharyngeal sphincter at the upper end of oesophagus.
- Lack of this coordination, i.e. failure of cricopharyngeal sphincter to relax when pharyngeal muscles are contracting causes hypopharyngeal diverticulum.

Pharyngitis

Acute Pharyngitis:

- Very common and occurs due to varied aetiological factors like viral, bacterial, fungal or others
- Viral causes are more common.
- Acute streptococcal pharyngitis (due to Group A beta *haemolytic streptococci*) has received more importance because of its aetiology in rheumatic fever and post-streptococcal glomerulonephritis
- May be secondary to sinonasal disease, caustic injury, chronic allergy



Pathogens:

- Typically viral; most common types are adenovirus, rhinovirus, and enterovirus (less common are mumps, herpes simplex, and rubella);
- Secondary bacterial infections are less common (GA streptococci, pneumococci, H. influenza)

Viral	Bacterial	Fungal	Miscellaneous
• Rhinoviruses • Influenza • Parainfluenza • Measles and chickenpox • Coxsackie virus • Herpes simplex • Infectious mononucleosis • Cytomegalovirus	• <i>Streptococcus</i> (Group A, beta-haemolyticus) • <i>Diphtheria</i> • <i>Gonococcus</i>	• <i>Candida albicans</i> • <i>Chlamydia trachomatis</i>	• Toxoplasmosis (parasitic, rare)

Clinical features:

- Pharyngitis may occur in different grades of severity.
- *Milder* infections present with discomfort in the throat, some malaise and low grade fever.
- Pharynx in these cases is congested but there is no lymphadenopathy.
- *Moderate* and *severe* infections present with pain in throat, dysphagia, headache, malaise and high fever.
- Pharynx in these cases shows erythema, exudate and enlargement of tonsils and lymphoid follicles on the posterior pharyngeal wall.
- *Very severe* cases show oedema of soft palate and uvula with enlargement of cervical nodes
- Cervical adenopathy in moderate and severe

(It is not possible, on clinical examination, to differentiate viral from bacterial infections)

(Viral infections are generally mild and are accompanied by rhinorrhoea and hoarseness while the bacterial ones are severe)

(Gonococcal pharyngitis is mild and may even be asymptomatic)

Diagnosis:

- Clinical exam, consider throat cultures, viral smears rarely indicated
- (It can detect 90% of Group A *Streptococci*)
- (*Diphtheria* is cultured on special media)
- (Swab from a suspected case of gonococcal pharyngitis should be cultured immediately without delay)
- (Failure to get any bacterial growth suggests a viral aetiology).

Treatment

- Supportive care (bed rest, hydration, humidity, lozenges, anesthetic sprays [cetacaine or xylocaine], iodine glyceride solutions, antipyretics, decongestants)
- Antibiotics for suspected bacterial infection antibiotics for suspected bacterial infections



Chronic Pharyngitis:

- Chronic inflammatory condition of the pharynx.
- Pathologically
- Characterized by hypertrophy of mucosa, seromucinous glands, subepithelial lymphoid follicles and even the muscular coat of the pharynx
- Two types:
 1. Chronic catarrhal pharyngitis
 2. Chronic hypertrophic (granular) pharyngitis

Etiology

1. Persistent infection in the neighbourhood

- In chronic rhinitis and sinusitis (PND).
- This causes hypertrophy of the lateral pharyngeal bands.
- Similarly, chronic tonsillitis and dental sepsis are also responsible for chronic pharyngitis and recurrent sore throats.

2. Mouth breathing Breathing

- Exposes the pharynx to air which has not been filtered, humidified and adjusted to body temperature thus making it more susceptible to infections. - Mouth breathing is due to:

- (i) Obstruction in the nose, e.g. nasal polypi, allergic or vasomotor rhinitis, turbinal hypertrophy, deviated septum or tumours,
- (ii) Obstruction in the nasopharynx, e.g. adenoids and tumours,
- (iii) Protruding teeth which prevent apposition of lips,
- (iv) Habitual, without any organic cause.

3. Chronic irritants

Excessive smoking, chewing of tobacco and pan, heavy drinking, highly spiced food can all lead to chronic pharyngitis.

4. Environmental pollution

Smoky or dusty environment or irritant industrial fumes may also be responsible for chronic pharyngitis.

5. Faulty voice production

Less often realised but an important cause of chronic pharyngitis in the faulty voice production.

Excessive use of voice or faulty voice production seen in certain professionals or in "pharyngeal neurosis" where person resorts to constant throat clearing, hawking or snorting, and that may cause chronic pharyngitis, especially of hypertrophic variety

Symptoms:

1. Discomfort or pain in the throat

(Especially noticed in the morning).

2. Foreign body sensation in throat

(Constant desire to swallow or clear his throat)

3. Tiredness of voice

Patient cannot speak for long and has to make undue effort to speak as throat starts aching.

The voice may also lose its quality and may even crack.

4. Cough

Throat is irritable and there is tendency to cough.

Mere opening of the mouth may induce retching or gagging

Signs:

Chronic catarrhal pharyngitis

- Congestion of posterior pharyngeal wall with engorgement of vessels
- Faucial pillars may be thickened.
- There is increased mucus secretion which may cover pharyngeal mucosa

Chronic hypertrophic (granular) pharyngitis

1. Pharyngeal wall appears thick and oedematous with congested mucosa and dilated vessels.
2. Posterior pharyngeal wall may be studded with reddish nodules (hence the term granular pharyngitis). These nodules are due to hypertrophy of subepithelial lymphoid follicles normally seen in pharynx
3. Lateral pharyngeal bands become hypertrophied.
4. Uvula may be elongated and appear oedematous



Dx:

Clinical history and exam, culture and biopsy if failed empiric therapies

Treatment:

- 1.** In every case of chronic pharyngitis, aetiological factor should be sought and eradicated.
- 2.** Voice rest and speech therapy is essential for those with faulty voice production. Hawking, clearing the throat frequently or any other such habit should be stopped.
- 3.** Warm saline gargles, especially in the morning, are soothing and relieve discomfort.

(Mandl's paint may be applied to pharyngeal mucosa.)

(Cautery of lymphoid granules is suggested. Throat is sprayed with local anaesthetic and granules are touched with 10-25% silver nitrate.

Electrocautery or diathermy of nodules may require general anaesthesia.)

Bacterial Infections

- Normal flora consists of gram-positive aerobic organisms (including alpha- and beta-hemolytic streptococci), and anaerobic organisms (Peptostreptococcus, Fusobacterium, Bacterioides)

Streptococcal Infection

- Most common (esp in kids) is group A beta-hemolytic (Streptococcus pyogenes)
- Other streptococci may be involved; group C and G have been linked to severe pharyngitis and complications (reactive arthritis)
- GABHS has an incubation period of 12 hrs to 4 days
- Fever, sore throat, dysphagia, exudate, neck nodes; rhinorrhea and cough are not common
- Uncommon in infants due to maternal IgG and a lack of pharyngeal receptors
- 2 rapid tests:
 1. Enzyme immunoassay rapid antigen test
 2. Optical immunoassay rapid antigen test; culture if rapid antigen is -ve due to the risk of rheumatic fever
- **Centor criteria** (modified)
 - Fever
 - Absence of cough
 - Presence of pharyngeal exudate
 - Cervical LNs
- Abx in pts with -ve throat culture results has little or no efficacy
- Penicillin (erythromycin or cephalosporins if allergic) is the treatment of choice; must be aware of beta-lactamase-producing organisms
- Complications:
 - Suppurative (OM, acute sinusitis)
 - Non-suppurative (GN, rheumatic fever)
 - Rheumatic heart disease
 - Grisel syndrome
 - Subluxation of atlantoaxial joint due to an infectious / inflammatory process, presents with atraumatic torticollis and history of H&N infection or procedure

MODIFIED JONES CRITERIA

Major

Carditis

Polyarthrititis

Minor (AFPEE)

Arthralgia

Fever

- **Rheumatic fever** → 2 major or 1 major and 2 minor criteria with evidence of preceding GABHS infection
- Pts with acute rheumatic fever is at high risk for recurrent attacks; abx prophylaxis
- Risk of GN is not reduced by abx use
- **Scarlet fever** → acute streptococcal pharyngotonsillitis accompanied by a rash and production of an **erythrogenic toxin**
 - Rash starts on the chest and trunk and spreads (face, palms, soles are spared); desquamation can occur
 - Strawberry tongue

Staphylococcal Infection

- S. aureus or salivarius
- Mucopurulent drainage, localized pustules

Diphtheroid Infection

- **Corynebacterium species** are rarely pathogenic except C. diphtheriae (gram +ve rods)
- Uncommon due to vaccinations
- Gray-black membrane firmly adherent to underlying tissue involving the upper aerodigestive tract; may lead to airway obstruction and sepsis
- Toxin-mediated; treatment is antitoxin; abx is used an adjuvant therapy



Pertussis

- **Bordetella pertussis** (gram -ve coccobacillus)
- Paroxysms of coughing accompanied by a loud inspiratory sound (whooping cough)
- 3 stages
 - **Catarrhal stage**: lasts 1-2 wks, low-grade fever and upper respiratory symptoms
 - **Paroxysmal stage**: characteristic coughing and mucopurulent exudate, no fever; 2-4 wks or longer
 - **Convalescent stage**: lasts 1-2 wks
- Prolonged self-limited disease; abx do not alter the course of the disease
- Rare due to immunization

Gonorrhea

- Neisseria gonorrhoeae (gram-negative diplococcus)
- Generally asymptomatic
- Culture on **chocolate agar** media for identification
- Tx: penicillin, tetracycline, cephalosporins, quinolones

Syphilis

- Treponema pallidum
- Clinical stages include primary, secondary, tertiary, congenital
- **Primary stage**
 - Painless solitary chancre at the site of inoculation
 - Inflammatory infiltrate with obliterative endarteritis is the histologic hallmark of this disease
 - Organism can be demonstrated with **Warthin-Starry staining**
 - Chancre heals spontaneously in 3-6 wks
- **Secondary (disseminated) stage**
 - Skin lesions and lymphadenopathy are seen in 90%
 - Pharyngotonsillitis with mucosal patches (painless, superficial erosions with a silver-gray appearance surrounded by red margins)
 - 1/3rd go into spontaneous remission; 1/3rd continue with latent disease, 1/3rd progress to tertiary disease

- **Tertiary syphilis**

- May occur years after initial infection
- Typically involves the CNS or the aorta
- Gummas → granulomatous lesion consisting of central coagulative necrosis surrounded by palisading macrophages and fibroblasts
- Non-specific nontreponemal antibody tests → Venereal Disease Research Laboratory (VDRL) test and rapid plasma reagin test; used as screening test
- Specific treponemal antibody test → FTA-ABS; used as confirmatory test
- Tx: penicillin (for allergic pts, use tetracycline or erythromycin)

Other Bacterial Infections

- *Salmonella typhimurium*
- *Fusobacterium nucleatum*; *necrophorum* (Lemierre syndrome)
- *Yersinia enterocolitica*
- *Arcanobacterium haemolyticum* (exanthematous eruption of the skin that mimics scarlet fever)
- *Moraxella catarrhalis*, *Francisella tularensis*, *Mycoplasma* species, *Neisseria meningitidis*, *Chlamydia* species
- *Coxiella burnetii* (rickettsial organism) may cause pharyngitis (typically causes Q fever)
- Identification by culture, immunoassay, latex agglutination, in situ hybridization, PCR

Viral Infections

Herpes Simplex Virus

- Type 1 (usually oral) and type 2 (usually genital)
- Primary HSV infection gingivostomatitis or rarely as acute pharyngitis
- Primary HSV most commonly in children 10 mos-3 yrs
- Malaise, fever, and pharyngitis (vesicular lesions that bleed easily, covered with black crusts or shallow tonsillar ulcers covered with gray exudates)
- Acute illness evolves over 7-10 days; followed by rapid regression of symptoms and lesions
- Rarely, disseminated form with CNS involvement occurs (mortality rates up to 80%)
- Virus can remain latent and can be reactivated under a variety of stimuli (resides in sensory neural ganglia)
- Tx: symptomatic and acyclovir especially for the immunocompromised

Measles

- **Rubeola** (morbillivirus)
- Coryza, conjunctivitis, **Koplik spots** (spotty exanthematous lesions on the buccal mucosa), LN, lymphoid hyperplasia
- Symptomatic treatment (self-limited); mortality from bacterial super-infections

Epstein-Barr Virus

- Member of the family Herpesviridae; infects B-cells
- Nasopharyngeal undifferentiated ca and African Burkitt lymphoma have been associated with EBV
- Causes 95% of infectious mononucleosis (mono-like syndromes caused by CMV, Toxoplasma gondii, rubella, hep A virus, HIV type 1, adenovirus)
- Mono-splenomegaly (50%); hepatomegaly (10%); both seen mostly at 2-4th wks of the disease (counsel pts regarding contact activities)
- Dx
 - Atypical lymphocytes; elevated liver enzymes
 - Demonstration of serum **heterophile antibodies** to either horse or sheep erythrocytes (mono-spot test); may be -ve in children < 10 yrs
 - At clinical presentation, **IgG and IgM to viral capsid antigen (VCA)** is present; also **Ab to early antigen complex** present
 - IgM to VCA disappear within 2-3 mos; early antigen complex Ab lasts 2-6 mos
 - IgG to VCA and anti-EBNA (EBV nuclear antigens) persist for life

CMV

- Member of the family Herpesviridae
- Congenital and acquired infections; most are asymptomatic
- Most effective and sensitive method to detect active CMV infection is urine culture with inoculation of human diploid cell culture

Human Immunodeficiency Virus Type 1

- Viral particles may be present in the pharynx and tonsils

Herpangina. It is caused by Group A coxsackie virus and mostly affects children. Characteristic features include fever, sore throat and vesicular eruption on the soft palate and pillars. Vesicles are small and surrounded by a zone of erythema.

Pharyngoconjunctival fever. It is caused by an adenovirus, and is characterised by sore throat, fever and conjunctivitis. There may be pain in abdomen, mimicking appendicitis.

Acute lymphonodular pharyngitis. It is usually caused by a coxsackie virus and characterised by fever, malaise and sore throat. White-yellow, solid nodules appear on the posterior pharyngeal wall in this type of pharyngitis.

Fungal infections

- Occurs in immunocompromised or those with chronic disease

Candidal Infections

- **Candida albicans** is most common (part of normal flora)
- HIV or post-radiation pts
- Gram-stain or periodic acid Schiff stain (or silver stain) of a smear or culture on **Sabouraud agar**
- Topical nystatin or oral ketoconazole or fluconazole; if systemic, amphotericin B may be used

Deep-Seated Mycoses

- Cryptococcus neoformans, Rhinosporidiosis seeberi, Histoplasma capsulatum, Blastomyces dermatitidis, Paracoccidioides brasiliensis

Granulomatous Diseases Causing Pharyngitis

- Granuloma-chronic inflammatory process with modified macrophages, other inflammatory cells, giant cells, and fibroblasts

Mycobacterium TB (M. bovis)

- Pharyngotonsillitis can occur from expectorations of infected sputum from pulmonary involvement or isolated occurrence (esp in poor socioeconomic regions)

Leprosy (Hansen disease)

- Mycobacterium leprae
- Nasal cavity then pharynx
- Classified as either lepromatous or tuberculoid
 - Intradermal injection shows early response and later response (Mitsuda reaction)
 - Mitsuda reaction is strongly +ve (> 5 mm) in tuberculoid leprosy; in lepromatous leprosy, the reaction is weak (0-2 mm)
- Nerve damage or thickening (great auricular nerve)
- **Foamy macrophages** (lepra cells) containing large numbers of bacilli found in lepromatous leprosy with no granulomas
- Tx: **dapsone**, clofazimine, and rifampin

Other

- Parasitic infections
- FB granulomas
- Crohn disease → pharyngeal involvement in 9%
- Systemic or discoid lupus erythematosus can involve the oropharynx

Other Causes of Pharyngitis

Radiation Pharyngitis

- Oral and pharyngeal mucosa has high cell turnover
- 16-22 cGy causes atrophic changes due to inhibition of cellular division
- Tx: sucralfate, diphenhydramine, antibacterial agents, topical steroids mixture oral concoctions; pilocarpine for xerostomia

Integumental Disorders

Stevens - Johnson syndrome

- Aka exudative erythema multiform
- Onset usually follows URI or use of certain meds (sulfonamides, anticonvulsants, barbiturates)
- Painful vesiculobullous lesions skin and mucosal lesions; commonly ulcerate, bleed and crust over
- Acute phase includes fever and prostration; 10% mortality due to pulmonary involvement
- Self-limited with skin and mucosal lesions resolving in few wks
- High (20%) recurrence rate
- Tx: symptomatic with fluids and electrolyte balance; treat secondary infections; withdrawal of the offending agent

Pemphigus

- Autoimmune mechanism
- Rapid appearance of vesicles and bullae; rupture leaves a raw eroded defects that bleed and cause pain
- **Nikolsky sign** (rubbing unaffected areas resulting in sloughing) is not seen in mucosal sites
- Periods of remissions and exacerbations; may result in early death
- Intercellular bridges between epithelial cells disappear to produce **acantholysis**; leads to clumps of epithelial cells (**Tzanck cells**) lying free within vesicular spaces
- Tx: steroids, immunosuppressive
- Familial benign chronic pemphigus → rare disorder that becomes less severe as pts age

Cicatricial Pemphigoid “benign mucous membrane pemphigoid”

- Vesiculobullous, autoimmune disease
- Lesions commonly occur on the conjunctiva and upper airway mucosa (rarely skin)
- No acantholysis; supportive tx with steroid or immunosuppressives for severe disease

Bullous Pemphigoid “parapemphigus”

- Most are > 60 yrs
- Skin lesions that start out as a rash; later vesiculobullous lesions appear
- Similar to cicatricial pemphigoid but oral mucosal lesions are rare

Epidermolysis Bullosa

- Spontaneous or post-traumatic onset
- Usually **autosomal recessive**, occurring in infancy or early childhood
- Rupture of bullae leaves raw, painful surfaces that heal by scarring
- Early mortality due to extensive scarring
- No cure; steroids show inconsistent results

Reflux Pharyngitis

- Associated with esophageal and laryngeal ca
- Most reliable finding is interarytenoid erythema
- Most accurate test is 24-hour esophageal pH monitoring

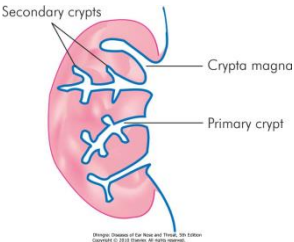
PFAPA

- Periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis
- Affects children (3 yrs); 5-day long history with symptoms occurring about every 28 days
- Steroids; tonsillectomy and H2-blockers can lead to remission
- Must exclude cyclic neutropenia

Idiopathic Pharyngitis

- Pharyngeal pain without obvious source
- PND-subclinical rhinopharyngitis; GERD associated
- Dietary (acidic or hot liquids) or personal habits (smoking)
- Meds (mouth washes and throat sprays)
- Thermal injury from crack or free-base cocaine

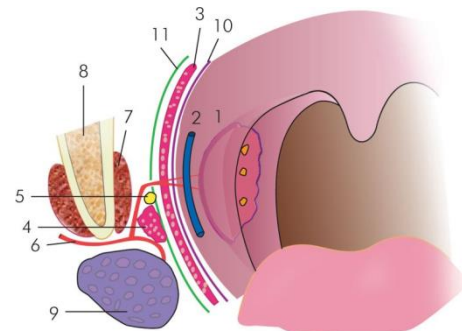
Tonsils Anatomy:

- Two in number, oval in shape.
 - Located in lateral wall of oropharynx between Anterior pillars (Palatoglossal fold) and Posterior pillars (Palatopharyngeal fold).
 - Deep, attached to fascia overlying superior constrictor muscle.
 - Extend upwards into the soft palate, downwards into the base of tongue and Anteriorly into palatoglossal arch.
 - Consists of:
 1. Epithelium continuous with oropharynx lining.
 2. Crypts: tube-like invaginations from epithelium.
 3. Lymphoid tissue
 - **Medial surface:**
 - Lined by antigen presenting Non-keratinising squamous epithelium which inhale and ingest antigens.
 - 10-30 Crypts, increase surface area for contact with foreign substance.
 - Crypts may be filled with cheesy material consisting of epithelial cells, bacteria and food debris which can be expressed by pressure over the anterior pillar.
 - **Crypta magna:**
 - o Intratonsillar cleft.
 - o Large and deep.
 - o Near the upper pole.
 - o Represents the ventral part of second pharyngeal pouch
- 
- **Lateral surface:**
 - Well-defined fibrous **capsule**.
 - Fibres of palatoglossus and palatopharyngeus muscles are attached to the capsule of the tonsil.
 - Loose areolar tissue:
 - o Between capsule and bed of tonsil
 - o Easy to dissect tonsil in the plane during tonsillectomy.
 - o Site for collection of pus in peritonsillar abscess.
 - Internal Carotid Artery runs 2 cm posterolateral to tonsil bed.
 - **Upper pole:**
 - Extends into soft palate.
 - Semilunar fold covers the medial surface and enclosing a potential space called supratonsillar fossa.
 - **Lower pole:**
 - Attached to the tongue.
 - Triangular fold extends from anterior pillar to the anteroinferior part of tonsil and encloses a space called anterior tonsillar space.
 - Tonsil is separated from tongue by Tonsillolinguual sulcus (**seat of carcinoma**).

- Bed of the tonsil:
- Formed by the superior constrictor and styloglossus muscles.
- Glossopharyngeal nerve and styloid process lie in relation to the lower part of tonsillar fossa.

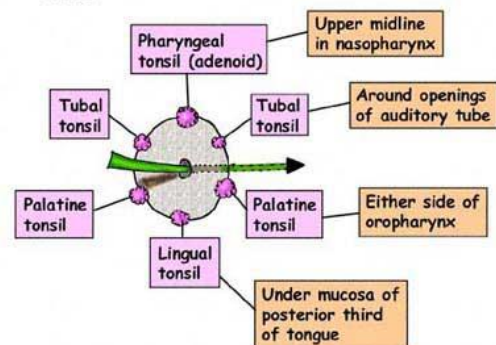
- Relations of the tonsil:

1. Capsule.
2. Loose areolar tissue containing paratonsillar vein.
3. Superior constrictor muscle.
4. Styloglossus .
5. Glossopharyngeal nerve.
6. Facial artery.
7. Medial pterygoid muscle.
8. Angle of mandible.
9. Submandibular salivary gland.
10. Pharyngobasilar fascia.
11. Buccopharyngeal fascia .



WALDEYER'S RING

An interrupted circle of protective lymphoid tissue at the upper ends of the respiratory and alimentary tracts



Functions of Tonsils:

- Part of Waldeyer's ring.
- Part of MALT (Mucosa-associated lymphoid tissue).
- Protective role and act as sentinels at the portal of air and food passage.
- Increase in size between 1-3 years old after exposure to antigens.
- Peak between 3-7 years old.
- Involutesc after puberty.
- Removed if become a seat of disease.

ANATOMIC AND PHYSIOLOGIC DIFFERENCES BETWEEN NORMAL ADENOIDS AND TONSILS		
	Adenoids	Tonsils
Anatomic location	Posterior wall nasopharynx May extend into posterior choanae	Lateral walls oropharynx Occasionally extends into nasopharynx or hypopharynx
Gross	Triangular shape; invaginated by deep folds, few or no crypts	Generally ovoid shape, sometimes bilobed Invaginated by 20-30 branching crypts
Microscopic	Three types of epithelium: Ciliated pseudostratified Columnar Squamous Transitional antigen (Ag) processing No afferent lymphatics	Specialized antigen (Ag) processing No afferent lymphatics
Physiologic	Mucociliary clearance Antigen processing Immune surveillance	Antigen processing, immune surveillance

- **Immunology:**
- Major immunologic organs of upper aerodigestive tract.
- Involved in both local immunity and immune surveillance.
- Acute and chronic antigenic stimulation causes a variety of changes, which return to normal post Adenotonsilectomy.
- No afferent lymphatics; direct antigen presentation; immune stimulation and modulation including production of memory cells
- No clear immune compromise post removal; but avoid if possible.
- All 5 immunoglobulin (Ig) isotypes are produced in the palatine tonsils.
- **IgA** is the most important product of the tonsillar immune system.

- **Arterial blood supply:**

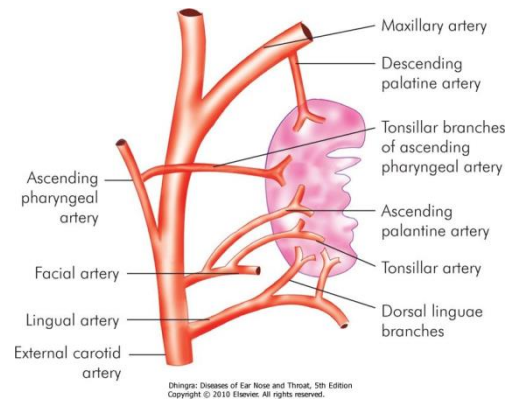
- From branches of External carotid Artery:

- **Upper Pole:**

1. Maxillary Artery:
→ Descending Palatine Artery.
2. Ascending Pharyngeal Artery.

- **Lower Pole:**

3. Facial Artery (**Main Supply**):
→ Ascending Palatine Artery.
→ Tonsillar Artery.
4. Lingual Artery → Dorsal lingual Artery.



- **Venous Drainage:**

- Lingual + Pharyngeal veins → Internal jugular vein.
- Paratonsillar vein is source of venous bleeding following tonsillectomy.

- **Lymphatic Drainage:**

- **No afferent lymphatics.**
- Drainage into Superior deep cervical + Jugular digastric LN.

- **Sensory innervation:**

- Glossopharyngeal nerve (CN-IX) and some branches of lesser palatine nerve. (Sphenopalatine ganglion)

- **Histology:**

- Four tonsillar zones for antigen presentation:

1. Reticular cell epithelium:

- Non-keratinizing squamous epithelium.
- Rich with Antigen-presenting cells (**M-cell**) which transport Antigen to Lymphoid Germinal center.

2. Extra-follicular Area:

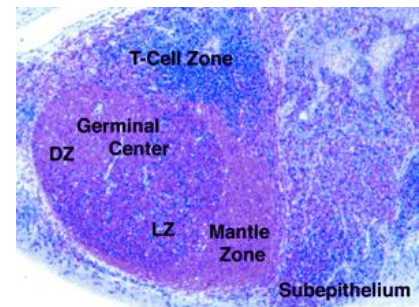
- Rich with (**T-cell**)

3. Lymphoid Follicle Mantle zone:

- Rich with (**Mature B-cell**)

4. Lymphoid Follicle Germinal centre:

- Rich with (**Active B-cell**)



ACUTE TONSILLITIS

1. Acute Superficial Tonsillitis:

- Part of generalized pharyngitis.
- Mainly in viral infections.

2. Acute Membranous Tonsillitis:

- Crypts exudation form a membrane on tonsil surface.

3. Acute Follicular Tonsillitis:

- Crypts infected and filled with pus.
- Yellowish spots over tonsils

4. Acute Parenchymatous Tonsillitis:

- Tonsil substance is affected.
- Tonsil is uniformly enlarged and red.

- Pathophysiology

- Viral infection with secondary bacterial invasion.
- Inflammation and loss of integrity of the crypt epithelium result in chronic cryptitis and crypt obstruction, leading to stasis of crypt debris and persistence of antigen.
- Most common pathogens:
 - Group A B-hemolytic streptococci.
 - Moraxella Catarrhalis.
 - Haemophilus influenzae.
 - Pneumococci.
 - Staphylococci

- Non-microbial causes of development chronic inflammation:

1. Extraesophageal reflux (EER)
 2. Presence of free radicals and other immunomodulators.
 3. Vibratory effects of chronic snoring.
- Viruses unlikely to cause chronic infections, but can obstruct crypts leading to infections.
 - EBV can cause significant hypertrophy.
 - Phases of Tonsillitis:
 - Tonsillar erythema → Exudative tonsillitis → Follicular tonsillitis → Cryptic tonsillitis (chronic infection)

- Clinical manifestations:

- School-age children mainly
- Rare in infants and above 50 years of age.
- Predominant symptoms:
 1. Sore throat.
 2. Difficulty in swallowing due to local pain.
 3. Trismus

4. Halitosis
5. Fever.
6. Earache either referred pain or result of acute otitis media which may occur as a complication.
7. Constitutional symptoms: headache, general body aches, malaise, constipation and abdominal pain due to mesenteric lymphadenitis.

- **Signs:**

- Tonsil is uniformly enlarged and red.
- breath is foetid and tongue is coated.
- Hyperaemia of pillars, soft palate and uvula.
- Tonsils are red and swollen with yellowish spots of purulent material presenting at the opening of crypts (acute follicular tonsillitis)
- Tonsils may have whitish membrane on the medial surface of tonsil which can be easily wiped away with a swab (acute membranous tonsillitis)
- Tonsils may be enlarged and congested so much so that they almost meet in the midline along with some oedema of the uvula and soft palate (acute parenchymatous tonsillitis).
- The jugulodigastric lymph nodes are enlarged and tender.

- **Recurrent Acute Tonsillitis:**

- At least 7 episodes in the past year or
- At least 5 episodes per year for 2 years or
- At least 3 episodes per year for 3 years

- **Diagnosis:**

- Clinical exam.
- Throat cultures
- GABHS rapid antigen test.
- Antistreptolysin-O (ASO)
- Monosopt test.
- Carrier state → Positive culture with no change in ASO titer.

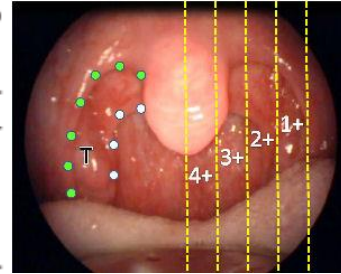
- **Differential Diagnosis:**

- Peritonsillar Abscess.
- Leukemia.
- Lymphoma.

- **Tonsils grading system:**

Table 1. Grade of obstruction of palatine tonsil according to Brodsky¹⁰.

Grade	Proportion of Tonsil in Oropharynx
0	Tonsil in Palatine Fossa
1	Tonsil occupying less than 25% of oropharynx
2	Tonsil occupying 25 - 50% of oropharynx
3	Tonsil occupying 50 - 75% of oropharynx
4	Tonsil occupying more than 75% of oropharynx



- **Complications:**

1. Chronic tonsillitis with recurrent acute attacks.
 - Due to incomplete resolution of acute infection.
 - Chronic infection may persist in lymphoid follicles of the tonsil in the form of microabscesses.
2. Peritonsillar abscess.
3. Parapharyngeal abscess.
4. Retropharyngeal abscess.
5. Cervical abscess due to suppuration of jugulodigastric lymph nodes.
6. Acute otitis media.
7. Rheumatic fever.
 - Seen in association with tonsillitis due to **Group A beta-haemolytic streptococci**.
8. Acute glomerulonephritis.
9. Subacute bacterial endocarditis.
 - Acute tonsillitis in a patient with valvular heart disease may be complicated by endocarditis.
 - It is usually due to **Streptococcus viridans** infection.

- **Management:**

1. Bed rest.
2. Hydration.
3. Oral hygiene.
4. Analgesia.
5. Antibiotics:
 - Most of the infections are due to streptococcus.
 - **Penicillin** is the drug of choice
 - **Clindamycin** if allergic to penicillin.
 - Should be continued for 7-10 days.
6. Tonsillectomy.

CHRONIC TONSILLITIS

- Polymicrobial infection.
- It may be a complication of acute tonsillitis.
- Pathologically, microabscesses walled off by fibrous tissue have been seen in the lymphoid follicles of the tonsils.
- Subclinical infections of tonsils without an acute attack.
- Mostly affects children and young adults, rarely occurs after 50 years.
- Chronic infection in sinuses or teeth may be a predisposing factor.

Types:

1. Chronic follicular tonsillitis:
 - Tonsillar crypts are full of infected cheesy material which shows on the surface as yellowish spots.
2. Chronic parenchymatous tonsillitis:
 - Tonsillar hyperplasia and may interfere with speech, deglutition and respiration.
 - Attacks of sleep apnoea may occur.
 - Long-standing cases develop features of cor pulmonale.
3. Chronic fibroid tonsillitis.
 - Tonsils are small but infected, with history of repeated sore throats.

Clinical Features:

1. Recurrent attacks of sore throat or acute tonsillitis.
2. Chronic irritation in throat with cough.
3. Bad taste in mouth and foul breath (halitosis) due to pus in crypts.
4. Thick speech, difficulty in swallowing and choking spells at night (when tonsils are large and obstructive).
5. Cervical lymphadenopathy.
6. Tonsilloliths.

Examination:

1. Tonsils may show varying degree of enlargement. Sometimes they meet in the midline (chronic parenchymatous type).
2. There may be yellowish beads of pus on the medial surface of tonsil (chronic follicular type).
3. Tonsils are small but pressure on the anterior pillar expresses frank pus or cheesy material (chronic fibroid type).
4. **Flushing of anterior pillars** compared to the rest of the pharyngeal mucosa is an important sign of chronic tonsillar infection.
5. **Enlargement of jugulodigastric lymph nodes** is a reliable sign of chronic tonsillitis. During acute attacks, the nodes enlarge further and become tender.

- **Management:**

1. Conservative treatment:
 - Attention to general health, diet, treatment of co-existent infection of teeth, nose and sinuses.
 - Long-term antibiotics against B-lactamase producing organisms and encapsulated organisms.
2. Tonsillectomy.

- **Complications:**

1. Tonsilloliths.
 - Calculus of the tonsil.
 - More often seen in adults.
 - Retention of debris of chronic tonsillitis blocked its crypts.
 - Inorganic salts of calcium and magnesium are then deposited leading to formation of a stone.
 - It may gradually enlarge and then ulcerate through the tonsil.
 - Give rise to local discomfort or foreign body sensation.
 - Diagnosed by palpation or gritty feeling on probing.
 - Treatment is simple removal of the stone or tonsillectomy.
2. Intratonsillar abscess.
 - Accumulation of pus within the substance of tonsil.
 - Follows blocking of the crypt opening in acute follicular tonsillitis.
 - Marked local pain and dysphagia.
 - Tonsil appears swollen and red.
 - Treatment is administration of antibiotics and drainage of the abscess if required.
 - Later tonsillectomy should be performed.
3. Tonsillar cyst.
 - Due to blockage of a tonsillar crypt and appears as a yellowish swelling over the tonsil.
 - Very often it is symptomless.
 - It can be easily drained

INDICATIONS FOR TONSILLECTOMY

○ **Obstruction**

1. Tonsillar hyperplasia with obstruction
2. Sleep-related disordered breathing:
 - a. Obstructive sleep apnea syndrome
 - b. Upper airway resistance syndrome
 - c. Obstructive hypoventilation syndrome
3. Failure to thrive (Not attributable to other causes)
4. Cor pulmonale (Not attributable to other causes)
5. Swallowing abnormalities (Not attributable to other causes)
6. Speech abnormalities
7. Orofacial/dental abnormalities
8. Lymphoproliferative disorder

○ **Infection**

1. Recurrent/chronic tonsillitis
2. Tonsillitis with:
 - a. Cervical nodes abscess
 - b. Acute airway obstruction
 - c. Cardiac valve disease
3. Persistent tonsillitis with:
 - a. Persistent sore throat
 - b. Tender cervical nodes
 - c. Halitosis
 - d. Tonsillolithiasis
4. Streptococcal carrier state unresponsive to medical therapy in a child or household at risk.
5. Peritonsillar abscess unresponsive to medical therapy or in a patient with recurrent tonsillitis or recurrent abscess.

○ **Neoplasia**

1. Suspected neoplasia, either benign or malignant

INDICATIONS FOR TONSILLECTOMY (Darrow, 2002)

○ Absolute:

1. Tonsillar hyperplasia with obstructive sleep apnea.
2. Failure to thrive (Not attributable to other causes)
3. Abnormal Dentofacial growth.
4. Persistent or recurrent tonsillar hemorrhage.
5. Suspected malignancy (unilateral tonsillar enlargement).

○ Relative:

1. Recurrent acute tonsillitis (documented 7 infections in 1 year, 5 infections in 2 years, or 3 infections in 3 years).
2. Peritonsillar abscess (2 attacks)
3. Chronic tonsillitis with swallowing difficulties.
4. Chronic tonsillitis with dysphagia.
5. Halitosis.
6. Streptococcus carrier unresponsive to medical management.

Tonsillectomy can be done as a part of another operation:

1. Palatopharyngoplasty which is done for sleep apnoea syndrome.
2. Glossopharyngeal neurectomy. Tonsil is removed first and then IX nerve is severed in the bed of tonsil.
3. Removal of styloid process.

CONTRAINDICATIONS FOR TONSILLECTOMY

1. Haemoglobin level less than 10 g%.
2. Presence of acute infection in upper respiratory tract, even acute tonsillitis. Bleeding is more in the presence of acute infection.
3. Children under 3 years of age. They are poor surgical risks.
4. Overt or submucous cleft palate.
5. Bleeding disorders, e.g. leukaemia, purpura, aplastic anaemia, haemophilia.
6. At the time of epidemic of polio.
7. Uncontrolled systemic disease, e.g. diabetes, cardiac disease, hypertension or asthma.
8. Tonsillectomy is avoided during the period of menses.

TABLE 84.7**COMPLICATIONS OF ADENOTONSILLAR DISEASE AND ADENOIDECTOMY AND TONSILLECTOMY**

Complication	Presentation	Management Options
Peritonsillar abscess	Sore throat/dysphagia Pharyngotonsillar bulge Trismus Drooling	Antibiotics (i.v.) Needle aspiration/I & D Immediate tonsillectomy
Acute airway obstruction secondary to T & A hyperplasia	Stridor/stertor Muffled/hyponasal voice Drooling Enlarged tonsils (and adenoids)	Nasopharyngeal airway Steroids (i.v.) Antibiotics (i.v.)
Hemorrhage	Bleeding from mouth or nose Frequent swallowing	Local control (cautery or vasoconstriction) Control in OR topical or when uncontrollable, by arterial ligation/embolization Evaluate for coagulopathy in selected cases
Airway obstruction	Occurs in first 4–24 h Palatal swelling Hypopharyngeal secretions	Nasopharyngeal airway Steroids (i.v.) Gentle suctioning
Dehydration	Poor oral intake Dry mucous membranes Lethargy	Control emesis if present i.v. Hydration Parental education Pain control prn
Persistent VPI after adenoidectomy	Hypernasal speech (lasting beyond 2-mo postop) Nasal regurgitation of fluids	Speech therapy Palate surgery Palatal prosthesis
Pulmonary edema after relief of airway obstruction ^a	Difficulty with oxygenation Frothy pink secretions from endotracheal tube	Positive end expiratory ventilation Lasix Morphine

i.v., intravenous; I & D, incision and drainage; OR, operating room; T & A, tonsillectomy and adenoidectomy; VPI, velopharyngeal insufficiency.

^aCan occur after laryngospasm or relief from, chronic or acute airway obstruction from enlarged tonsils or adenoids.

- **SURGICAL TECHNIQUE**

- **Coblation**

- Mechanism of action differs in that tissue ablation occurs as a result of dissociation of organic molecules rather than the conventional heat-based processes described.
- Minimize thermal damage to adjacent tissue structures.
- The coblation technique, unlike previous radiofrequency, uses bipolar current to create a plasma field, which can then split tissue.
- Creation of the field occurs at temperatures of only 60° C to 70° C, which is less than those produced with conventional radiofrequency and electrocautery.
- The literature indicates an improved quality of life and decreased pain associated with coblation.
- Bleeding rates do not appear to differ between conventional electrosurgical tonsillectomy and coblation.

- **Harmonic Scalpel**

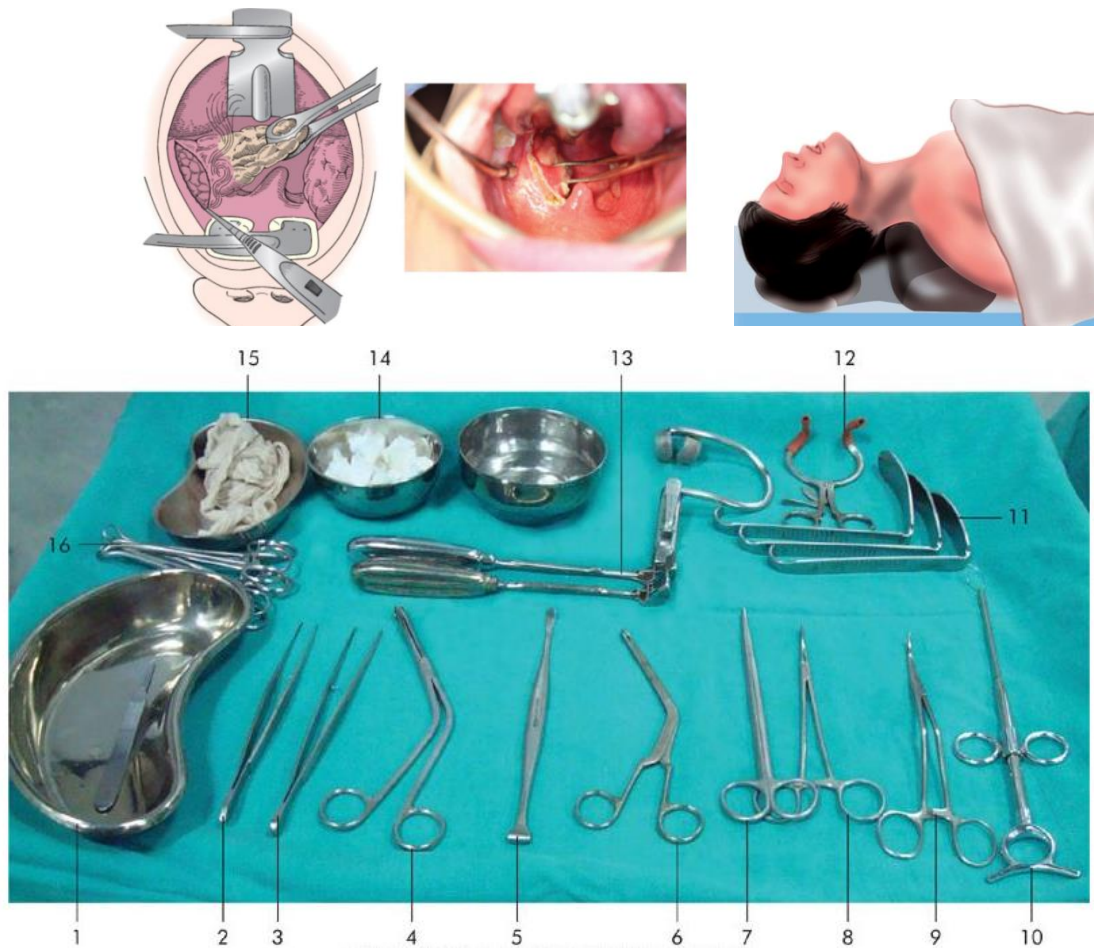
- Ultrasonic technology is used to cut and coagulate tissue at temperatures lower than those associated with electrocautery and lasers.
- The decreased thermal damage is thought to lead to decreased postoperative pain.
- There are limited studies with the harmonic scalpel.
- Several indicate decreased postoperative pain, although other studies showed an increase in the postoperative pain rate, and still others found
- no difference between traditional electrocautery and the harmonic scalpel technique in terms of intraoperative blood loss or postoperative hemorrhage.
- Use of the harmonic scalpel for tonsillectomy may be a promising technique in the future.

- **Laser Ablation**

- The use of lasers is not routine for the performance of tonsillectomy.
- Initially, they were involved the use of argon plasma coagulation.
- Good results were obtained in comparison to conventional electrocautery tonsillectomy, and there was a reported reduction in operative time.
- Subsequently, the CO2 laser as well as KTP laser were incorporated to control bleeding.
- The contact diode laser has also been reported to decrease postoperative pain rates when compared with traditional monopolar cautery.

- **Powered Instruments**
- The majority of reports involve partial tonsillectomy or intracapsular tonsillectomy.
- There have been reports of lower postoperative hemorrhage rates, as well as rapid recovery, in young children after the use of powered instruments for intracapsular tonsillectomy.
- This technique does not appear to be feasible for complete tonsillectomy and is therefore listed only for informational purposes.
- **Monopolar Cautery**
- The consensus of the majority of studies is that electrocautery dissection is associated with less operative time and intraoperative blood loss compared with sharp techniques.
- This technique, however, may cause increased postoperative morbidity with respect to pain and return to normal diet and activity.
- **Cold (Sharp) Dissection**
- The early techniques of performing tonsillectomy with cold dissection followed by suture ligation was initially used by all otolaryngologists approximately 20 years ago. Now, however, it is used in only 10% of cases nationally.
- Decreased postoperative pain.
- In summary of the techniques listed,
- Electrocautery appears to be the most commonly performed.
- Electrocautery groups appear to have decreased surgery, anesthesia, and operating room times.
- There are no reported differences in hemorrhage rates, but there does appear to be a need for significantly more analgesic doses after surgery.
- The other articles cite a slightly higher rate of hemorrhage with a "steel tonsillectomy technique".
- **Tonsillotomy:**
- Also called partial tonsillectomy or intracapsular tonsillectomy.
- It is partial removal of tonsils in the presence of specific indications:
 - o Children
 - o Presence of OSA
 - o No history of recurrent/chronic tonsillitis
- The size of tonsils is dramatically reduced but the tonsillar capsule is not dissected from the muscles and nerve endings are spared.
- No risk of bleeding.
- No pain postoperatively.
- Patient should be warned that tonsillar remnant may cause recurrent tonsillitis in the future and tonsillectomy may be necessary.

- **Steps of Surgery:**
- Under GA with endotracheal intubation.
- Rose's position (supine position with head extended by placing a pillow under shoulders and a rubber ring placed under the head)
- Hyperextension should always be avoided.
- Boyle-Davis mouth-gag.
- Grasp the tonsil with curved Allis forceps and retract it medially and slightly inferiorly to place the mucosa of the anterior pillar under tension.
- Make an incision in the anterior pillar just lateral to the mucosal reflection onto the tonsillar surface
- The anterior pillar should be preserved.
- It is necessary to dissect the mucosa and underlying soft tissue from the tonsil capsule.
- Entry into the tonsil at the superior pole is a technical error that leads to difficulty in further dissection of the tonsil because of an inability to identify the plane between the capsule and the surrounding soft tissue.
- Once the capsule of the superior pole has been identified, the tonsil is retracted inferiorly.
- Countertraction on the soft palate by suction held by the assistant often makes this dissection easier and assists in identification of the correct plane of dissection.
- Vessels can frequently be identified as they enter the tonsil capsule and can be controlled with electrocautery before being transected.
- As the tonsil is dissected inferiorly and freed from the surrounding muscular attachments, the remaining mucosa of the anterior and posterior pillars must be incised.
- Postoperative discomfort and deformity can be minimized by keeping the line of the mucosal incision immediately adjacent to the tonsil, thereby preserving as much mucosa as possible.
- When the inferior pole of the tonsil has been reached, the tonsil can easily be amputated by electrodissection.
- A gauze sponge is placed in the fossa and pressure applied for a few minutes.
- Care must be taken to avoid leaving excessive tonsil tissue at the inferior pole, as well as avoid unnecessary excessive dissection into the hypopharynx and tongue base.
- Pharynx is irrigated with saline solution.
- The mouth gag should be closed and opened once or twice to ensure that bleeding is not being controlled merely by mouth gag compression.
- If no bleeding is evident, the pharynx is suctioned and the patient is returned to the anesthesia staff for awakening and extubation.



(1) Knife in kidney tray, (2) & (3) Toothed and non-toothed Waugh's forceps, (4) Tonsil holding forceps, (5) Tonsil dissector and anterior pillar retractor, (6) Luc's forceps, (7) Scissor, (8) Curved artery forceps, (9) Negus artery forceps, (10) Tonsillar snare, (11) Boyle Davis mouth gag with three sizes of tongue blades, (12) Doyen's mouth gag, (13) Adenoid curette, (14) Tonsil swabs, (15) Nasopharyngeal pack, (16) Towel clips.

○ **Post-OP Management:**

- Intravenous fluids are continued until the patient awakens sufficiently to begin oral intake.
- Routinely beginning a liquid diet with progression to a soft diet is standard.
- Patients who are unable to tolerate an adequate oral diet, have excessive vomiting, or have evidence of bleeding should not be discharged on the same day but rather be observed until they stabilize.
- Dietary restrictions.
- Analgesia
- Antibiotics (No evidence)

- **Criteria for admission post T&A:**

1. OSA
2. Craniofacial syndromes
3. < 3 yrs
4. Live > 60 min away
5. Potential for neglect
6. Complex medical problems
7. Complications: emesis, dehydration, bleeding & obstruction

Complications:

- Intra-Op:

1. Bleeding.
2. Injury to surrounding structures (teeth, tonsillar pillars, uvula, soft palate, tongue or superior constrictor muscle) due to bad surgical technique.

- Post-OP:

1. Bleeding.
2. Infection of tonsillar fossa may lead to parapharyngeal abscess or otitis media.
3. Dehydration due to pain.
4. Aspiration of blood and Atelectasis.
5. Pulmonary edema caused by sudden relief of long-standing airway obstruction.
6. Scarring in soft palate and pillars.
7. Tonsillar remnants due to inadequate surgery, may get repeatedly infected.
8. Hypertrophy of lingual tonsil.
9. C1-C2 subluxation from hyperextension (Down syndrome)
10. Eagle syndrome.

Post-Tonsillectomy Bleeding:

- **Types:**

- 1. **Primary Bleeding (<24h):**

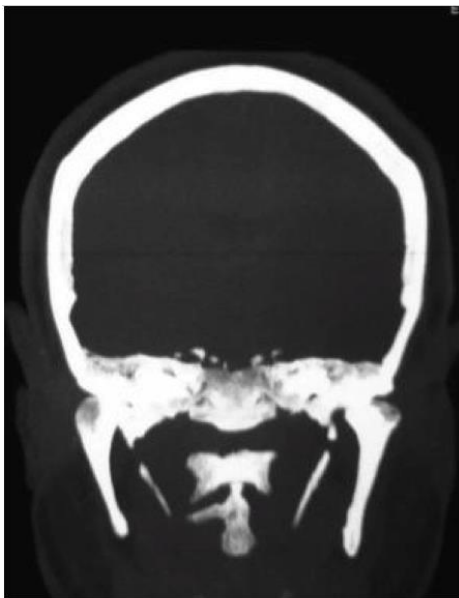
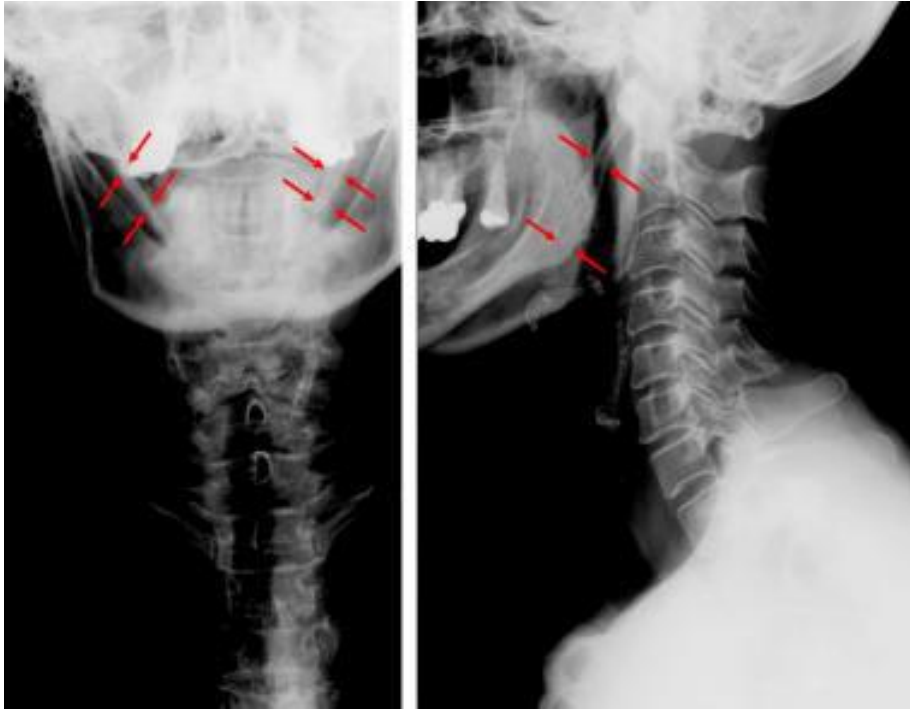
- Due to inadequate homeostasis intra-OP.
 - Controlled by removal of the clot, application of pressure or vasoconstrictor.
 - Presence of a clot prevents the clipping action of the superior constrictor muscle on the vessels which pass through it.
 - If above measures fail, ligation or electrocoagulation of the bleeding vessels can be done under general anaesthesia.

- 2. **Secondary Bleeding (>24h):**

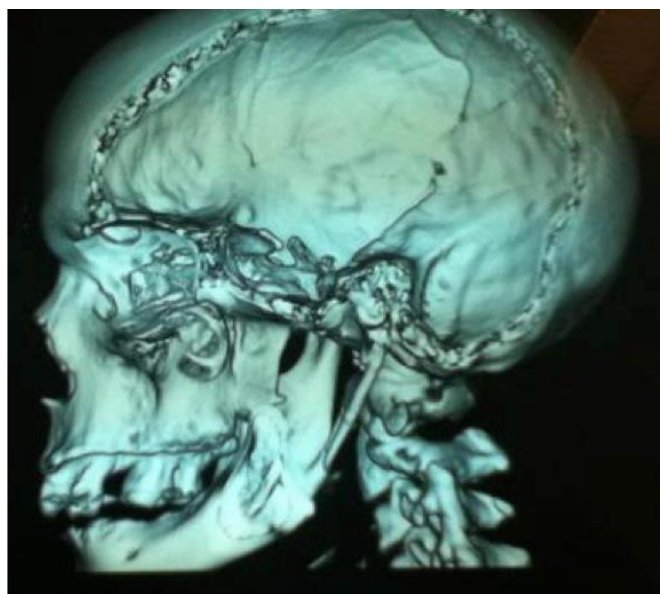
- Usually 5-10 days postop.
 - Due to eschar sloughing off.
 - Due to infection and eschar sloughing off.
 - Controlled by removal of the clot, application of pressure or vasoconstrictor.
 - Administration of systemic Antibiotic.
 - For profuse bleeding, general anaesthesia is given and bleeding vessel is electrocoagulated or ligated.

Eagle syndrome:

- Recurrent pain in the oropharynx and face due to an elongated styloid process or calcified stylohyoid ligament.
- Typically occurs after pharyngeal trauma or tonsillectomy
- Incidence is about 0.16%.
- Normal length of styloid process is 3.0 cm.
 - Styloid process length in Eagle syndrome is 4-5 cm.
- Clinical picture:
 - Ipsilateral dull and persistent pharyngeal pain, centered in the ipsilateral tonsillar fossa, which can be referred to the ear and exacerbated by rotation of the head.
 - Other symptoms include dysphagia, sensation of foreign body in the throat, tinnitus, or cervicofacial pain
- Diagnosis:
 - Plain x-ray
 - CT scan (Best).
 - Relief of discomfort by LA injection in Anterior pillar (diagnostic).
- Treatment:
 - Conservative
 - Surgery (styloidectomy):
 - Intraoral (transpharyngeal) or cervical approach.
 - For severe cases only.



CT scan: calcification of the stylohyoid ligament.



Three-dimensional reconstruction CT scan.

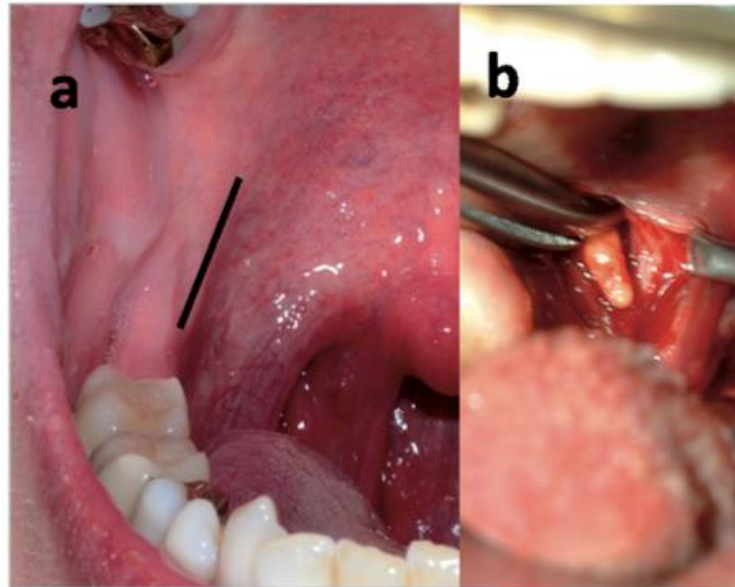


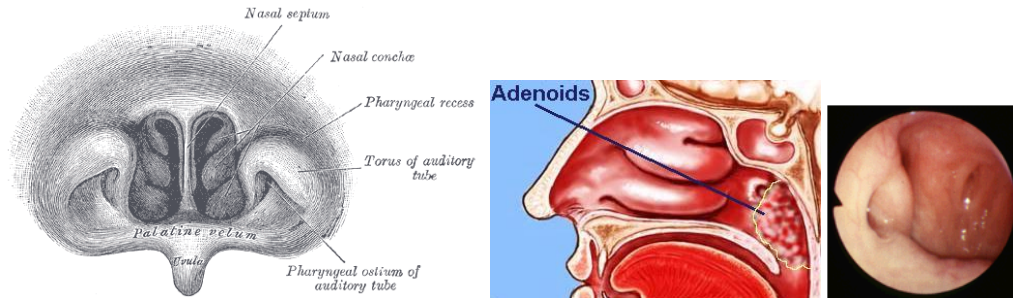
Fig. 2. Schematic presentation of the retromolar, paratonsillar approach (a) and the tip of the elongated styloid process (b). After blunt dissection the styloid process could be identified and easily prepared to the skull base by using a round hypophysis ring curette. After the resection near the skull base by a small luer bone rongeur or neurosurgical bone punch the mucosal defect was closed primarily.



Fig. 5 – Extraoral approach to styloid process.

Adenoid:

- Lymphoid tissue at junction of the roof and posterior wall of the nasopharynx.
- Laterally, the adenoids end in a depression known as the fossa of Rosenmüller, medial to the opening of the eustachian tube and torus tubarius.
- Inferiorly, adenoid tissue extends to the superior margin of the superior constrictor muscle and forms what is known as Passavant's ridge.



- Ridges separated by deep clefts.
- No crypts and No capsule
- Forms the bulk of the lymphoid tissue (Waldeyer's ring)
- Present at birth.
- Physiological enlargement in childhood up to the age of 6 in response to antigen presentation
- Atrophy at puberty and almost completely disappears by age of 20.
- Nasopharyngeal adenoid tissue serves as a **mechanical obstruction** to the eustachian tube orifice, thereby leading to difficulty in ventilation of the middle ear mucosa, and a **reservoir of bacteria**, which may contribute to episodes of otitis media.
- Most commonly cultured bacteria:
 1. Haemophilus influenzae,
 2. Group A beta-hemolytic Streptococcus
 3. Staphylococcus aureus
 4. Moraxella catarrhalis
 5. Streptococcus pneumoniae.
- **Histology:**
 1. Pseudostratified ciliated columnar epithelium. (**Respiratory epithelium**)
 2. Transitional epithelium.
 3. Stratified squamous epithelium.
- **Chronically infected adenoid have less clearing ciliated mucosa and more antigen-presenting squamous mucosa**

- **Arterial Blood supply:**
 - From branches of External carotid Artery:
 - 1. **Ascending Pharyngeal Artery (Mainly)**
 - 2. **Ascending Palatine Artery** (branch of Facial artery)
 - 3. **Pharyngeal Artery** (branch of Maxillary artery)
- **Venous Drainage:**
 - Pharyngeal veins → Facial and internal jugular veins.
- **Causes of Adenoid hypertrophy:**
 - 1. Physiological enlargement in childhood.
 - 2. Recurrent attacks of Rhinosinusitis.
 - 3. Chronic tonsillitis
 - 4. Allergic rhinitis.
- **Sensory innervation:**
 - Glossopharyngeal (CN-IX) and Vagus nerve. (CN-X)
 - Referred pain to ear/throat.
- **Clinical Classification of Diseases of the Adenoids:**
- **Acute adenoiditis**
 - Rhinorrhea (purulent), nasal obstruction, fever, OM
 - Sicker than with typical URI
- **Recurrent acute adenoiditis**
 - **4 or more attacks over 6/12.**
 - Consider immunodeficiency or other comorbidity.
 - May consider Abx prophylaxis [daily low-dose (one-half to one-third the full dose) or episodic prophylaxis.
 - Tough to distinguish from recurrent sinusitis (X-ray to assess sinuses)
- **Chronic adenoiditis**
 - Persistent nasal discharge; halitosis; PND; chronic congestion
 - Tough to differentiate from sinus disease; OM may help
 - Think of Extraesophageal reflux
- **Obstructive adenoid hyperplasia**
 - Chronic nasal obstruction (associated with snoring and obligate mouth breathing); rhinorrhea; hyponasal voice.
- **Clinical Features:**
 - Depend on the available space in the nasopharynx and size of the adenoid mass.

Nasal Symptoms:

- Bilateral nasal obstruction (**Most common**)
- Nasal discharge due to choanal obstruction and associated chronic rhinitis.
- Sinusitis due to persistence of nasal discharge and infection.
- Snoring

- Hyponasality.
- Epistaxis.
- OSA

Aural Symptoms:

- Eustachian tube obstruction leading to retracted TM and OME and conductive hearing loss.
- Recurrent AOM due to spread of infection via the eustachian tube.

General Symptoms:

- Mouth breathing.
- Nocturnal cough due to post nasal drip.
- Disturbed sleeping.
- Fail to thrive because respiration and feeding cannot take place simultaneously.
- Pulmonary hypertension and Cor pulmonale. due to long-standing nasal obstruction.

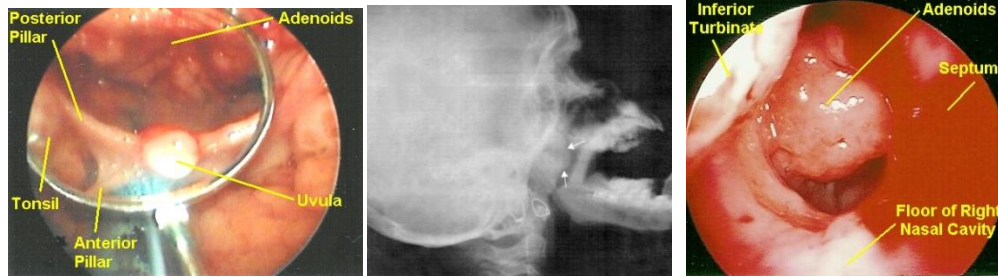
- Adenoid face due to chronic nasal obstruction and mouth breathing:

1. Hitched up upper lip with incompetent lip seal.
2. Prominent and crowded upper teeth.
3. Retrognathic mandible.
4. Elongated face with dull expression.
5. Dark circles under eyes
6. Hypoplastic maxilla
7. High arched palate
8. Elevated nostrils.



- **Diagnosis:**

1. Mirror:
 - Need cooperation.
2. Lateral neck x-ray:
 - Neck extension during nasal inspiration.
 - Evaluate size of adenoids and the available nasopharyngeal air space.
3. Flexible endoscope:
 - Reliable and dynamic diagnostic method (Best)



- **Adenoid Grading System:**

1. **Grade 0:** Absent
2. **Grade 1:** Obstruction < 25%
3. **Grade 2:** Obstruction 25-50%
4. **Grade 3:** Obstruction 50-75%
5. **Grade 4:** Obstruction > 75%



-
- **Management:**
- 1st and 2nd grades of obstructions:
 - o Nasal obstructions are usually due to either allergic or inflammatory pathology.
 - o Decongestant nasal drops and antihistamine can improve the condition without need for surgery.
- 3rd and 4th grades of obstructions:
 - o Symptoms are marked
 - o Adenoidectomy is always recommended

Nasopharyngitis

- **Acute infection** of the nasopharynx may be an isolated infection confined to this part only or be a part of the generalised upper airway infection.
- Caused by viruses (common cold, influenza, para-influenza, rhino or adenovirus) or bacteria (especially streptococcus, pneumococcus or Haemophilus influenzae).

- **Chronic infection** is associated with chronic infections of nose, paranasal sinuses and pharynx.
- Commonly seen in heavy smokers, drinkers and those exposed to dust and fumes.

- **Clinical Features:**

- Dryness and burning of the throat above the soft palate.
- Pain and discomfort localized to the back of nose with some difficulty on swallowing.
- In severe infections, Fever and enlarged cervical lymph nodes.
- Examination of nasopharynx reveals congested and swollen mucosa often covered with whitish exudate.
- **In chronic conditions**, postnasal discharge and crusting with irritation at the back of nose is the most common complaint.
- Patient has a constant desire to clear the throat.

- **Treatment:**

- Mild cases clear up spontaneously.
- In severe cases with general symptoms, systemic antibiotic may be necessary.
- In children, there is associated adenoiditis which causes nasal obstruction, and requires nasal decongestant drops.
- Chronic infections of the nose, paranasal sinuses and oropharynx should be attended to.
- Excessive smoking and drinking should be corrected.
- Preventive measures should be taken to avoid dust and fumes.
- Alkaline nasal douche helps to remove crusts and mucopus.
- Steam inhalations are soothing.

- **Indications of Adenoidectomy:**

1. Airway obstruction causing:
 - Snoring
 - Mouth breathing
 - Speech problems and hyponasality
 - OSA
 - Orofacial/dental abnormalities
 - Cor pulmonale
2. Recurrent/Chronic infection: (**Done independent of Adenoid size**)
 - Adenoiditis
 - OME
 - Sinusitis

- **Contraindications of Adenoidectomy:**

1. Acute URTI.
2. Coagulation abnormalities.
3. Cleft palate or submucous palate: **Risk of velopharyngeal Insufficiency**

- **Partial adenoidectomy:**

- Removing the upper part of the adenoid for relief of nasal obstruction while leaving the lower portion of the adenoid intact to ensure velopharyngeal competence in patients with submucous cleft palate.
- Done by suction diathermy or Transnasal endoscopic approach.

Pre-OP Evaluation:

- Examination of the oropharynx to rule out any cleft abnormalities and signs of a submucosal cleft palate (**bifid uvula, zona pellucida, posterior palatal notching**).



- **Steps of Surgery:**

- Under GA with endotracheal intubation.
- Rose's position (supine position with head extended by placing a pillow under shoulders and a rubber ring placed under the head)
- Hyperextension should always be avoided.
- Boyle-Davis mouth-gag.
- Soft palate retraction by rubber catheter.
- Nasopharynx examination by mirror.
- Adenoid curette is placed against the posterior edge of the vomer distance from the eustachian tube orifice and torus tubarius.
- The curette is depressed against the vomer edge, and with the thumb used as a fulcrum, the blade is pushed over the posterior nasopharyngeal wall.
- Nasopharyngeal packing with moist adenoid sponges.
- **Persistent bleeding** is generally a result of retained fragments of adenoid tissue which can be controlled intra-op by:
 1. Removal of remaining tissue by curette or suction-cautery.
 2. Packing soaked with hydrogen peroxide for 10 mins
 3. Otrovon nasal drops.
 4. Repeat Packing for longer time to ensure hemostasis.
 5. If bleeding is not controlled, posterior nasal pack is left for 24 hours.



Methods of Adenoidectomy:

1. Curette.
 - Simple
 - Higher risk of bleeding
 - Higher residual adenoid tissue
2. Suction-cautery:
 - Quick
 - Good homeostasis with low blood loss.
 - Used under direct vision.
 - More painful with bad smell post-op
3. Microdebrider.
 - Quick
 - Less bleeding
 - Precise tissue removal
 - Improved visualization through endoscopic approach.
4. Coblation:
 - Quick
 - Less bleeding

- **Post-OP Management:**

- Minimal postoperative care.
- No dietary restrictions.
- Analgesia
- Antibiotics ??
- Otrivin nasal drops and normal saline drops.

- **Complications:**

1. Postoperative bleeding which can be controlled by otrivin nasal drops and by cauterization in OR if persistent.
 2. Velopharyngeal insufficiency VPI and hypernasality in patients with palatal abnormalities.
 3. Injury to posterior tip of inferior turbinate causing massive bleeding.
 4. Injury to the eustachian tube orifice and torus tubarius which can be recognized by the appearance of cartilage in the adenoid specimen.
 5. Nasopharyngeal stenosis due to scarring.
 6. Recurrence due to regrowth of adenoid tissue left behind.
 7. Atlanto-axial joint subluxation (Grisel's Syndrome)
- Placement of the curette under direct visualization will reduce the likelihood of injury to adjacent structures.

- **Postadenoidectomy hypernasality:**
- VPI, observed in 0.03-0.06% of cases, occurs as a result of incomplete closure of the palate to the posterior and lateral nasopharyngeal wall, where the adenoids had previously been located
- VPI is observed **transiently** in more than half the patients undergoing an adenoidectomy and usually resolves in 2-4 weeks.
- Persistent VPI (>3 mo) occurs in 1 in 1500-3000 adenoidectomies.
- Persistent VPI occurs more often in children who have generally decreased muscle tone or a known palatal abnormality.
- Hypernasality after adenoidectomy can also occur in children with **normal palatal function** who have:
 - o Enlarged tonsils or
 - o Prominent remaining adenoid tissue on posterior pharyngeal wall
- Incomplete removal of the adenoid tissue should be avoided and enlarged tonsils should be removed at the time of adenoidectomy to prevent the risk for postoperative hypernasality.
- Some recommend performing a partial adenoidectomy, leaving the inferior portion of the adenoid pad, in patients at high risk for VPI
- Treatment initially consists of speech therapy for as long as 12 months, depending on the severity of the VPI.
- Surgery is required in 50% of persistent cases.

- **Grisel's syndrome:**
- Non-traumatic subluxation of the atlanto-axial joint.
- Results from a pathological relaxation of the ligaments surrounding the joint following an inflammatory process (URTI, Tonsillitis) or otolaryngological surgical procedures.
- Children are most frequently affected and classically have torticollis associated with neck stiffness or pain upon neck movement.
- **Risk factors:**
 - o Down syndrome.
 - o Excessive passive rotation and hyperextension of patient's head.
 - o Use of Monopolar suction cautery during adenoidectomy.
- **Diagnosis:**
 - o **Lateral plain cervical x-ray:**
 - Not sensitive.
 - Diagnosis is made if atlanto-dens interval of **> 5 mm.**
 - o **CT and MRI:**
 - More sensitive method of diagnosis.
- **Treatment:**
 - o **Broad spectrum prophylactic Antibiotic.**
 - o **Cervical collar.**
 - o **Orthopaedic or neurosurgical consultation.**

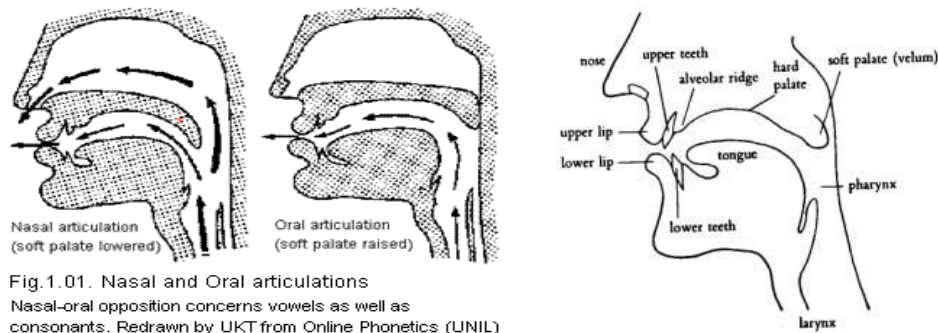


(1) Knife in kidney tray, (2) & (3) Toothed and non-toothed Waugh's forceps, (4) Tonsil holding forceps, (5) Tonsil dissector and anterior pillar retractor, (6) Luc's forceps, (7) Scissor, (8) Curved artery forceps, (9) Negus artery forceps, (10) Tonsillar snare, (11) Boyle Davis mouth gag with three sizes of tongue blades, (12) Doyen's mouth gag, (13) Adenoid curette, (14) Tonsil swabs, (15) Nasopharyngeal pack, (16) Towel clips.

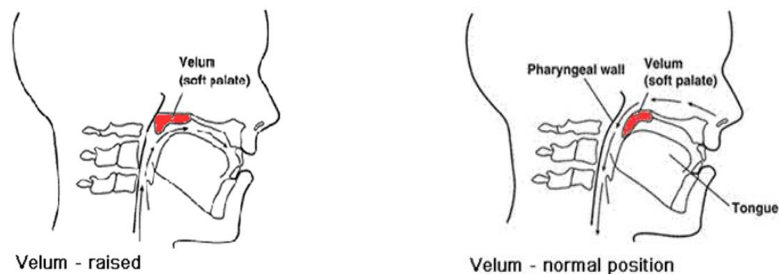
Velopharyngeal Insufficiency:

- **Anatomy**
- The soft palate is commonly referred to as the Velum.
- Acts as a sphincteric valve to separate the oral and nasal cavities during speech and swallowing.
- Velopharynx (VP):
- Anteriorly: the velum
- Laterally: lateral pharyngeal walls
- Posteriorly: posterior pharyngeal wall

- In speech production, the velopharynx is considered an articulator, as are the jaw, tongue, lips, pharynx, and larynx.
- In English, **ALL** phonemes are produced with oral airflow that requires VP closure to exclude nasopharyngeal airflow.
- **Exception** of /m/, /n/, and /ng/ which require an open nasopharyngeal valve and nasal airflow to produce these sounds.



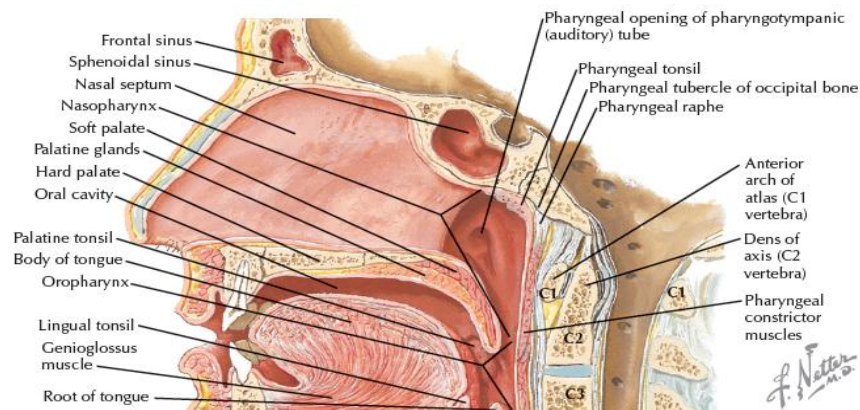
- **Functions of VP valve:**
- **Closure:**
 1. Prevent the nasal escape of air and sound during production of oral sounds (most consonants and all vowels)
 2. Prevent nasal regurgitation during swallowing
- **Opening:**
 1. Permit nasal consonant production /m/, /n/, and /ng/
 2. Permit nasal respiration.



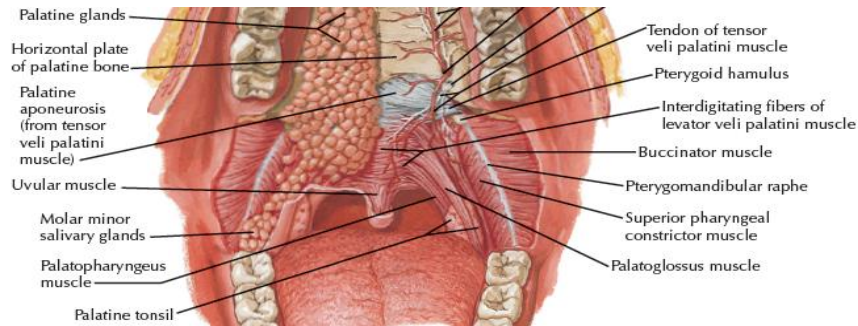
- **VP Closure (VPC):**
- At the level of the VP isthmus, located along the inferior aspect of the adenoid bed.
- Elevation and retraction the of soft palate postero-superiorly.
- Simultaneous contraction of the lateral and posterior pharyngeal walls that move medially and anteriorly about 10 mm.

Six muscles comprise the velopharyngeal sphincter:

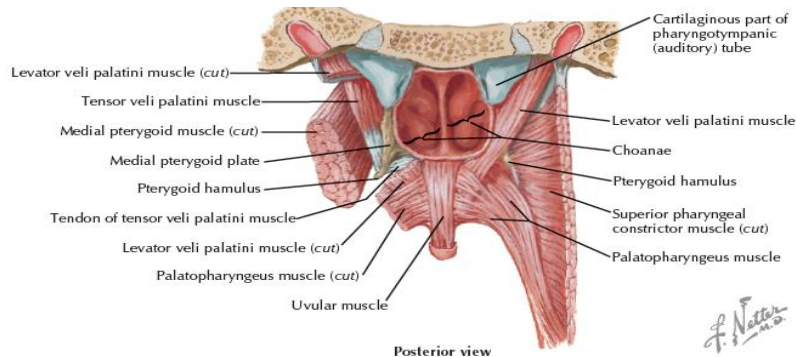
1. Tensor veli palatini
2. Levator veli palatini
3. Musculus uvulae
4. Palatoglossus
5. Palatopharyngeus:
6. Superior constrictor



- **Tensor veli palatini:**
- Tenses the soft palate and pulls it laterally.
- Opens the auditory tube during swallowing.
- Innervated by mandibular branch of the **Trigeminal nerve**.
- **Levator veli palatini:**
- **Main muscle** mass of the velum.
- LVP fibers interdigitate in the midline with the contralateral LVP to form the VP sling.
- Pulls the velum in a posterosuperior direction
- Major elevator of the velum
- Innervated by **Pharyngeal plexus**.
- **Palatoglossus:**
- Lowers the velum
- Elevates the tongue upward and backward to constrict the pillars
- Innervated by **Pharyngeal plexus**.



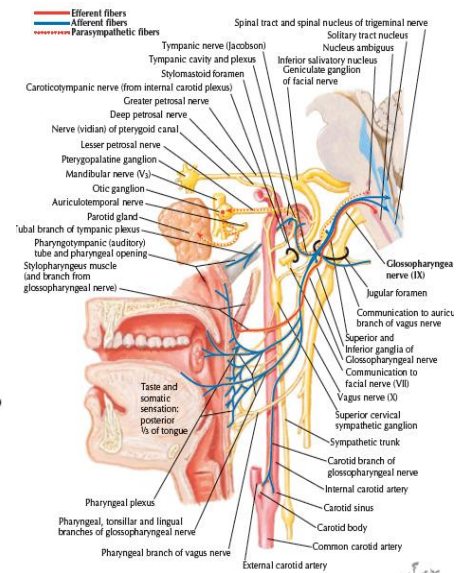
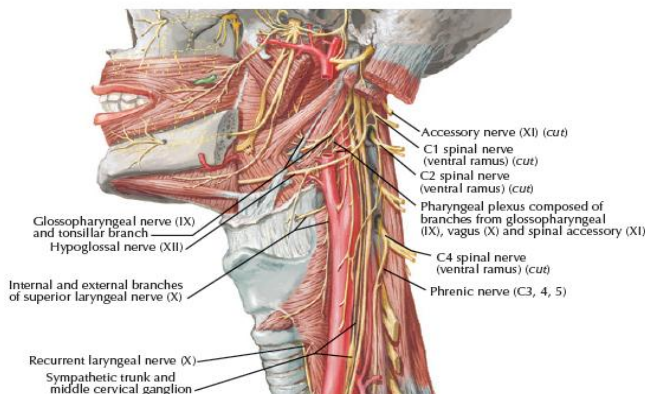
- **Palatopharyngeus:**
- Narrows the velopharyngeal orifice by adducting the posterior pillars and constricting the pharyngeal isthmus.
- Raises the larynx.
- Lowers the pharynx.
- Innervated by **Pharyngeal plexus.**
- **Musculus uvulae:**
- Elevate the uvula
- Add bulk to the VP to occlude the VP port during speech and deglutition.
- Innervated by **Pharyngeal plexus.**
- **Superior constrictor:**
- Medial movement of the lateral aspects of the pharyngea walls.
- May function to draw the velum posteriorly
- Innervated by **Pharyngeal plexus.**



- **Passavant's ridge:**
- 20% of population
- Posterior wall movement seen in some individuals during speech or swallow, caused by a similar movement of the uppermost fibers of the superior constrictor and palatopharyngeus.



- **Levator veli palatini** → elevation and posterior motion of the velum
- **Palatoglossal + palatopharyngeus muscles** → pull the palate down
- **Musculus uvulae** → adds bulk to the dorsal side of the velum.
- **Uppermost fibers of the superior constrictor + palatopharyngeus** → lateral wall motion.
- **Pharyngeal plexus:**
 - Vagus nerve – Motor
 - Cranial part of Accessory nerve - Motor
 - Glossopharyngeal nerve - Sensory
 - Superior cervical sympathetic ganglion.
 - Motor Supply to **ALL** the muscles of the pharynx (**except** stylopharyngeus, by the glossopharyngeal nerve).
 - Motor Supply to **ALL** the muscles of the soft palate (**except** tensor veli palatini, by the mandibular division of the trigeminal nerve)
 - Sensory innervation to oropharynx and hypopharynx



- **Velopharyngeal inadequacy:**
 - Failure of the sphincteric mechanism due to structural or neurologic impairment or mechanical interference leading to loss of normal functions of the velopharyngeal valve
 - 3 etiologic categories:
 1. Insufficiency
 2. Incompetence
 3. Mislearning

- **Insufficiency:**
- **Structural defects** that result in insufficient tissue to accomplish closure
- Ex: Cleft palate

- **Incompetence:**
- Impairment of motor control secondary to neurologic **dysfunction**, such as paresis or paralysis.
- Ex: Post skull base surgery and tumors around the jugular foramen and vagus nerve. (CNS) impairment from brainstem stroke.

- **Mislearning:**
- Etiologic factors independent of structural defects or neuromotor pathology.

- **Structural Abnormalities**
- **Cleft palate (most common)**
- Congenital short palate
- Occult submucous cleft palate
- Absent musculus uvulae.
- Hypertrophic tonsils by impeding palate closure
- Post tonsillectomy or other pharyngeal surgery causing tethering of velum by scarring
- Post adenoidectomy.

- **Neuromuscular Abnormalities**
- **Structurally intact VP**
- Muscular dystrophies
- Myasthenia gravis
- Down syndrome
- CVA
- Palatal paralysis

- An adenoidectomy can create or unmask VPD in any of these settings.
- Large adenoid can compensate for a short palate or a poorly mobile palate.
- With removal of the adenoidal tissue, the mechanism of compensation is eliminated.
- Velopharyngeal inadequacy has been reported to occur after 1 in 1500 adenoidectomies.

- **The characteristic speech patterns of VPD :**
- 1. Increased transmission through the nasal cavity
- 2. Hypernasality due to excessive nasal resonance.
- 3. Nasal emission due to turbulent airflow through the nasal cavity
- 4. Weak pressure consonants: due to inability to generate sufficient oral pressure due to nasal leak
- 5. Nasal regurgitation

- **VPI ASSESSMENT**

- The diagnosis of VPI is typically ascertained by:

1. Routine historical intake
2. Physical examination
3. Combination of perceptual speech, instrumental evaluation.

- **Detailed history** is elicited for any known syndrome or neurologic abnormalities.

- Inquiry about prior surgical intervention within the oral cavity or pharynx.

- **Physical examination** includes detailed inspection of the oral and nasal cavities.

- Attention to the velar height, palatine tonsil size, tongue, palatal and uvular appearance, nasal mucosa, turbinates and septum.

- Attention to the OP searching for signs of a submucosal cleft palate (bifid uvula, zona pellucida, posterior palatal notching)

- Physical examination always overlooks an occult cleft palate and thus other instrumental evaluation is needed to make this diagnosis.

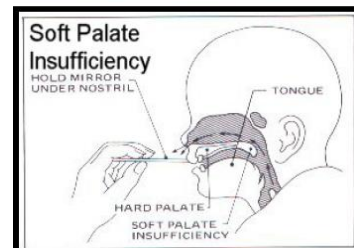
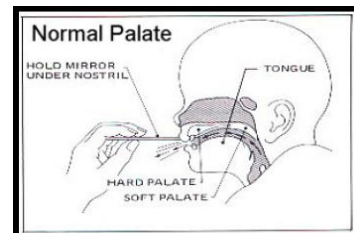
- **Perceptual Speech Evaluation**

- **The gold standard of VPI evaluation.**

- Listening to the spontaneous or prompted production of specific pressure phonemes while monitoring for misarticulations, hypernasal speech, nasal rustle, and facial grimacing.

- Facial grimacing is a compensatory action to narrow the external nares to decrease nasal air emission during non-nasal speech.

- Use of the dental mirror placed under the nares during vowel production, may also assist in determining the amount of visible nasal air emission as shown by fogging of the mirror.



- **Speech Videofluoroscopy**

- A small amount of barium is introduced through the nose to coat the velopharynx.

- Under fluoroscopy, the child repeats or reads a set of phoneme-specific speech tasks.

- **Limitations:**

- Radiation
- Cooperation
- Difficult to evaluate postsurgical changes.

- **Speech Endoscopy**
- Flexible nasopharyngoscope is introduced into the velopharynx.
- Most useful tool for evaluation.
- A series of words, phrases, and sentences are spoken while
- Endoscopy allows assessment of the static anatomy and visualization of the VP closure

- **Four primary closure patterns.**

- These patterns are best understood in terms of the residual gap orientation on incomplete closure of the velopharynx.

- **Coronal pattern**

- Coronally oriented gap
- **Most common (55%)**
- Major contribution to closure is from the palate (velum)

- **Sagittal pattern**

- Sagittally oriented gap
- Least common (10-15%)
- Major contribution to closure is from the lateral walls.

- **Circular pattern**

- Circular central gap
- 10% to 20% of the population
- Major contribution to closure is from the velum and the lateral walls.

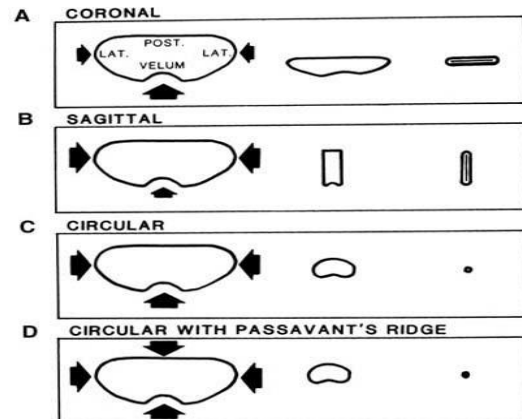
- **Circular pattern with passavant's ridge**

- 15% to 20% of the population
- Motion of the velum and lateral walls with Passavant's ridge
- Knowing where the "gap" is should allow the clinician to decide how best to obturate that gap.

- **Management:**

- **Speech therapy**

- Not appropriate for abnormal structure only (e.g., VPD with intact articulation after adenoidectomy or tumor resection)
- Provide compensatory strategies, if the structural abnormality cannot be corrected
- Mild VPD
- Post-op to learn to use the new structure



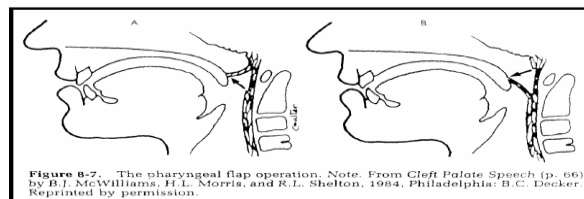
- **Prosthetic Management**
- **Palatal lift** pushes the palate up and back to contact the posterior pharyngeal wall,
- Not good for short palate
- **Obturator** places a round piece of acrylic into the velopharynx



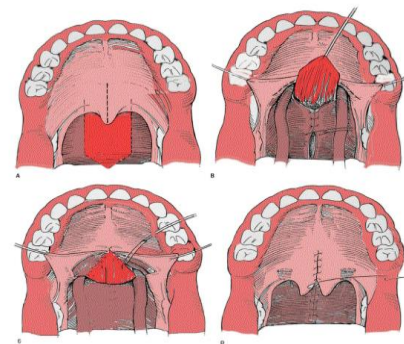
- **Surgical Management:**

1. **Pharyngeal Flap**

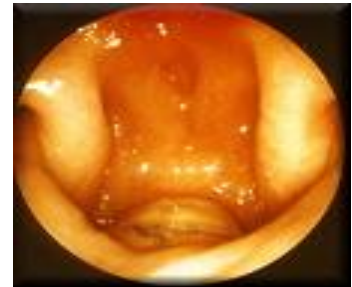
- Indicated in case of good lateral wall motion and poor velar motion (**Sagittal** or **circular** closure patterns)
- The most widely performed surgical procedure to improve VPI.
- Tissue from the posterior pharyngeal wall is attached to the soft palate, creating a midline subtotal obstruction of the oral and nasal cavities with 2 small lateral openings.
- Pharyngeal flaps can be based superiorly (most common) or inferiorly.
- Inferiorly based posterior wall flap was initially described .
- Modification to this procedure was done by using a superiorly based flap after founding that an inferiorly based flap tended to contract and tether the palate downward over time, worsening the patient's VPI.



- The soft palate is incised in the sagittal midline from the uvula toward the junction of the soft and hard palate.
- The superiorly based pharyngeal flap is elevated off the prevertebral fascia.
- The flap is inset to the soft palate and sutured to the nasal side of the soft palate with interrupted sutures.
- The donor site is partially closed with 3-0 Vicryl sutures.
- The oral side of the soft palate is then closed with interrupted sutures

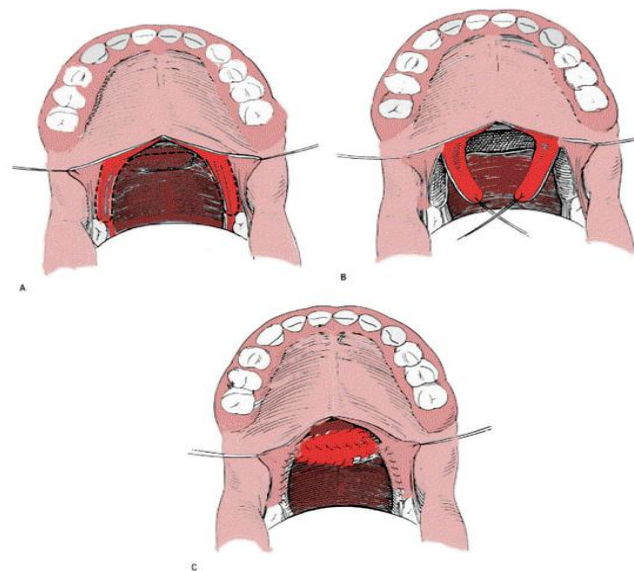
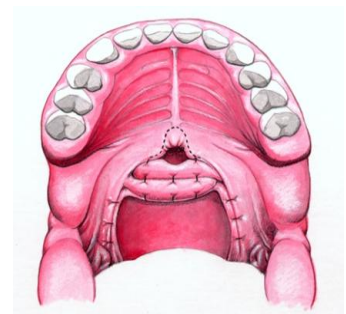


- **Outcome:**
- 90% resolution of hypernasality.
- Failure to improve is related to inadequate flap width either by design or due to contracture.
- A flap that is too wide narrows the lateral ports and produces hyponasal speech.
- **Postoperative airway obstruction** is most likely to occur within the first 24 hours and resolved within 2 days in more than 90% of patients.



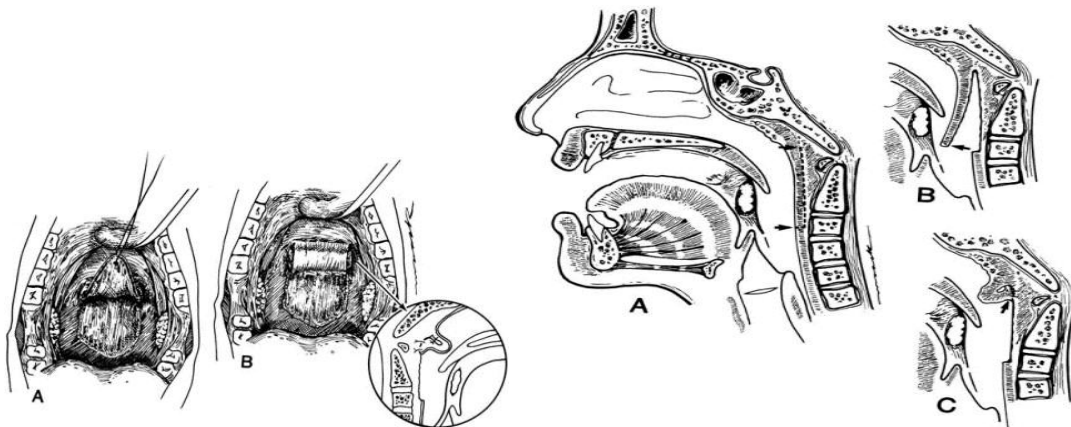
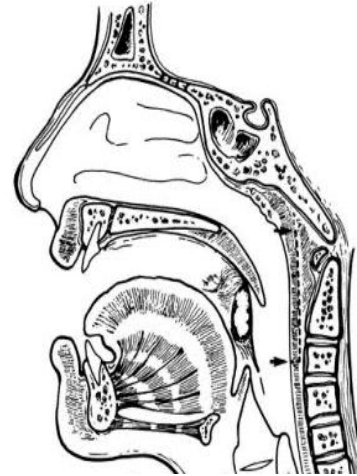
2. **Sphincter Pharyngoplasty**

- Patients with good velar elevation but poor lateral wall motion are good candidates for sphincter pharyngoplasty.
- Ideal for a **coronal** closure, and useful in the **circular** closure.
- Narrow the central velopharyngeal orifice and minimizing airflow through the nose during speech.
- It accomplishes this by rearranging palatopharyngeus myomucosal flaps raised from the posterior tonsillar pillars, which are transposed to the posterior pharyngeal wall and approximated on the midline.
- This procedure tightens the central orifice without creating lateral ports.
- **Decrease risk of post-op airway obstruction.**



3. Posterior Pharyngeal Wall Augmentation

- Indicated mainly for very small posterior midline gap.
- **Persistent postadenoidectomy VPI** is also an appropriate indication for this procedure
- A wide variety of implantable materials have been tried.
- Substances are typically placed within the retropharyngeal space just deep to the superior constrictor muscle but superficial to the pharyngobasilar fascia.
- Most commonly fat and calcium hydroxylapatite.
- Outcome data are scant. varying degrees of success have been reported.
- **Rolled posterior pharyngeal wall:**
- Raising a U-shaped posterior pharyngeal wall myomucosal flap to the tubercle of the atlas. The tip of the flap is connected by deep bites to the base of the flap. By securing the knots, the flap is rolled on itself, creating a soft tissue bulge or augmentation.

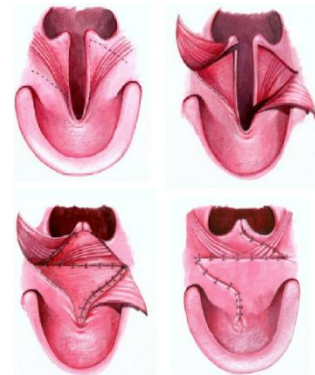


4. Palatal Lengthening

- Goals of this procedure are to lengthen the palate, occlude a small gap in the velopharyngeal port, and retrodisplace the palate in a more physiologically normal place.

5. V-Y pushback procedure

6. Furlow double-opposing Z-plasty palatoplasty



Snoring and Obstructive Sleep Apnea (OSA):

- **Physiology of Sleep:**
- Average adult needs 8 hours of sleep to feel rested.
- Sleep is entered through NREM sleep.
- Normal sleep consists of 4-6 cycles of NREM sleep alternating with REM sleep every 90-120 minutes.
- First two cycles are predominantly NREM.
- Later stages (early am) are predominantly REM.
- Normal sleep is divided into:
 - **Non-rapid eye movement (NREM) sleep:**
 - Forms 80% of total sleep.
 - Occurs in four stages:
 - Stage N1:
 - Constitutes 5% of sleep.
 - Transition from wakefulness to sleep.
 - Characterized by **theta waves**.
 - Person can be easily aroused from this stage.
 - Stage N2:
 - Constitutes 50% of sleep.
 - Characterized by **sleep spindles or 'K' complexes**, and decrease in muscle tone.
 - Stage N3:
 - Constitutes 10% of sleep.
 - Characterized by **delta waves**.
 - It is deep most restful sleep.
 - Dominates the first third of the night's sleep.
 - **Rapid eye movement (REM) sleep:**
 - Forms 20% of total sleep.
 - **OSA occurs in this stage due to decreased muscular tone.**
 - Dominates the last third of the night's sleep.
 - Characterized by rapid eye movements, increased autonomic activity with erratic cardiac and respiratory movements.
 - EEG shows **sawtooth waves**.
 - Dreaming occurs in this stage but muscular activity is decreased so that dreams are not enacted.

Table 54-3. Differences between non-REM and REM sleep

	Non-REM	REM
Duration	75-80% of sleep	20-25% of sleep
Eye movements	No eye rolling	Rapid conjugate eye movements
Autonomic activity	Less autonomic activity gives slow heart rate, low BP, slow and steady respiration	Increased autonomic activity with fluctuations in BP, heart rate and respiration
Brain activity	Minimal	Brain is active (REM sleep is also called activated brain in a paralysed person)
Muscular activity	Functional but less	Decreased. Since muscles are relaxed, snoring and OSA occurs in this stage.
EEG	Passes from alpha to delta waves from stage I to IV	Mixed frequency, low voltage waves with occasional bursts of saw-tooth waves
Dreaming	No	Yes

- **Snoring:**
- Sound generated by vibration of pharyngeal soft tissues.
- Muscles of pharynx are relaxed during sleep and cause partial obstruction.
- Breathing against obstruction causes vibrations of uvula, soft palate, tonsillar pillars and base of tongue producing sound.
- 80% of snoring sound comes from the palate.
- Sound as loud as 90 dB has been recorded during snoring.
- Louder during inspiration than expiration.
- Affects at 40% of men and 20% of women.
- Often accompanies sleep-disordered breathing with significant positive correlation between severity of OSA and snoring intensity.
- Isolated snoring:
 - o Habitual audible snoring occurs with AHI < 5 events/hour without daytime symptoms.
 - o Not associated with arousals, desaturations, airflow limitation or arrhythmias.
 - o Doesn't require polysomnography for diagnosis.
- Causes of snoring:
 - o In children, the most common cause is adenotonsillar hypertrophy.
 - o In adults, cause of snoring can be one of the following:

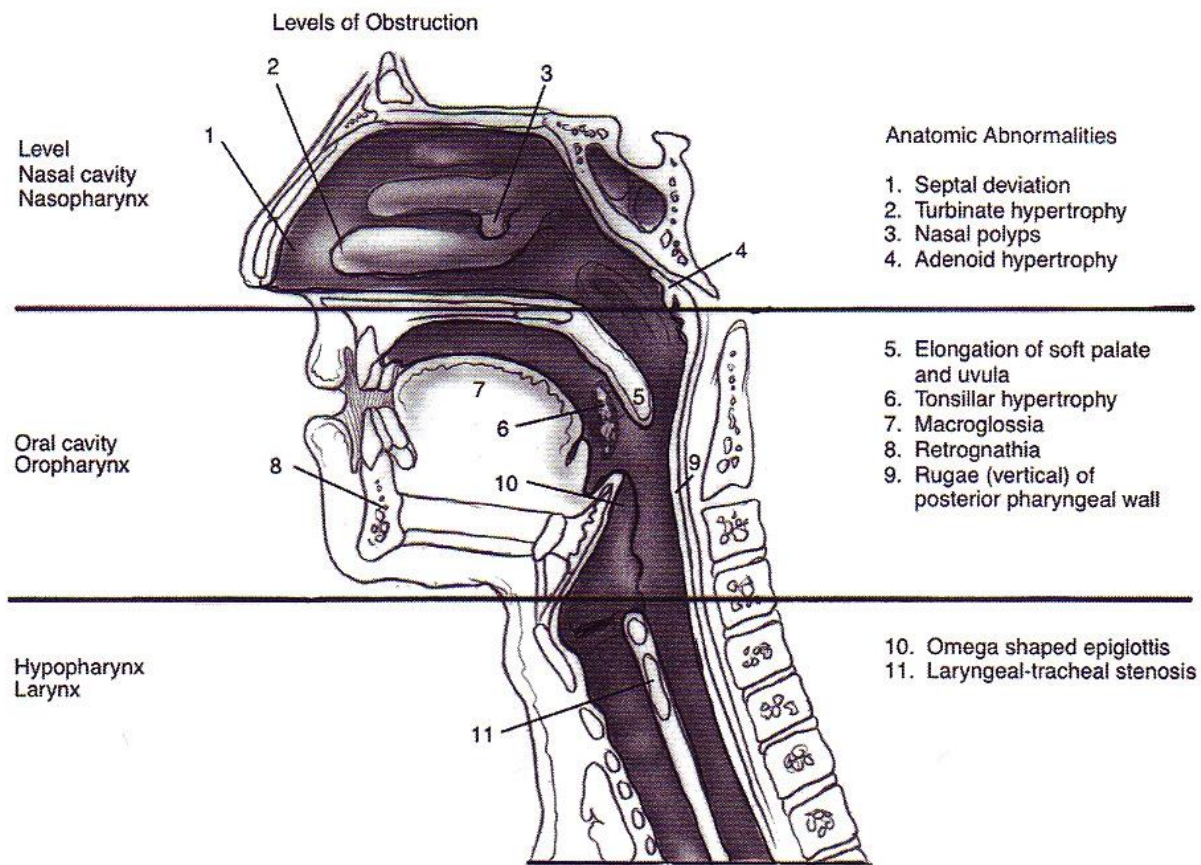
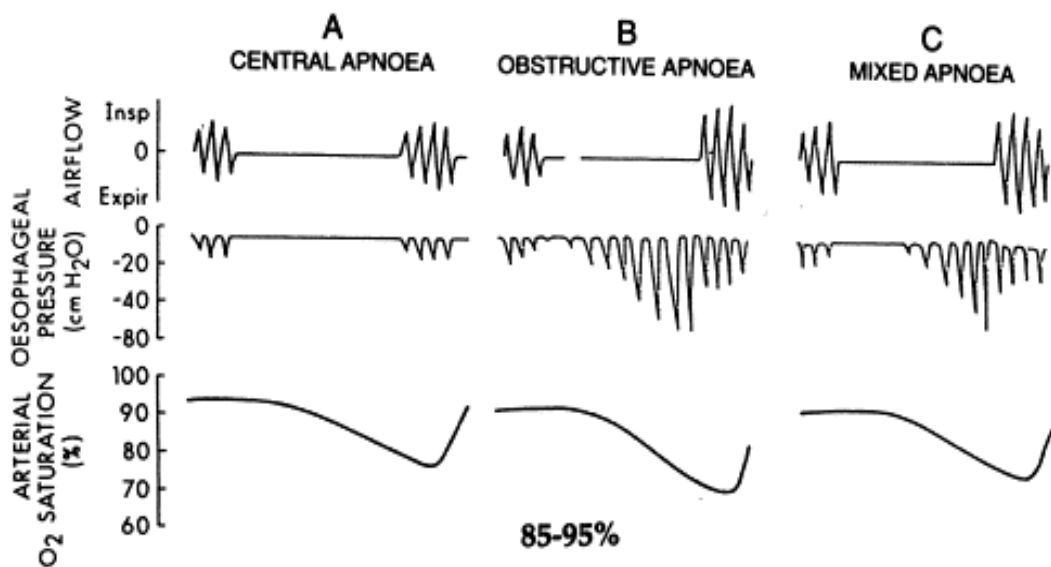
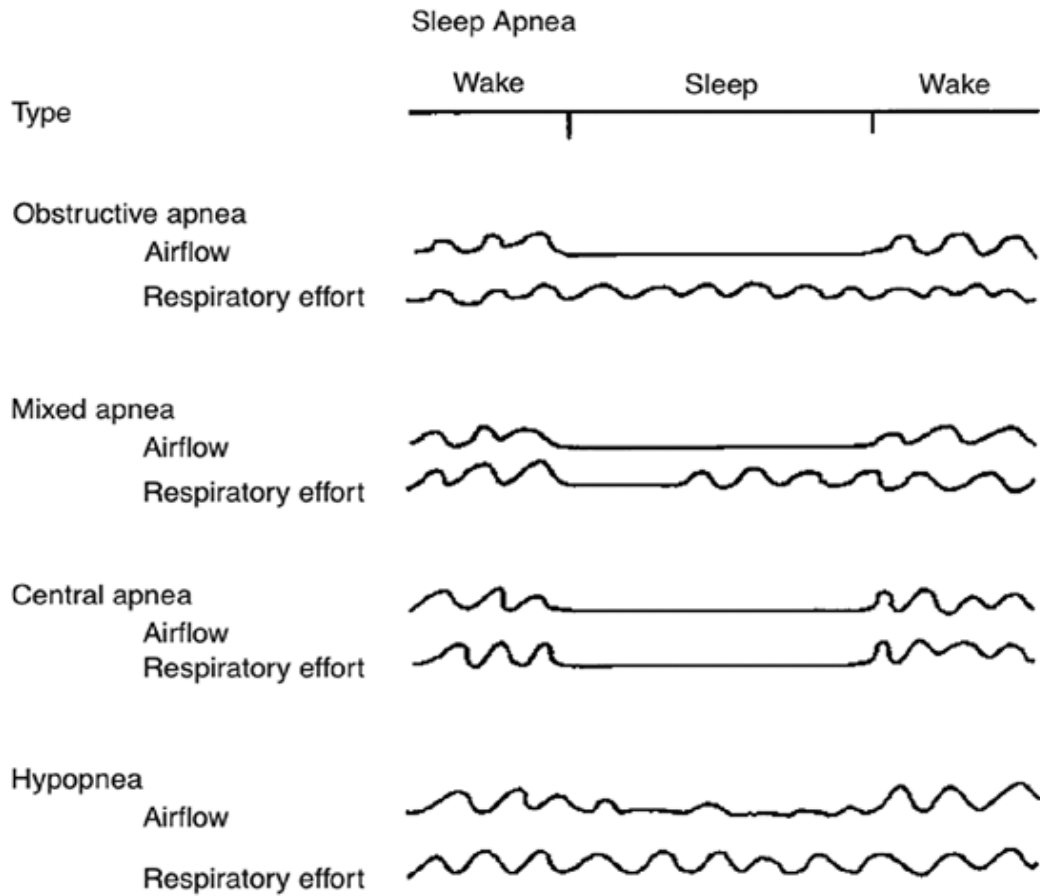


Figure 46.2 Levels and sites of possible airway obstruction in patients with sleep disorders.

- Definitions:
 - **Apnea:**
 - Cessation of airflow.
 - Lasting for ≥ 10 seconds.
 - Leading to arousal.
 - **Hypopnea:**
 - Reduction of $\geq 30\%$ in airflow.
 - Lasting for ≥ 10 seconds.
 - Associated with $\geq 4\%$ oxygen desaturation.
 - **OR**
 - Reduction of $\geq 50\%$ in airflow.
 - Lasting for ≥ 10 seconds.
 - Associated with:
 - $\geq 3\%$ oxygen desaturation.
 - **OR**
 - Arousal.
 - **Apnea-hypopnea index (AHI):**
 - Number of (apneas and hypopneas) per hour of total sleep time.
 - **Respiratory effort-related arousal (RERA):**
 - Respiratory effort that does not meet criteria for an apnea or hypopnea.
 - Lasting for ≥ 10 seconds.
 - Leading to arousal.
 - **Respiratory disturbance index (RDI):**
 - Number of (apneas, hypopneas and RERAs) per hour of total sleep time.
- Types of sleep apnea:
 - **Obstructive:**
 - Breathing cessation with presence of respiratory effort.
 - Collapse of the upper airway resulting in cessation of air flow.
 - Most common type.
 - **Central:**
 - Breathing cessation with absence of respiratory effort.
 - Airways are patent but brain fails to signal the muscles to breath.
 - Causes includes:
 - Opioid
 - Neuromuscular disorders
 - Brainstem lesions.
 - **Mixed:**
 - Combination of both type.
 - Breathing cessation with absence of respiratory effort then followed by severe effort



- **Obstructive sleep apnea (OSA):**
- Sleep disorder that involves cessation or significant decrease in airflow in the presence of breathing effort.
- Common medical condition with significant adverse consequences.
- Prevalence in USA is 10%.

- Pathophysiology:
 - o Collapse of pharyngeal airway during sleep due to relaxation of the pharyngeal muscles.
 - o Oropharynx is the major area of airway obstruction leading to OSA.
 - o **Nasal obstruction is rarely the sole cause of OSA:**
 - It worsen the OSA by:
 - Increasing airway resistance.
 - Leading to mouth breathing which increases upper airway collapsibility.
 - Surgical correction of nasal obstruction allow for decreased CPAP levels to be used for subsequent treatment.

- Consequences of untreated OSA:
 - o Overall increase in morbidity and mortality.
 - o Increase risk of:
 - **Stroke**
 - **Cardiovascular events:**
 - Hypertension
 - Coronary heart disease
 - Congestive heart failure
 - Arrhythmias
 - Pulmonary hypertension
 - **Obesity-Hypoventilation Syndrome (Pickwickian syndrome):**
 - Obesity
 - OSA
 - Chronic Hypercapnia
 - **Metabolic syndrome:**
 - Obesity
 - Insulin Resistance
 - Hypertension
 - Dyslipidemia
 - **Gastroesophageal reflux disease**
 - **Fatal motor vehicle accidents.**

- Risk factors of developing OSA:

○ **Structural factors:**

- Soft tissue hypertrophy.
- Anatomic variations:
 - Facial elongation
 - Posterior facial compression
 - Retrognathia
 - Micrognathia
 - Mandibular hypoplasia
 - Inferior displacement of the hyoid
- Syndromes:
 - **Down syndrome:**
 - OSA in 60% of patients.
 - Anatomical consideration and tendency for obesity.
 - Persistent OSA in 80% post adenotonsillectomy.
 - Pierre Robin syndrome
 - Marfan syndrome
 - Prader-Willi syndrome

○ **Non-structural factors:**

- Obesity(BMI>35)
- Male gender
- Age (>65 Years)
- Postmenopausal state
- Alcohol and Sedative use
- Smoking
- Supine sleep position
- Familial factors
- Hypothyroidism
- Stroke

- Clinical picture:

○ **Nocturnal symptoms:**

- Loud snoring and bothersome to others.
- Witnessed apnea, choking or gasping episodes that arouse the patient from sleep.
- Restless sleep with frequent turning during the night.
- Nocturnal sweating
- Nocturnal enuresis
- Insomnia

○ **Daytime symptoms:**

- Excessive daytime sleepiness.
- Morning fatigue or irritability.
- Morning headaches
- Decreased cognitive function.
- Depression.
- Personality or mood changes.
- Decreased libido and impotence.

- Physical examination:

○ **General examination:**

▪ **Body mass index (BMI):**

- Calculated by dividing body weight in Kg by height in m².
- BMI does not actually measure body fat.
- Over-estimates adiposity on patients with more lean body mass (athletes).
- Increase risk when BMI > 35
- Classes:
 1. Underweight: **BMI < 18.5**
 2. Normal weight: **BMI 18.5 - 24.9**
 3. Overweight **BMI 25 - 29.9**
 4. Class 1 obesity: **BMI 30 - 34.9**
 5. Class 2 obesity: **BMI 35-39.9**
 6. Class 3 obesity: **BMI > 40**

▪ **Collar size:**

- Neck circumference at level of cricothyroid membrane.
- Better predictor of OSA than BMI.
- Increase risk of OSA when collar size:
 - **> 42 cm** in males
 - **> 37 cm** in females.

▪ **Waist-hip ratio:**

- Waist circumference at its narrowest part just above the belly button.
- Hip circumference at its widest part of the buttock or hip.
- Assess central abdominal obesity.
- Better predictor of ventilation abnormalities than BMI.
- Increase risk of ventilation abnormalities when waist-hip ratio:
 - **> 0.9** in males
 - **> 0.85** in females.

▪ **Facial characteristics:**

- Anatomical features predispose to airway obstruction.
- Mandible and maxilla position and size.
- Dysmorphic features.

○ **Throat examination:**

- Tonsil size
- Tongue size and position
- Elongation of palate and uvula
- Crowding of oropharynx
- Lingual tonsils size
- **Friedman score:**
 - Modified Mallampati score.
 - Performed with mouth open and without protrusion of tongue.
 - Classes:
 - **Class 1:**
 - Full visibility of hard palate, soft palate, uvula and tonsils.
 - **Class 2:**
 - Full visibility of hard palate, soft palate, uvula and upper pole of tonsils.
 - **Class 3:**
 - Full visibility of hard palate, soft palate and base of uvula.
 - **Class 4:**
 - Full visibility of hard palate only.

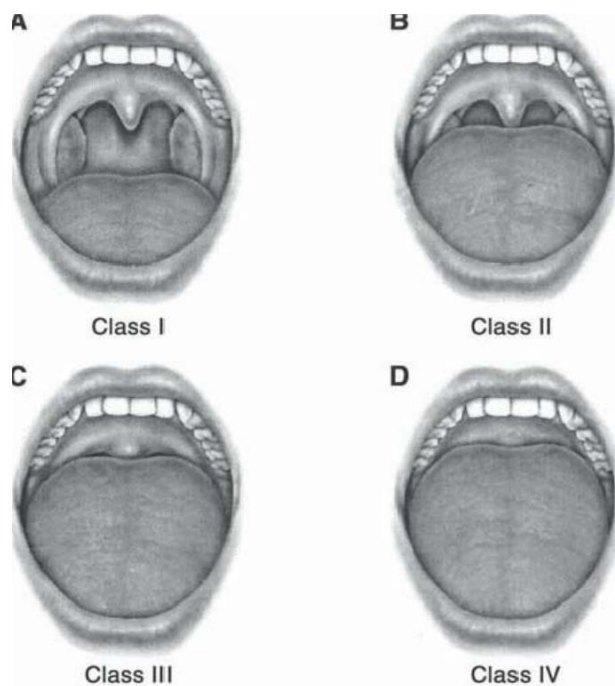


Figure 135.7 Friedman modification of Mallampati classification. **A:** Class I: Full visibility of tonsils, uvula, soft palate, and hard palate. **B:** Class II: Visibility of hard palate, soft palate, upper portion of tonsils, and uvula. **C:** Class III: Soft palate, hard palate, and base of uvula are visible. **D:** Class IV: Only hard palate is visible.

- **Flexible scope examination:**
 - **Nasal examination:**
 - External deformity
 - Adequacy of nasal valve
 - Septal position
 - Turbinate size
 - Presence of polyps
 - **Nasopharyngeal examination:**
 - Presence of adenoid.
 - **Oropharyngeal and hypo-pharyngeal examination:**
 - Position and the size of the tonsils.
 - Size of lingual tonsils.
 - **Müller maneuver:**
 - Used to trigger airway collapse.
 - Performed in awake patient by generating negative pressure with inhalation against closed glottis (reverse Valsalva) with closed nose and mouth while the flexible scope is introduced into the oropharynx at 2 levels:
 1. Level of soft palate (Retropalatal)
 2. Level of base of tongue (Retrolingual)
 - Useful for guiding surgical decision when area of collapse is only retropalatal.
 - Doesn't predict the surgical outcomes after UPPP due to potential underestimation of degree of collapse at retrolingual level (better assessed with DISE).
 - **Modified Müller maneuver:**
 - Grading of degree of airway collapse and reduction in cross sectional area:
 - **1+:** < 25% reduction.
 - **2+:** < 50% reduction.
 - **3+:** < 75% reduction.
 - **4+: > 75% reduction (Required surgical intervention at that level).**
 - **Laryngeal examination:**
 - Evaluate presence of laryngeal masses.
 - Laryngomalacia.
 - Status of vocal folds.

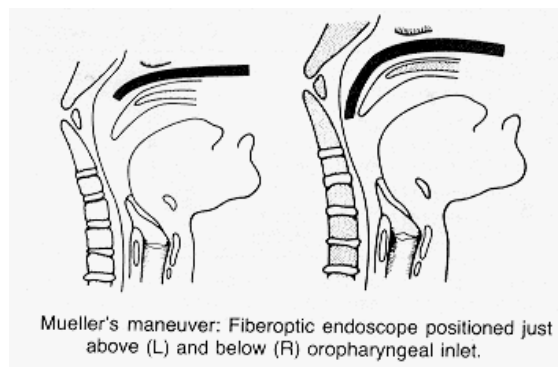




Fig. 27.3. Progressive collapse at the velopharyngeal level during a Müller maneuver



FIGURE 18-4. Fiberoptic view of the hypopharyngeal airway before the Müller maneuver. (From Troell RJ, Riley RW, Powell NB, Li K. Surgical management of the hypopharyngeal airway in sleep disordered breathing. *Otolaryngol Clin North Am* 1998;13:983.)

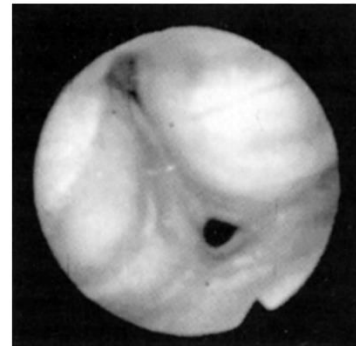


FIGURE 18-5. Fiberoptic view of hypopharyngeal collapse during the Müller maneuver. (From Troell RJ, Riley RW, Powell NB, Li K. Surgical management of the hypopharyngeal airway in sleep disordered breathing. *Otolaryngol Clin North Am* 1998;13:983.)



Upper airway endoscopy. (A) Retropalatal airway during Müller's maneuver. (B) Retropalatal airway during passive inspiration. (C) Retroglottal airway during Müller's maneuver. (D) Retroglottal airway during passive inspiration.

○ **Imaging:**

- Not frequently used.
- Poor sensitivity for diagnosing OSA.
- Provide limited information about obstruction during sleep as most of the radiologic techniques are performed in awake patients.
- **Cephalometric radiograph:**
 - Most frequently used imaging modality.
 - Available and relatively low cost.
 - Provide information on bony skeleton and overlying soft tissues.
 - Findings in OSA:
 - Reduction in mandible length
 - Inferior displacement of the hyoid
 - Narrow posterior airway space
 - Long soft palate

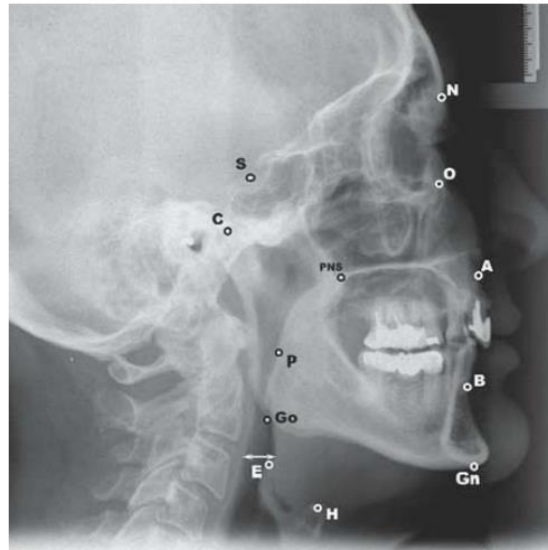


Figure 135.10 Lateral x-ray cephalogram. The cephalometric landmarks used in this study. *S* (sella), midpoint of sella; *N* (nasion), anterior point at the frontonasal suture; *ANS* (anterior nasal spine), most anterior point of the anterior nasal spine; *PNS* (posterior nasal spine), most posterior point of hard palate; *A* (subspinal point), deepest anterior point in concavity of the maxilla; *B* (supramental point), deepest anterior point in concavity of the mandible; *Gn* (gnathion), most inferior point of the mandibular symphysis; *H* (hyoid), most anterosuperior point of hyoid bone; *E* (epiglottis), base of epiglottis; *Go* (gonion), most posterior and inferior point of the angle of the mandible; *P* (soft palate), most inferior and posterior point of the soft palate; *MP* (mandibular plane), mandibular plane joining gonion with gnathion; *SNA* (deg), angle between nasion–sella line (NSL) and the line from *A* to *N* (measures projection, anterior or posterior of maxilla); *SNB* (deg), angle between NSL and the line from *B* and *N* (measures position of mandible); *ANB* (deg), angle between the line from *A* to *N* and the line from *B* to *N* (measures position of maxilla with mandible); *PNS–ANS*, linear distance between *PNS* and *ANS*; *ANS–H*, linear distance between *ANS* and *H*; *ANS–E*, linear distance between *ANS* and *E*; *PNS–P*, linear distance between *PNS* and *P* (measures length of velum of palate); *MP–H*, linear distance between the mandibular plane and *H*.

- Diagnosis of OSA:

○ **Epworth Sleepiness Scale (ESS):**

- Widely used tool to **assess daytime sleepiness**.
- OSA is suspected if **ESS score > 10**.

EPWORTH SLEEPINESS SCALE	
Please answer the following questions based on this scale:	
0.	Would never fall asleep
1.	Slight chance of dozing
2.	Moderate chance of dozing
3.	High chance of dozing

<u>Situation</u>	<u>Chance of Dozing</u>
Reading	_____
Watching TV	_____
Sitting in a public place (e.g., theater or meeting place)	_____
Driving a car, stopped at a traffic light	_____
As a passenger in a car for an hour without a break	_____
During quiet time after lunch without alcohol	_____
Lying down to rest when circumstances permit	_____
Total Score: _____	

Epworth Score <10 = Normal

- **Polysomnography (PSG):**

- Gold standard for diagnosis of OSA in Adults.

- Types:

- **In-center PSG:**

- Fully attended overnight sleep study.
- Minimum of seven parameters are recorded.
- Components:
 - 1. Electro-encephalogram (EEG):**
 - For sleep staging.
 - 2. Electro-oculogram (EOG):**
 - Measure eye movements.
 - 3. Electromyogram (EMG):**
 - Submental and tibial.
 - Assess for movement or atonia.
 - 4. Electro-cardiogram (ECG):**
 - Asses for any heartbeat changes.
 - 5. BP monitor:**
 - Asses for any blood pressure changes.
 - 6. Pulse oximetry:**
 - Detect any desaturation.
 - 7. Nasal/Oral airflow:**
 - Detect any reduction in airflow.
 - 8. Respiratory effort monitor:**
 - Detect presence or absence of thoraco-abdominal respiratory effort.
 - 9. Body-position monitor:**
 - Video recording of movements.
 - 10. Microphone:**
 - Auditory recording of snoring.
 - 11. Esophageal pressure:**
 - Optional.
 - Useful for detection of RERAs.

- **Portable PSG:**

- Home sleep testing (HST).
- Components:
 - **Pulse oximetry**
 - **Nasal/Oral airflow**
 - **Respiratory effort monitor**
 - **Heart rate**
- RDI in these devices is the number of apneas and hypopneas per hour of recording time.
- Many patients can be successfully diagnosed with sleep apnea using portable monitoring devices.

- **Indications of PSG in Children with OSA:**
 - **Adenotonsillar hypertrophy with presence of:**
 - Obesity
 - Down syndrome
 - Craniofacial anomalies
 - Neuromuscular disorders
 - Sickle cell disease
 - Mucopolysaccharidoses
 - **Discordance between tonsillar size and OSA symptoms.**
 - **Persistent OSA symptoms after adenotonsillectomy.**

**TABLE
135.16**

**INDICATIONS FOR THE USE
OF PORTABLE MONITORING
IN ADULTS—AASM GUIDELINES**

Patients with a high pretest probability of moderate to severe OSA
 Patients for whom in-laboratory PSG is not possible by virtue of immobility, safety, or critical illness
 To monitor response to non-CPAP treatments for OSA including oral appliances, upper airway surgery, and weight loss

**TABLE
135.17**

**CONTRAINDICATIONS FOR THE
USE OF PORTABLE MONITORING**

Patients with significant comorbid medical conditions (moderate to severe pulmonary disease, neuromuscular disease, congestive heart failure recent cerebrovascular accident, nocturnal hypoxemia requiring oxygen therapy)
 Patients suspected to have other sleep disorders (CSA, periodic limb movement disorder, insomnia, parasomnias, circadian rhythm disorders, or narcolepsy)
 General screening of asymptomatic populations

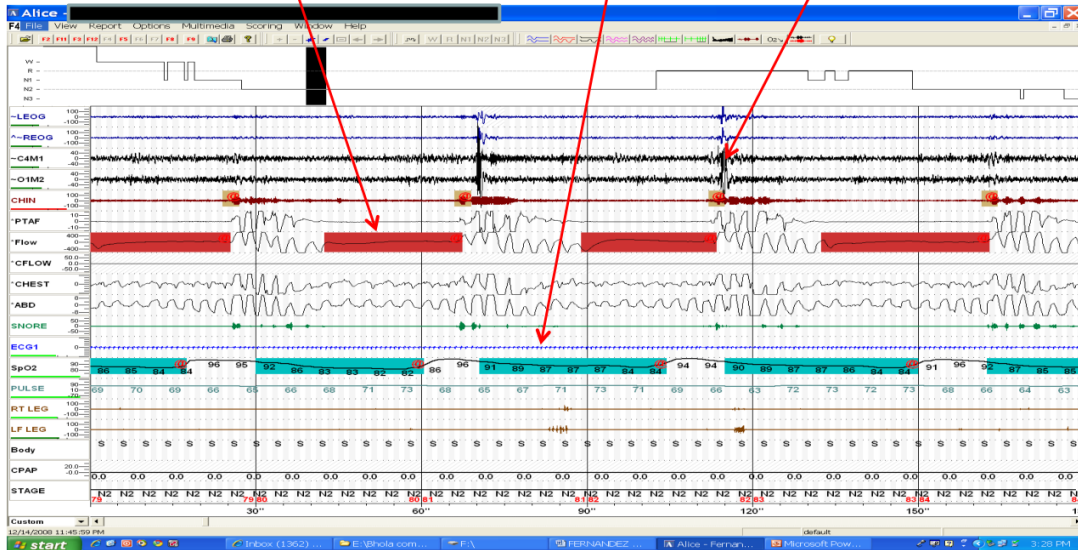
**TABLE
135.15**

CLASSIFICATION OF SLEEP STUDIES

Type 1	≥7 physiologic parameters monitored and sleep technician in attendance
Type 2	≥7 physiologic parameters monitored, unattended
Type 3	4–7 physiologic parameters monitored, unattended
Type 4	≥3 physiologic parameters monitored, unattended

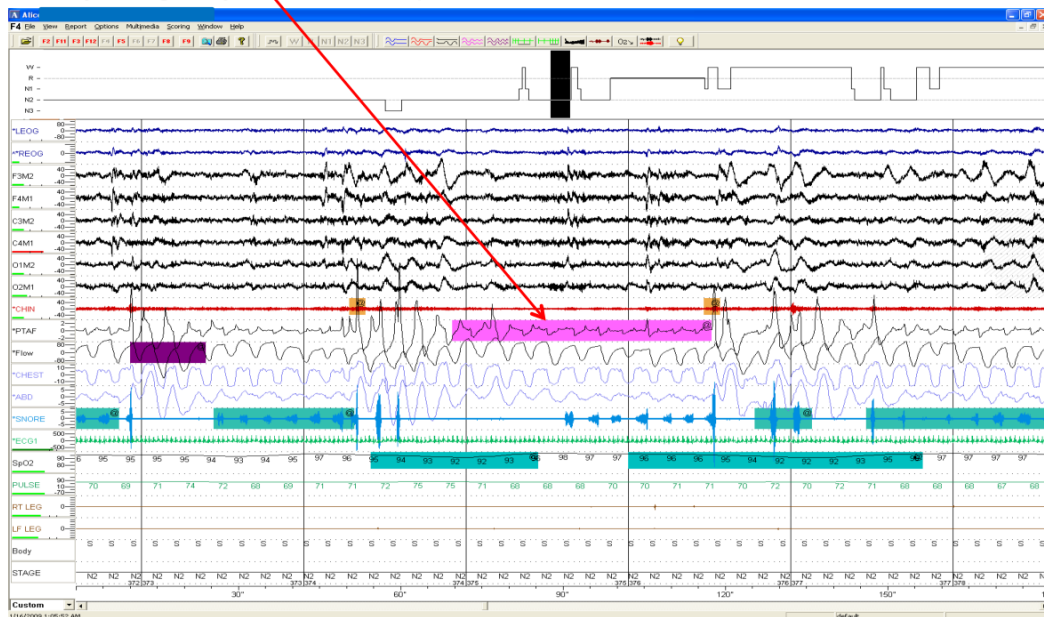
Obstructive Apnea

Complete airway collapse causing cessation of airflow at nose and mouth for at least 10 secs, with drop in oxygen level and arousal from sleep



Hypopnea:

- Partial airway collapse causing shallow breathing (airflow reduced by at least 30-50%) during sleep, lasting 10 seconds or longer, usually associated with a fall in blood oxygen saturation.
- Physiologically same as an apnea



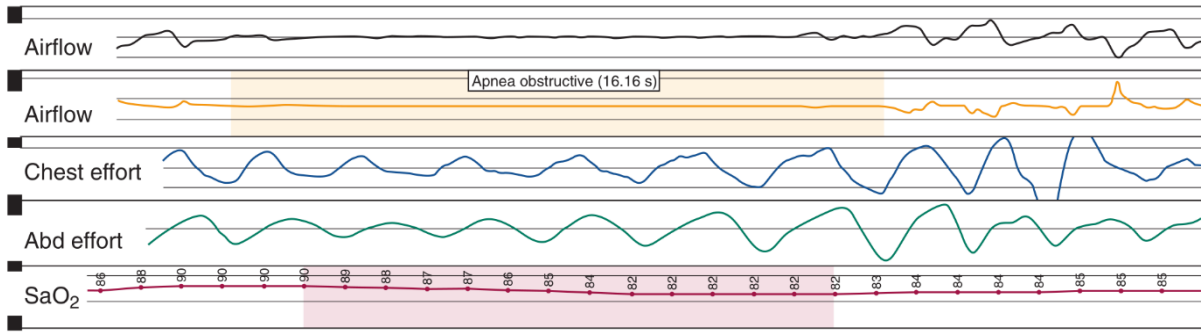


FIGURE 18-8. Polysomnographic tracing of an obstructive apnea. Abd, abdominal.

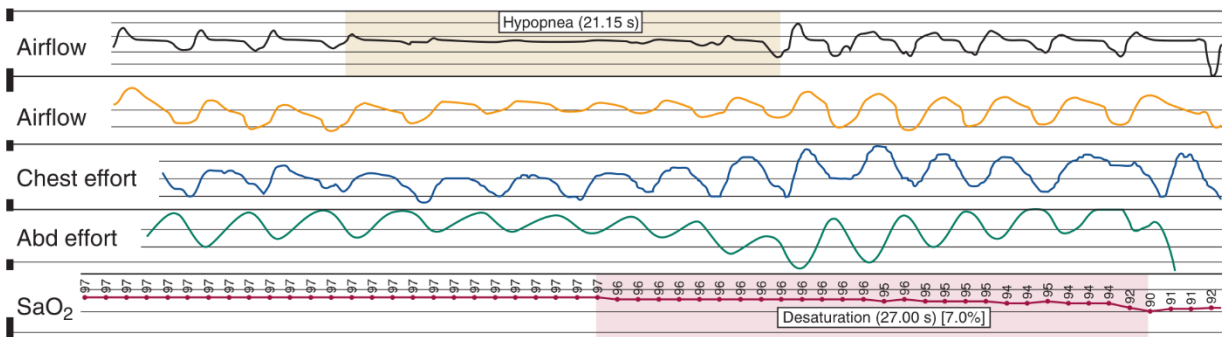


FIGURE 18-9. Polysomnographic tracing of a hypopnea. Abd, abdominal.

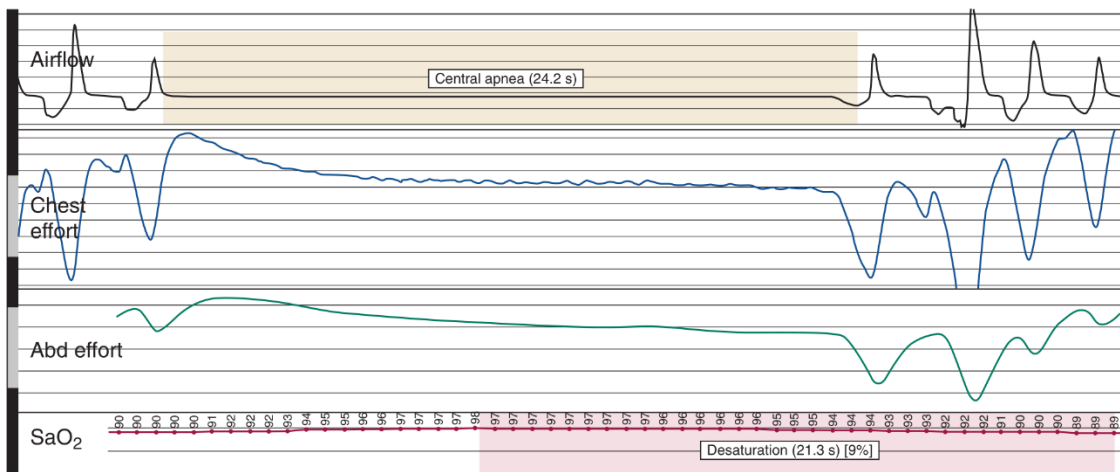
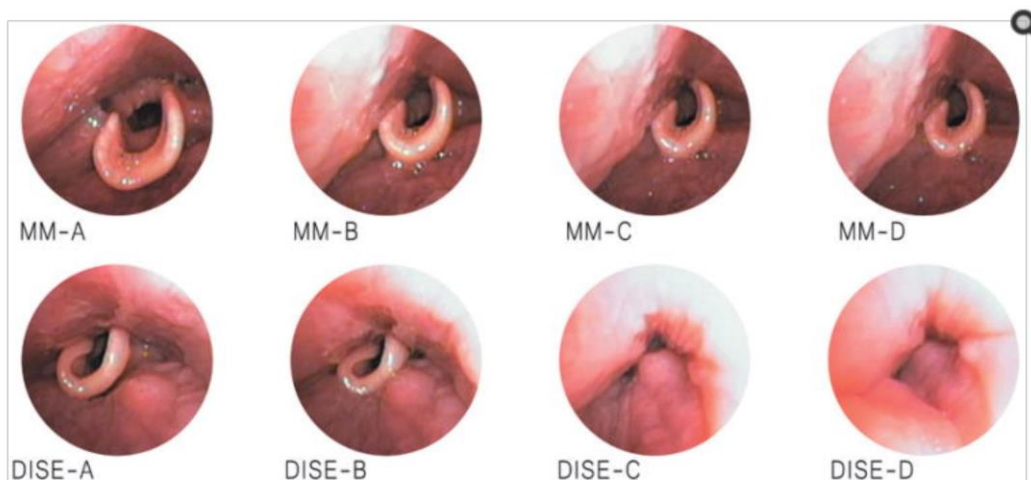


FIGURE 18-10. Polysomnographic tracing of a central apnea. Abd, abdominal. Abd, abdominal.

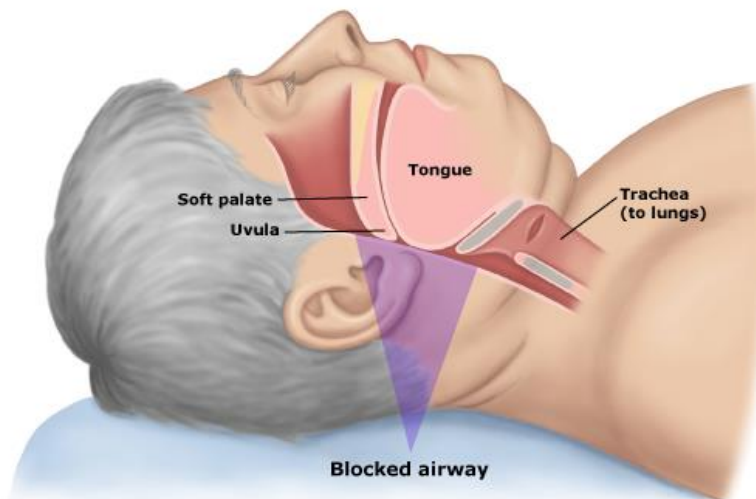
- **Severity of OSA in ADULTS:**
 - **Mild: 5-15** respiratory events/hour.
 - **Moderate: 15-30** respiratory events/hour.
 - **Severe: > 30** respiratory events/hour.
- **Severity of OSA in CHILDREN:**
 - **Mild: 1-5** respiratory events/hour.
 - **Moderate: 5-10** respiratory events/hour.
 - **Severe: > 10** respiratory events/hour.
- **Changes in sleep cycle as observed by PSG in patients with OSA:**
 1. **No reduction in total sleep time.**
 2. **Prolonged stage 1 NREM sleep:**
 - 30-50% of total sleep time.
 3. **Marked reduction in stage 3-4 NREM sleep:**
 - Slow-wave sleep.
 4. **OSA occurs in REM sleep:**
 - Decreased muscular tone.
- **Drug-Induced Sleep Endoscopy (DISE):**
 - Fiberoptic nasopharyngoscopy evaluation of site of airway collapse during pharmacologically induced sleep.
 - More physiological assessment of upper airway collapse.
 - More accurate tool than Mueller maneuver and awake fiberoptic nasopharyngoscopy for assessing location, severity, and pattern of airway obstruction during sleep.
 - Most commonly performed using combination of midazolam and propofol for sedation in OR.



An example of the endoscopic disparity in apparent degree of severity of retrolingual collapse occurring during inspiration on FNMM (MM-A through MM-D) and DISE (DISE-A through DISE-D) same patient. MM-A and DISE-A = beginning inspiration; MM-D and DISE-D = end inspiration. DISE, drug-induced sleep endoscopy; FNMM, fiber-optic nasal endoscopy with Müller's maneuver.

Treatment of OSA:

- Requires a comprehensive long-term treatment plan and follow-up.
- Should be approached in a stepwise manner.
- Algorithm for OSA treatment:
 - **Mild OSA (AHI 5-15):**
 - Behavior modification.
 - If no improvement consider CPAP or surgery.
 - **Moderate OSA (AHI 15-30):**
 - Behavior modification and CPAP.
 - If no improvement consider surgery.
 - **Severe OSA (AHI >30):**
 - Behavior modification and CPAP.
 - Consider surgery as adjunct to improve CPAP compliance.
 - If BMI > 40 refer for bariatric surgery.
 - Consider tracheostomy as last resort.
- Behavioral Treatment:
 - **Weight reduction:**
 - **First-line of therapy in overweight ADULTs with mild OSA.**
 - Recommended for all overweight patients.
 - Has significant beneficial effect on OSA.
 - 10% loss in body weight result in 25% decrease in AHI.
 - Bariatric surgery result in 75% reduction in RDI.
 - **Positional therapy:**
 - Head of bed elevation and lateral recumbent position.
 - Avoid supine position as it is associated with relapse of the tongue against the posterior oropharyngeal wall.
 - **Life style-modification:**
 - Avoid excessive caffeine, alcohol or nicotine consumption.



- **Medical Treatment:**

○ **Positive Airway Pressure (PAP):**

- Gold standard and first line of treatment for ADULTs with moderate to severe OSA.
- Acts as pneumatic splint prevents upper airway collapse by providing constant positive intraluminal pressure during inspiration and expiration.
- Optimum airway pressure for device to open the airway is determined during sleep study.
- Indications of PAP in pediatric OSA:
 - Craniofacial anomalies
 - Neuromuscular weakness
 - Obesity
 - Persistent OSA after adenotonsillectomy
- Types:
 - **Continuous Positive Airway Pressure (CPAP):**
 - Provides fixed level of positive airway pressure during both inspiration and expiration.
 - **Bilevel Positive Airway Pressure (BIPAP):**
 - Provides independent inspiratory positive airway pressure and expiratory positive airway pressures.
 - Expiratory positive airway pressure is LOWER than inspiratory positive airway pressure.
 - **Autotitrating CPAP (APAP):**
 - Pressure required to maintain airflow and control vary throughout the night.



Figure 136.2 PAP interfaces: Examples of the three most common categories of mask interfaces for positive pressure therapy are depicted: (A) nasal pillows, (B) nasal mask, and (C) full-face mask.

- Advantages of PAP:
 - Reduction in AHI
 - Improvement in objective and subjective sleepiness
 - Improvement in overall quality of life
 - Reduction in risk of cardiovascular events
 - Reduction in risk of motor vehicle accidents
- Disadvantages of PAP:
 - **Side effects:**
 - Mask leaks
 - Rhinitis
 - Claustrophobia
 - **Non-Compliance:**
 - 50% patients of OSA using PAP.
 - Use of BIPAP or APAP didn't show an increase in compliance.
 - Increase compliance by:
 - Humidification
 - Topical nasal steroids
 - Upper airway surgery (nasal surgeries, UPPP).
- Contraindications of PAP:
 - No respiratory drive
 - CSF leak (cribriform plate injury)
 - Pneumocephalus
 - Pneumothorax
- **Oral Appliances:**
 - Second line of treatment for ADULTs with mild to moderate OSA who do not tolerate CPAP.
 - Act by increasing the posterior oropharyngeal airway.
 - Less effective than CPAP.
 - High rate of patient adherence up to 77%.
 - Examples:
 - Mandibular Repositioning Appliance
 - Tongue Retaining Device
 - Thermoplastic Splints
 - Side effects:
 - Tooth and jaw muscle pain
 - Difficulty chewing in the morning
 - Excessive salivation.

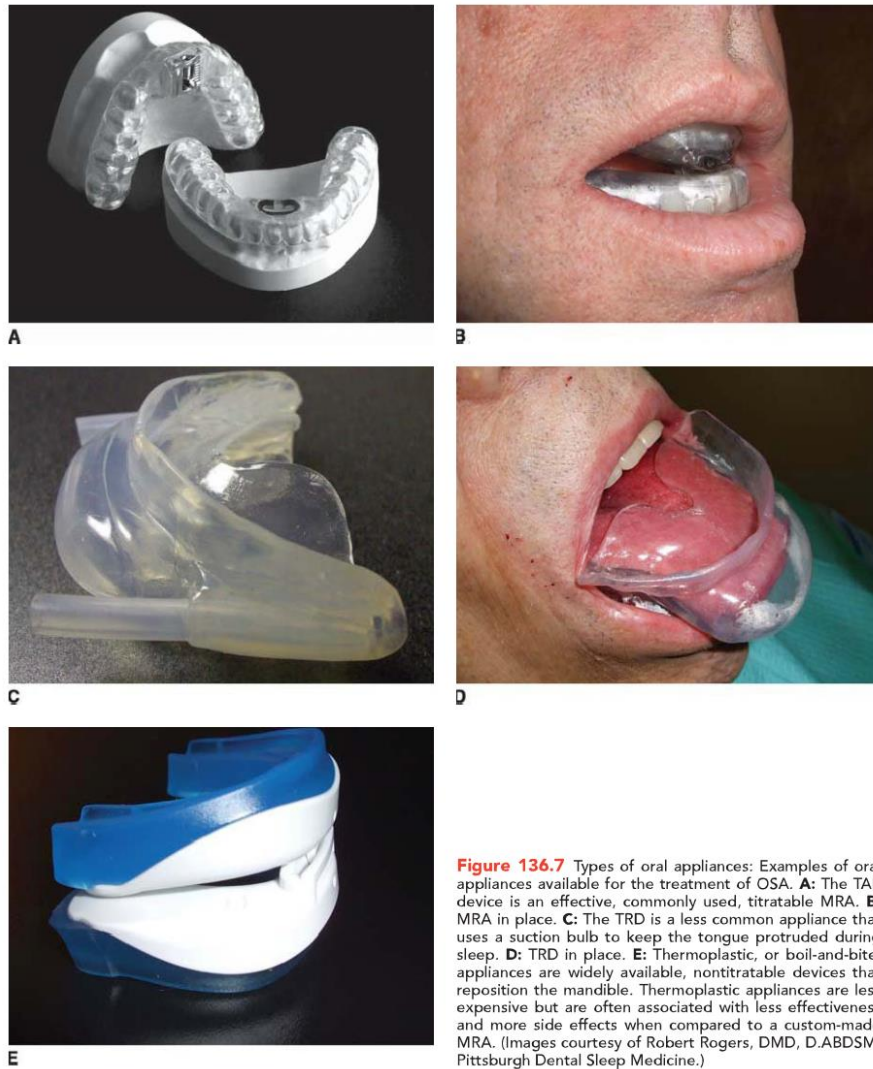


Figure 136.7 Types of oral appliances: Examples of oral appliances available for the treatment of OSA. **A:** The TAP device is an effective, commonly used, titratable MRA. **B:** MRA in place. **C:** The TRD is a less common appliance that uses a suction bulb to keep the tongue protruded during sleep. **D:** TRD in place. **E:** Thermoplastic, or boil-and-bite, appliances are widely available, nontitratable devices that reposition the mandible. Thermoplastic appliances are less expensive but are often associated with less effectiveness and more side effects when compared to a custom-made MRA. (Images courtesy of Robert Rogers, DMD, D.ABDSM, Pittsburgh Dental Sleep Medicine.)

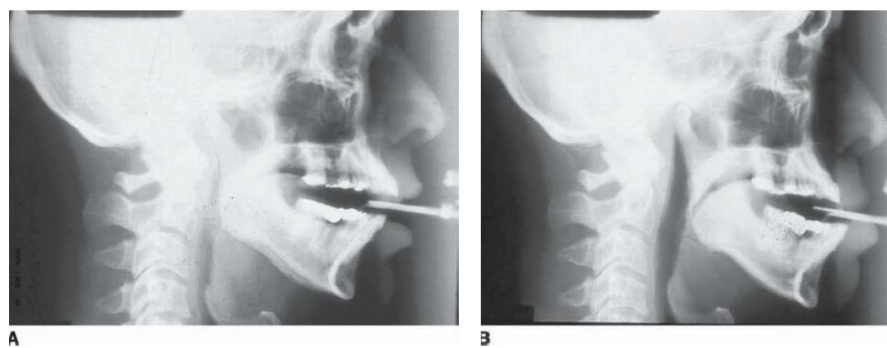


Figure 136.8 Effect of oral appliances on the airway: Lateral cephalometric radiograph in a patient with OSA, before (A) and after (B) application of a MAD. Note the enlargement of the pharyngeal airway with the oral appliance at both the retrolingual and retropalatal portions of the airway. (Images courtesy of Robert Rogers, DMD, D.ABDSM, Pittsburgh Dental Sleep Medicine.)

- **Nasal Steroids:**
 - **Not recommended as a single intervention for ADULTs with OSA.**
 - Recommended for pediatric OSA in presence of coexisting rhinitis and adenotonsillar hypertrophy.

- **Surgical Treatment:**

- Goal of airway reconstruction is to:
 - Improve airway patency and flow.
 - Improve CPAP tolerance.
- Requires multimodality approach including multiple staged surgical procedures and possible continued CPAP use.
 - In adults, OSA is not caused by a single anatomic obstruction.
- **Adenotonsillectomy is treatment of choice in children over CPAP.**
- Important factors before surgical treatment:
 - Patient's wishes
 - CPAP tolerance
 - Severity of disease
 - Comorbidities
 - Site and severity of upper airway collapse:
 - Surgically correctable anatomy (favorable) versus more difficult anatomical findings (unfavorable).

TABLE 5–3. Assessment of Favorable Versus Unfavorable Surgically Correctable Anatomy

Favorable	Unfavorable
Tonsillar Hyperplasia	Obesity
Adenoid Hyperplasia	Jowling or “Rounded Face”
Hyperplastic Lingual Tonsils	(Parapharyngeal Fat, Masseter Muscle Hypertrophy, East Asians)
Elongated Soft Palate	Retrognathia/Micrognathia
Nasal Septal Deformities	Macroglossia
Velopharynx > 11 mm	Nasopharyngeal Stenosis or Collapse
Uvular Hypertrophy	Bulky Base of Tongue
Isolated Velopharyngeal Collapse	Large Neck Circumference
	Lateral/Posterior Pharyngeal Wall Collapse
	High Arched Palate

- Success in OSA surgery:
 - 50% reduction in the RDI
 - RDI <20
- Indications of surgical treatment:
 - 1. Adenotonsillar hypertrophy in children.**
 - 2. Failed CPAP trial.**
 - 3. CPAP noncompliance.**
 - 4. Reliving of Snoring.**
- Relative contraindications:
 - 1. Morbid obesity**
 - 2. Severe cardiopulmonary disease**
 - 3. Unfavorable anatomy**

Box 18-6. SURGICAL TREATMENT OPTIONS

Nasal Surgery

Nasal septoplasty
Inferior turbinate reduction
Adenoidectomy
Nasal tumor or polyp resection
Nasal valve reconstruction

Palatal Surgery

Palatal radiofrequency ablation
Pillar implants
Injection snoreplasty
Tonsillectomy
Uvulopalatopharyngoplasty/Z-palatoplasty
Transpalatal advancement pharyngoplasty

Hypopharyngeal Surgery

Lingual tonsillectomy
Partial midline glossectomy
Tongue base radiofrequency ablation
Mandibular osteotomy and genioglossal advancement
Hyoid myotomy and suspension
Tongue-suspension suture
Maxillomandibular osteotomy and advancement

○ **Nasal Surgeries:**

- Selection of a nasal procedure is based on the pathology.
- Isolated nasal procedures are unlikely to improve OSA.
- Nasal obstruction should be address as initial step in OSA management to facilitate better CPAP adherence.
- Advantages:
 - Improve nasal airway
 - Decrease mouth breathing
 - Decrease nasopharyngeal snoring
 - Improve CPAP compliance by decreasing pressure requirements (best surgical method to improve CPAP compliance).

- **Oropharyngeal Surgeries:**
 - Most common type of surgeries for OSA.
 - Considered in patients with:
 - Significant oropharyngeal (velopalatine) crowding.
 - To improve snoring.
 - Tonsillectomy and/or adenoidectomy:
 - **First line of treatment of Pediatric OSA.**
 - Doesn't required pre-op PSG to confirm OSA,
 - Tonsillectomy is effective in treatment of OSA in 60-70% children with tonsillar hypertrophy.
 - Tonsillectomy is effective in ONLY 10-25% of obese children with OSA.
 - In adults, limited outcome of simple tonsillectomy.
 - Uvulopalatopharyngoplasty (UPPP):
 - Most common surgery for OSA in adults.
 - Reconstruct and reduce posterior edge of the soft palate and uvula combined with a tonsillectomy.
 - Should be considered in patients with one level of obstruction limited to oropharyngeal area.
 - Average success rates up to 50% for OSA.
 - Success rates up to 80% for snoring.
 - **Favorable criteria for UPPP with 70% success rate:**
 - Tonsil size (Grade 3-4).
 - Friedman score (Grade 1-2).
 - BMI < 40
 - Disadvantages:
 - Risk of air leak with CPAP due to reduction of the soft palate and uvula which can worsen compliance.
 - Long-term results are unfavorable possibly due to multiple sites of obstruction.
 - Painful recovery
 - Risk of post-op VPI in 10-15%.
 - Lateral Pharyngoplasty:
 - Techniques that expand the lateral diameter.
 - Palatal Stiffening Procedures:
 - Includes implants, sclerosing injections, and radiofrequency ablation procedures.
 - Can be combined with uvulectomy.
 - Office-based procedures.
 - Limited to improve snoring.

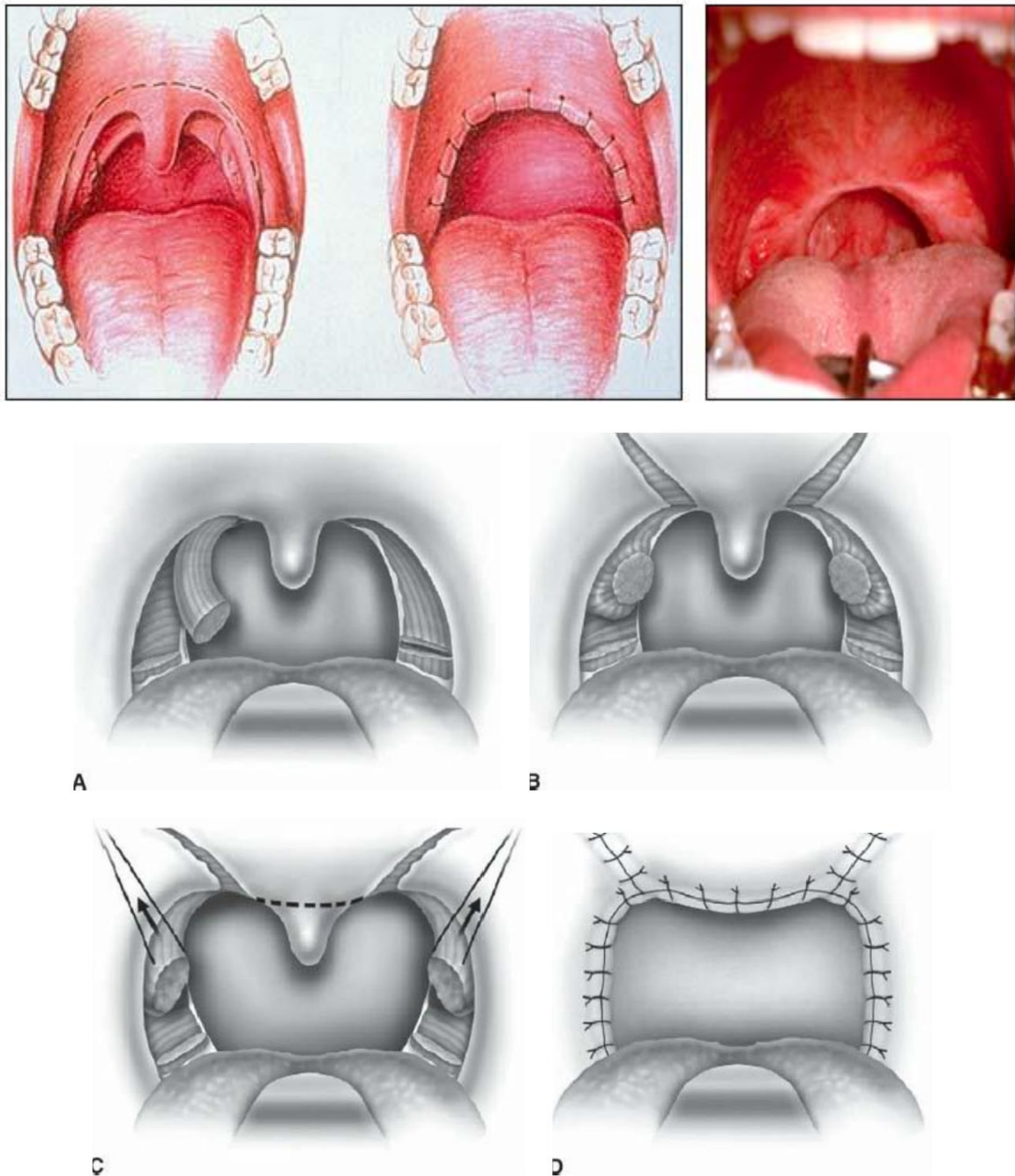


Figure 137.4 Expansion sphincter pharyngoplasty. **A:** After tonsillectomy is completed (if necessary), the palatopharyngeus muscles are isolated and transected at the inferior aspect. **B:** Mucosal incisions or submucosal tunnels are made up toward the hamulus anteriorly. **C:** A figure-eight tendon suture is placed in the cut end of the muscle to suspend it up toward the anterior soft palate and hamulus. **D:** The incisions are closed. The uvula may be preserved in some patients. (Reprinted from Pang KP, Woodson BT. Expansion sphincter pharyngoplasty in the treatment of obstructive sleep apnea. *Oper Tech Otolaryngol Head Neck Surg* 2006;17(4):223–225, with permission.)

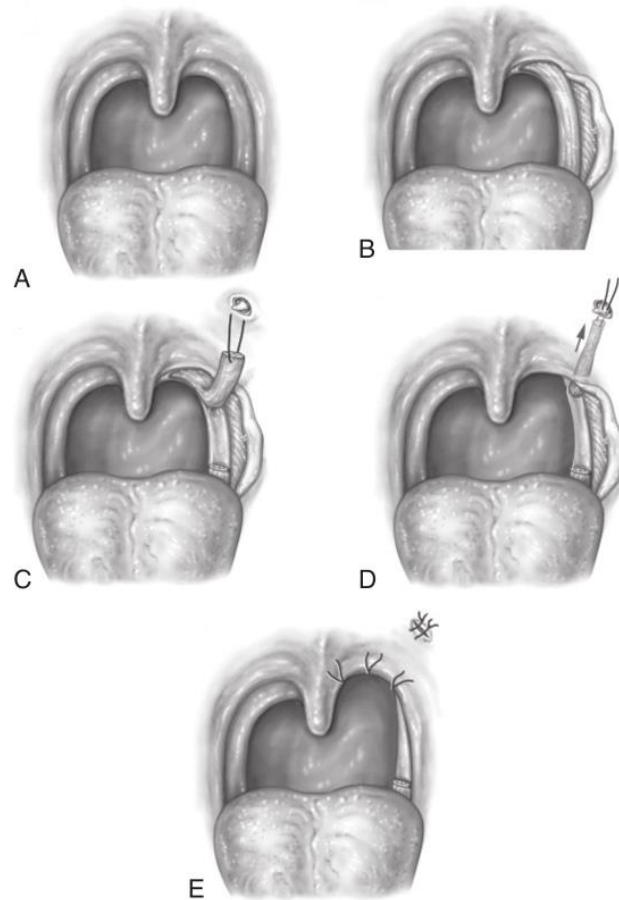
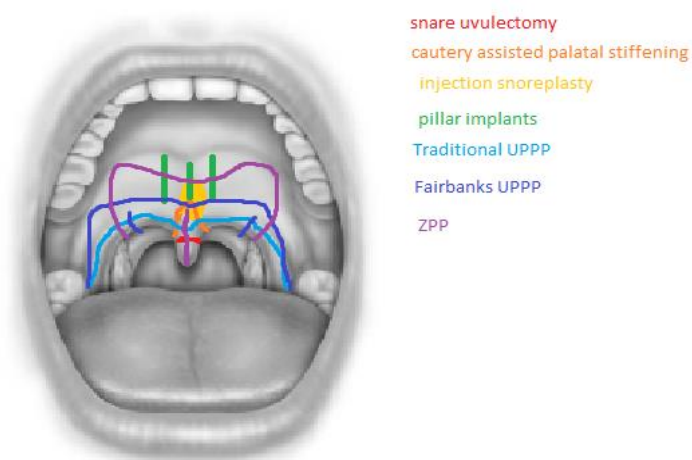


FIGURE 18-15. Expansion sphincter pharyngoplasty technique. **A**, Preoperative view of the oropharynx. **B**, Exposure of the palatopharyngeus (vertical fibers). **C**, Elevation of the palatopharyngeus. **D**, Rotation and tunneling of the palatopharyngeus toward the hamulus. **E**, Suture suspension and approximation. (From Woodson BT, Sittton M, Jacobowitz J. Expansion sphincter pharyngoplasty and palatal advancement pharyngoplasty: airway evaluation and surgical techniques. *Oper Techn Otolaryngol* 2012;23:6.)



- **Hypopharyngeal Surgeries:**
 - Considered in patients with:
 - Significant hypopharyngeal obstruction
 - Relative macroglossia
 - Base of tongue obstruction,
 - Retroflexed epiglottis
 - Tongue Base Reduction, Lingual Tonsillectomy:
 - Various reduction techniques to address the retrolingual space including:
 - Laser
 - Coblation
 - Radiofrequency
 - Cautery
 - Complications:
 - Risk of dysphagia
 - Injury to the lingual artery
 - Lingual hematoma
 - Partial Midline Glossectomy:
 - Reduce midline soft tissue medial to the neurovascular bundle.
 - Complications:
 - Risk of dysphagia
 - Injury to the lingual artery
 - Lingual hematoma
 - Genioglossal Advancement:
 - Repositions the tongue anteriorly by advancing a segment of mandible with the genioglossus attached.
 - Does not move the teeth
 - Limited anterior advancement
 - Risk of relaxation of tongue base.
 - Genioglossal Suspension:
 - Tongue base is suspended anteriorly to mandible using various material including sutures.
 - Simple technique.
 - Limited anterior advancement
 - Risk of breakage and relaxation of tongue base
 - Hyoid Myotomy and Suspension:
 - Hyoid bone is repositioned anteriorly by releasing infrahyoid muscles and suspending to the lingual surface of the mandible or to the thyroid cartilage.
 - Risk of breakage
 - Risk of mental nerve injury
 - Epiglottoplasty:
 - Stabilizes the epiglottis to the tongue base.
 - Risk of aspiration

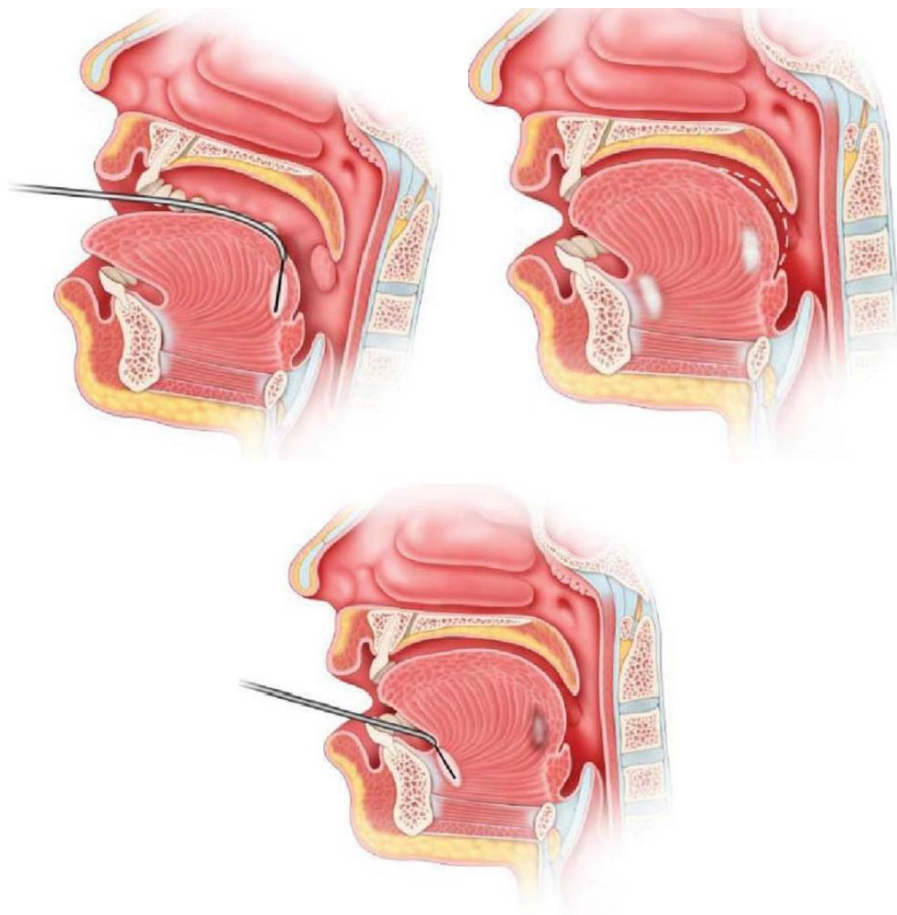
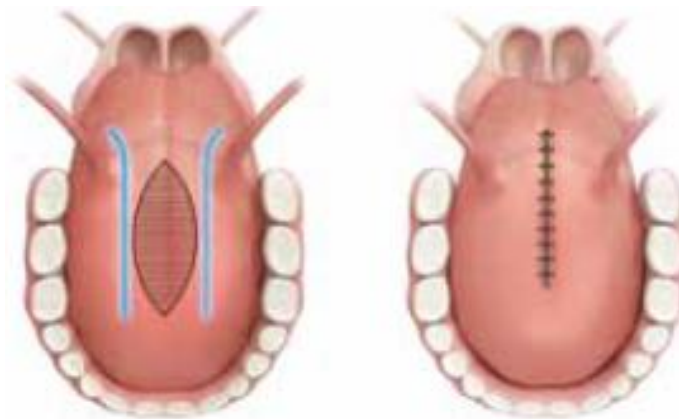


Figure 138.7 Submucosal tongue radiofrequency. Figures show the creation of tissue injury in the tongue base (**left**) and ventral tongue (**middle**), both of which heal with the development of fibrosis at these sites (**right**). (Reprinted with permission of www.sleep-doctor.com)



Midline glossectomy – part of the bulky tongue can be removed to improve breathing.

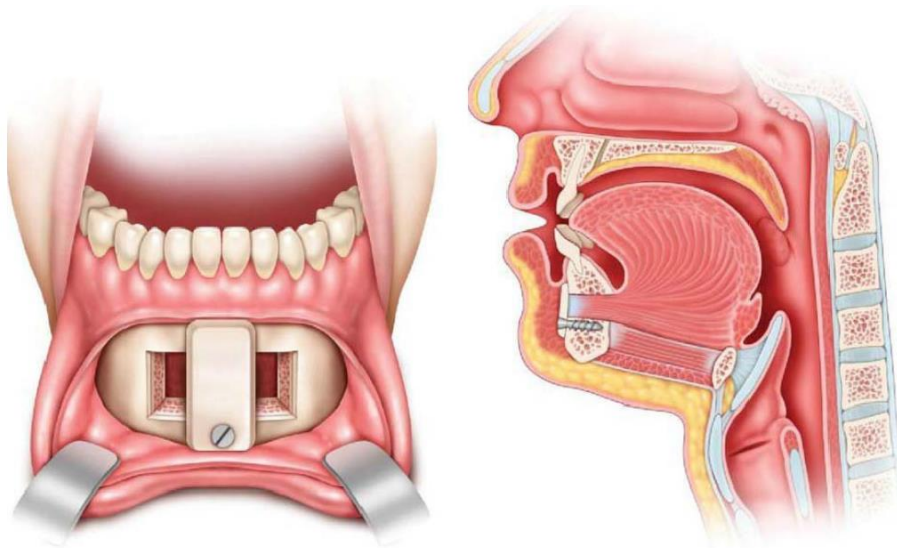


Figure 138.5 Genioglossus advancement procedure. On the **left**, an anterior view shows the mandibular osteotomy with fragment advanced, rotated, and secured inferiorly. On the **right**, a sagittal view demonstrates the advanced position of the genioglossus muscle insertion. (Reprinted with permission of www.sleep-doctor.com)

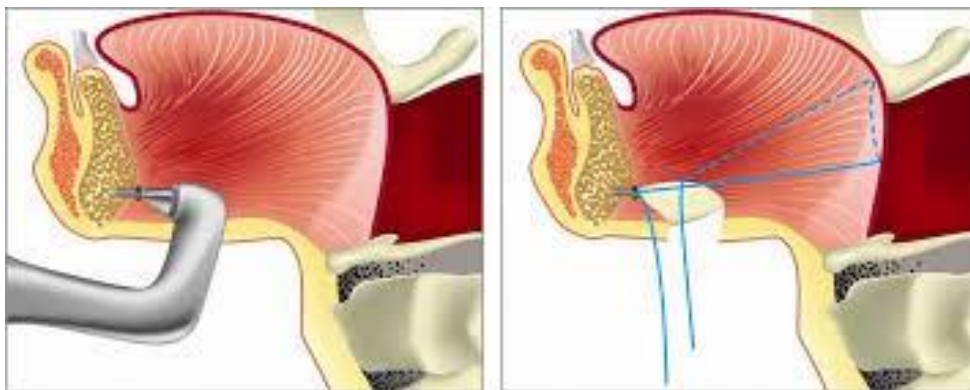
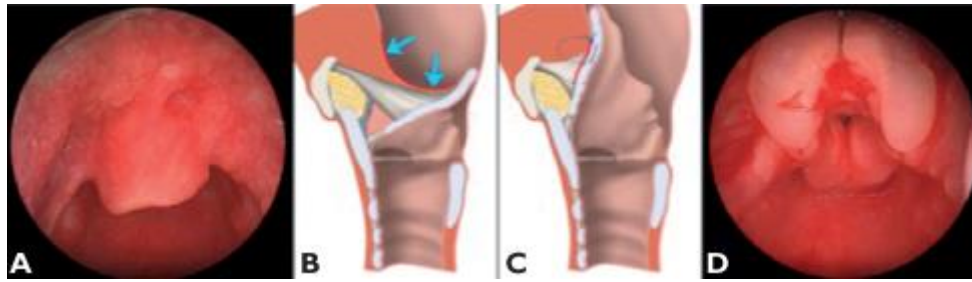


Figure 138.6 Hyoid suspension using a thyrohyoid technique. An anterior view (**left**) and sagittal view (**right**) demonstrate the use of sutures to secure the hyoid bone to the superior border of the thyroid cartilage. (Reprinted with permission of www.sleep-doctor.com)



- **Mandibular and Midface Advancement Surgeries:**
 - Considered in patients with:
 - obstruction involving the retrolingual and retropalatal airway.
 - Le Fort I maxillary osteotomy with rigid plate fixation and a bilateral sagittal split mandibular osteotomy.
 - More effective in addressing the retrolingual/palatal airway than soft tissue oropharyngeal and hypopharyngeal techniques.
 - Can be performed in 1 stage.
 - Requires prolonged recovery.
 - May change facial appearance.
 - Requires maxillomandibular fixation.

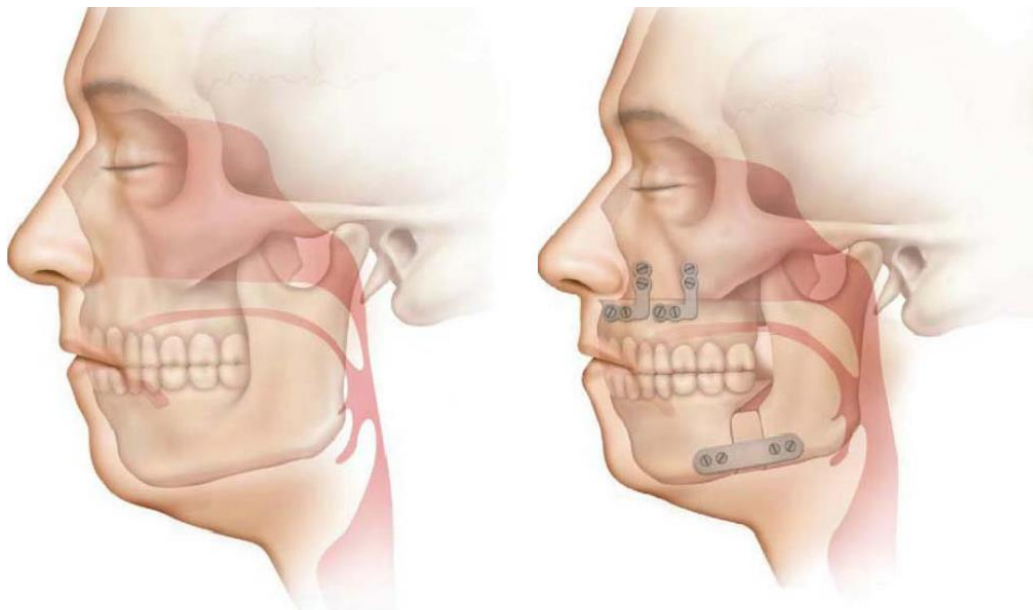


Figure 138.9 Maxillomandibular advancement. Preoperative (**left**) and postoperative (**right**) sagittal images showing the changes in the facial skeleton and pharyngeal airway. (Reprinted with permission of www.sleep-doctor.com)

○ **Tracheotomy:**

- Considered in patients with:
 - Severe OSA with failed CPAP and other surgical treatment.
 - Morbid obese.
 - Complicated OSA (Cor pulmonale).
- Traditional gold standard of surgical management of OSA.
- Most effective surgical treatment for sleep apnea.
- Completely bypassing the collapsed portion of airway.
- Associated with psychosocial problems, perceived inconvenience and morbidity.

TABLE 18-3. European Respiratory Society Task Force 2011 Recommendations for Obstructive Sleep Apnea

Recommended	Not Recommended
Weight reduction	Positional therapy (except in carefully selected patients)
Oral appliances (mild to moderate OSA)	Apnea-triggered muscle stimulation
Intranasal corticosteroids	Tongue-retaining devices
Adenotonsillectomy (pediatric OSA)	Drug therapy
Tonsillectomy (adults with tonsillary hypertrophy)	Nasal dilators
Uvulopalatal flap with tonsillectomy	Nasal surgery as single intervention
Maxillomandibular advancement	Uvulopalatopharyngoplasty (except in carefully selected patients)
Distraction osteogenesis	Laser-assisted UPP Radiofrequency surgery of the soft palate Uvulopalatal flap as single intervention Pillar implants (except in carefully selected patients) Radiofrequency surgery of the tongue base Hyoid suspension as a single intervention Laser midline glossectomy/tongue suspension Genioglossus advancement as a single procedure Multilevel surgery as first-line therapy

From Randerath WJ, Verbraecken J, Andreas S, et al. Non-CPAP therapies in obstructive sleep apnoea. *Eur Respir J* 2001;37((May)5): 1000-1028.

OSA, obstructive sleep apnea; UPP, uvulopharyngoplasty.

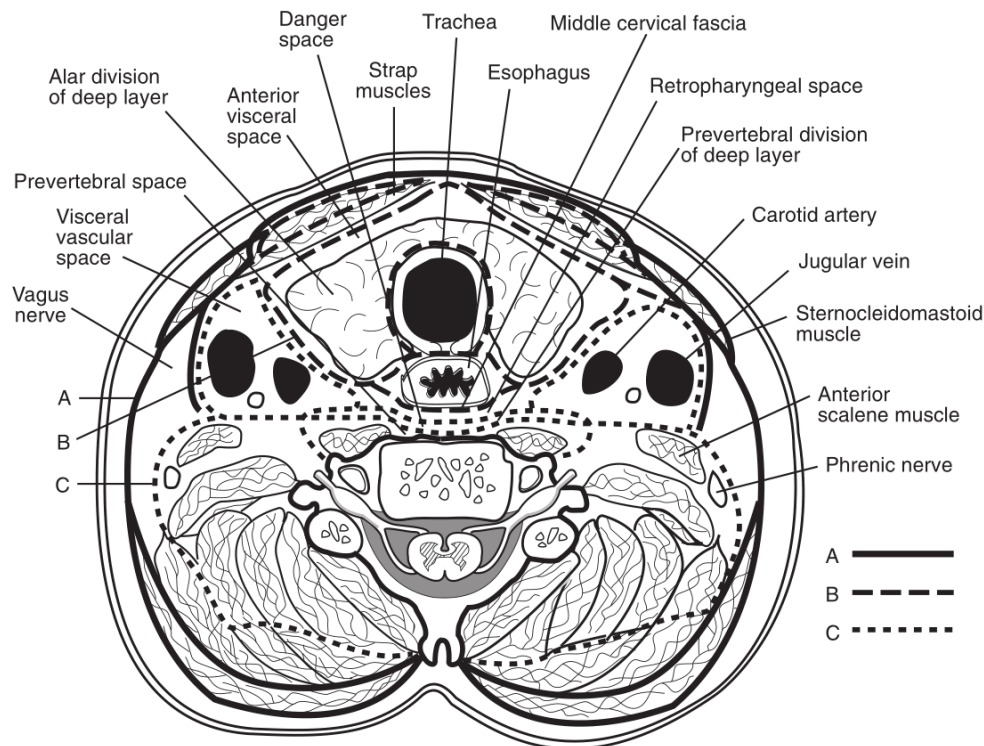
- **Post-operative Management of OSA:**
- Increased post-op airway obstruction secondary to:
 - o Upper airway edema
 - o Post-anesthesia sedation and narcotic pain medications.
- Post-op ICU should be considered in multilevel surgery.
- Nasal CPAP is used for first post-op night to avoid NPPE.
- Narcotic analgesics should be avoided.
- Recommended to use CPAP for first 2 weeks in patients with severe OSA.
- Post-op PSG to be done after 3 months to evaluate response to surgery.

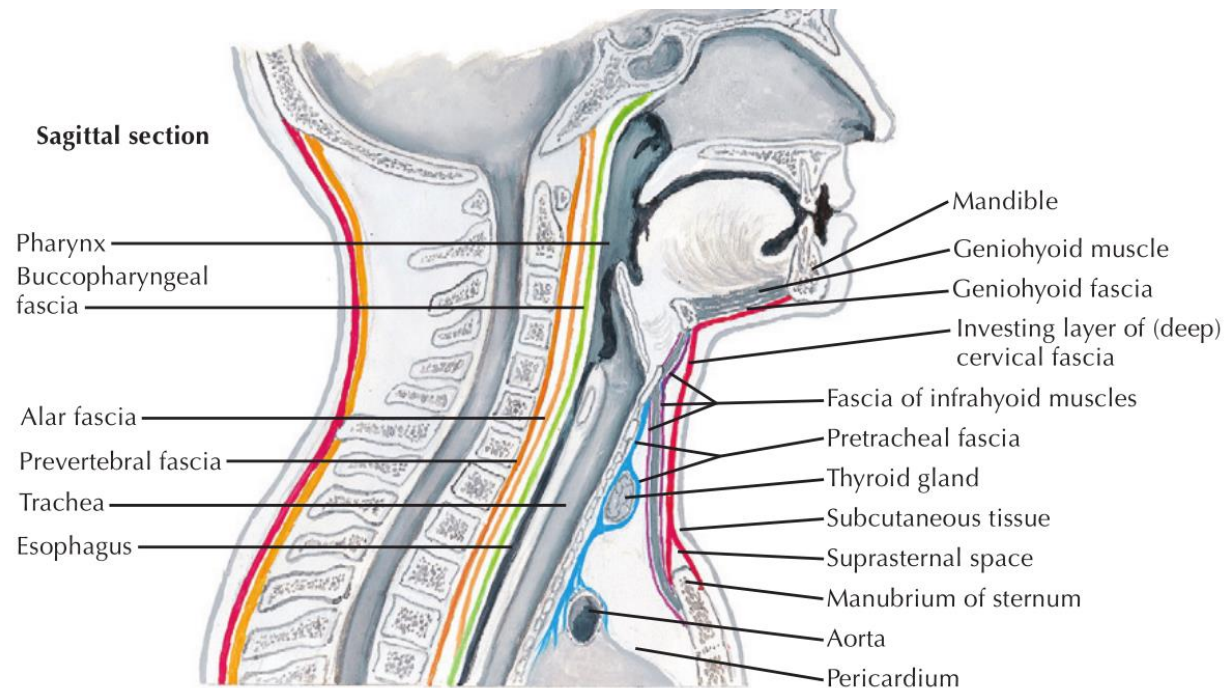
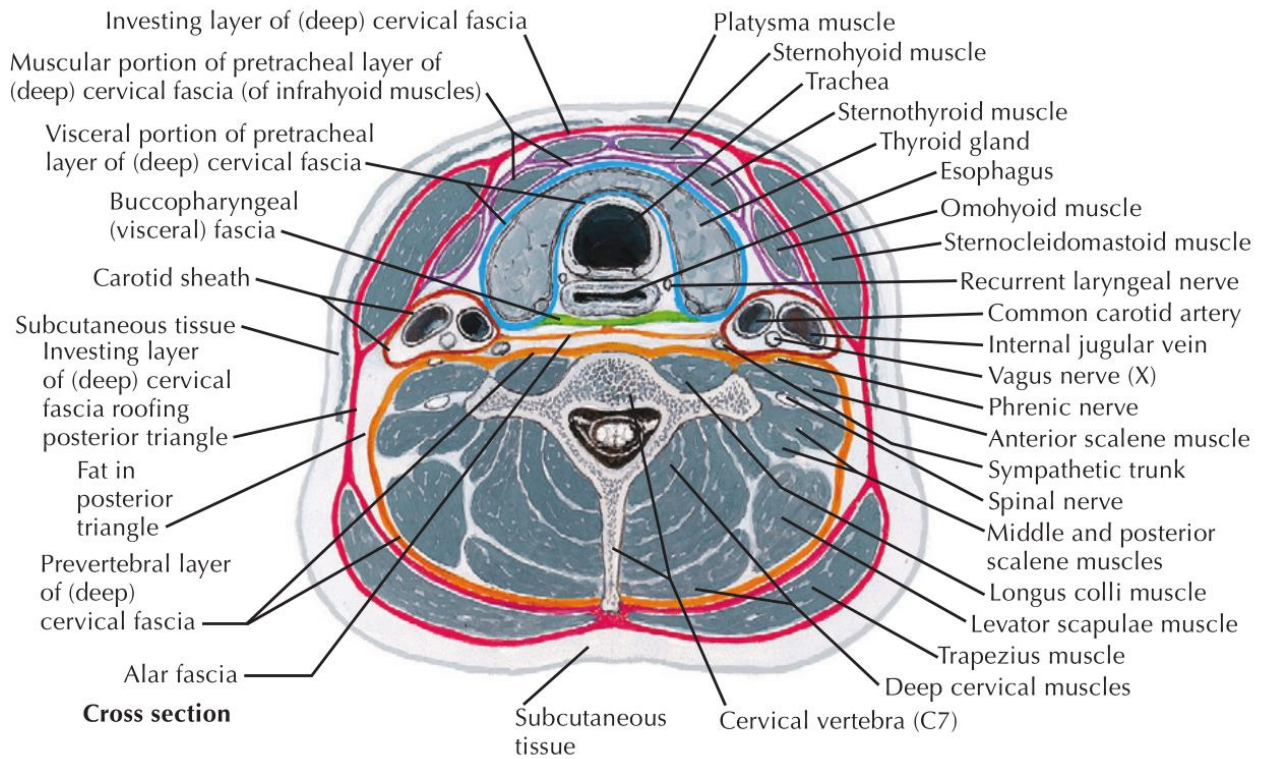
- **Negative Pressure Pulmonary Edema (NPPE):**
 - o Post-obstructive pulmonary edema.
 - o Types:
 - **Type I:**
 - Occurs immediately after acute airway obstruction.
 - Laryngeal spasm-induced pulmonary edema.
 - Incidence of 10%.
 - **Type II:**
 - Occurs after relief of chronic upper airway obstruction.
 - **Incidence of 40%.**
 - o Pathophysiology:
 - **Type I:**
 - Acute upper airway obstruction leads to excessive inspiratory efforts to overcome the obstruction.
 - This generates highly negative intra-pleural and alveolar pressures which causes fluid to move out of the pulmonary capillaries and into the interstitial and alveolar spaces.
 - **Type II:**
 - In chronic upper airway obstruction there is a modest level of Auto positive end-expiratory pressure (PEEP) with an increase of end expiratory lung volume.
 - When this chronic obstruction is relieved acutely, Auto PEEP will disappear, lung volumes and pressure return to normal, creating a negative intra-pulmonary pressure and transudation of fluids in the lung interstitium and alveoli.
 - o Clinical picture:
 - Decreased O2 saturation.
 - Pink frothy sputum.
 - Chest crackles and occasionally wheezes.
 - Chest radiograph show diffuse interstitial and alveolar infiltrates.
 - **Symptoms usually develop within 1 h of the precipitating event.**

- Treatment:
 - **In case of Upper airway obstruction (TYPE I):**
 - Relieve the airway obstruction (PEEP, ETT or tracheostomy).
 - **In case of acute relapse of long standing OSA (Type II):**
 - Patients should have a longer period of observation in the PACU or ICU.
 - Oxygenation with positive pressure (PEEP).
 - IVF restriction.
 - Diuretics (Furosemide 1mg/kg).
 - If no improvement, intubation with positive pressure (PEEP).

Cervical Fascia Anatomy:

- Fascia is a band of connective tissue that surrounds structures, giving rise to potential tissue spaces and pathways that allow infection to spread.
- Cervical Fascia is divided into:
 1. **Superficial Fascia**
 2. **Deep Fascia:**
 1. Superficial layer of Deep fascia
 2. Middle layer of Deep fascia
 3. Deep layer of Deep fascia
 4. Carotid Sheath
- Superficial Cervical Fascia:
 - o Lies immediately under skin.
 - o Extends from the zygomatic process to the thorax and axilla.
 - o Includes superficial musculoaponeurotic system (SMAS).
 - o Contents:
 - Fat
 - Platysma
 - Facial muscles of expression





- Deep Cervical Fascia:

- Lies deep to the superficial fascia.
- Aids muscle movements.
- Provides passageways for nerves and vessels.
- Provides attachment for some muscles.
- There is no deep fascia in the face, which allows free spread of fluid.
- Divided into:

1. Superficial (Investing) Layer of Deep Fascia:

- Forms Stylomandibular ligament which separates the parotid and submandibular glands.
- Contents:
 - Trapezius
 - SCM
 - Masseter
 - Submandibular and parotid glands

2. Middle Layer of Deep Cervical Fascia:

- **Muscular Division:**
 - Contents:
 - Strap muscles.
- **Visceral Division:**
 - Contributes to formation of:
 - Pretracheal fascia
 - Buccopharyngeal fascia
 - Contents:
 - Pharynx
 - Larynx
 - Trachea
 - Esophagus
 - Thyroid and Parathyroids
 - Pharyngeal constrictors
 - Buccinator muscle.

3. Deep Layer of Deep Cervical Fascia:

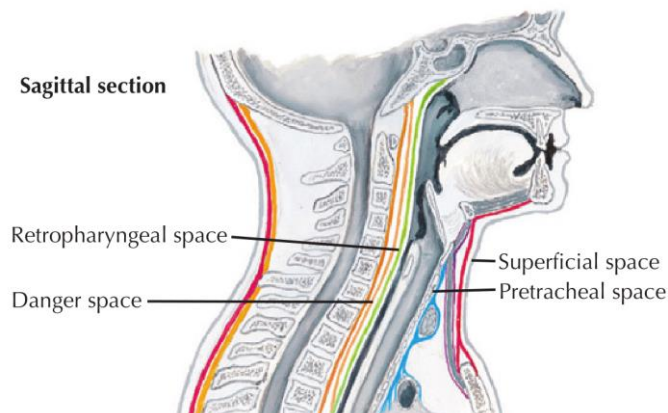
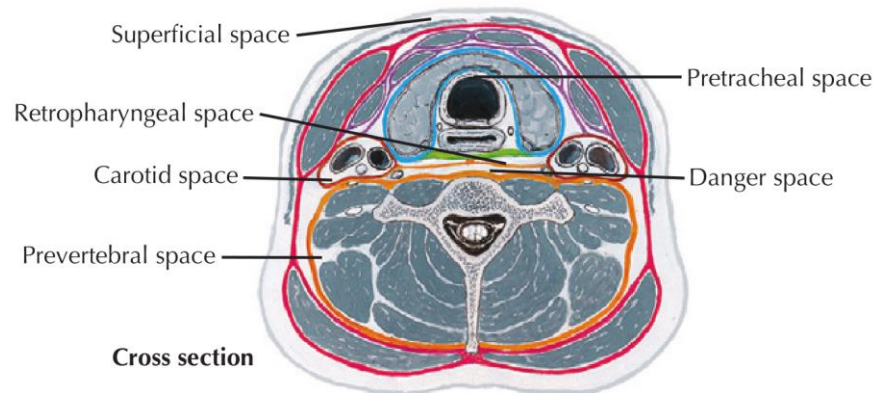
- Divided into:
 - **Alar Fascia:**
 - Content:
 - Cervical sympathetic trunk.
 - **Prevertebral Fascia:**
 - Contents:
 - Paraspinus muscles
 - Cervical vertebrae

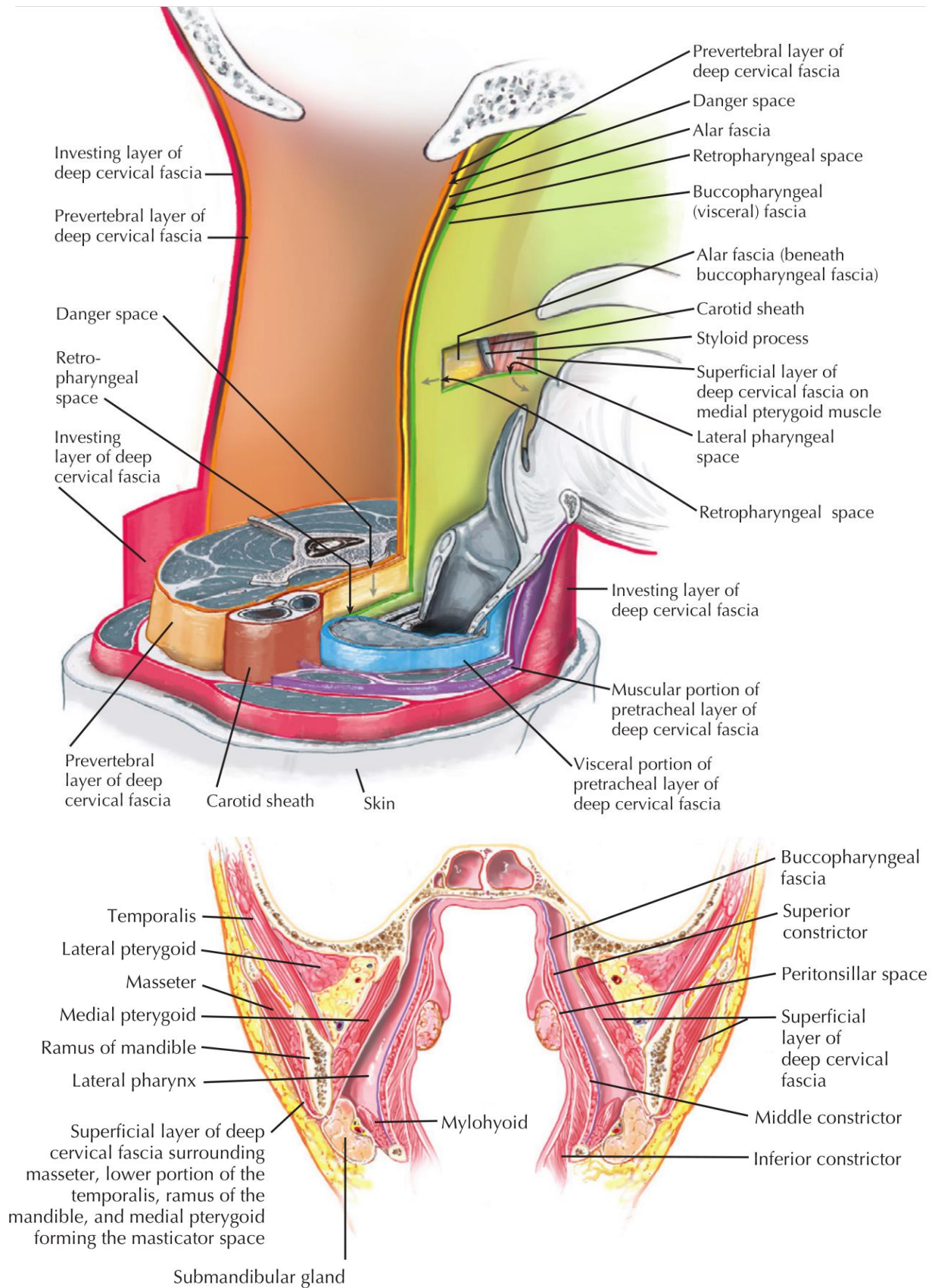
4. Carotid Sheath:

- Composed of all layers of Deep cervical fascia.
- Contents:
 - Common carotid artery
 - Internal jugular vein
 - Vagus nerve
 - Ansa cervicalis

Deep Neck Spaces Anatomy:

- Layers of Deep fascia "create" potential fascial spaces.
- All are filled by loose areolar connective tissue.
- Hyoid bone is the most important anatomic structure in the neck that limits the spread of infection.
- Infections spread by the path of least resistance to reach the fascial spaces.
- Deep Neck Spaces are divided into:
 - **Suprahyoid spaces:**
 - Parapharyngeal space
 - Submandibular and Sublingual spaces
 - Parotid space
 - Masticator space
 - Peritonsillar space
 - Temporal space
 - **Infrahyoid spaces:**
 - Anterior visceral space
 - Suprasternal space
 - **Entire neck length spaces:**
 - Retropharyngeal space
 - Danger space
 - Prevertebral space
 - Carotid space





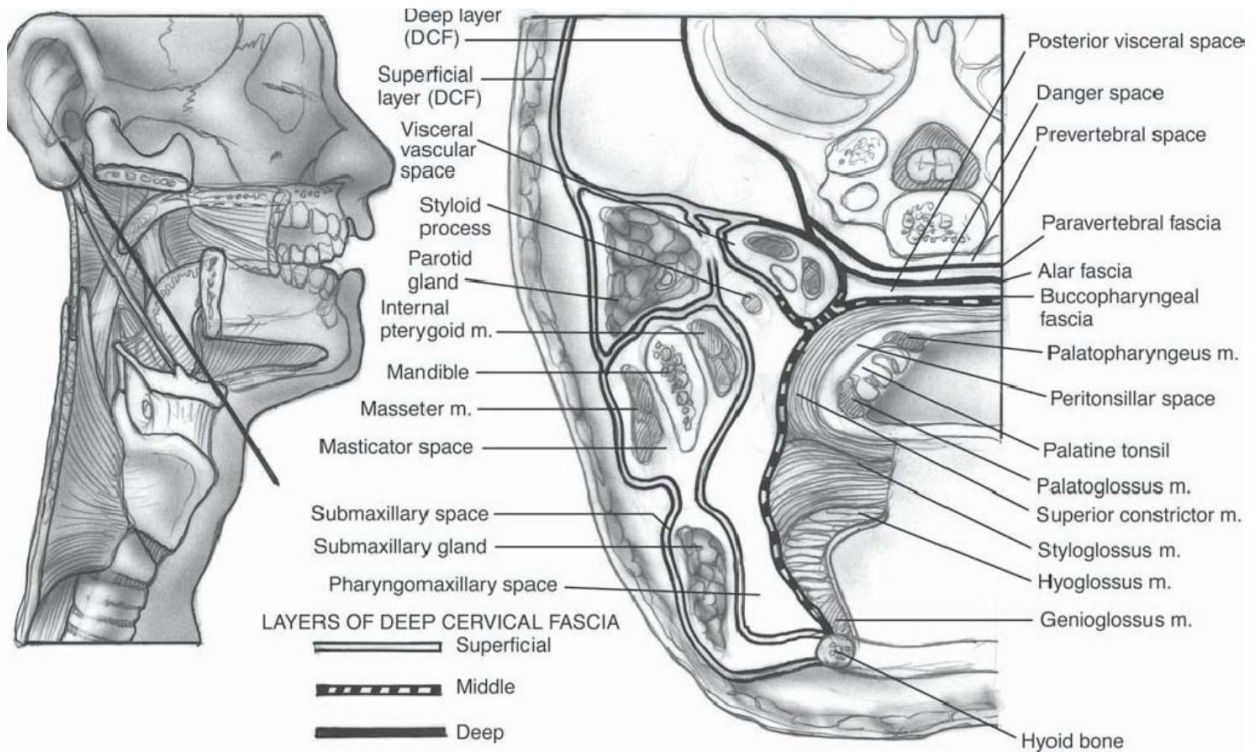
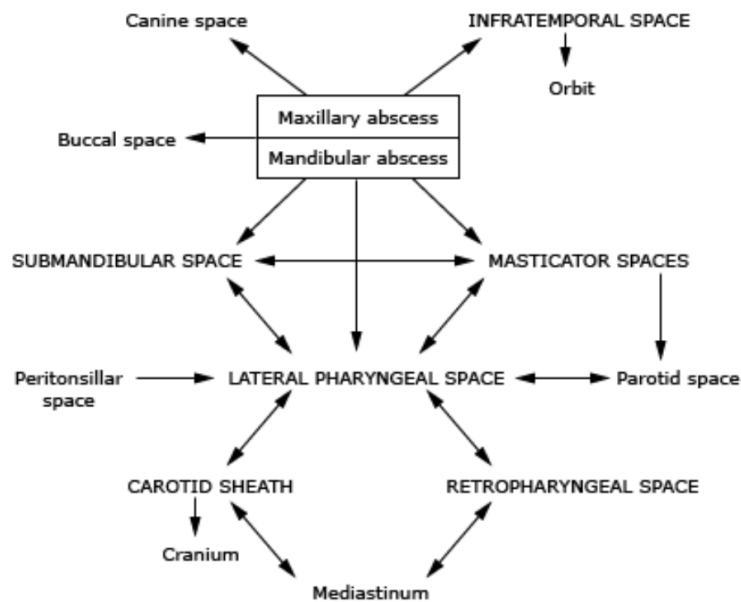
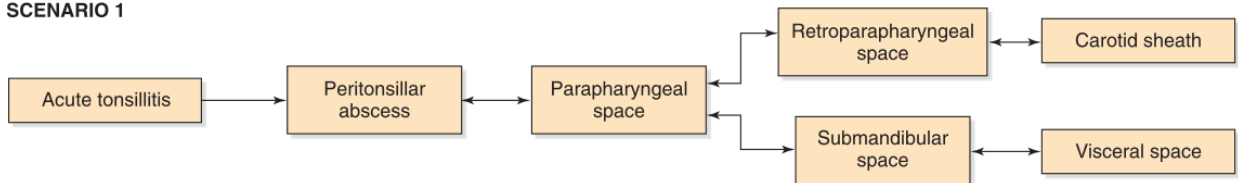


Figure 55.4 Oblique section through the neck shows the anatomic relations of the spaces limited to above the hyoid bone to the spaces that transverse the entire neck. The important relation between the parapharyngeal (pharyngomaxillary) space and the other spaces is evident.

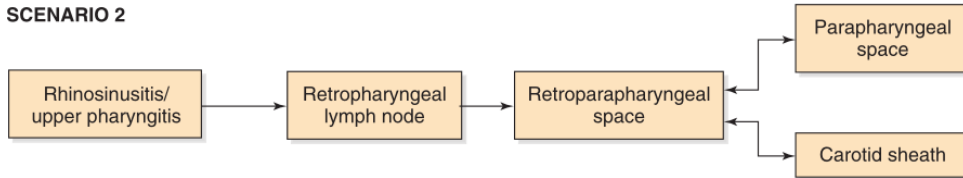
Potential pathways of extension in deep cervical fascial space infections



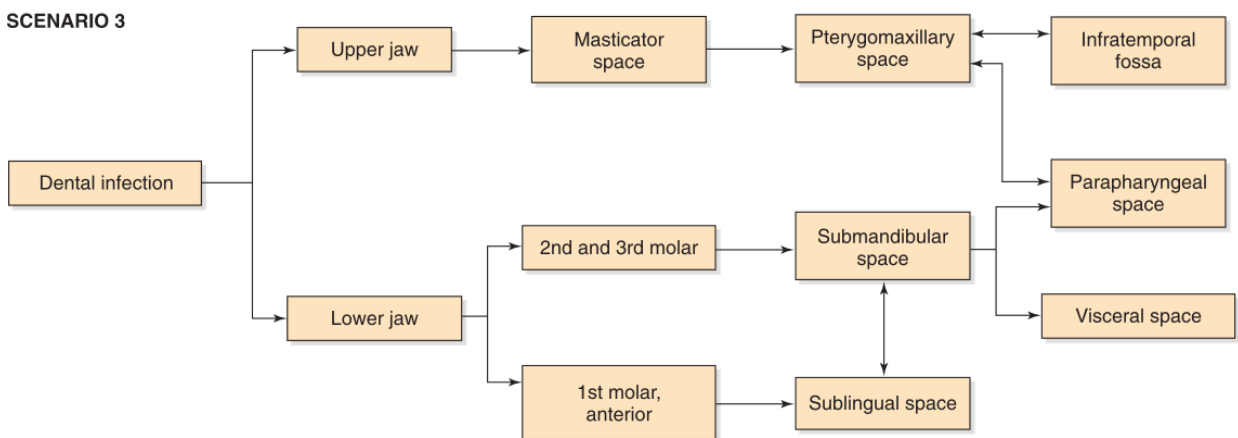
SCENARIO 1



SCENARIO 2



SCENARIO 3



SCENARIO 4

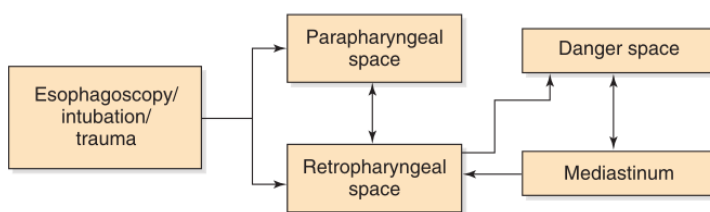


FIGURE 10-2. Common pathways of spread in deep neck infection.

Deep Neck Infections:

- Infrequent entity in post-penicillin era.
- Result in serious or fatal complications if not recognized and treated early.
- Proper management begins with securing the airway.
- Despite recent advances in imaging and conservative medical management, surgical drainage remains a mainstay of treatment.
- Etiology:
 - o **Infections:**
 - Oropharyngeal infections:
 - Most common cause of deep neck infections in children.
 - Dental infections:
 - Most common cause of deep neck infections in adults.
 - **Mandatory to look for a potential dental source in any patient with deep neck space abscess.**
 - Acute mastoiditis:
 - Progress to Bezold abscess.
 - Acute rhinosinusitis:
 - Lead to retropharyngeal lymph nodes suppuration in children.
 - Acute Sialadenitis
 - Acute lymphadenitis
 - Cellulitis
 - o **Infected congenital neck masses:**
 - Branchial cleft cysts, TGDC or laryngoceles.
 - Suspected in recurrent deep neck infections in children.
 - o **Surgical instrumentation:**
 - Bronchoscopy.
 - Esophagoscopy.
 - o **Trauma:**
 - Needle injection associated IV drug use.
 - Oropharyngeal trauma in children (lollipops, pencil).
 - o **Malignancy:**
 - Necrotic malignant lymph nodes.
- Bacteriology:
 - o **Polymicrobial**, representing oropharyngeal flora.
 - o Most common Aerobic:
 - **Strep. Viridans**
 - Strep. Pyogenes (Group A beta-hemolytic streptococcus)
 - Staph. Epidermis
 - Staph. Aureus
 - o Most common Anaerobic:
 - Peptostreptococcus
 - Fusobacterium
 - Bacteriodes

- Children:
 - Staph. Aureus
 - Methicillin-resistant Staphylococcus aureus (MRSA)
- Immunocompromised:
 - Gram-negative (Haemophilus, E.coli , Pseudomonas)
 - Methicillin-resistant Staphylococcus aureus (MRSA)
 - Klebsiella pneumoniae (uncontrolled DM)
- Atypical and granulomatous (Less common):
 - Actinomyces
 - Tuberculous
 - Cat scratch
 - Barumella henselae

TABLE 10-1. Normal Oral Flora	
Aerobic Bacteria	Anaerobic Bacteria
Gram-Positive Cocci	
<i>Streptococcus</i>	<i>Streptococcus</i> <i>Peptococcus</i> <i>Peptostreptococcus</i>
Gram-Negative Cocci	
<i>Neisseria</i>	<i>Veillonella</i>
Gram-Positive Bacilli	
Diphtheroids	<i>Clostridium</i> <i>Actinomyces</i> <i>Eubacterium</i> <i>Lactobacillus</i>
Gram-Negative Bacilli	
<i>Haemophilus</i> <i>Eikenella</i>	<i>Prevotella</i> <i>Bacteroides</i> <i>Fusobacterium</i> <i>Porphyromonas</i>

Modified from Peterson LJ. Odontogenic infections. In Cummings CW, ed: *Otolaryngology—head and neck surgery*, vol 2, ed 2. St Louis: Mosby; 1993.

- Clinical Evaluation:

○ **History:**

- Inflammatory symptoms:
 - Pain, fever, swelling and redness
- Local symptoms:
 - Dysphagia
 - Odynophagia
 - Drooling
 - Hot potato voice
 - Hoarseness
 - Dyspnea
 - Trismus
 - Ear pain
- Recent events:
 - Dental work
 - Upper airway surgery or intubation
 - IV drug use
 - Sinusitis
 - Pharyngitis
 - Otitis
 - Blunt or penetrating soft tissue trauma
- Immunocompromised conditions:
 - High risk of deep neck infections with atypical pathogens.

○ **Physical exam:**

- Vital signs and airway evaluation
- Complete Head and Neck examination
 - Trismus indicates spread of inflammation to parapharyngeal, pterygoid, or masseteric spaces.
- Flexible fiberoptic evaluation:
 - Important to identify an evolving airway obstruction before it occurs.
- Complete Cranial nerve examination.
- Orbital examination
- Dental examination

- Investigation:

○ **Blood tests:**

▪ CBC with differential:

- Elevated WBC with a predominance of neutrophils.
- Daily measures of WBC is helpful in monitoring patient's response to treatment.
- Steroid-related leukocytosis will make it difficult to monitor treatment response.
- Lack of initial leukocytic response may indicate:
 - Viral illness
 - Immunodeficiency
 - Tumor

▪ Serum electrolytes:

- Assess glucose level and hydration status.

▪ Others:

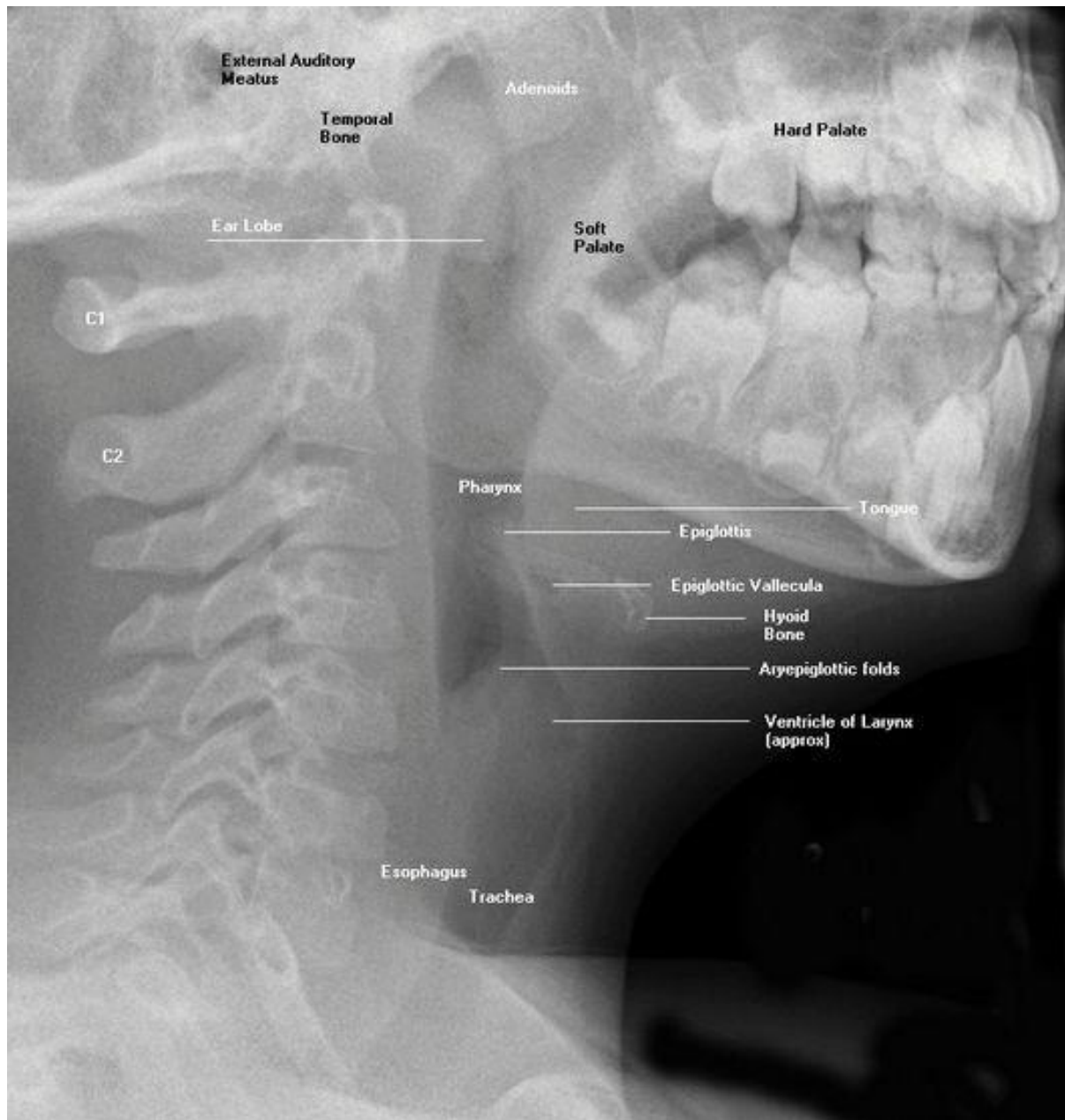
- Directed based on medical history or examination.
- Includes:
 - Blood culture
 - HIV screening
 - TB skin test

○ **Imaging:**

▪ Plain films:

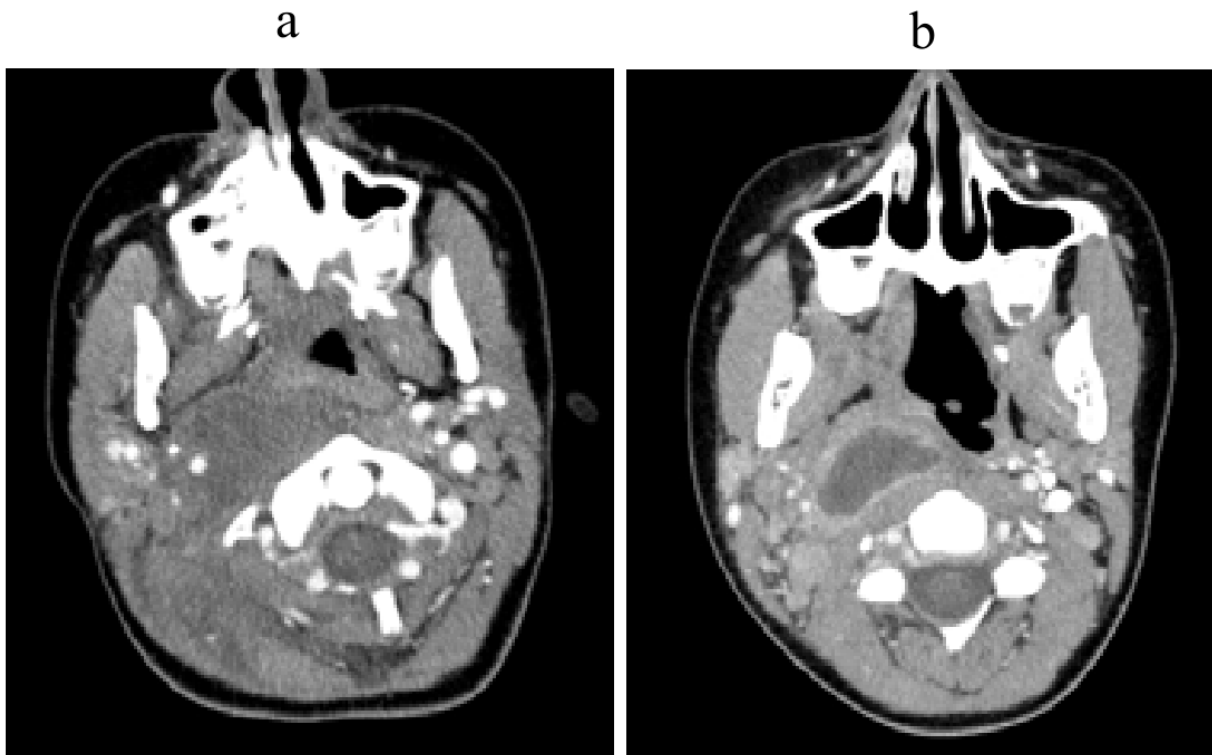
- Limited value for diagnosing deep neck infections.
- Lateral neck films is used as screening tool for retropharyngeal infection or epiglottitis.
 - Sensitivity of 80%.
- Chest radiography is indicated in cases of dyspnea, tachycardia, and/or cough to rule out aspiration and/or mediastinitis.





- Ultrasound:
 - Noninvasive tool with lack of radiation.
 - Preferred for pediatric patients.
 - Fluid levels of abscess can be seen if large and superficial enough, and lack of visualization does not rule out the possibility of abscess.
 - Guides diagnostic or therapeutic needle aspirations.
 - Limited value in:
 - Parapharyngeal or retropharyngeal infections
 - Significant neck edema or phlegm

- CT with IV Contrast:
 - Gold standard imaging.
 - Better assessment of airway and deep neck spaces.
 - Abscess usually appears on CT as:
 - Well-formed hypodense (fluid density) surrounded by complete ring enhancement.
 - Phlegm is soft tissue edema and the stage just before abscess formation and appears on CT as:
 - Diffuse hypodense (soft-tissue density) thickening with incomplete ring enhancement.
 - Sometimes, difficult to reliably differentiate between phlegm and purulent abscess.
 - **25% of CT-suspected abscesses are found to be non-drainable phlegm after I&D (False positive).**
 - Decision for neck exploration should be made based on clinical evaluation and not only with radiological imaging.



- MRI:
 - Superior soft tissue differentiation.
 - Not routinely indicated for deep neck infections **EXCEPT:**
 - Intracranial extension.
 - Pre-vertebral involvement.
 - Parotid space involvement.
 - Suspicion of vascular complications:
 - Suppurative vascular thrombi.
 - Pseudoaneurysm.
 - Rupture.
 - Sequences:
 - **T1:** Central HYPO-intensity
 - **T1 C+:** Central HYPO-intensity with peripheral enhancement.
 - **T2:** Central HYPER- intensity

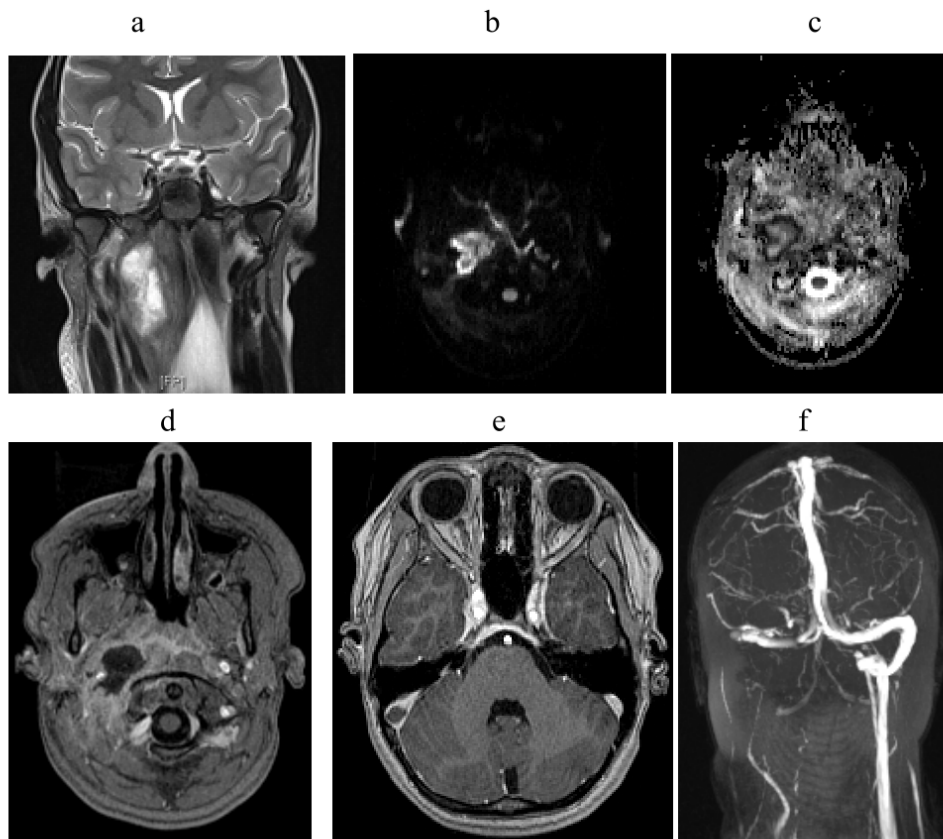


Figure 2: MRI on day 5 shows enlarged lymph node in right RPS (T2W) with restricted diffusion in the periphery (2b and 2c, DWI and ADC map). Post Gadolinium enhancement scan shows central necrosis and diffuse surrounding soft tissue enhancement/phlegmon (2d, GdT1W). There is non-occlusive thrombus in the right sigmoid sinus on the MRV which was acquired with Gd (2e, GdT1W). MRV also demonstrates lack of blood flow in the right sigmoid to jugular vein (2f).

- Management:

○ **Airway Management:**

- Safe and secure airway maintenance is the most important initial therapeutic objective.
- Hypoxia and asphyxia are the most common causes of mortality rather than uncontrollable sepsis.
- Airway complications should be anticipated with involvement of:
 - Floor of mouth
 - Parapharyngeal space
 - Retropharyngeal space
- Patients with mild respiratory symptoms should be admitted in **ICU** for close monitoring.
- Early intervention should be made for patients with progressive airway symptoms:
 - **Fiberoptic intubation:**
 - Nasal or oral intubation.
 - Done while patient is setting in upright position.
 - Instruments for surgical airway should be available.
 - **Awake tracheostomy:**
 - Done under LA in controlled situation.
 - In case of failed fiberoptic intubation.
- Conventional ET intubation should be avoided in large abscess due to:
 - Risk of laryngospasm in awake patient
 - Risk of abscess rupture with subsequent aspiration.

○ **Medical Management:**

- Medical management alone without surgical intervention can resolve deep neck infections in 60% of cases.
- Trial of 48 hour of medical management before any surgical intervention is indicated in:
 - **All stable pediatric patients with deep neck infections.**
 - Even sizable collections respond favorably with medical management.
 - **Stable adults with the following criteria:**
 1. Abscess cavities **< 2.5 cm** in diameter.
 2. Involving **one** single neck space.
- Repeat imaging and/or surgical intervention are necessary in patients who fail to improve or who worsen during the observation period.

▪ **IVF Hydration:**

- Most patients benefit from 1-2 L isotonic IVF infusion.
- Signs of dehydration includes:
 - Tachycardia
 - Dry mucous membranes
 - Decreased skin turgor

▪ **IV Antibiotics:**

- Should be started empirically with broad spectrum antibiotics covering the mixed oropharyngeal flora:
 - **Gram-positive cocci**
 - **Gram-negative rods**
 - **Anaerobes**
- Antibiotic coverage is expanded to cover **Pseudomonas** in cases of otologic infection, sinus infection or nosocomial infections (**Ciprofloxacin 400 mg Q12**).
- Then it should be directed according to the culture and sensitivity of drained abscess.
- IV antibiotics will be continued until patient is afebrile for 48 hours then switched to oral medication for additional 2 weeks.
- Patients who require surgical intervention need 48 hours of IV antibiotics post-op before discharge home on oral therapy.

• **Clindamycin:**

- Monotherapy
- Resistance in 5-10% of MRSA-related pediatric deep neck infections.
- IV Dose:
 - 15 mg/kg per dose Q8 in children
 - 600 mg Q6-8 in adults
- Oral Dose:
 - 15 mg/kg per dose Q8 in children
 - 300-450 mg Q6 in adults

• **Moxifloxacin:**

- Monotherapy
- Used in patients with odontogenic infections from *Eikenella corrodens* which is resistant to clindamycin, metronidazole and macrolides.
- IV and Oral Dose:
 - 400 mg daily in adults
- Contraindications:
 - Children
 - Pregnant

Box 10-1. FIRST-LINE ANTIBIOTIC ALTERNATIVES FOR DEEP NECK INFECTION

Community-Acquired Infection (Gram-Positive Cocci, Gram-Negative Rods, Anaerobes)

- Ampicillin-sulbactam 1.5-3.0 g IV q6h
- Clindamycin (if penicillin allergic) 600-900 mg IV q8h
- Moxifloxacin (if *Eikenella* is suspected) 400 mg daily

Compromised Patients/Nosocomial Infection (*Pseudomonas*; MRSA): Pseudomonal and Gram-Negative Alternatives

- Ticarcillin-clavulanate 3.0 g IV q6h
- Piperacillin-tazobactam 3.0 g IV q6h
- Imipenem-cilastatin 500 mg IV q6h
- Ciprofloxacin (if penicillin allergic) 400 mg IV q12h
- Levofloxacin (if penicillin allergic) 750 mg IV q24h

MRSA

- Clindamycin 600-900 mg IV q8h *plus* vancomycin 1 g IV q12h
- Trimethoprim-sulfamethoxazole 10 mg/kg/day divided q8h (if clindamycin-resistant) *plus* vancomycin 1g IV q12h

Necrotizing Fasciitis (Mixed Gram-Positive and Expanded Anaerobes)

- Ceftriaxone 2 g IV q8h *plus* clindamycin 600-900 mg IV q8h *plus* metronidazole 500 mg IV q6h

IV, intravenous; MRSA, methicillin-resistant *Staphylococcus aureus*.

▪ **IV Steroids:**

- Significantly reduces hospitalization hours, throat pain, fever, and trismus in comparison with treatment involving only parenteral antibiotics in patients with **peritonsillar abscess**.
- Single high dose of steroids (Methylprednisolone 2-3 mg/kg up to 250 mg).
- Also it helps to improves airway edema in patients with respiratory symptoms.

○ **Surgical Management:**

- Antibiotic availability in pus-filled spaces is limited by poor vascularity.
- Surgical drainage should be done only if the cellullitic process has localized into a discrete abscess.
- Premature incision into an area of cellulitis area may worsen the situation by breaking down the natural defenses and hastening the spread of infection.
- Indications of surgical intervention:
 - **Presence of complications:**
 - Airway compromise.
 - Septicemia
 - **Stable adult with following criteria:**
 1. Abscess cavities > 2.5 cm in diameter.
 2. Involving multiple neck spaces.
 3. Presence of gas in the neck.
 - **Failure to respond for 48 hours of empiric medical therapy.**
- Principles of surgical management:
 - Treatment depends on open incision and dependent drainage
 - Incisions should be placed to allow for gravity-dependent drainage when possible.
 - Important to open all primary and secondary spaces.
 - Fluid/tissue sample should be obtained for tissue staining, culture and sensitivity testing.
 - Thorough therapeutic irrigation of infected body cavity should be performed with sterile saline.
 - Larger cavities will remain open and be packed with antimicrobial iodoform, eventually healing by secondary intention.
 - Smaller cavities can be closed loosely with placement of Penrose drain to prevent the re-accumulation.
 - Involved teeth should be extracted ideally at time of incision and drainage.
- Methods of drainage:
 - **Needle Aspiration:**
 - Performed bed side or with US guidance.
 - Utilizing 18 gauge needle with negative pressure.
 - Indications:
 - Small abscesses contained within the confines of a lymph node.
 - Abscess caused by suspected congenital cysts.
 - Suspected granulomatous diseases (TB).

- **Trans-oral Incision and Drainage:**
 - Drain insertion is avoided due to risk of aspiration.
 - Indications:
 - Peritonsillar abscess.
 - Retropharyngeal abscess.
 - Parapharyngeal abscess just beneath lateral pharyngeal wall.
 - Shallow sublingual abscess.
 - Buccal abscess.
 - Masticator abscess.
- **Trans-cervical Incision and Drainage:**
 - Indications:
 - Parapharyngeal abscess.
 - Submandibular abscess.
 - Ludwig angina.
 - Prevertebral abscess
- Complications of Deep Neck Infections:
- Typically with delayed treatment or in immunocompromised patients.
 - **Airway obstruction.**
 - **Infectious complications:**
 - **Spreading of infection to other deep spaces.**
 - **Aspiration pneumonia:**
 - If abscess ruptures into the airway.
 - **Necrotizing cervical fasciitis:**
 - Rapid spread of infection throughout fascial planes.
 - Potentially lethal with high mortality rate **(30%)**.
 - Mainly affecting immunocompromised patients (DM, HIV).
 - Caused by polymicrobial odontogenic infection:
 - Clostridium, aerobes, anaerobes and MRSA.
 - Clinical picture:
 - Extremely toxic appearance with spiking fevers.
 - Ischemic discoloration of skin with crepitus.
 - CT findings:
 - Subcutaneous emphysema and gas formation
 - Non-loculated hypodense areas without peripheral enhancement.
 - Management:
 - ICU for unstable patients.
 - Broad spectrum IV antibiotics.
 - Correction causes of immunocompromise.
 - Multiple surgical debridements in OR:
 - For nonviable tissues until bleeding is reached.
 - Wound left open for 2nd look after 48 hours.
 - Adjuvant hyperbaric oxygen

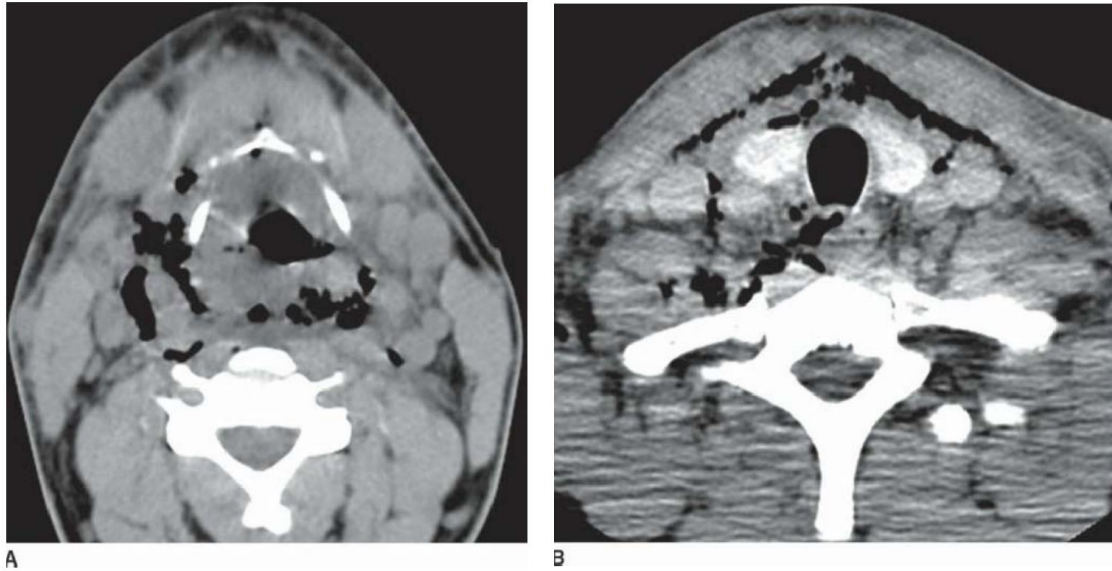
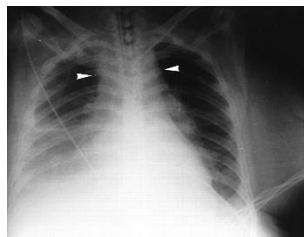


Figure 55.9 Necrotizing fasciitis. Axial unenhanced CT images at the level of the hyoid bone (A) and thyroid gland (B) show extensive gas formation throughout the deep fascial compartments of the neck. There is also extensive infiltration of the fat planes, which distinguishes this case of necrotizing fasciitis from perforation of the upper aerodigestive tract.

▪ Mediastinitis:

- Lethal with high mortality rate (40%).
- Extension from deep neck infections are rare.
- Infection spread through:
 - Retropharyngeal space
 - Danger space
 - Prevertebral space
 - Anterior visceral space
- Clinical picture:
 - Progressive chest pain and dyspnea.
 - Along with neck swelling.
- Imaging (CT or plain film):
 - **Widened mediastinum**
 - Fluid collections
 - Pneumomediastinum
 - Pneumothorax
- Management:
 - ICU for unstable patients.
 - Broad spectrum IV antibiotics.
 - Surgical drainage for mediastinal collection.



- **Vascular complications:**

- **Lemierre Syndrome:**

- Internal jugular thrombophlebitis.
- Caused most commonly by anaerobic gram-negative bacillus:
 - ***Fusobacterium necrophorum*.**
- Pathophysiology:
 - Bacterial endotoxin spread via tonsillar veins to internal jugular system.
 - Induces platelet aggregation and septic thrombus formation.
- Suspected in patients with:
 - Antecedent pharyngitis
 - Persistent fever despite antimicrobial therapy.
 - Lateral neck tenderness
 - Septic pulmonary other distant emboli.
- Findings on contrasted CT:
 - Intraluminal filling defects of IJV
 - Distended veins with enhancing walls
 - Swelling of adjacent soft tissues.
- Treatment:
 - Prolonged antibiotic therapy (4-6 weeks).
 - Anticoagulation (controversial).
 - Drainage of causative abscess.
 - Ligation of IJV for refractory cases.

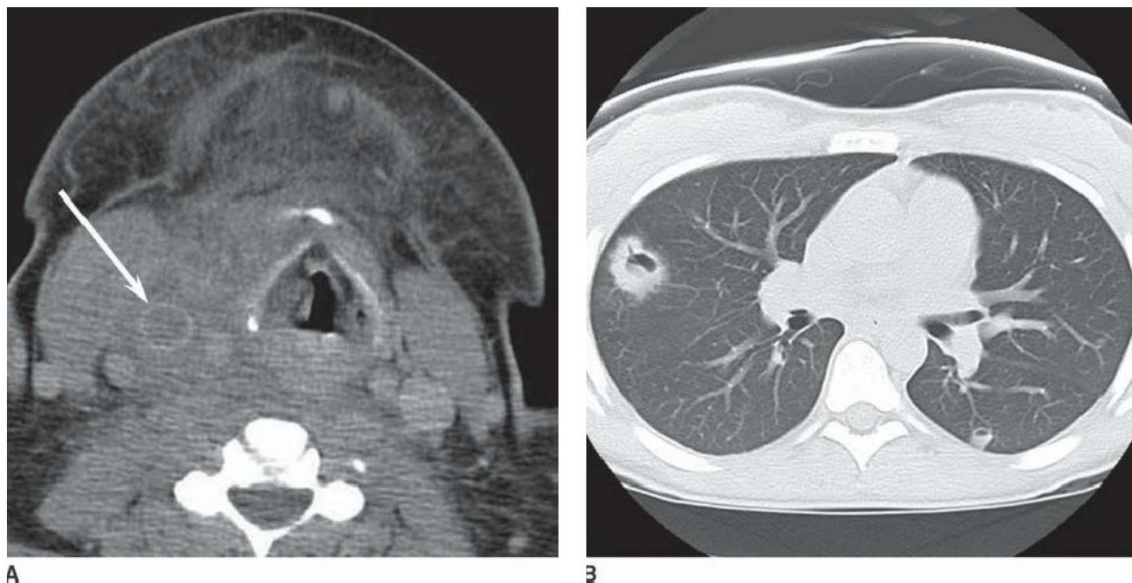


Figure 55.8 Internal jugular thrombophlebitis with Lemierre syndrome. **A:** Axial unenhanced CT shows hypodensity of the right IJV (arrow) with extensive surrounding fat infiltration. On contrast-enhanced CT, the vein would fail to enhance like the other vessels. Ultrasound may also be used to document disruption of venous flow. **B:** Axial CT on lung windows shows multiple cavitary lung masses with ill-defined borders, representing lung abscesses from infected emboli (Lemierre syndrome).

▪ **Cavernous Sinus Thrombosis:**

- Lethal with high mortality rate **(30%)**.
- Pathophysiology:
 - Caused by retrograde spread of infection from upper teeth or paranasal sinuses via valveless ophthalmic venous system to cavernous sinus.
- Clinical picture:
 - Fever
 - Lethargy
 - Orbital pain
 - Proptosis
 - Reduced extraocular mobility
 - Dilated pupil with sluggish papillary light reflex.
- Diagnosis:
 - Confirmed with Brain MRI with contrast.
 - Demonstrates dural enhancement in region of cavernous sinus.
- Treatment:
 - Prolonged antibiotic therapy (4-6 weeks).
 - Anticoagulation (controversial).
 - Drainage of causative abscess.

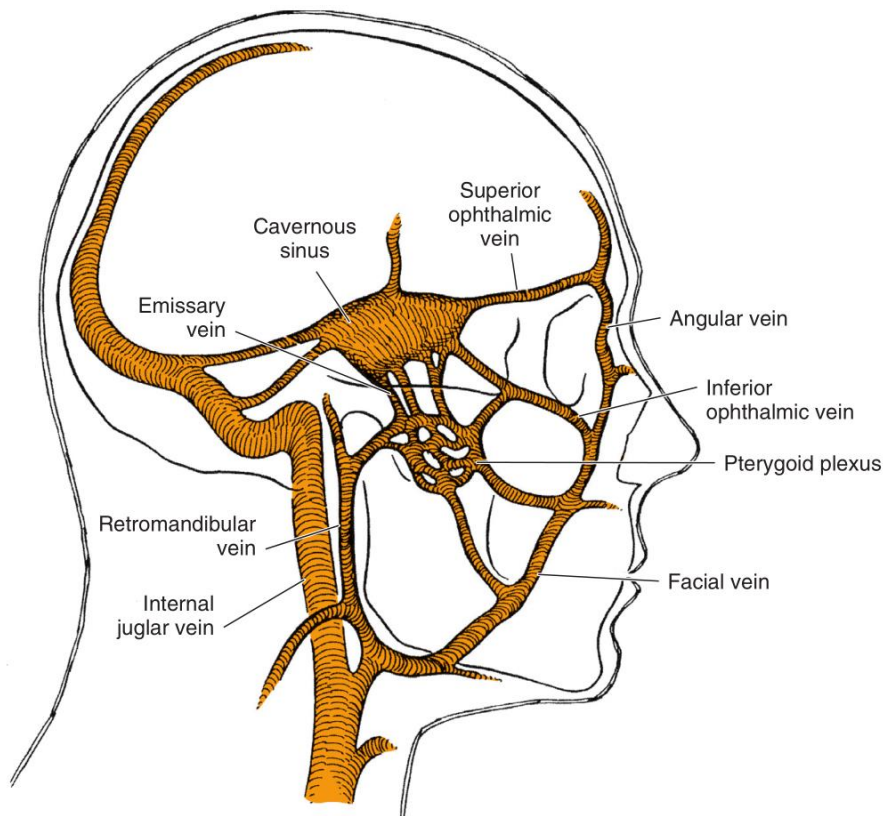


FIGURE 10-9. Infection can spread to the cavernous sinus from the jaw, facial soft tissues, and sinuses via the valveless inferior and superior ophthalmic venous plexus.

- **Carotid Artery Pseudoaneurysm or Rupture:**
 - Rare.
 - Spread of infection from retropharyngeal or parapharyngeal spaces to the carotid space.
 - Pathophysiology:
 - Pulsatile neck mass
 - Horner syndrome (CN IX to XII palsies).
 - Expanding hematoma or neck ecchymosis
 - Bright red blood from nose or mouth.
 - Treatment:
 - Immediate surgical ligation of common carotid artery.
 - If time permits, endovascular stenting or vessel occlusion in interventional radiology suite.
- **Neurological complications:**
 - **Nerve palsies:**
 - Caused by spread of infection to upper carotid space and parapharyngeal space.
 - Clinical picture:
 - CN IX to XII palsies.
 - Horner's syndrome:
 - Post-ganglionic sympathetic chain palsy.
 - Triad:
 - Ptosis
 - Anhydrosis
 - Miosis
 - **Meningitis.**
 - **Intracranial abscess.**

Peritonsillar Abscess (Quinsy):

- Most common deep neck infection in children and adolescents **(50%)**.
- Occurs most frequently in young adults from 15-30 year old.
- Peritonsillar space:
 - o **Boundaries:**
 - Tonsillar capsule medially and superior constrictor muscle laterally.
 - Anterior pillar anteriorly and posterior pillar posteriorly.
 - o **Contents:**
 - Loose connective tissue
 - Tonsillar branches of Lingual, Facial and Ascending pharyngeal vessels.
 - o **Communicating Spaces:**
 - Parapharyngeal space
- Pathophysiology:
 - o Results from:
 - Suppurative complication of acute tonsillitis with extension into peritonsillar space.
 - Obstruction of Weber glands (small salivary glands in the soft palate just superior to the tonsil and connected to the surface of the tonsil by a duct).
 - o Begins as a **peritonsillar cellulitis** and progresses to **peritonsillar abscess**, initially at superior pole of the tonsil.
 - o Delayed drainage of abscess result in parapharyngeal space extension.
- Bacteriology:
 - o Polymicrobial.
 - o Predominant bacterial species:
 - Streptococcus pyogenes (group A streptococcus)
 - Oral anaerobes
- Clinical picture:
 - o High fever
 - o Sore throat
 - o Odynophagia
 - o Unilateral otalgia:
 - Referred pain via CN-IX.
 - o Muffled (hot-potato) voice
 - o Drooling of saliva
 - o Trismus:
 - Irritation of pterygoid muscles which are in close proximity to the superior constrictor.
 - o Ipsilateral soft palate fullness or edema.
 - o Deviation of the uvula towards the unaffected side.
 - o Torticollis:
 - Patient keeps the neck tilted to the side of abscess.
 - o Cervical lymphadenopathy:
 - Jugulodigastric lymph nodes.

Peritonsillar abscess



A large unilateral abscess is visible in the pharynx of a patient examined in the Emergency Department. Prominent swelling of the anterior pillar and soft palate is present.

Courtesy of Lawrence B Stack, MD.

- Risk factors:
 - Recurrent infections
 - Smoking
- Diagnosis:
 - Made clinically without need for laboratory tests or imaging.
 - Diagnosis is confirmed by collection of pus at the time of drainage.
- Lab investigations:
 - Needed to know level of illness and direct the medical therapy.
 - Includes:
 - **CBC with differential:**
 - Elevated WBC with a predominance of neutrophils.
 - **Serum electrolytes:**
 - If the patient's oral intake has been decreased.
 - **Culture and sensitivity:**
 - Gram stain, culture (aerobic and anaerobic), and susceptibility testing of abscess fluid if a drainage procedure is performed.
 - Help guide antimicrobial therapy in immunocompromised patients or those with complications or extension of infection.

- Imaging:

- Not necessary to make the diagnosis of peritonsillar abscess.

- Indications:

1. Inadequate examination secondary to trismus.
2. Suspecting spread of infection to parapharyngeal space.
3. Suspecting other differential diagnosis (Epiglottitis and retropharyngeal abscess).

- Modalities:

- **Lateral neck radiographs:**

- Obtained initially to exclude epiglottitis or retropharyngeal abscess if these conditions have not been excluded clinically.

- **Intra-oral US:**

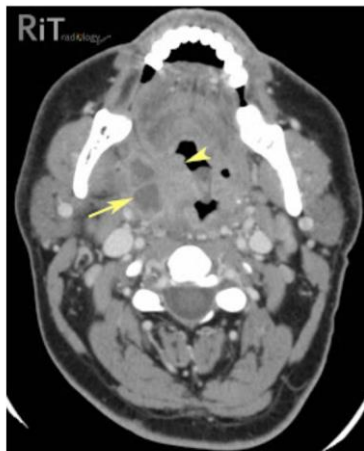
- Distinguishes peritonsillar abscess from cellulitis and to guide needle aspiration.
 - May be limited by trismus.

- **CT with IV contrast:**

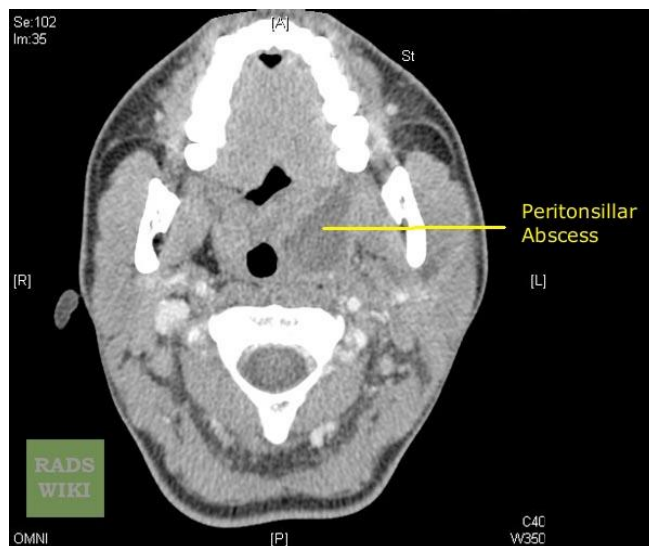
- **Preferred imaging modality.**
 - Distinguishes peritonsillar abscess from cellulitis and demonstrates the spread of infection to contiguous deep neck spaces.
 - Should be omitted in children with respiratory distress, and evaluation should be done in the operating room, where if necessary, an artificial airway can be established.

- Findings:

- Peritonsillar abscess appears as a hypodense mass with ring enhancement.
 - 25% of CT-suspected abscesses are phlegmonous and not drainable by I&D.
 - Peritonsillar cellulitis include soft tissue swelling, loss of the fat planes, and lack of ring enhancement.



Axial CT image shows a multilocular low density collection (arrow) beneath the enlarged right tonsil (arrowhead), which is displaced medially.



- Differential diagnosis:

- Major considerations in the differential diagnosis:

- **Epiglottitis:**

- More rapidly progressive.
- Occurs in younger children.

- **Retropharyngeal abscess or cellulitis:**

- Occurs in younger children.
- Neck stiffness is a prominent feature.
- Trismus is less common.

- **Parapharyngeal abscess:**

- Examination may reveal bulging behind the posterior tonsillar pillar rather than superior to the tonsil.
- Induration and swelling below the angle of the mandible.

- Treatment:

- **Medical:**

- **Hydration with IVF**
- **IV Analgesia**
- **IV Steroid**
- **IV Antibiotics (Clindamycin)**

Infection	Usual causative organisms	Antimicrobial regimens	
		Normal host	Compromised host ^Δ
NOTE: Coverage for methicillin-resistant Staphylococcus aureus (MRSA) should be included for those with risk factors ^o			
Peritonsillar abscess (Quinsy)	Group A streptococcus (S. pyogenes), Fusobacterium spp, Peptostreptococcus spp, and other oral anaerobes ^Δ	Ampicillin-sulbactam 3 g IV Q 6 h	Cefepime 2 g IV Q 12 h PLUS metronidazole 500 mg IV Q 6-8 h OR monotherapy with: ● Imipenem 500 mg IV Q 6 h or ● Meropenem 1 g IV Q 8 h or ● Piperacillin-tazobactam 4.5 g IV Q 6 h
		OR	
		Penicillin G 2-4 MU IV Q 4-6 h plus Metronidazole 500 mg IV Q 6-8 h	
		OR	
		Clindamycin 600 mg IV Q 6-8 h	

- **Surgical:**

○ **Drainage of Abscess:**

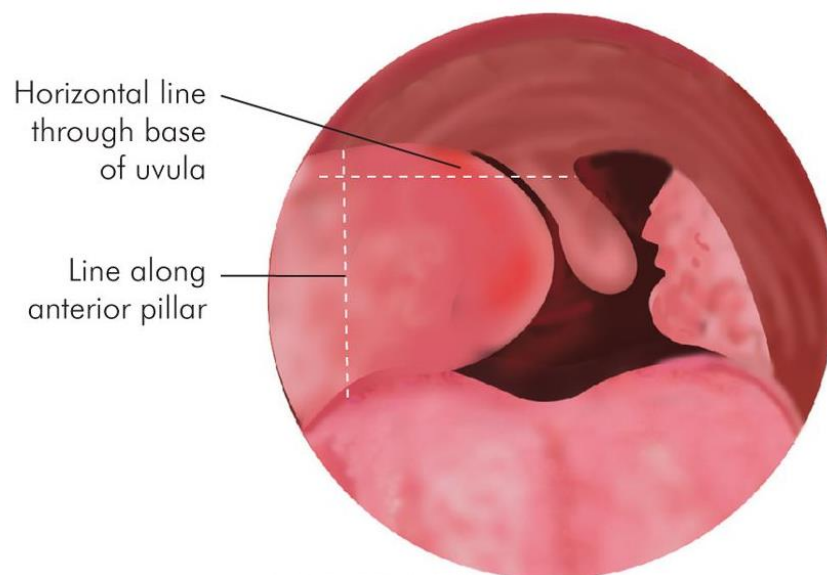
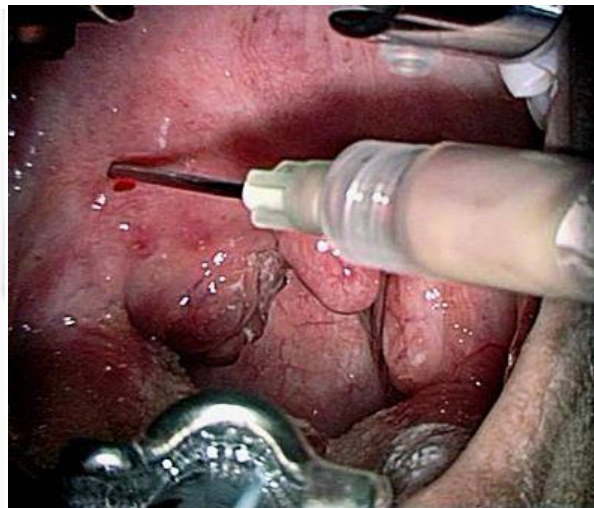
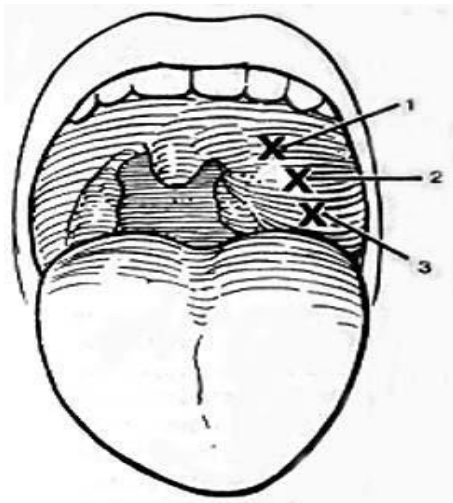
▪ Indications:

1. Peritonsillar abscess (Not cellulitis).
2. No improvement after 24 hours of medical management.

▪ Methods:

• **Needle Aspiration:**

- Less invasive and less painful.
- 95% successful in acute resolution.
- Done using topical anesthesia.
- 18-gauge spinal needle with a 10-mL syringe.
- At point of maximum bulge above upper pole of tonsil or just lateral to point of junction of anterior pillar with a line drawn through the base of uvula.
- 3-point aspiration (0.5 cm inferior and lateral).



- **Incision and Drainage:**
 - More definitive.
 - More painful and causes more bleeding.
 - Done using topical anesthesia.
 - GA is usually required for young children.
 - Use a small curved mosquito and #11 blade with the sharp edge facing the curve of the mosquito to protect the rest of the oral cavity and give you depth control.
 - Vertical stab incision is made just enough to get the tip of the mosquito in to open the abscess.
 - If still no puss comes out, and the patient is sick consider a neck CT with contrast to rule out extension to parapharyngeal space.
- **Hot (Quinsy) Tonsillectomy:**
 - Indication:
 1. Uncooperative children
 2. Significant upper airway obstruction.
 3. Unresponsive cases
 - Risks:
 - Abscess rupture during intubation.
 - Excessive bleeding intraoperatively.
- **Interval Tonsillectomy:**
 - Done after 4 weeks after resolution of infection.
 - Indication:
 - Recurrent tonsillitis.
 - Previous attack of peritonsillar abscess.
- **Response to Treatment:**
 - Successful treatment is defined by symptomatic improvement within 24 hours of intervention.
 - Failure of improvement indicates:
 - Complications
 - Infection with unusual organisms
 - Revaluation with:
 - Repeat CT with contrast to look for extension of infection.
 - Repeat surgical intervention.
 - Broadening antimicrobial therapy.
 - Recurrence rate of 10-15%.

- Complications of peritonsillar abscess:

- Early diagnosis and prompt, appropriate management of peritonsillar infection is critical to avoiding complications.

▪ **Airway obstruction.**

▪ **Infectious complications:**

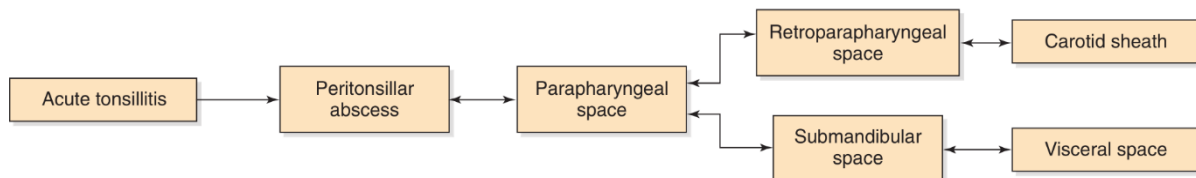
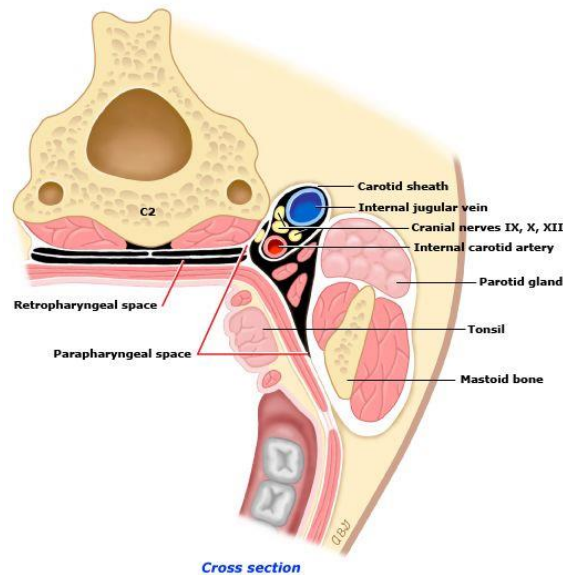
- **Spreading of infection to other deep spaces :**
 - Parapharyngeal space.
- **Aspiration pneumonia**
- **Necrotizing cervical fasciitis**
- **Mediastinitis**
- **Septicemia.**

▪ **Vascular complications:**

- **Lemierre Syndrome**
- **Cavernous Sinus Thrombosis**
- **Carotid Artery Pseudoaneurysm or Rupture**

▪ **Neurological complications:**

- **Nerve palsies.**
- **Meningitis.**
- **Intracranial abscess.**



Submandibular Abscess:

- Submandibular space:

- Composed of two spaces divided by Mylohyoid muscle, and continuous with each other along the posterior free border of Mylohyoid muscle:

▪ **Sublingual space:**

- Boundaries:
 - Between floor of mouth and mylohyoid muscle.
 - From tongue base posteriorly to mandible anteriorly.
- Contents:
 - Sublingual gland
 - Deep part of submandibular gland
 - Wharton's duct
 - Hypoglossal nerve (CN XII)
 - Lingual nerve
- Communicating spaces:
 - Opposite sublingual space
 - Submandibular space
 - Parapharyngeal space.

▪ **Submandibular/Submylohyoid/ Submaxillary space:**

- Boundaries:
 - Between mylohyoid muscle and superficial layer of deep cervical fascia.
 - From hyoid posteriorly to mandible anteriorly.
- Contents:
 - Submandibular gland
 - Anterior digastric muscle.
 - Facial artery
 - Submandibular lymph nodes
- Communicating spaces:
 - Opposite submandibular space
 - Sublingual space
 - Parapharyngeal space.

Submandibular space

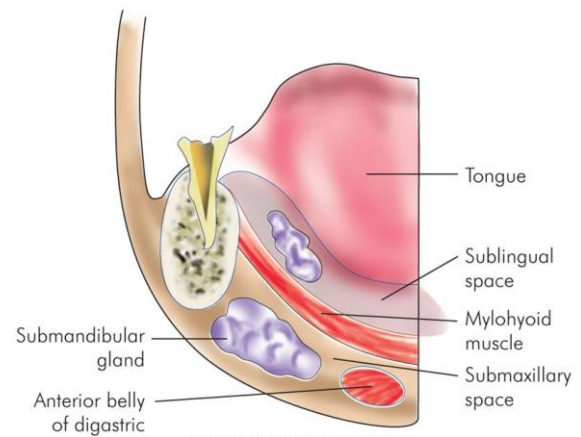
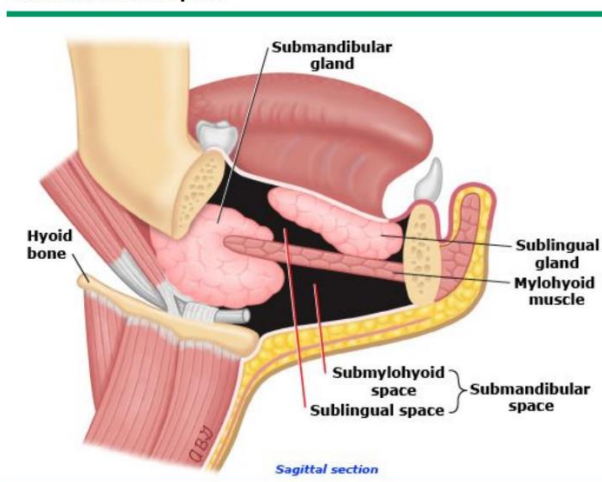


Figure 51.1 Anatomy of submandibular space.

- Pathophysiology:

○ Results from:

▪ **Dental infections:**

- Most common cause (80%).
- Sublingual space infection:
 - Caused by infection from **Incisors to 1st Molar**.
 - Root of these teeth lie above the attachment line of mylohyoid muscle.
- Submandibular space infection:
 - Caused by infection from **2nd and 3rd Molars**.
 - Root of these teeth lie below the attachment line of mylohyoid muscle.

▪ **Submandibular sialadenitis**

▪ **Suppurative lymphadenitis**

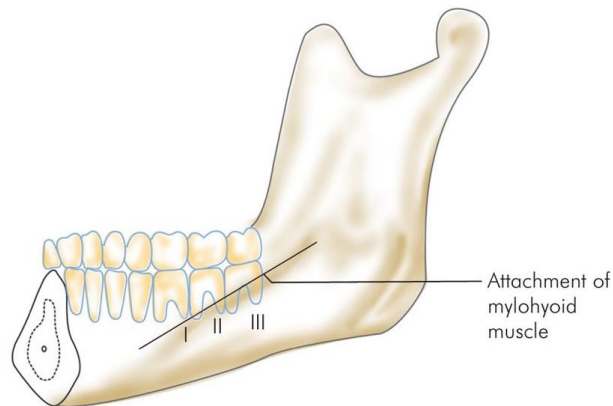
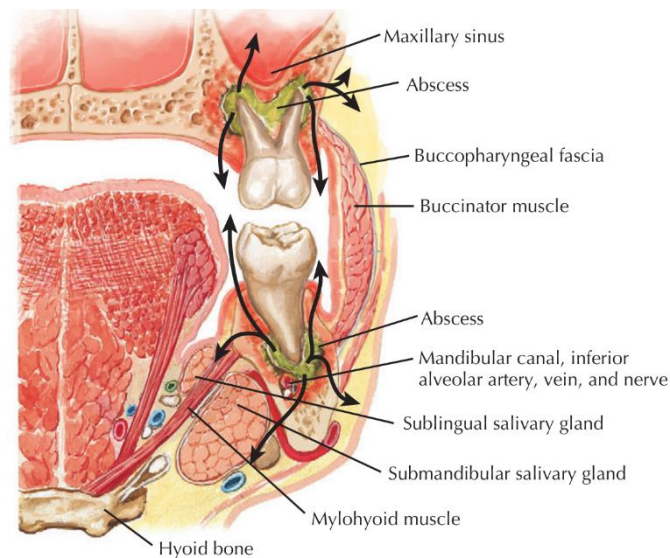


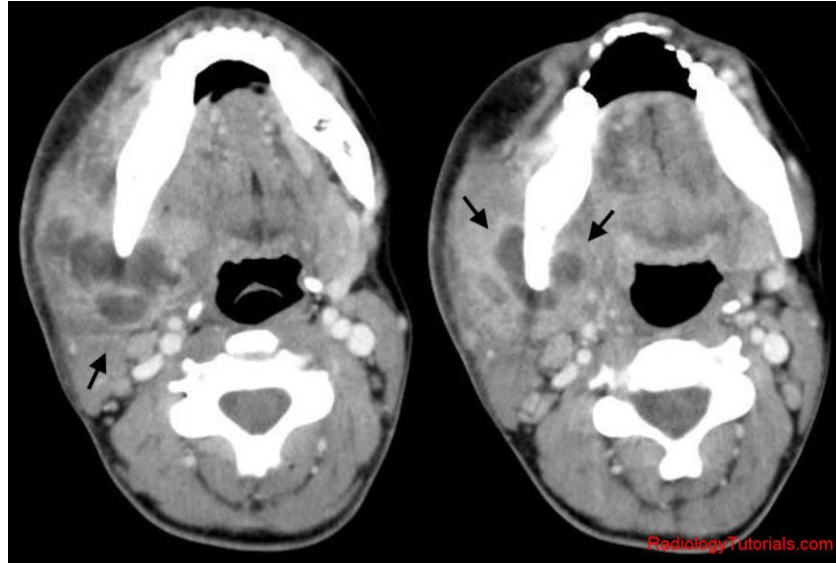
Figure 51.2 Roots of molar teeth project below, and those of premolars above the attachment of mylohyoid muscle.



Coronal section posterior to 1st molar tooth (anterior view)

- Clinical picture:

- Fever
- Sore throat
- Odynophagia
- Trismus
- Cervical lymphadenopathy



- Treatment:

- **Medical:**

- Hydration with IVF
- IV Analgesia
- IV Steroid (+/-)
- IV Antibiotics (**Clindamycin**)

Infection	Usual causative organisms	Antimicrobial regimens	
		Normal host	Compromised host ^A
NOTE: Coverage for methicillin-resistant <i>Staphylococcus aureus</i> (MRSA) should be included for those with risk factors ^o			
Submandibular space infections (Ludwig's angina)	Viridans and other streptococci, <i>Staphylococcus</i> spp, <i>Peptostreptococcus</i> spp, <i>Bacteroides</i> spp, and other oral anaerobes ^A	Ampicillin-sulbactam 3 g IV Q 6 h OR	Cefepime 2 g IV Q 12 h PLUS metronidazole 500 mg IV Q 6-8 h OR monotherapy with: ● Imipenem 500 mg IV Q 6 h or ● Meropenem 1 g IV Q 8 h or ● Piperacillin-tazobactam 4.5 g IV Q 6 h
		Penicillin G 2-4 MU IV Q 4-6 h plus Metronidazole 500 mg IV Q 6-8 h OR	
		Clindamycin 600 mg IV Q 6-8 h	

- **Surgical:**

○ **Drainage of Abscess:**

- US guided aspiration for localized abscess.
 - Trans-cervical incision and drainage:
 - Standard approach.
 - Done with horizontal submandibular cervical incision.
 - Allows **dependent** drainage of both compartments of the submandibular space.
 - Evacuation of pus followed by saline irrigation and drain insertion.
 - Trans-oral incision and drainage:
 - Utilized in selected patients with shallow sublingual collection.
 - Done with incision located at anterior and lateral portion of floor of mouth, parallel and lateral to submandibular duct.
- **Tooth Extraction:**
- Eliminating the source of infection in odontogenic causes.

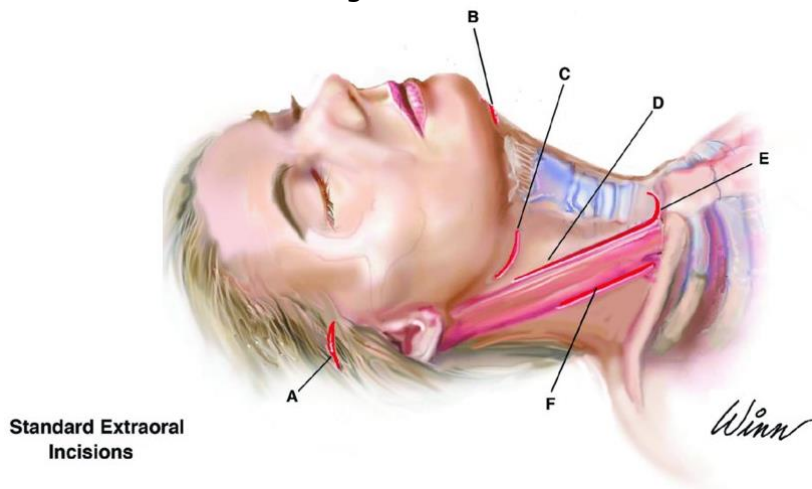
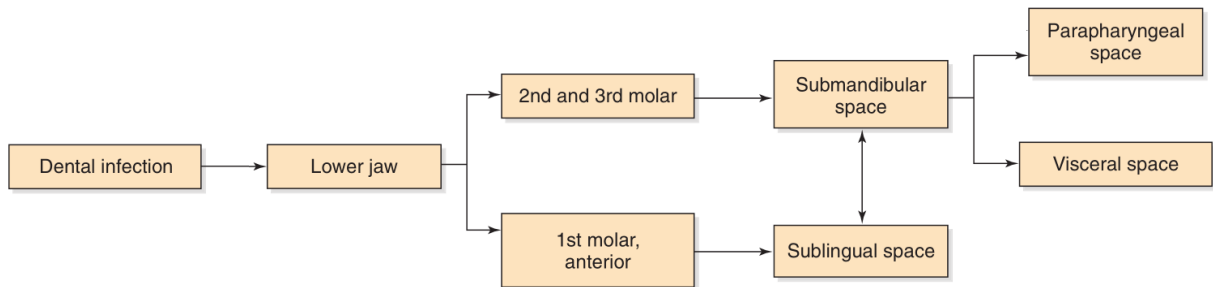


Figure 10.3. (A) Gillies incision; (B) submental incision; (C) submandibular incision; (D) anterior sternocleidomastoid muscle (SCM) incision; (E) transcervical mediastinum incision; and (F) posterior SCM incision.



- Complications of submandibular abscess:

- **Airway obstruction.**
- **Infectious complications:**
 - **Spreading of infection to other deep spaces:**
 - Bilateral submandibular (Ludwig's Angina).
 - Parapharyngeal space.
 - **Aspiration pneumonia**
 - **Necrotizing cervical fasciitis**
 - **Mediastinitis**
 - **Septicemia.**
- **Vascular complications:**
 - **Lemierre Syndrome**
 - **Cavernous Sinus Thrombosis**
 - **Carotid Artery Pseudoaneurysm or Rupture**
- **Neurological complications:**
 - **Nerve palsies.**
 - **Meningitis.**
 - **Intracranial abscess.**



Ludwig's Angina:

- Bilateral cellulitis of both compartments of submandibular space.
- No lymphatic involvement or abscess formation.
- Most commonly due to infection of 2nd or 3rd mandibular molars.
- Clinical picture:
 - o Most common in children
 - o Odynophagia
 - o Trismus (if parapharyngeal spread)
 - o Swelling of floor of mouth
 - o Swelling and displacement of tongue superiorly and posteriorly.
 - o Bilateral submandibular and submental swelling and tenderness (woody-hard feel).
 - o Risk of airway obstruction.

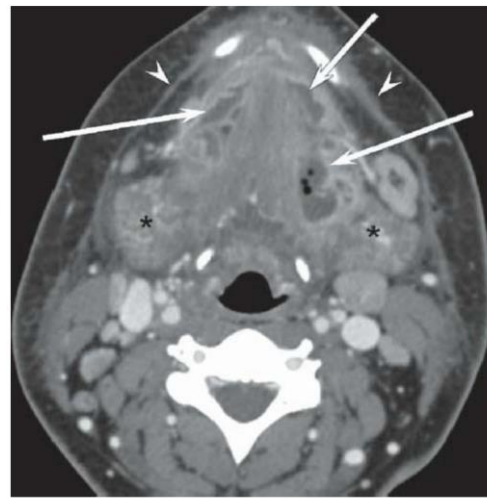
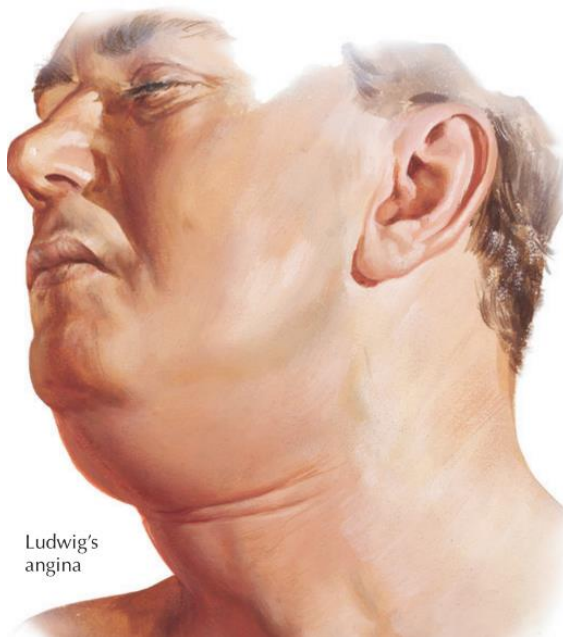


Figure 55.14 Ludwig angina. Axial contrast-enhanced CT shows diffuse infiltration of the fat planes in the submandibular, submental, and subcutaneous spaces. The platysma muscles (arrowheads) are thickened. The submandibular glands (asterisks) are inflamed. There is an extensive abscess (arrows) throughout the submandibular and submental spaces, with air locules indicating anaerobic infection.



FIGURE 10-4. A patient with Ludwig angina with swelling of the bilateral upper neck (A) and bilateral sublingual spaces causing the tongue to obstruct the upper airway (B).

- Treatment of Ludwig's Angina:
 - **Medical Treatment:**
 - **Early stage (Stable without airway compromise):**
 - Close monitoring of airway in ICU.
 - IV Antibiotics (**Clindamycin**)
 - IV Steroids (+/-)
 - **Stable with signs of airway compromise:**
 - Secure safe airway:
 - Fiberoptic oral or nasal intubation in semi-seated position with instruments for surgical airway immediately on hand.
 - Awake tracheostomy.
 - IV Antibiotics (**Clindamycin**)
 - IV Steroids (+/-)
 - **Surgical Treatment:**
 - **Incision and Drainage (I&D):**
 - Indications:
 1. Patients with abscess
 2. Patients with impending complications
 3. No improvement after 48 hours of IV Antibiotics.
 - Earlier surgical intervention improves outcomes.
 - Typically, no discrete abscess is encountered but rather a malodorous "dirty dishwater" fluid.
 - Trans-cervical approach with midline horizontal incision above the hyoid and extended laterally to angle of the mandible.
 - Evacuation of puss followed by irrigation and drain insertion.
 - **Tooth extraction:**
 - Should be done to eliminate source of infection.

TABLE 53.2 ANTIBIOTICS FOR ODONTOGENIC INFECTIONS		
	Oral	Parenteral
Very effective	amoxicillin/clavulanic acid penicillin clindamycin metronidazole (anaerobes only)	ampicillin/clavulanic acid penicillin clindamycin metronidazole (anaerobes only)
Effective	erythromycin cephalexin tetracycline	cefazolin cefoxitin carbapenems
Not effective	sulfa quinolone	gentamicin tobramycin amikacin Third generation cephalosporins

Buccal Abscess:

- Buccal space:

○ **Boundaries:**

- Buccinator muscle medially and cheek skin laterally.
- Zygomatic arch superiorly and lower border of mandible inferiorly.

○ **Contents:**

- Buccal fat pad
- Stensen's duct
- Transverse facial vessels
- Anterior facial vessels

○ **Communicating Spaces:**

- Submandibular space
- Masticator space
- Infratemporal space

- Pathophysiology:

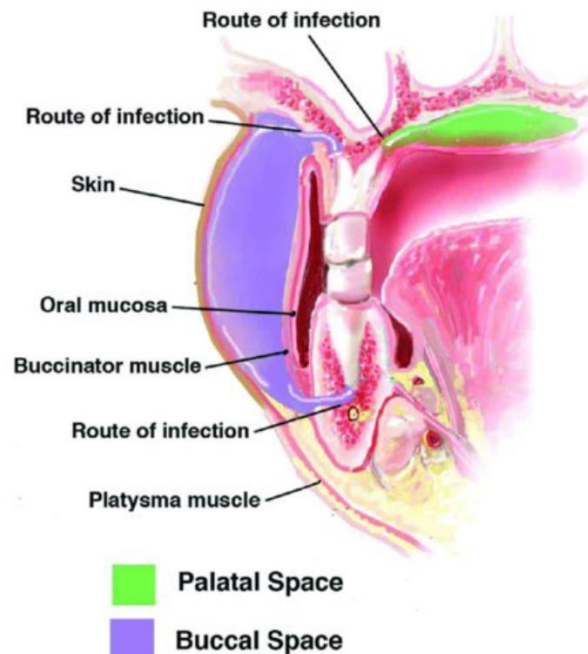
○ Results from:

▪ **Dental infections:**

- Most common cause.
- Infection of any molars.

▪ **Infected facial tissue fillers:**

- Patients with an atypical course of facial inflammation should be questioned about a history of cosmetic procedures.
- Period between injection and infection varies from 1 week to 6 years.



- Clinical picture:

- Fever
- Anterior Facial swelling:
 - May extend up to eyelid (preseptal) and orbicularis oris.
- Trismus:
 - Indicates posterior spread of infection to masticator space.

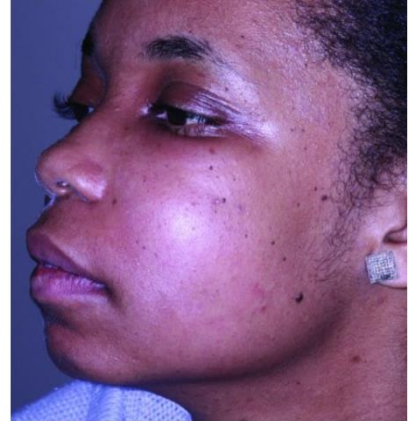
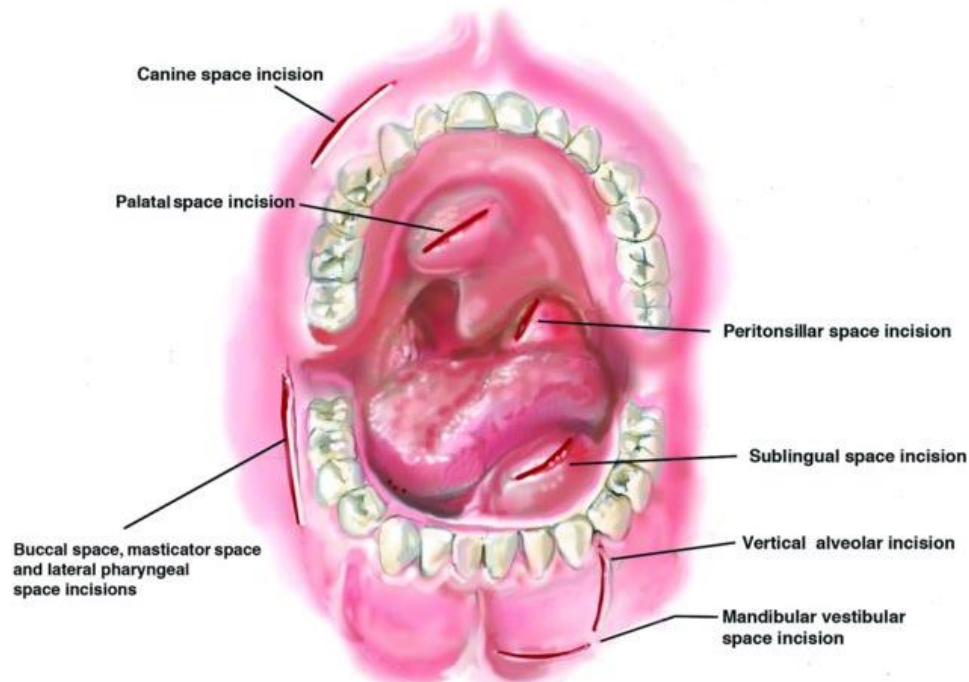


Figure 10.2. Typical presentation of a buccal space abscess with marked cheek edema anterior to the masseter.

- Treatment:

- **Antibiotics (Clindamycin)**
- **Drainage of Abscess:**
 - Aspiration for localized abscess.
 - Trans-oral incision and drainage:
 - Accomplished via horizontal mandibular or maxillary vestibular incision with blunt dissection parallel to facial nerve through buccinators.
 - Extra-oral incision and drainage:
 - Incision inferior to point of fluctuance.
 - Submandibular incision can be done to avoid facial scars.
- **Tooth Extraction:**
 - Eliminating the source of infection in odontogenic causes.

Intraoral Incisions



Masticator Abscess:

- Masticator space:

○ **Compartments:**

- Masseteric space:
 - Between masseter muscle and ramus of the mandible.
- Pterygomandibular space:
 - Between pterygoid muscles and ramus of the mandible.
- Superficial Temporal space:
 - Between superficial temporal fascia and temporalis muscle
- Deep Temporal space:
 - Between temporal fascia and temporal bone.

○ **Contents:**

- Muscles of mastication
- Body and ramus of mandible
- Inferior alveolar nerve
- Internal maxillary artery

○ **Communicating Spaces:**

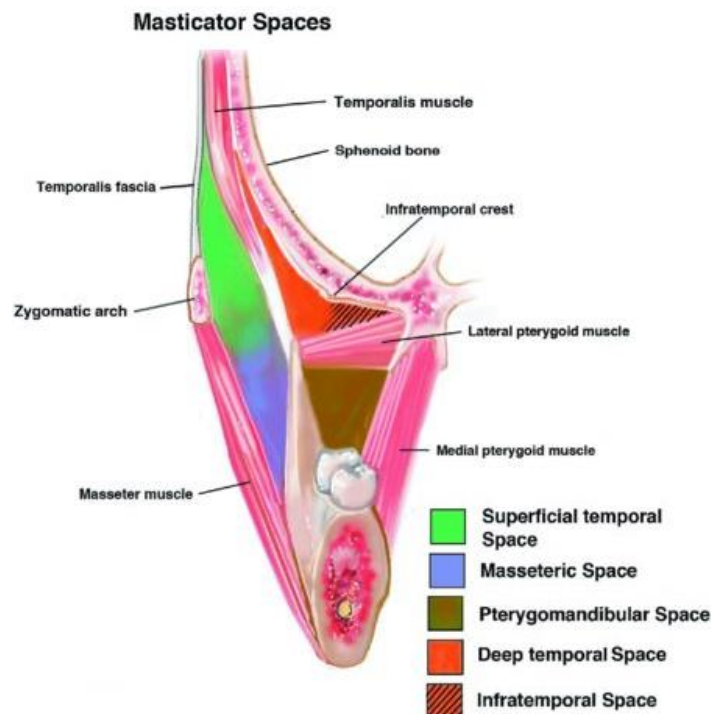
- Parapharyngeal space
- Pterygomaxillary space
- Buccal space
- Parotid space

- Pathophysiology:

○ Results from:

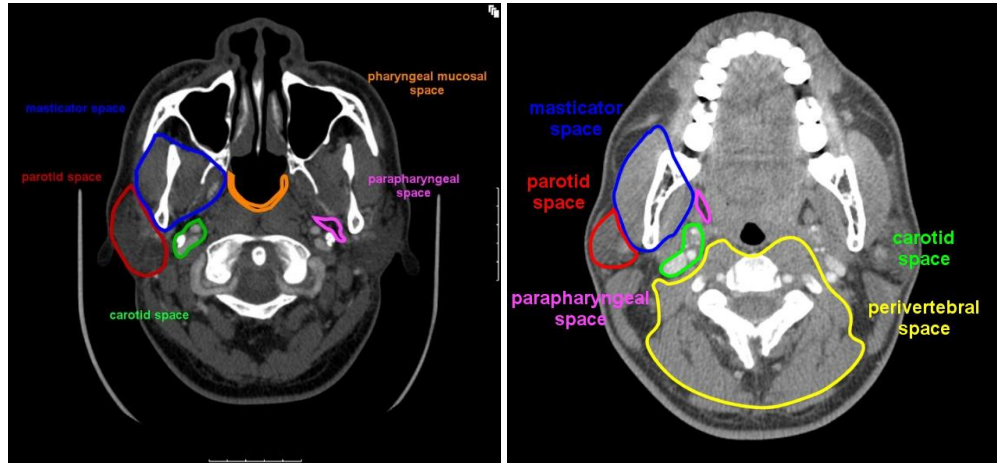
▪ **Dental infections:**

- Most common cause.
- Infection of 3rd mandibular molar.



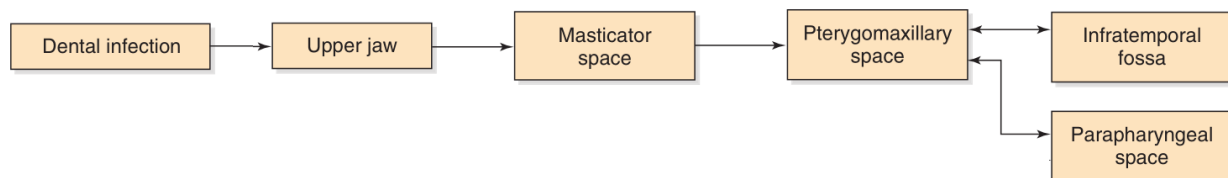
- Clinical picture:

- Fever
- Facial swelling over mandibular ramus
- Trismus



- Treatment:

- **Antibiotics (Clindamycin)**
- **Drainage of Abscess:**
 - Aspiration for localized abscess.
 - Trans-oral incision and drainage:
 - Not practical due to the presence of trismus and the inability to establish dependent drainage.
 - Trans-cervical incision and drainage:
 - Submandibular approach to allow for gravity dependent drainage.
 - After entering the submandibular space, blunt dissection is continued posteriorly through the pterygomasseteric sling to enter the masseteric space located between the body of the masseter and the lateral ramus of the mandible.
 - Once the inferior border of the mandible is reached, blunt dissection proceeds posteriorly through the pterygomasseteric sling to enter the pterygomandibular space located between the body of the medial pterygoid muscle and the medial ramus of the mandible.
- **Tooth Extraction:**
 - Eliminating the source of infection in odontogenic causes.



Parotid Abscess:

- Parotid space:
 - **Boundaries:**
 - Parotid fascia laterally and parapharyngeal space medially.
 - **Contents:**
 - Parotid gland
 - CN-VII
 - Posterior facial vein
 - External carotid artery
 - Periparotid lymph nodes
 - **Communicating Spaces:**
 - Parapharyngeal space
 - Masticator space
- Pathophysiology:
 - Results from parotid infections.
 - Parotid fascia adheres tightly to the gland resulting in formation of firm capsule which make differentiation of abscess versus cellulitis is impossible on physical examination (No fluctuation).
 - Parotid fascia is weaker on the medial side leading to the spread of infection into parapharyngeal space.
- Clinical picture:
 - Fever
 - Facial swelling over parotid region and angle of mandible
 - Trismus
 - Pus from stenson's duct on pressure over the parotid
- Treatment:
 - **Supportive (Hydration, sialagogues, oral hygiene, warm compresses, massage)**
 - **Antibiotics (Clindamycin)**
 - **Drainage of Abscess:**
 - Aspiration for localized abscess.
 - Trans-cervical incision and drainage:
 - Modified Blair incision to reach parotid gland.
 - Abscess is bluntly opened working parallel to the branches of the VIIth nerve.
 - Risk of facial nerve injury.



Parapharyngeal Abscess:

- Parapharyngeal space:

- **Boundaries:**
 - Inverted pyramid from skull base to **hyoid bone**.
 - Buccopharyngeal fascia covering superior constrictor muscle medially and medial pterygoid muscle and deep portion of parotid gland laterally.
- **Compartments:**
 - Pre-styloid:
 - Anterior to styloid process.
 - Contains:
 - Fat
 - Deep lobe of parotid gland
 - Lymph nodes.
 - Internal maxillary artery
 - Inferior alveolar nerve
 - Lingual nerve
 - Auriculotemporal nerve
 - Post-styloid:
 - Posterior to styloid process.
 - Contains (neurovascular bundle):
 - Carotid artery
 - Internal jugular vein
 - Sympathetic chain
 - CN-IX
 - CN-X
 - CN-XI
 - CN-XII
- **Communicating Spaces:**
 - Retropharyngeal space
 - Peritonsillar space
 - Submandibular space
 - Masticator space
 - Parotid space
 - Carotid space

- Pathophysiology:

- Potentially life-threatening infection due to risk of complications:
 - Carotid sheath involvement
 - Airway impingement
 - Bacteremic dissemination.
- Results from:
 - **Dental infections (Most common)**
 - **Peritonsillar abscess**
 - **Pharyngeal infection**
 - **Parotid gland infection**
 - **Submandibular infection**

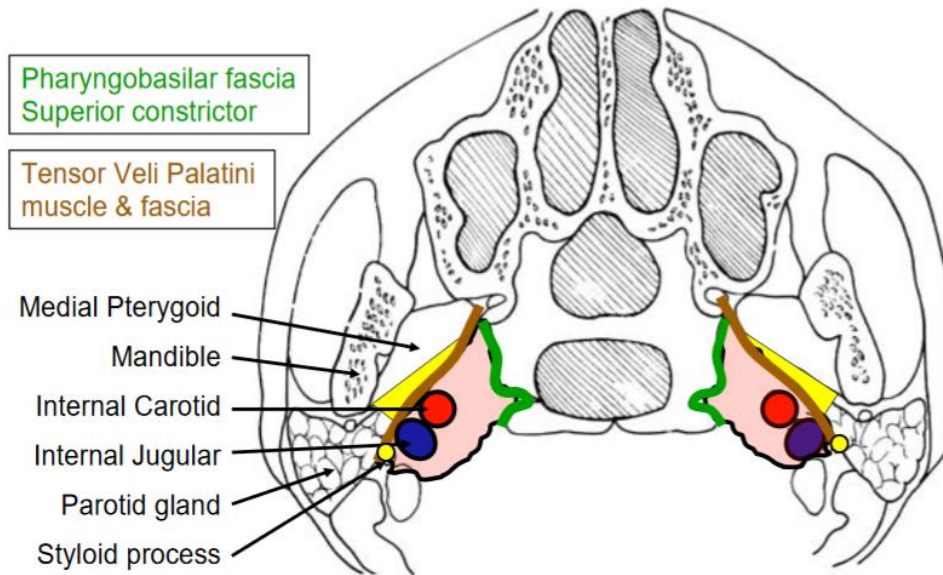


Figure 57: Schematic axial view of prestyloid (yellow) and poststyloid (pink) parapharyngeal spaces

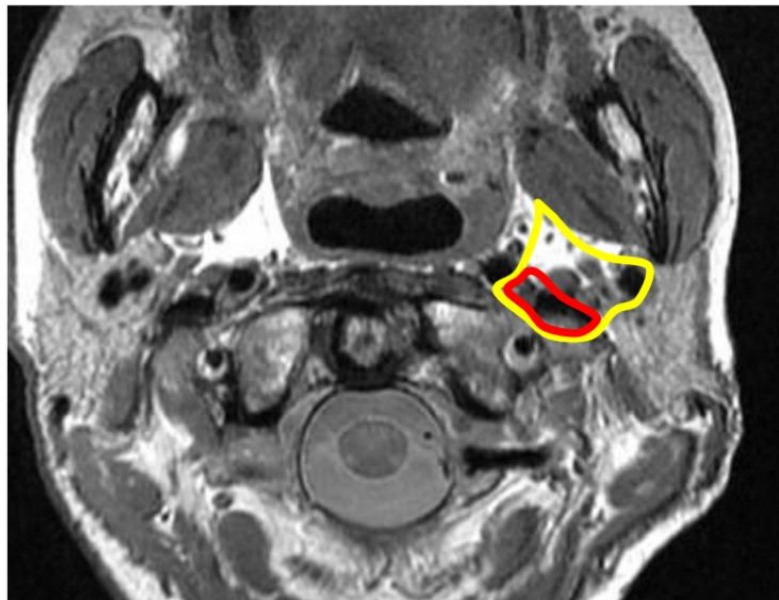


Figure 58: Parapharyngeal space (yellow outline) and carotid space/poststyloid space (red outline)

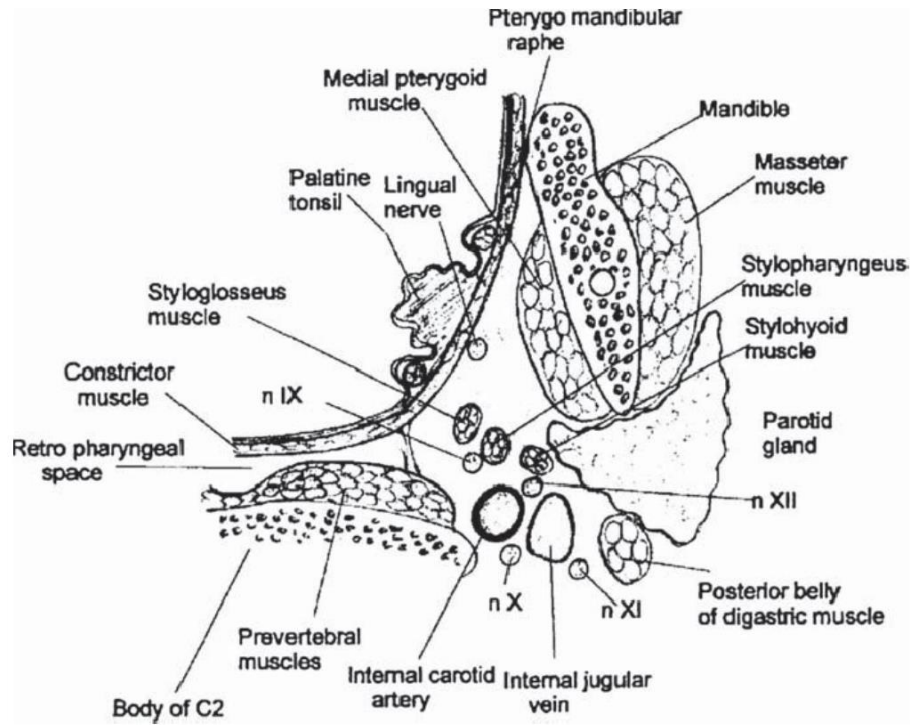
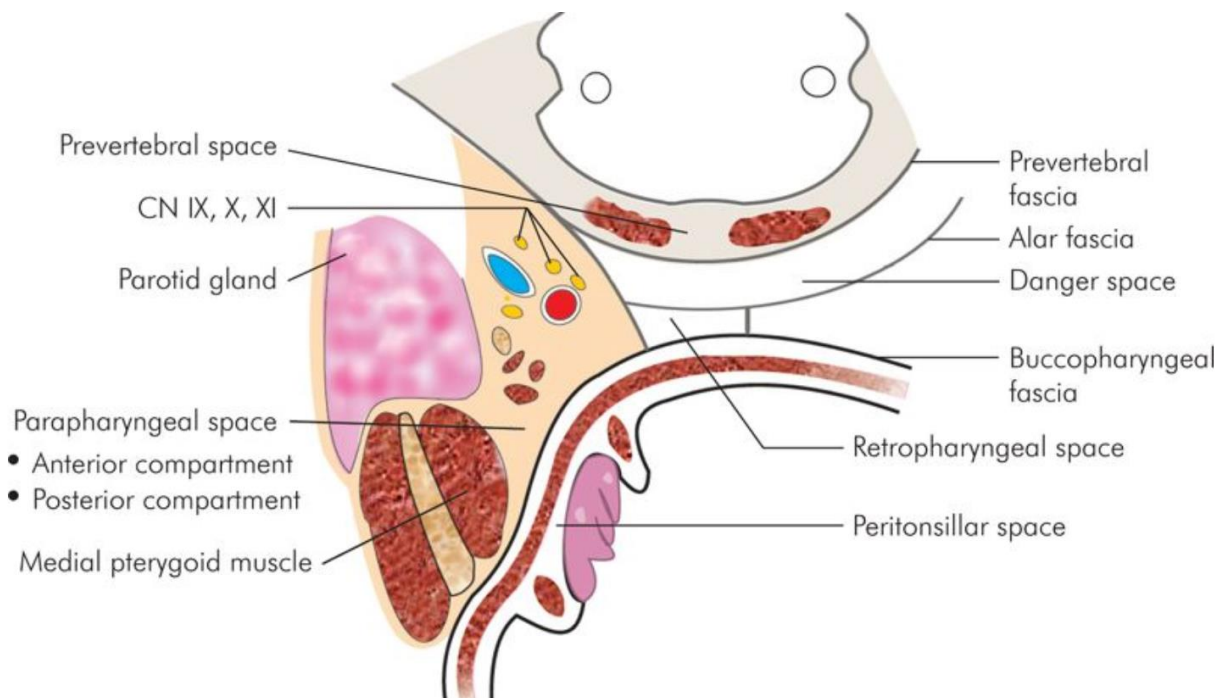


Figure 55.5 Schematic representation of the parapharyngeal space in an axial view at the C2 level.



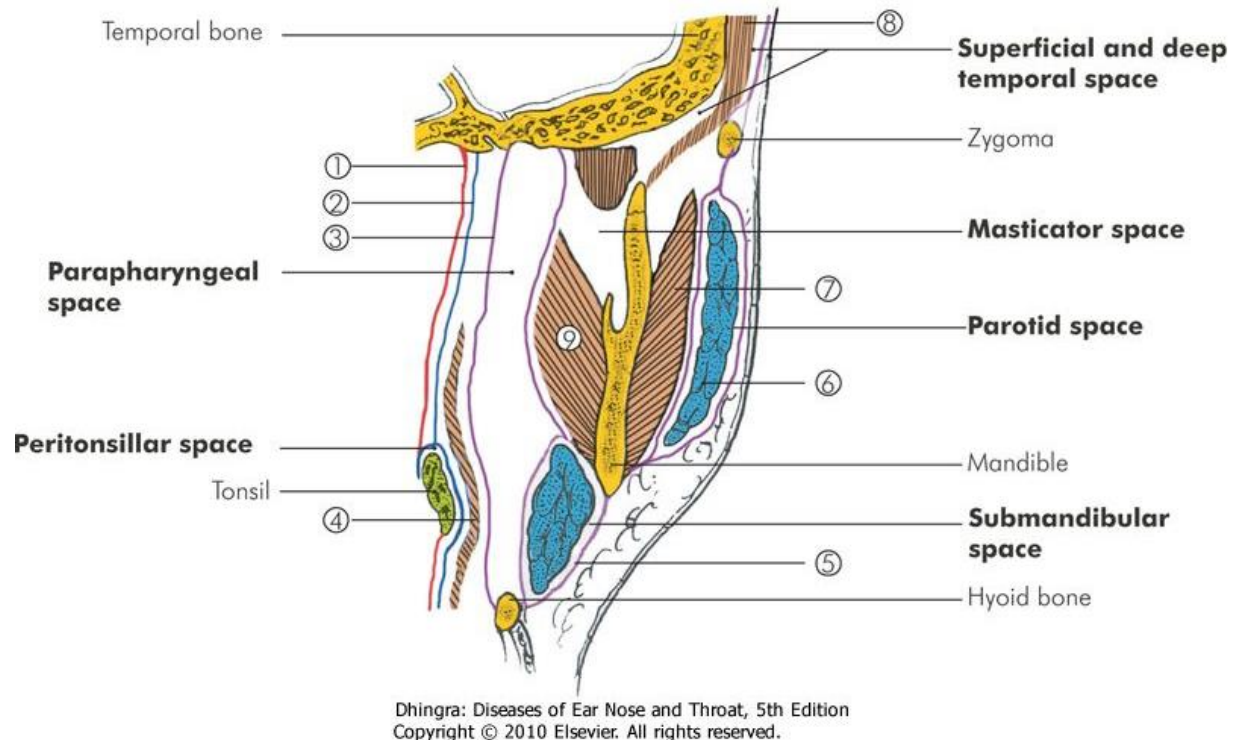


Figure 51.7 Spaces of head and neck seen in coronal section. Mucosa (1), pharyngobasilar fascia (2), buccopharyngeal fascia (3), superior constrictor muscle (4), superficial layer of deep cervical fascia enclosing submandibular gland (5), parotid gland (6), masseter muscle (7), temporalis muscle (8), medial pterygoid muscle (9).

- Clinical picture:

- Symptoms and signs of the primary source of infection.
- Cardinal clinical features of parapharyngeal space infections:
 - **Trismus (due to spasm of medial pterygoid muscle)**
 - **Torticollis (due to spasm of prevertebral muscles)**
 - **Induration and swelling below the angle of the mandible**
 - **Medial bulging of the pharyngeal wall**
 - **Systemic toxicity with fever and rigors**

- Treatment:

- **Antibiotics (Clindamycin)**
- **Drainage of Abscess:**
 - US guided aspiration for localized abscess.
 - Trans-oral incision and drainage:
 - Incising anterior pillar lateral to the tonsil.
 - Indicated for localized abscess medial to great vessels.
 - Trans-cervical incision and drainage:
 - Modified apron incision used to expose submandibular area and upper parapharyngeal space.
 - Hockey stick incision (Anterior border of SCM) used to expose entire parapharyngeal space.
- **Tooth Extraction:**
 - Eliminating the source of infection in odontogenic causes.

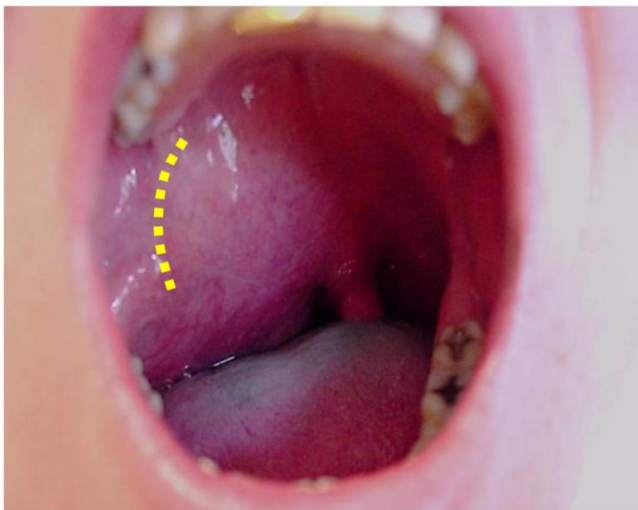


Figure 61: Typical clinical picture of a prestyloid PPS abscess and incision

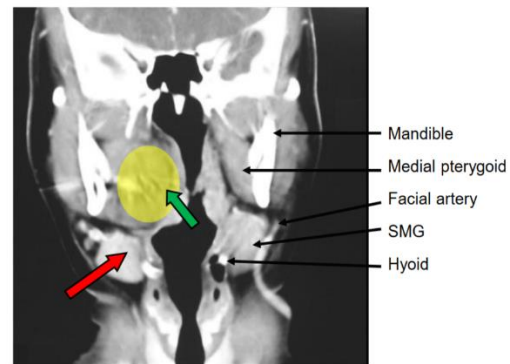


Figure 62: Anatomical relations of PPS abscess and drainage approaches either via oral cavity (green arrow) by incising the anterior tonsillar pillar lateral to the tonsil or via submandibular triangle above hyoid by dissecting bluntly with a finger posterior to the submandibular gland (red arrow)

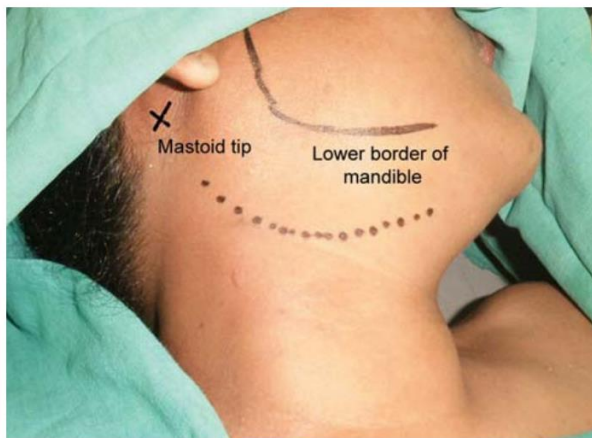


Fig. 2: The outline of modified apron incision

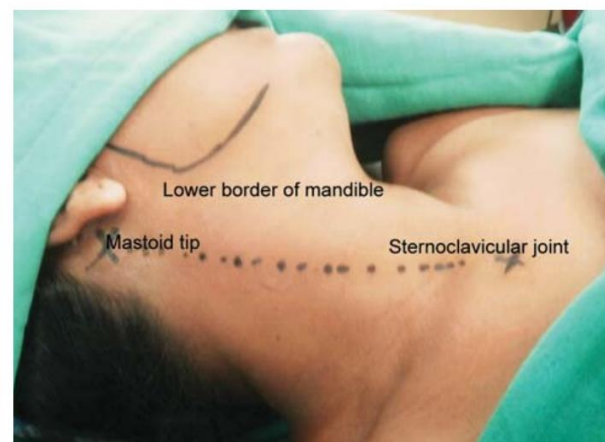


Fig. 3: The outline of 'hockey stick incision'

Retropharyngeal Abscess:

- Retropharyngeal space:
 - **Boundaries:**
 - From skull base to **carina in superior mediastinum (T2)**.
 - Buccopharyngeal fascia anteriorly and Alar fascia posteriorly.
 - Midline fascia separates retropharyngeal space into two lateral compartments.
 - **Contents:**
 - Nodes of Rouviere:
 - Drains sinonasal and nasopharynx.
 - Frequently disappear by age of 5 year.
 - **Communicating Spaces:**
 - Parapharyngeal space
 - Danger space
 - Superior mediastinum
- Pathophysiology:
 - Most commonly in children.
 - Results from:
 - **Penetrating trauma:**
 - **Most common in adults.**
 - Chicken bones or following instrumentation.
 - **URTI and sinusitis:**
 - **Most common in children.**
 - Suppuration of retropharyngeal lymph nodes.
- Clinical picture:
 - Fever
 - Odynophagia
 - Torticollis
 - Trismus (**NOT COMMON**)
 - Cervical lymphadenopathy
 - Unilateral (off-midline) posterior pharyngeal wall bulging.
- Lateral neck film:
 - Used for screening.
 - Not very sensitive or specific.
 - Should be done with head extension.
 - **General Rule for diagnosis:**
 - Prevertebral thickening **> 1/2 width of corresponding vertebral body.**
 - **Retropharyngeal infection in Adult:**
 - **C2:** prevertebral thickening **> 7 mm**
 - **C6:** prevertebral thickening **> 22 mm**
 - **Retropharyngeal infection in Pediatric:**
 - **C2:** prevertebral thickening **> 7 mm**
 - **C6:** prevertebral thickening **> 14 mm**

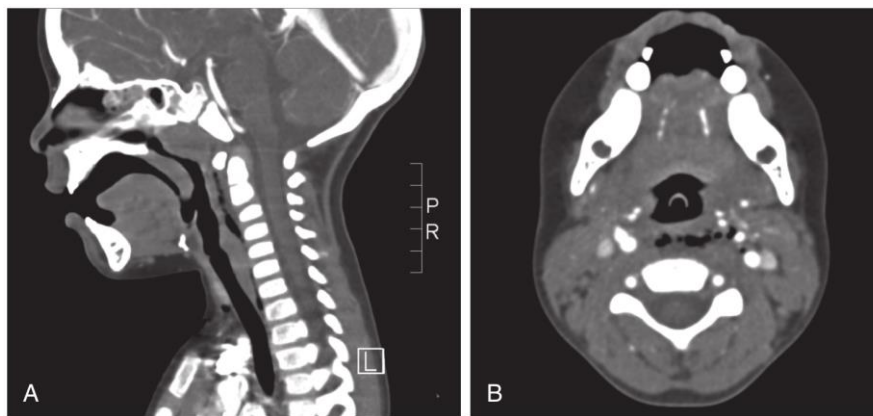
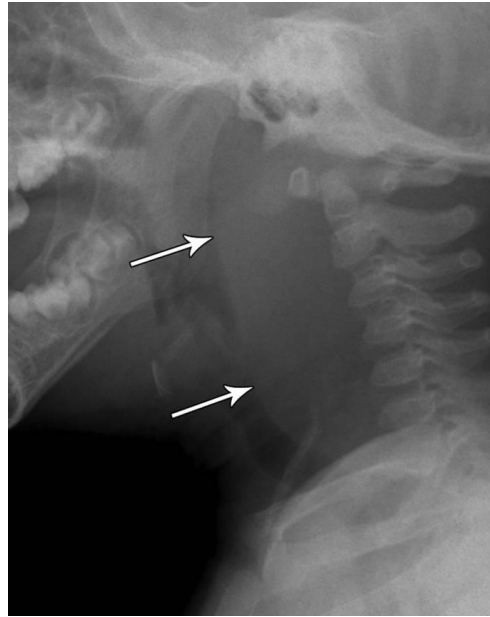
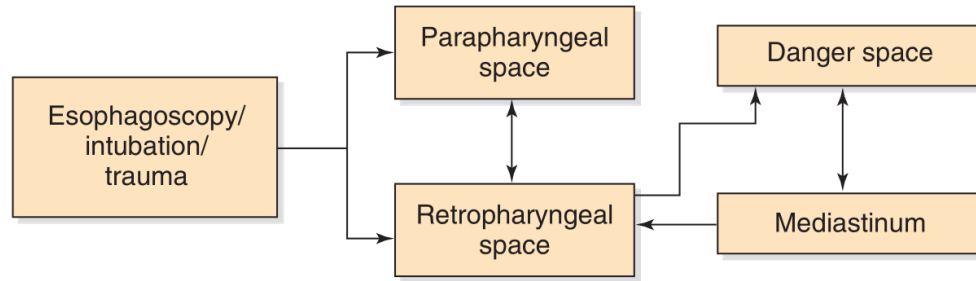


FIGURE 10-8. A penetrating trauma introduced air and secretions into the retropharyngeal space as seen on sagittal (A) and axial (B) computed tomography scan; the result was a deep neck infection.

- Treatment:

- **Antibiotics (Clindamycin)**
- **Drainage of Abscess:**
 - US guided aspiration for localized abscess.
 - Trans-oral incision and drainage:
 - Indicated for localized visualized abscess.
 - Vertical incision in the most fluctuant area of the abscess in the posterior pharyngeal wall.
 - Head should be down (Trendelenburg position) and suction should be available to prevent aspiration of pus.
 - Small ellipse of mucosa is excised to slow healing process and allow time for drainage before wound contraction.
 - Trans-cervical incision and drainage:
 - Indicated for diffuse abscess involving multiple spaces.
 - Hockey stick incision (Anterior border of SCM) used to expose entire retropharyngeal space.



Danger Space Infections:

- Danger space:
 - **Boundaries:**
 - From skull base to **diaphragm**.
 - Alar fascia anteriorly and prevertebral fascia posteriorly.
 - **Contents:**
 - Loose areolar tissue (infections spreads fast)
 - Cervical sympathetic trunk.
 - **Communicating Spaces:**
 - Retropharyngeal space
 - Superior Mediastinum
 - Abdominal cavity
- Results mainly from spread of infection from retropharyngeal space or penetrating injuries.
- Similar clinical picture, and treatment of retropharyngeal infections **except** of midline posterior pharyngeal wall swelling.

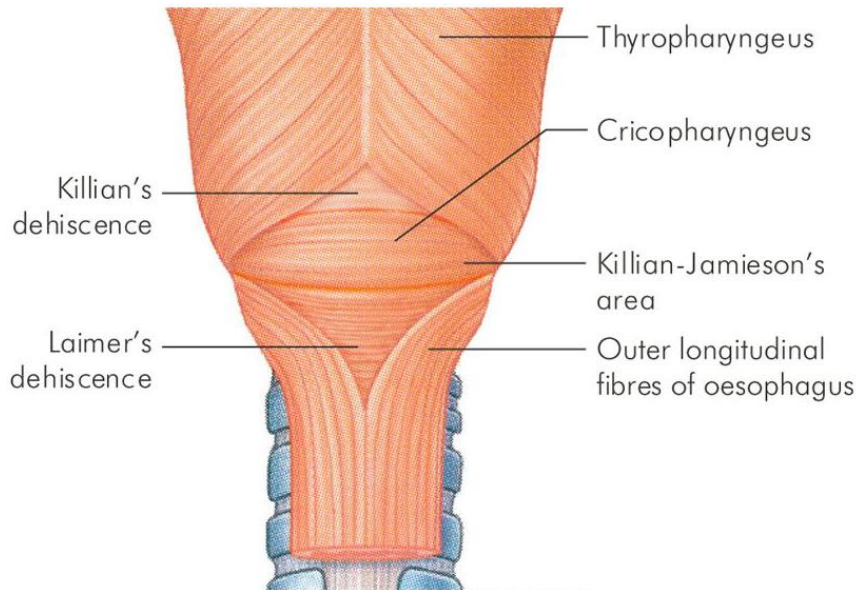
Prevertebral Infections:

- Prevertebral space:
 - **Boundaries:**
 - From skull base to **coccyx**.
 - Prevertebral fascia anteriorly and vertebral bodies posteriorly.
 - **Contents:**
 - Dense areolar tissue
 - Vertebral vessels
 - Phrenic nerve
 - Branchial plexus
 - Paraspinal, prevertebral and scalene muscles
 - **No Communicating Spaces (Closed space).**
- Results mainly from spread of infection from spine (**Pott's Abscess** tuberculous osteomyelitis of the spine) or penetrating injuries.
- Similar clinical picture, and treatment of retropharyngeal infections **except** of midline posterior pharyngeal wall swelling.

Neck Space	Boundaries	Contents	Communicating Spaces
Peritonsillar	• Medial-palatine tonsil • Lateral superior constrictor muscle	• Loose connective tissue • Tonsillar branches of the lingual, facial, and ascending pharyngeal vessels	• Parapharyngeal
Parapharyngeal	• Superior: base of middle fossa • Inferior: hyoid bone • Anterior: pterygomandibular raphe • Posterior: prevertebral fascia • Medial: superior constrictor • Lateral: deep lobe parotid, medial pterygoid	<i>Prestyloid</i> • Fat • Lymph nodes • Internal maxillary artery • Auriculotemporal, lingual, inferior alveolar nerves • Pterygoid muscles • Deep lobe parotid <i>Poststyloid</i> • Carotid • Internal jugular • Superior sympathetic nerve • Cranial nerves IX, X, X, and XII	• Peritonsillar • Submandibular • Visceral • Retropharyngeal • Carotid • Masticator • Parotid
Infratemporal fossa	• Superior: sphenoid and temporal skull, fossa medial to zygomatic arch • Anterior: infraorbital fissure, maxilla • Lateral: ramus and coronoid of mandible • Medial: lateral pterygoid plate with tensor and levator palatine muscles	• Pterygoid muscles • Temporalis tendon • Internal maxillary artery • Pterygoid venous plexus • Mandibular nerve (V3) with otic ganglion	• Temporal fossa • Pterygomaxillary fossa
Pterygomaxillary fossa	• Superior: sphenoid body, palatine bone • Anterior: posterior wall or maxillary antrum • Posterior: pterygoid process, greater wing of sphenoid • Medial: palatine bone • Lateral: temporalis muscle via pterygomaxillary fossa	• Maxillary nerve (V2) • Sphenopalatine ganglion • Internal maxillary artery	• Infratemporal fossa • Parapharyngeal space • Masticator space • Temporal fossa
Temporal fossa	• Superior: temporal line of skull • Inferior: zygomatic arch • Lateral: temporalis fascia • Medial: pterion skull	• Temporalis muscle • Temporal fat pad	• Infratemporal fossa • Pterygomaxillary fossa
Parotid	• Medial: parapharyngeal space • Lateral: parotid fascia	• Parotid gland • Facial nerve • External carotid artery • Posterior facial vein	• Parapharyngeal • Temporal fossa • Masticator
Masticator	• Medial: fascia medial to pterygoid muscles • Lateral: fascia overlying masseter	• Masseter muscle • Pterygoid muscles • Ramus and posterior body of mandible • Inferior alveolar nerve • Internal maxillary artery	• Parotid • Pterygomaxillary • Parapharyngeal
Submandibular	• Superior: floor of mouth • Inferior: digastric muscle • Anterior: mylohyoid and anterior belly digastric muscle • Posterior: posterior belly of digastric and stylomandibular ligament • Medial: hyoglossus and mylohyoid • Lateral: skin, platysma, mandible	• Sublingual and submandibular glands • Wharton duct • Lingual nerve • Lymph nodes • Facial artery and vein • Marginal branch of cranial nerve VII	• Parapharyngeal • Visceral space
Visceral	• Superior: hyoid bone • Inferior: mediastinum • Anterior: superficial layer of deep cervical fascia • Posterior: retropharyngeal space; prevertebral fascia • Lateral: parapharyngeal space	• Pharynx • Esophagus • Larynx • Trachea • Thyroid gland	• Submandibular • Parapharyngeal • Retropharyngeal
Carotid sheath	• Anterior: sternocleidomastoid muscle • Posterior: prevertebral space • Medial: visceral space • Lateral: sternocleidomastoid muscle	• Carotid artery • Internal jugular vein • Vagus nerve • Ansa cervicalis nerve	• Visceral space • Prevertebral • Parapharyngeal
Retropharyngeal	• Superior: base of skull • Inferior: superior mediastinum • Anterior: pharynx, esophagus • Posterior: alar fascia • Medial: midline raphe of superior constrictor • Lateral: carotid sheath	• Lymph nodes • Connective tissue	• Carotid sheath • Superior mediastinum • Parapharyngeal space • Danger space
Danger	• Superior: base of skull • Inferior: diaphragm • Anterior: alar fascia of deep layer of deep cervical fascia • Posterior: prevertebral fascia of the deep layer of the deep cervical fascia	• Loose areolar tissue	• Retropharyngeal • Prevertebral • Mediastinum
Prevertebral	• Superior: base of skull • Inferior: coccyx • Anterior: prevertebral fascia • Posterior: vertebral bodies • Lateral: transverse process of vertebrae	• Dense areolar tissue • Paraspinal, prevertebral, and scalene muscles • Vertebral artery and vein • Brachial plexus • Phrenic nerve	• Danger space

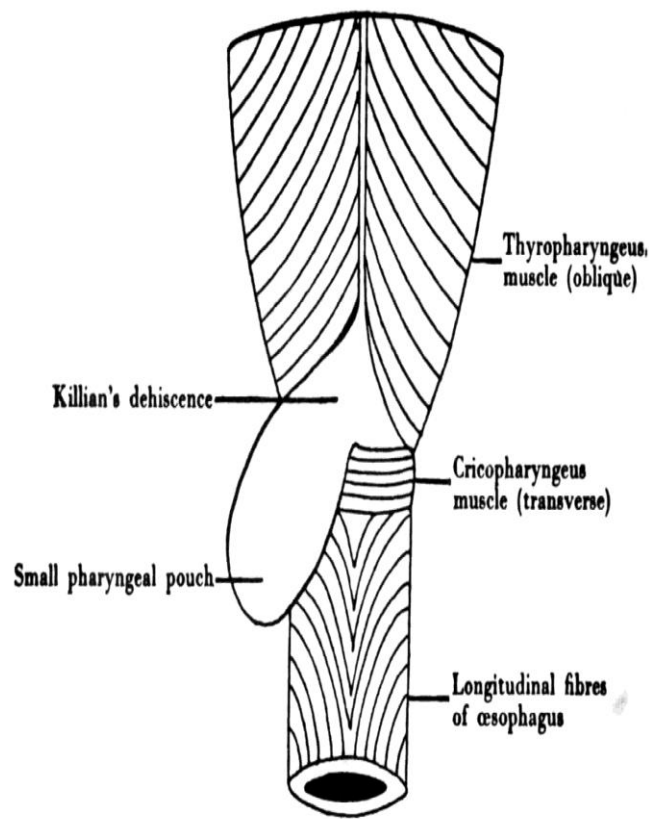
PHARYNGEAL POUCH

- Also called **hypopharyngeal diverticulum** or **Zenker's diverticulum**
- Herniation of mucosa through muscular wall
- It is a pulsion diverticulum where pharyngeal mucosa herniates through the Killian's dehiscence (a weak area between two parts of the inferior constrictor)
- True: all layers; false: only mucosa/submucosa



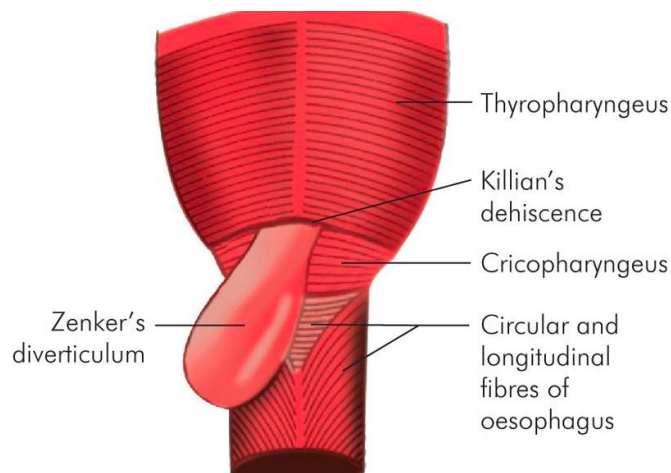
Aetiology:

- Exact cause is not known.
- It is probably due to spasm of cricopharyngeal sphincter or its incoordinated contractions during the act of deglutition (**created from elevated intraluminal pressure**).
- It is usually seen M>W; 6-9th decade
- Herniation of pouch starts in the midline.
- It is at first behind the oesophagus and then comes to lie on its left.
- Mouth of the sac is wider than the opening of oesophagus and food preferentially enters the sac



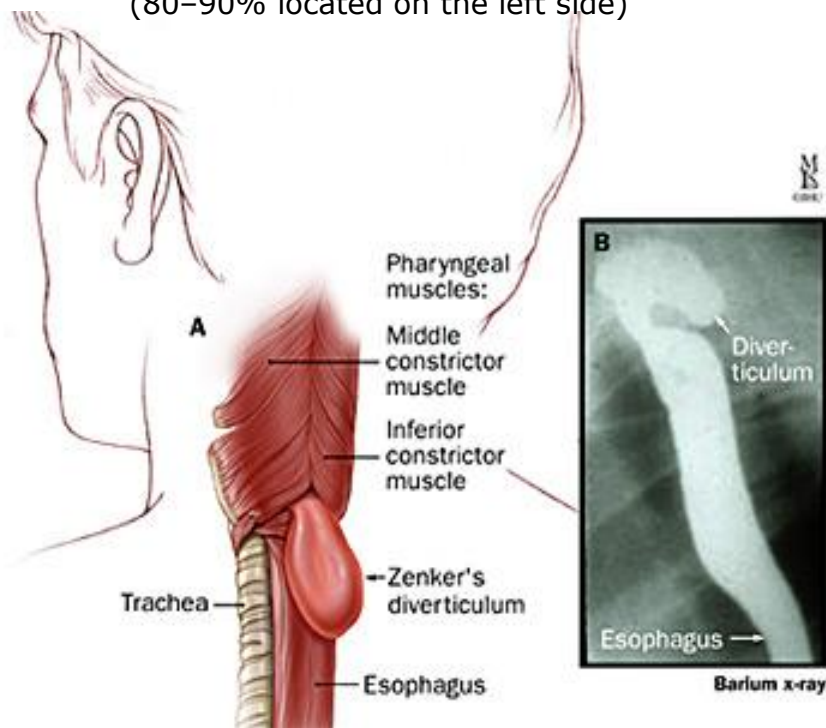
Clinical Features:

- Dysphagia is the prominent feature.
- It appears after a few swallows when the pouch gets filled with food, and presses on the oesophagus.
- Gurgling sound is produced on swallowing. (Bryce sign: gurgling sound on external pressure)
- Undigested food may regurgitate at night, when patient is recumbent, causing cough and aspiration pneumonia.
- Malodorous breath
- Patient is often malnourished due to dysphagia.
- Patients with pharyngeal pouch may have associated hiatus hernia.
- Rarely carcinoma can develop in long-standing cases of pharyngeal pouch
- Exam: usually normal; may feel crepitus in neck



Diagnosis

- Barium swallow (size, sac)
- Endoscopy
- (80–90% located on the left side)



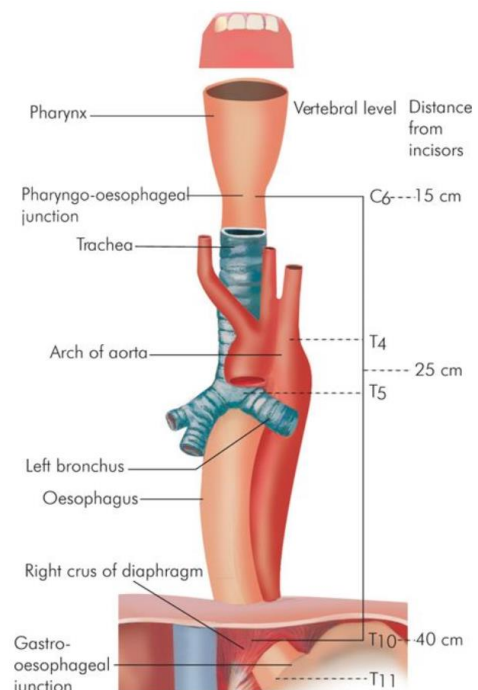
Treatment:

- Small – nothing
- Endoscopic division for symptomatic ones the new standard
- Some prefer open approach, and its occasionally needed if endoscopic exposure not possible (neck incision and excision)
- Small symptomatic ones may do well with cricopharyngeal myotomy alone
- Diverticulopexy also an option (tack up to prevertebral fascia)
open if >1.5cm
- Procedure: **endoscopic myotomy with diathermy DOHLMAN PROCEDURE** (The partition wall between the oesophagus and the pouch is divided by *diathermy* through an endoscope. This is done in poor risk debilitated patients)
- Surgical complications: RLN injury; leak; mediastinitis; recurrence; hematoma; wound infection; stenosis; death

Esophageal Anatomy:

- Muscular tube connecting the pharynx to the stomach.
- 25 cm in length in adults.
- Extends from lower end of pharynx (C6) to the cardiac end of stomach (T11).
- Composed of five layers:
 1. Non-keratinized Stratified squamous epithelium.
 2. Muscularis mucosae.
 3. Submucosa.
 4. Inner circular muscular layer.
 5. Outer longitudinal muscular layer.
- **No serosal layer.**
- Muscle is striated in upper third, mixed in middle third and smooth in lower third of esophagus.
- Esophagus has three segments:
 1. **Cervical:**
 - Situated anterior to the vertebrae and posterior to the trachea.
 - Supplied by branches of Inferior thyroid artery.
 2. **Thoracic:**
 - Situated adjacent to vital structures.
 - Its upper third has an intimate relationship with the trachea, and its lower third lies close to the aorta.
 - Supplied by aortic esophageal arteries or terminal branches of bronchial arteries.
 3. **Abdominal:**
 - Measured 3 cm in length and connects with the gastroesophageal junction.
 - Supplied by left gastric artery and a branch of the left phrenic artery.
- Normal Esophageal Constrictions:
 1. First one is at **15 cm** from upper incisors at cricopharyngeal sphincter.
 2. Second one at **25 cm** from upper incisors where it is crossed by aortic arch and left main bronchus.
 3. Third one at **40 cm** from upper incisors where it pierces the diaphragm.

Foreign bodies in the esophagus can be held up at these constrictions.



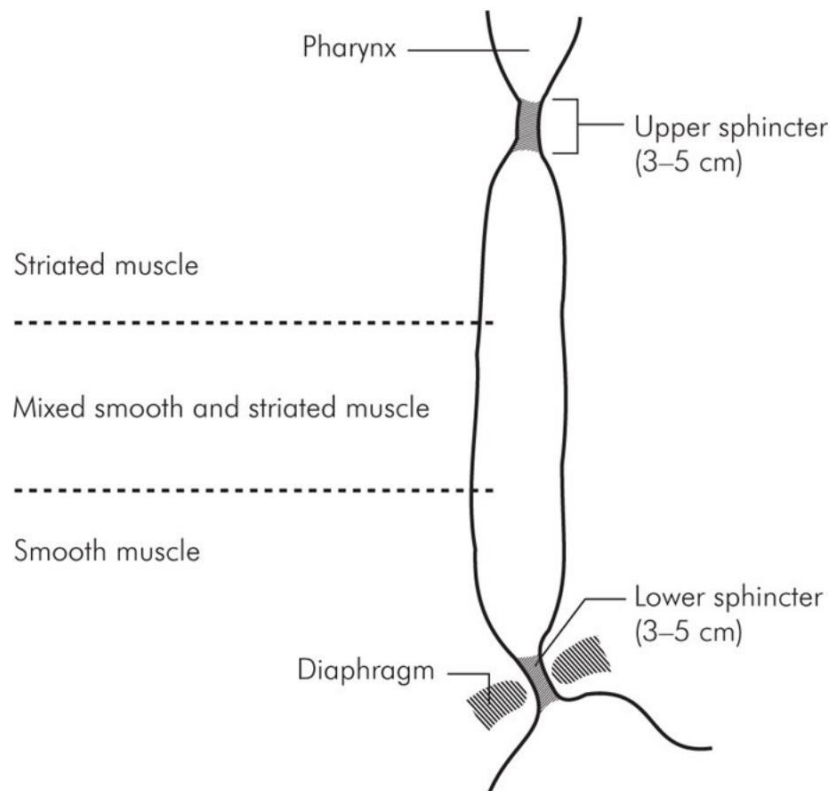
- Esophageal Sphincters:

- 1. Upper Esophageal Sphincter (UES):**

- High pressure zone.
- Starts at the upper border of esophagus.
- 3-5 cm in length.
- Inner circular layer of muscle thickens at level of cricoid and blends with cricopharyngeal muscle.
- Functions during the act of swallowing.

- 2. Lower Esophageal Sphincter (LES):**

- Situated at lower portion of esophagus.
- 3-5 cm in length.
- No single distinct muscle responsible for LES has been identified.
- Resting tone is 15-45 mm Hg.
- Functions to prevent gastroesophageal reflux.
- Relaxes in response to swallowing.



Physiology of Swallowing

- The act of swallowing is divided into three phases:

1. Oral
2. Pharyngeal
3. Esophageal

1. Oral Phase:

- The food which is placed in the mouth is chewed, lubricated with saliva, converted into a bolus and then propelled into the pharynx by elevation of the tongue against the palate.

2. Pharyngeal Phase:

- It is initiated when the bolus of food comes into contact with pharyngeal mucosa.
- A series of reflex actions take place carrying the food past oro- and hypopharynx into the esophagus.
- The communications into nasopharynx, oral cavity and larynx are cut off.
 - Closure of nasopharynx:
 - Soft palate contracts against the Passavant's ridge on the posterior pharyngeal wall and completely cuts off the nasopharynx from the oropharynx.
 - Closure of oropharyngeal isthmus:
 - The entry of food back into oral cavity is prevented by contraction of tongue against the palate and sphincteric action of palatoglossal muscles.
 - Closure of larynx:
 - Aspiration into the larynx is prevented by temporary cessation of respiration, closure of laryngeal inlet by contraction of aryepiglottic folds, closure of false and true cords, and rising of larynx under the base of tongue.
 - The role of epiglottis in providing protection to larynx is not clear but it is seen to deflect backwards when food passes into the pyriform fossae.
 - Contraction of pharyngeal muscles and relaxation of cricopharyngeus:
 - Relaxation of cricopharyngeus muscles is so timed and synchronous that food passes from pharynx into the oesophagus during contraction of pharyngeal muscles.

3. Esophageal Phase:

- After food enters the esophagus, the cricopharyngeal sphincter closes and the peristaltic movements of esophagus take the bolus down the stomach.
- LES relaxes well before peristaltic wave reaches and permits fluids to pass.
- Bolus of food is passed by contraction of peristaltic waves and then the sphincter closes.

Esophageal Disorders:

Gastro-Esophageal Reflux Disease (GERD):

- Common condition caused by relaxation of LES which allows stomach contents to reflux into the unprotected lining of the esophagus.
- 5-10% of the population have daily reflux.
- Other causes of GERD:
 - Pregnancy, hiatus hernia, scleroderma, excessive use of tobacco and alcohol, and drugs that relax the smooth muscle (anticholinergic, beta-adrenergic drugs and calcium-channel blockers).
- Clinical picture:
 - Commonest symptom is heart burn, exacerbated by stooping or consuming a heavy meal.
 - Less common symptoms include atypical chest pain, dysphagia, nocturnal asthma and hoarseness.
 - Long standing reflux can result in esophageal ulceration and stricture formation.
- Complications:
 - Esophagitis, esophageal ulceration, stricture formation.
 - Barrett's esophagus (precancerous):
 - Normal squamous epithelium of esophagus is replaced by columnar epithelium as a result of continuous inflammation.
 - Aspiration pneumonia, Asthma, Bronchiectasis.
 - Laryngitis, hoarseness, laryngeal granulomas, Carcinoma larynx.
 - Pharyngitis, Sinusitis, Otitis media with effusion.
- Diagnosis:
 - PH Monitoring (Gold standard)
 - Barium swallow
 - Manometry
 - Esophagoscopy and biopsy (R/O Eosinophilic esophagitis or Malignancy).
- Treatment:
 - **Behavioral:**
 - Elevation of the head of bed at night.
 - Avoiding food at least 3 hours before bed time.
 - Avoiding smoking, alcohol, caffeine, chocolates, mints and carbonated drinks.
 - **Medical:**
 - Proton pump inhibitors: Pantoprazole
 - H2-Blocker: Ranitidine
 - Pro-kinetics: Metaclopramide
 - Cytoprotectants: Sucralfate
 - Buffer: CaCO₃
 - **Surgical:**
 - Nissen's fundoplication.

Eosinophilic Esophagitis:

- Chronic, immune/antigen-mediated, esophageal disease characterized clinically by symptoms related to esophageal dysfunction and histologically by eosinophil-predominant inflammation.
- Affects both children and adults.
- Men are more commonly affected than women.
- Most commonly found among young boys and men.
- Pathophysiology:
 - Esophagus is normally devoid of eosinophils.
 - However, it is immunologically active organ that is capable of recruiting eosinophils in response to a variety of stimuli.
 - Eosinophils play an integral role in remodeling of esophageal tissues, which is observed histologically as subepithelial fibrosis.
 - There is a strong association of eosinophilic esophagitis with allergic conditions such as food allergies, environmental allergies, asthma, and atopic dermatitis.
- Clinical picture:
 - **Children**
 - Vomiting (Most common)
 - Feeding difficulty
 - Abdominal pain
 - Dysphagia
 - Food impaction
 - **Adult:**
 - Dysphagia to solid food (Most common)
 - Food impaction
 - Chest pain
 - GERD symptoms
 - Upper abdominal pain
- Diagnostic Criteria:
 - **Clinical:**
 - Symptoms related to esophageal dysfunction (dysphagia, GERD symptoms, feeding intolerance, FTT).
 - Lack of responsiveness to high dose PPI (2mg/kg/day x 8 weeks).
 - **PH monitoring:**
 - Normal result (exclude GERD).
 - **Esophageal Endoscopy:**
 - Linear furrows
 - Esophageal rings (Trachealization)
 - White plaques (Eosinophilic microabscesses)
 - Small-caliber esophagus
 - Esophageal strictures
 - **Esophageal Biopsy:**
 - Multiple biopsies from distal esophagus and mid or proximal esophagus.
 - **≥15 eosinophils/HPF**

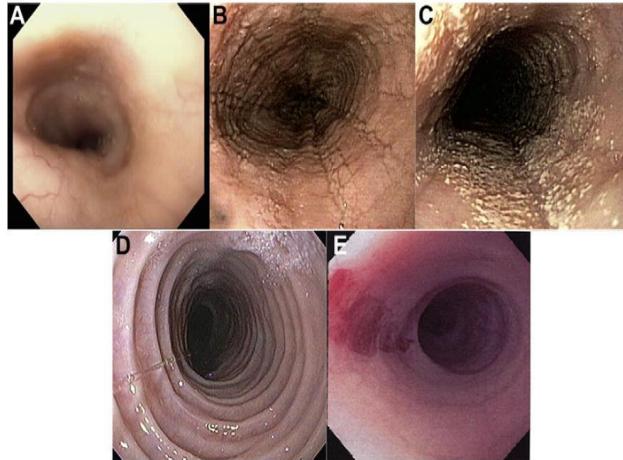


FIG E1. Endoscopic features of EoE. A, Normal esophagus. B, Esophageal furrowing. C, White mucosal plaques. D, Esophageal ring trachealization. E, Small-caliber esophagus with mucosal tearing after endoscopy.

- Treatment:
 - **Dietary Therapy (Elemental diet):**
 - **Effective first-line treatment.**
 - Elemental diet means placing the patient on an elemental formula, which eliminates all potential food allergens.
 - Risk of nutritional deprivation and should be administered under the supervision of a dietician.
 - **Medical Therapy:**
 - Proton Pump Inhibitors:
 - Undefined anti-inflammatory mechanisms.
 - Reduces acid production in patients with co-existent GERD.
 - Topical steroids:
 - Sprayed into the patient's mouth and then swallowed.
 - Patients should not inhale when the medication is being delivered and they should not eat or drink for 30 minutes following administration
 - Treatment for eight weeks.
 - Systemic steroids:
 - Indicated in patients with severe disease not controlled with topical steroids.
 - **Surgical Therapy:**
 - Esophageal dilation:
 - Reserved for patients who failed conservative therapy
 - Required as initial therapy in patients with high-grade strictures
 - Dilation should be performed carefully due to risk of deep mucosal tears and esophageal perforation.
 - Multiple dilations are often required to attain a goal esophageal diameter of 15 to 18 mm.

Esophageal Stricture

- Loss of lumen area within the esophagus.
- Normal esophageal diameter is 20 mm.
- Dysphagia generally occurs when diameter less than 15 mm.
- **Causes:**
 - **Intrinsic causes:**
 - Peptic Acid
 - Pill-induced
 - Chemical/Iye
 - Post-NG intubation
 - Infectious esophagitis
 - Sclerotherapy-induced
 - Irradiation-induced
 - Esophageal/gastric malignancies
 - Systemic inflammatory disease
 - **Extrinsic causes:**
 - Anomalous vessels and aneurysms
 - Pulmonary/mediastinal malignancies
 - Metastatic submucosal infiltration
- **Diagnosis:**
 - Contrast Esophagogram (Barium swallow).
 - Endoscopy.
- **Treatment:**
 - Esophageal dilation:
 - Rigid Savary-Gilliard dilators.
 - Balloon dilators.

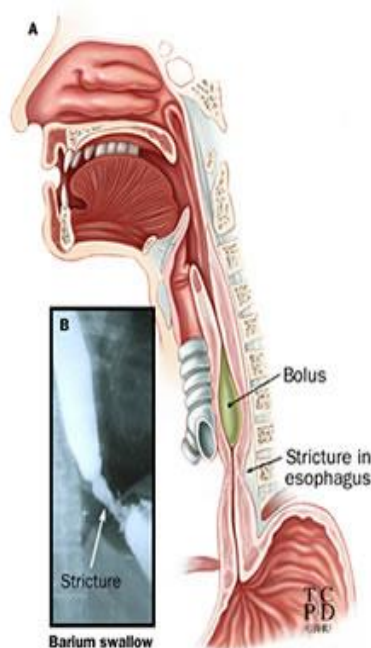
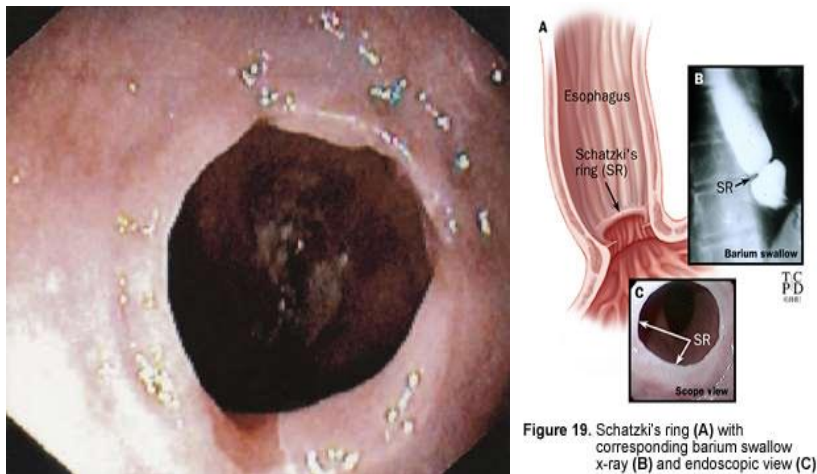


Figure 11. Esophageal stricture showing obstruction of food bolus with corresponding barium swallow.

Esophageal Rings/Webs:

- Rings:
 - Circumferential involve mucosa or muscle.
 - Most commonly occur in distal esophagus.
 - Schatzki's ring occurs at gastro-esophageal junction.
- Webs:
 - Involve only part of esophageal lumen.
 - Always involve mucosa only.
 - Most commonly occur in proximal esophagus.
 - Closely associated with iron deficiency.
- Diagnosis:
 - Contrast Esophagogram (Barium swallow).
 - Endoscopy.
- Treatment:
 - Esophageal dilation:
 - Rigid Savary-Gilliard dilators.
 - Balloon dilators.



Plummer-Vinson Syndrome:

- Triad:
 - Dysphagia
 - Iron-deficiency anemia (Glossitis, angular stomatitis, koilonychias, achlorhydria).
 - Upper esophageal web.
- There is atrophy of the mucous membrane of the alimentary tract.
- Predominantly, it affects females past 40 years.
- 10% risk of having post-cricoid carcinoma.
- It also predisposes to the development of carcinoma in the tongue, buccal mucosa, pharynx, esophagus and the stomach.

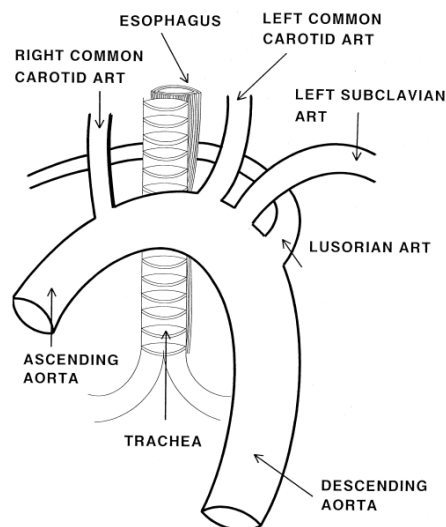
Esophageal Perforation:

- Potentially life threatening clinical situation.
- High morbidity and mortality rate of at least 20%.
- Delayed diagnostic work up may hinder timely & appropriate treatment with a negative effect on patient outcome.
- Contributing factors:
 - Difficulty of accessing the esophagus
 - Lack of strong serosal layer
 - Unusual blood supply of esophagus
 - Proximity of vital structures
- Causes:
 - Iatrogenic:
 - Most common cause.
 - During endoscopy or surgery.
 - Suspect perforation in all patients complaining of pain in the neck or interscapular region post-esophagoscopy.
 - Spontaneous rupture:
 - Boerhaave's syndrome.
 - Caused by straining or vomiting.
 - GERD with ulceration
 - Swallowing a foreign object or caustic chemicals.
 - Trauma
 - Tumors
- Clinical picture:
 - Chest pain:
 - Cardinal symptom
 - Acute and sudden in onset.
 - Radiating to the back or to the left shoulder.
 - Vomiting
 - Shortness of breath
 - Dysphagia
 - Subcutaneous emphysema
 - Bacterial Mediastinitis:
 - Tachycardia and Fever.
 - Develops rapidly within 24-48 hours of perforation.
 - May cause cardiopulmonary collapse and multi organ failure.
- Diagnosis:
 - **Chest X-ray:**
 - Pneumo-mediastinum
 - Subcutaneous emphysema
 - Mediastinal air-fluid levels
 - Pneumothorax
 - **Contrast Esophagogram:**
 - Should be done in any patient with suspected perforation.
 - Water-soluble contrast agent (Gastrografin) should be the initial study of choice (sensitivity of 60-75%).
 - Barium study should be done afterward if Gastrografin study showed no evidence of perforation (Sensitivity of 90%).

- Treatment:
 - **Early perforations of cervical esophagus can be managed by conservative measures:**
 - Careful observation.
 - Nil per orally
 - Appropriate analgesics
 - IV antibiotics
 - Proton pump inhibitor
 - Parenteral nutrition
 - **Surgical:**
 - Temporary endoscopic esophageal stents.
 - Endoscopic clips for perforation closure.
 - Endoscopic vacuum sponge therapy.
 - Myotomy along with defect covered with a fundoplication.
 - Repair over a T- tube is an alternative treatment.

Dysphagia Lusoria:

- Caused by aberrant right subclavian artery which arises from left side of the aortic arch.
- Compress the posterior esophagus (20% anterior compression).
- Diagnosis:
 - **Barium esophagogram:**
 - Indentation at level of 3rd and 4th thoracic vertebrae.
 - **CTA/MRA:**
 - Evaluate vascular system.
- Treatment:
 - Symptoms usually respond to changes in diet to small soft consistency.
 - Surgery relieves the obstruction by re-anastomosing the aberrant artery to the ascending aorta.



Achalasia:

- Most common cause of primary esophageal motility disorders.
- Characterized by an absence of peristalsis and failure of relaxation of lower esophageal sphincter.
- Etiology is unknown.
- Primary defect being described variously as neurogenic, myogenic and hormonal.
- Clinical picture:
 - Dysphagia to solids and liquids.
 - Vomiting, chest pain and weight loss.
- Diagnosis:
 - **Barium Esophagogram:**
 - Dilated esophagus
 - Obstruction at esophagogastric junction.
 - Rat tail (bird-beak) appearance.
 - **Esophageal Manometry:**
 - Confirm the diagnosis.
 - Diagnosis criteria:
 - High pressure(>30mmHg) LES zone
 - Failure of LES to relax
 - Absence of propulsive peristalsis
 - In-coordinated esophageal contractions.
 - **Endoscopy:**
 - Exclude benign stricture or any development of carcinoma which is a common complication of this disorder.
- Treatment:
 - **Medical:**
 - Nitrates and calcium channel blockers.
 - **Surgical:**
 - Botox injection.
 - Pneumatic dilation.
 - Heller's myotomy.



Diffuse Esophageal Spasm:

- Presence of simultaneous and repetitive contractions in esophageal body.
- Unlike achalasia, some normal peristalsis remains.
- Characterized by severe chest pain and dysphagia.
- Primarily involve the lower two-thirds of the esophagus.
- Diagnosis:
 - **Barium Esophagogram:**
 - Corkscrew esophagus.
 - **Esophageal Manometry:**
 - Confirm the diagnosis.
 - Presence of sequential peristaltic contraction waves associated with occasional multipeaked high- amplitude, long duration contractions.
- Treatment:
 - **Medical:**
 - Nitrates and calcium channel blockers.
 - **Surgical:**
 - Pneumatic dilation of lower one third of esophagus can be of benefit.



Scleroderma:

- Systemic collagen disorder primarily neural, but secondarily weakening the smooth muscles of the lower two-thirds of esophagus and LES.
- Most common cause of secondary esophageal motility disorders
- Dysphagia may precede cutaneous lesions.
- Barium swallow shows absence of peristalsis in distal two-thirds of the esophagus due to mural fibrosis.
- Many of these patients have hiatus hernia, or reflux esophagitis and may develop stricture in distal part of the esophagus due to recurrent inflammation.



Cricopharyngeal Spasm:

- Caused by failure of the upper esophageal sphincter to relax properly.
- There is in-coordination between relaxation of the upper esophageal sphincter and simultaneous contraction of the pharynx.
- Common causes are cerebrovascular accidents, Parkinson's disease, bulbar polio, multiple sclerosis and muscular dystrophies.
- Treatment:
 - Botox injection.
 - Myotomy.

Leiomyoma:

- Most common benign esophageal tumor.
- Accounts for two-thirds of all the benign neoplasms.
- Arises from the smooth muscle and grows in the wall of esophagus.
- Dysphagia is produced when tumor exceeds the diameter of 5 cm.
- Barium swallow shows an ovoid filling defect.
- Endoscopy reveals a submucosal swelling.
- Treatment is enucleation of the tumor.

Mucosal polyps, lipomas, fibromas and haemangiomas:

- Benign esophageal tumors
- They are often pedunculated and present in the esophageal lumen.
- Endoscopic removal is avoided because of the danger of esophageal perforation.
- Treatment is surgical excision by esophagotomy

Angioedema

- Swelling of the deep dermis, subcutaneous or submucosal tissue, or mucous membranes as a result of vascular leakage.
- Usually nonpitting and nonpruritic.
- Associated with pain.
- Involved skin often shows no change in color.
- Most commonly observed affecting the lips and eyes (periorbital).
- Other commonly involved areas may include the face, hands, feet, and genitalia.
- **Urticaria:**
 - Swelling of superficial layer of dermis (Papillary dermis) only.
 - Hypersensitivity Reaction.
 - Not involving subcutaneous or mucosa.
 - Associated mainly with pruritus.
 - No pain.

Types of Angioedema:

1. Allergic Angioedema:

- Type 1 hypersensitivity reaction.
- IgE/histamine-mediated angioedema.
- Often associated with urticaria.
- Typically occurs after 30 min to 2 hours after exposure to allergen
 - Food , Drugs , Insect bite.

2. Pseudo-allergic Angioedema:

- Non-IgE Mediated.
- Caused by a non-allergic or non-immunologic reaction.
- Presentation is very similar to those of allergic angioedema.
- Typical examples are angioedema induced by NSAIDs (Aspirin).

3. Hereditary Angioedema:

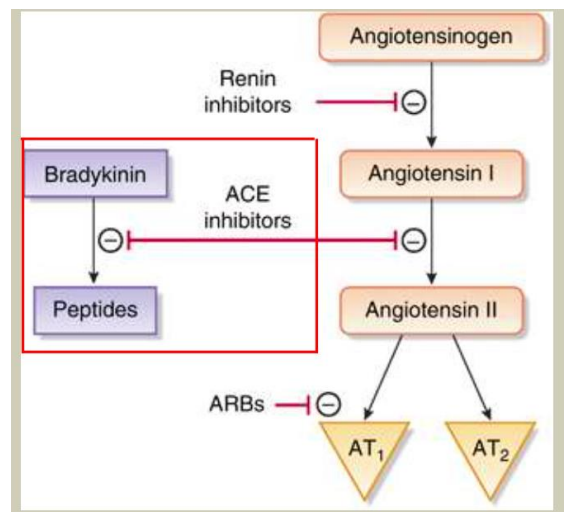
- Autosomal dominant disorder.
- Bradykinin-mediated Angioedema.
- Non-Allergic Angioedema (Non-IgE/histamine Mediated).
- Not associated with urticaria.
- Most patients report a family history of disease.
- Caused by abnormality in **C1-INH** and **C4**:
 - Lead to increase in Bradykinin level.
- Types of Hereditary Angioedema:
 - **TYPE 1:** C1-INH Deficiency (Most common).
 - **TYPE 2:** C1-INH Dysfunction
 - **TYPE 3:** Normal C1-INH but Dysfunction C1 Esterase.

4. Acquired Angioedema:

- Bradykinin-mediated Angioedema.
- Non-Allergic Angioedema (Non-IgE/histamine Mediated).
- Not associated with urticaria.
- Caused by accelerated consumption of C1-INH or the production of autoantibodies to C1-INH.
 - Lead to increase in Bradykinin level.
- Onset is usually in the fourth decade of life.
- Associated with autoimmune diseases and lymphoproliferative disorders.

5. ACE inhibitor–induced Angioedema:

- Bradykinin-mediated Angioedema.
- Non-Allergic Angioedema (Non-IgE/histamine Mediated).
- Not associated with urticaria.
- Caused by ACE inhibitors (Captopril):
 - Anti-Hypertension medications.
 - Inhibit conversion of Angiotensin-I to Angiotensin-II.
 - Inhibit break down of Bradykinin.
 - Lead to increase in Bradykinin level.



6. Physically–induced Angioedema:

- Non-Allergic Angioedema (Non-IgE/histamine Mediated).
- Caused by physical agents, such as cold, heat pressure, vibration, and ultraviolet radiation.

7. Idiopathic Angioedema:

- Exact mechanism is unclear.
- Most common cause of AE (40%).

- Important clinical information should include the following:
 - Onset of angioedema:
 - Acute (< 6 weeks).
 - Chronic (>6 weeks).
 - Associated urticaria:
 - Allergic angioedema.
 - History of recent use of new food or medicine:
 - Allergic angioedema.
 - History of recent use of NSAID:
 - Pseudo-allergic angioedema.
 - History of recent use of ACE-inhibitors:
 - ACE inhibitors-induced angioedema.
 - Family history of angioedema:
 - Hereditary angioedema.
 - History of Recurrent angioedema:
 - Hereditary angioedema.
 - ACE inhibitors-induced angioedema.
- Clinical picture:
 - **Oropharyngeal Angioedema:**
 - Involving lips, tongue or uvula.
 - **Laryngeal Angioedema:**
 - Stridor and airway obstruction.
 - **Peripheral Angioedema:**
 - Involving feet and hands.
 - **Abdominal Angioedema:**
 - Involving intestinal mucosa.
 - Lead to abdominal pain.
 - **Urogenital Angioedema:**
 - Involving scrotum.



Approach for Angioedema:

- Most cases can be managed well with outpatient treatment alone
- Inpatient care for angioedema is usually not necessary when timely treatment is administered.
- Indications of ICU admission for observation +/- early intubation:
 1. Laryngeal involvement (Stridor).
 2. Tongue edema.
 3. Multiple site involvement.
- **Angioedema with Urticaria:**
- Occurs due to Allergic IgE-Mediated angioedema.
- Acute treatment:
 - **Anti-Histamine (1st line):**
 - H1 Blocker: Diphenhydramine 50mg IV.
 - H2 Blocker: Ranitidine IV.
 - **Steroid:**
 - Indicated for moderate-to-severe cases.
 - Hydrocortisone 200mg IV.
 - Methylprednisolone 40-60 mg IV.
 - Prednisolone 40-60 mg OD x 5 days.
 - **Epinephrine (1:1000):**
 - 0.5 mg IM Q5 minutes PRN.
 - Indicated in patients with impeding airway obstruction (involvement of larynx or tongue).
- Chronic treatment:
 - **Anti-Histamine (1st line):**
 - Daily dose of non-sedating anti-histamine.
 - If uncontrolled symptoms:
 - Consider doubling the dose of antihistamine and/or adding a second antihistamine.
 - Consider further increase in dose of antihistamine up to 4 times the recommended dose.
 - Doxepin (Tricyclic antidepressant) has potent H1-blocking activity and has been used off label to treat recalcitrant angioedema.
 - If infrequent symptoms:
 - Consider PRN dose of antihistamine.
 - **Steroid:**
 - Indicated for more severe symptoms not controlled with antihistamine alone.
 - Prednisolone 40-60 mg OD x 5 days.
 - **Instructions:**
 - ACE inhibitor should be stopped and avoided in future, even if the patient is not taking it.
 - Angiotensin 2 Blocker (Losartan), can be used safely.
 - Epinephrine autoinjector should be considered if there is a history of significant angioedema affecting the upper airway.

- **Angioedema without Urticaria:**
- Occurs due to Non-Allergic IgE-Mediated angioedema.
 - Hereditary angioedema.
 - ACE inhibitors-induced angioedema.
- **Labs should be requested to exclude C1-INH abnormalities:**
 - C4 Level
 - C1-INH level
 - C1-INH function
- Antihistamines, corticosteroids and epinephrine have limited or no value in the treatment of these patients.
- **Acute treatment:**
 - **C1-INH concentrate:**
 - Infusion of C1-INH 20 U/kg.
 - Shorten the duration of acute attack.
 - Relief of symptoms within 1 hour of administration.
 - **Fresh Frozen Plasma (FFP):**
 - 2 units of FFP, which contains C1-INH, has been shown to be helpful in certain patients.
 - However, FFP also contains substrates that may worsen angioedema attacks.
 - If this treatment is used, be ready to intubate or perform a tracheostomy, if necessary.
 - **Ecallantide:**
 - Potent, selective, reversible inhibitor of plasma kallikrein.
 - Suppresses bradykinin generation.
 - FDA approved for treatment of acute HAE attacks.
 - **Icatibant:**
 - Bradykinin B2 receptor antagonist.
 - FDA approved for treatment of acute HAE attacks.
- **Chronic treatment:**
 - **Prophylaxis:**
 - Long-term prophylaxis is indicated if on-demand acute treatment is inadequate to minimize disease.
 - C1-INH concentrate
 - Danazol
 - **Instructions:**
 - ACE inhibitor should be stopped and avoided in future, even if the patient is not taking it.
 - Angiotensin 2 Blocker (Losartan), can be used safely.
 - Consider administering short-term preprocedural prophylaxis, particularly in cases involving dental or intraoral surgery and bronchoscopy or endoscopy; endotracheal intubation; and manipulation of the upper airway or pharynx.
 - Family members of HAE patients should be screened so that appropriate therapy can be available for treatment.

Neck Trauma

Penetrating Neck Trauma

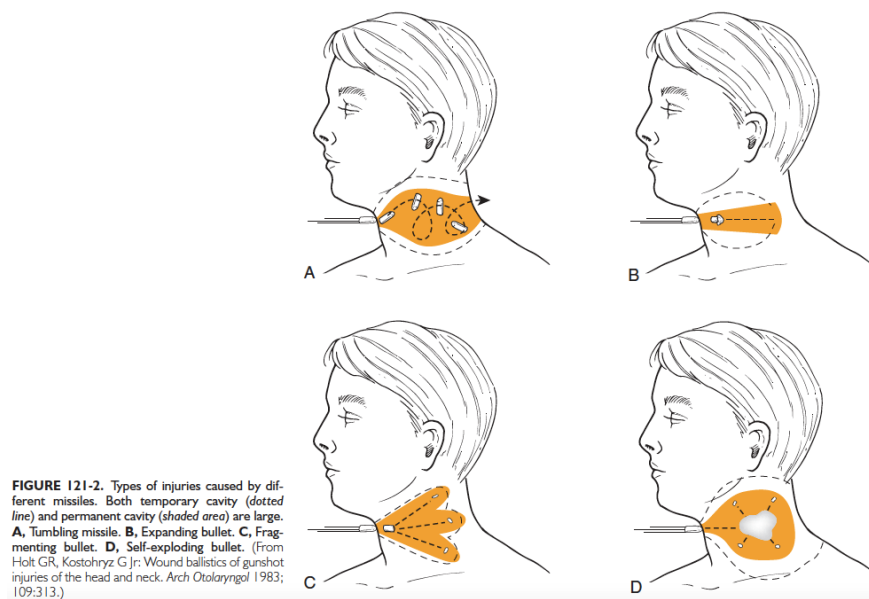
- Comprises 5% to 10% of all trauma cases.
- All penetrating neck wounds are potentially dangerous and require emergency treatment
 - Most civilian centers currently practice selective neck exploration, with mortality rates ranging 3-6% for low-velocity penetrating neck trauma (LVPNT)
 - U.S. military surgeons have treated high-velocity penetrating neck trauma (HVPNT) patients with selective neck exploration and have reported mortality rates equivalent to civilian mortality rates for LVPNT
 - **Knife** is the most common cause.

Mechanism of injury:

Table 7.1. Historical Categories of Missiles and Types of Penetrating Neck Wounds

Categories of Missiles Resulting in Penetrating Neck Wounds	Types of Penetrating Neck Wounds	
Knives	Low Velocity (<610m/s)	High Velocity (>610m/s)
Single Projectiles	▪ Stab wounds ▪ Handgun wounds ▪ Long-range (>5 m victim-to-weapon range) birdshot wounds	▪ Close-range (<5 m victim-to-weapon range) birdshot wounds
Multiple Projectiles	▪ Shotgun pellets ▪ Improvised explosive devices (IEDs) ▪ Grenades ▪ Mortars ▪ Rocket	▪ Close-range buckshot wounds ▪ Rifle wounds ▪ Wounds from bombs, IEDs, grenades, mortars, and rockets

< = less than; > = more than; m/s = meters per second.



Anatomy

1. Vital Structures in the Neck

- Airway (pharynx, larynx, trachea, and lungs).
- Blood vessels (carotid arteries, innominate artery, aortic arch vessels, jugular veins, and subclavian veins).
- Nerves (spinal cord, brachial plexus, cranial nerves, and peripheral nerves).
- Gastrointestinal tract (pharynx and esophagus).

2. Skeletal Anatomy:

- Mandible.
- Hyoid.
- Styloid process.
- Cervical spine.

3. Muscular Landmarks:

- **Platysma** muscle—**Penetration of the platysma muscle defines a deep injury** in contrast to a superficial injury.
- **Sternocleidomastoid** muscle—The sternocleidomastoid muscle also serves as a valuable landmark, since this large, obliquely oriented muscle divides each side of the neck into anterior and posterior triangles.
- **Anterior** triangle—The anterior triangle contains airway, major vasculature, nerves, and gastrointestinal structures, while the **posterior** triangle contains the spine and muscle.

4. Neck Zones:

- Neck is commonly divided into three distinct zones:

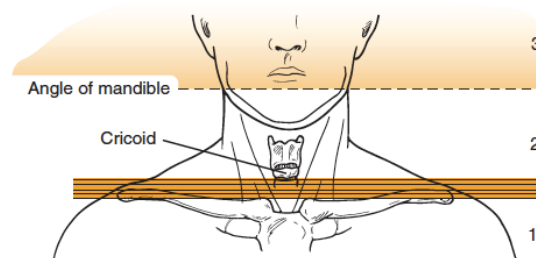
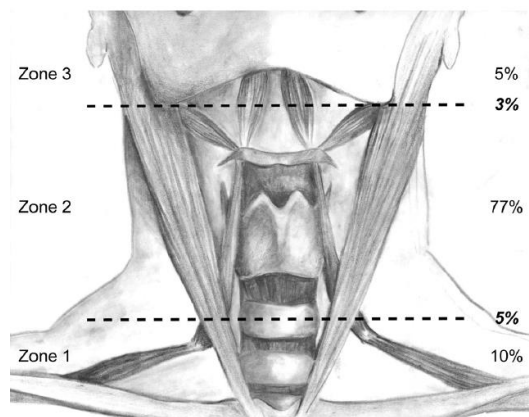


FIGURE 121-3. The three zones of the neck are seen on this frontal view. The shaded area represents the portion that some authors consider zone I but that others label zone II. (From Carducci B, Lowe RA, Dalsey W: Penetrating neck trauma: consensus and controversies. *Ann Emerg Med* 1986;15:208.)



Zone I:

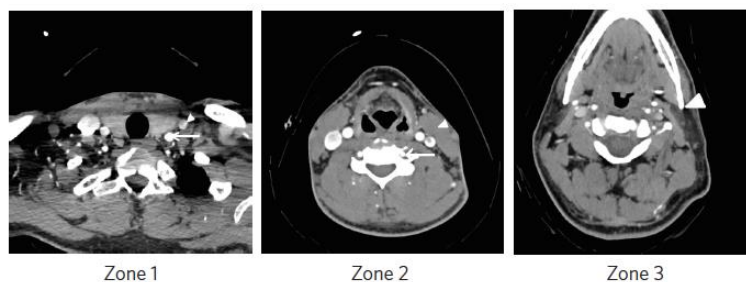
- Most **caudal** anatomic zone.
- Inferiorly by the clavicle/sternal notch
- Superiorly by the horizontal plane passing through the inferior cricoid cartilage.
- Mortality is high.
- Due to the sternum, surgical access to Zone I may require **sternotomy** or thoracotomy.
- Structures within this zone include the:
 - **Vessels:** arch of aorta, Proximal common carotid arteries, Vertebral arteries and subclavian vessels, innominate vessels
 - **Aerodigestive:** Trachea, esophagus, lung apices
 - Nerves: Recurrent laryngeal and vagus nerves, brachial plexus, spinal cord
 - Thoracic duct.

Zone II:

- **Middle** anatomic zone.
- Between the horizontal plane passing through the inferior **cricoid** cartilage and the horizontal plane passing through the angle of the **mandible**.
- Vertically or horizontally oriented **neck** exploration incisions provide straightforward surgical access to this zone,
- Contains:
 - **Vessels:** common Carotid arteries, internal and external carotid, jugular veins.
 - **Aerodigestive:** hypopharynx, and larynx, proximal esophagus, Proximal trachea.
 - **Nerves:** Recurrent laryngeal and vagal nerves and other cranial nerves, Spinal cord, sympathetic chain

Zone III:

- Most cephalic anatomic zone.
- Between the horizontal plane passing through the angle of the mandible and the **skull** base.
- **High associated mortality at the skull base.**
- Surgical access to it may require craniotomy, as well as mandibulotomy or maneuvers to anteriorly displace the mandible.
- Anatomic structures within Zone 3 include the:
 - **Vessels:** Internal carotid, extracranial carotid (terminal branches) and vertebral arteries, prevertebral vein plexus, Jugular veins.
 - Aerodigestive: oral cavity, pharynx
 - Nerves: Cranial nerves IX–XII, (trunk of VII), Sympathetic trunk, Spinal cord.



Classification by injured structure:

- Larynx and trachea 10%
- Pharynx and esophagus 10%
- Vascular structures:
 - Internal jugular vein 9%
 - Internal & Common carotid arteries 7%
 - Subclavian artery 2%
 - External carotid artery 2%
 - Vertebral artery 1%

Vascular Injuries:

- The incidence of vascular injuries is higher in **Zone 1 and Zone 3** penetrating neck trauma injuries.
- However, in **Zone 2**, the vessels are not fixed; therefore, they are more easily displaced by concussive forces, and the rate of vascular injury is lower.

Esophageal injury:

- For **Zone 1 and for some Zone 2** penetrating neck injuries, it is imperative that esophageal injuries be ruled out with **endoscopic examination** and, possibly, **swallow studies**.
- Missed esophageal injuries occur because up to **25%** of penetrating esophageal injuries are occult and asymptomatic.

Box 121-1. SIGNS AND SYMPTOMS OF PENETRATING NECK TRAUMA

Airway

Respiratory distress
Stridor
Hemoptysis
Hoarseness
Tracheal deviation
Subcutaneous emphysema
Sucking wound

Vascular System

Hematoma
Persistent bleeding
Neurologic deficit
Absent pulse
Hypovolemic shock
Bruit
Thrill
Change of sensorium

Nervous System

Hemiplegia
Quadriplegia
Coma
Cranial nerve deficit
Change of sensorium
Hoarseness

Esophagus/Hypopharynx

Subcutaneous emphysema
Dysphagia
Odynophagia
Hematemesis
Hemoptysis
Tachycardia
Fever

Diagnostic Evaluation:

1. Computed Tomographic Angiography (CTA):

- Initial procedure of choice to evaluate cervical vasculature and aerodigestive tract in patient with penetrating neck trauma.
- Signs of Probable Injury on CTA
 - Hematoma.
 - Subcutaneous air adjacent to the carotid sheath.
 - Intravenous contrast extravasation, filling defect.
 - Missile tracts in close proximity to vital structures.
 - Irregular vessel margin or caliber.
- CTA may have a 2% incidence of non-diagnostic studies due to the artifact from bullet fragments and metallic foreign bodies.

2. Contrast Swallow Studies:

- Used for evaluation of Hypopharyngeal and esophageal injuries.
- Less accurate than endoscopy.

	Gastrograffin	Barium
Consistency	Thin	Thick
Risk of aspiration	Common	Rare
Risk of mediastinitis	No	Yes
Leak detection	Large leaks	Can detect small injury

3. Pan-endoscopy (Laryngoscopy/ Bronchoscopy/Esophagoscopy):

- Performed in OR under general anesthesia.
- It is recommended that both rigid and flexible esophagoscopy be performed to rule out occult esophageal injuries.
- Rigid esophagoscopy may provide a better view of the proximal esophagus near the cricopharyngeal muscle.
- Flexible esophagoscopy, with its magnification on the viewing screen and ability to insufflate, gives excellent visualization of more distal esophageal anatomy.

TABLE 121-5. Accuracy of Selective Evaluation Techniques

Procedure	Indications	Contraindications	Accuracy (%)
Angiography	Wounds near a vessel in zone I or III	Expanding hematoma Profound shock Uncontrolled bleeding	98.5
Barium swallow	Hematemesis Drooling Dysphagia Vocal cord paralysis	Intubation Saliva in wound Unstable patient	90.0
Esophagoscopy	Suspected injury unconfirmed by barium swallow Intubation Laryngeal or tracheal injury Vascular injury in zone II or III	None	86
Direct laryngoscopy and bronchoscopy	Vocal cord paralysis Hoarseness Tenderness or crepitation over the larynx Subcutaneous emphysema Hemoptysis	None	100

From Miller RH, Duplchain JK: Penetrating wounds of the neck. *Otolaryngol Clin North Am* 1991;24:15.

Approach for Management of Neck Trauma:

- **Based on of Patient's Symptoms at Presentation:**
 - **Unstable symptomatic patients:**
 - Should be explored immediately in the OR.
 - Hard Signs:
 1. Refractory shock
 2. Uncontrolled bleeding
 3. Neurologic deficit
 4. Airway obstruction
 5. Expanding hematoma
 - **Stable symptomatic patients:**
 - Zone I and III:
 - CT angiography (CTA) is done to define the surgical approaches.
 - Hemorrhage is the leading cause of death for penetrating neck injuries in Zone I and III.
 - Zone II:
 - Choice of treatment remains controversial.
 - Some advice for immediate surgical exploration as the surgical approach is easy (in contrast to Zone I & III).
 - Others advise for CT angiography (CTA) followed by surgical exploration if positive results.
 - **Asymptomatic patients:**
 - Evaluated with diagnostic studies (CTA), and if pathologic findings are discovered during this workup, are taken to the operating room for neck exploration.
 - If asymptomatic patients have a negative diagnostic workup showing no neck pathology, then they will be observed.
- **Vascular** injury may result in active hemorrhage, expanding hematoma, vascular bruit, and pulse deficit.
- **Airway** injury may cause subcutaneous emphysema, hoarseness, stridor, and respiratory distress.
- **Esophageal** injury is often asymptomatic and may result in leakage of saliva, subcutaneous emphysema, bleeding from the esophageal inlet, and ultimately neck or mediastinal abscess.
- **Nerve** injury may result in cranial nerve deficits or hemiparesis. These fixed neurologic deficits may not require immediate neck exploration in an otherwise stable patient.

Hemodynamic and neurologic status should always be monitored closely for at least 48 to 72 hours.

Emergency Room Management

- The basic principles of trauma management apply to all patients with penetrating neck trauma (**ABCDE**).
- Accordingly, airway management is the first priority
- The overall prevalence of cervical spine fracture in patients with isolated facial trauma is (5-8%).
- Spinal stabilization should be maintained until cleared clinically and/ or radiographically.
- Most patients can be carefully intubated trans-orally or trans-nasally.
- Surgical airway is indicated in:
 - Bleeding obscuring the airway.
 - Edema (oral cavity/pharynx).
 - Laryngeal trauma.
- Deeply probing open neck wounds below the platysma muscle should be avoided in the emergency room.
- Two large-bore intravenous lines should be placed.
- Tetanus toxoid should be administered if the status is unknown or outdated.
- To avoid air embolism, the patient should be supine or in Trendelenburg's position.
- If possible, initial radiographic survey in the trauma bay should include **chest x-ray and cervical spine x-rays**.
- Prophylactic **antibiotics** and **NGT** suction placement should also be considered.

TABLE 121-4. Mandatory vs. Selective Management of Neck Wounds

Consideration	Mandatory	Selective
Diagnosis	Potential life-threatening injuries can be missed preoperatively; routine exploration can miss some injuries.	Most major injuries can be diagnosed by the preoperative workup.
Skill and resources	Selective management requires more skill, manpower, experience, and judgment; additional special diagnostic procedures are also required.	Selective care will reduce unnecessary explorations by a surgeon inexperienced with trauma.
Hospital stay	Length of stay is similar for observation and negative exploration.	No advantage of negative exploration has been found.
Delay	If occult injuries are delayed, morbidity and mortality will increase.	Delay has not been shown to significantly increase morbidity and mortality of occult injuries.
Patient care	Active observation requires concept of continuous collaboration among trauma team members and reduces unnecessary surgical exploration.	Selective management emphasizes the availability of experienced trauma medical staff for monitoring the patient.

Modified from Obeid F, Haddad G, Horst H, et al: A critical reappraisal of a mandatory exploration policy for penetrating wounds of the neck. *Surg Gynecol Obstet* 1985;160:517.

Management of Vascular Injuries:

- Start with compression (external compression or with 18-20 Foley Cath).
- Options of managements:
 - 1. Anticoagulation with heparin:**
 - Indicated in blunt injury.
 - 2. Endovascular intervention:**
 - Covered stent graft: Pseudoaneurysm, lacerations.
 - Embolization or coiling: Pseudoaneurysm.
 - Endovascular occlusion: Injured vertebral arteries.
 - Balloon occlusion test prior to ICA ligation.
 - 3. Surgical control:**
 - Ligation:
 - Ligation of ICA or common carotid is indicated in patients on coma or with high carotid injury.
 - Balloon occlusion test is useful if patient is stable.
 - Proximal ligation or angiographic embolization of vertebral artery can be used if the contralateral vertebral artery is intact.
 - Repair (revascularization):
 - Preferred in common carotid injuries.
 - Saphenous vein graft may be used.

Management of Hypopharyngeal and Esophageal Injuries:

- Best detected by combination of esophagoscopy and esophagram in symptomatic patients.
- Injection of air or methylene blue in the mouth may aid in localizing injuries.
- Alarm signs requiring Neck exploration:
 - Subcutaneous emphysema
 - Hematemesis
 - Hypopharyngeal blood
- Hypopharyngeal injuries (Above arytenoids):
 - Managed by conservative measures for 5-7 days (NPO, NGT, IV Antibiotics).
- Esophageal injuries:
 - Managed surgically within 24 hours by watertight closure in 2-layer fashion (muscle and mucosa) and placement of drain.
 - Divergent and drainage (late exploration).
 - Controlled fistula with T-tube or exteriorization of low non-repairable wounds
 - All patients should be NPO for 5-7 days.

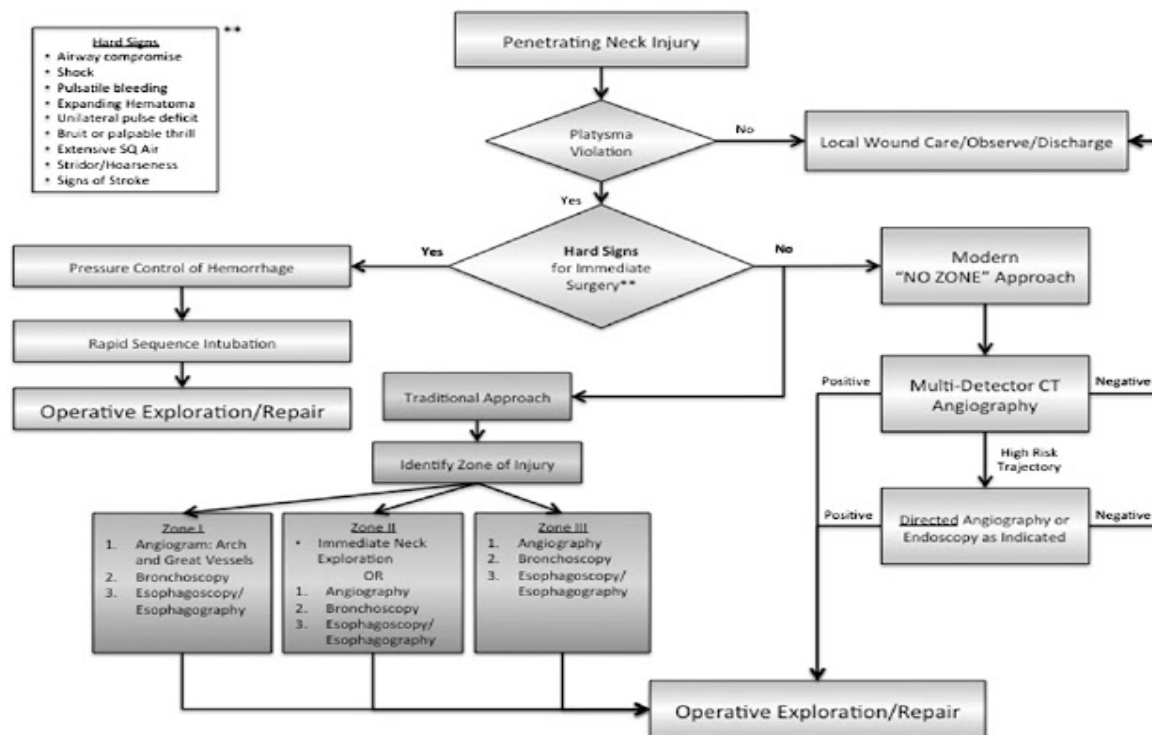
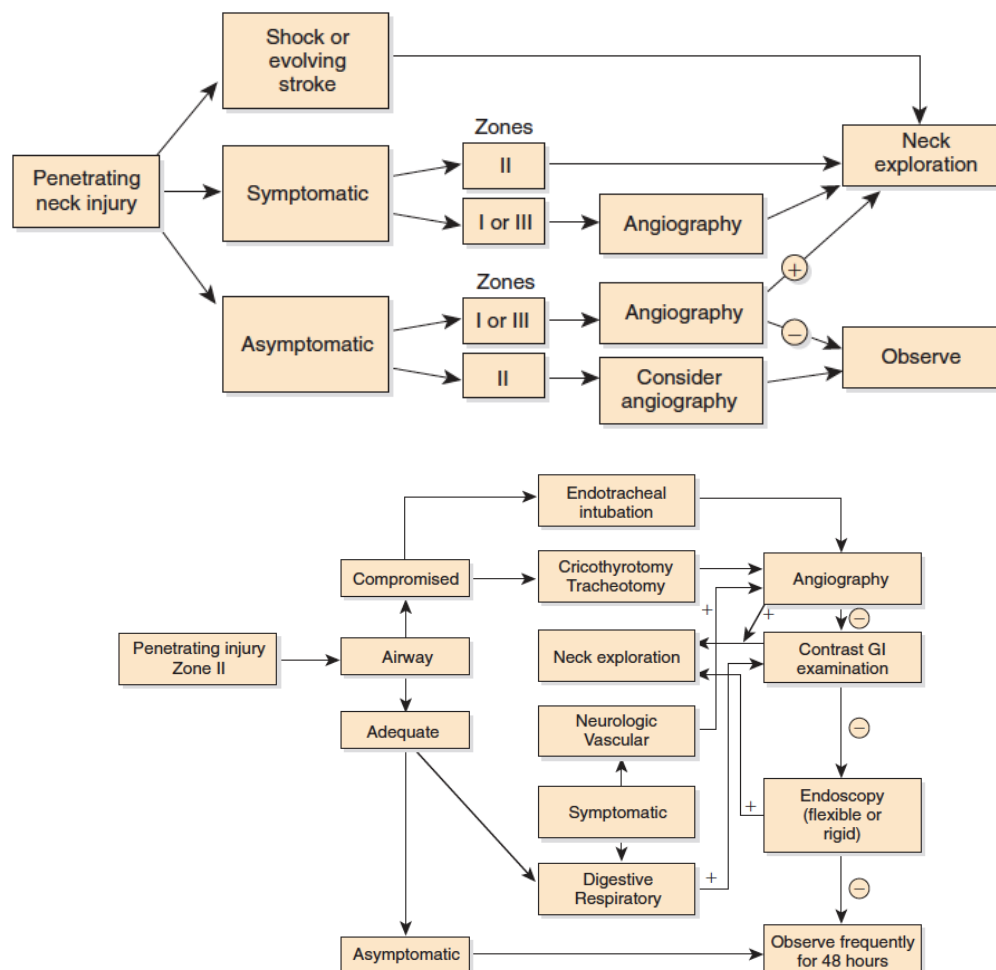


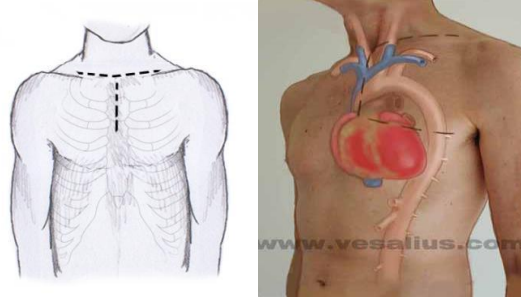
FIG. 1. Algorithm for the modern management of penetrating neck trauma.



Neck Exploration:

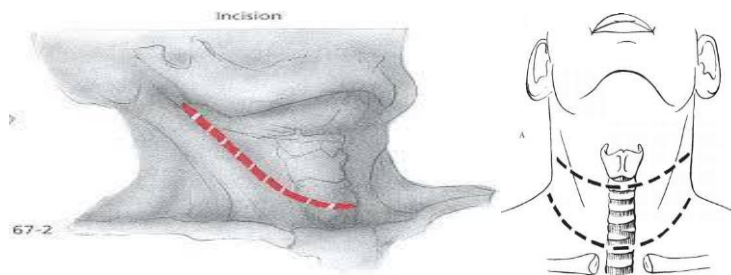
• Zone I:

- Median sternotomy with extension to an anterior sternocleidomastoid incision or supraclavicular incision with or without clavicular head resection.



• Zone II:

- Anterior sternocleidomastoid incision (most common approach).
- Cervical collar incision may provide access to both sides of the neck, with the potential to extend along the anterior sternocleidomastoid muscle.



• Zone III:

- Subluxation, dislocation, or resection of the mandible may be necessary to gain operative vascular control.
- Endovascular techniques have become a useful adjunct and an addition to the armamentarium available for the management of the acutely injured patient

TABLE 121-4. Mandatory vs. Selective Management of Neck Wounds

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Diagnosis	Potential life-threatening injuries can be missed preoperatively; routine exploration can miss some injuries.	Most major injuries can be diagnosed by the preoperative workup.
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Modified from Obeid F, Haddad G, Horst H, et al: A critical reappraisal of a mandatory exploration policy for penetrating wounds of the neck. *Surg Gynecol Obstet* 1985;160:517.

Injury	Investigation
Vascular	Doppler US CT angiogram 4 vessel angiogram (carotid & vertebral) Neck exploration
Esophageal	Contrast esophagogram Esophagoscopy Neck exploration
Laryngeal	CT scan Laryngotracheoscopy Neck exploration

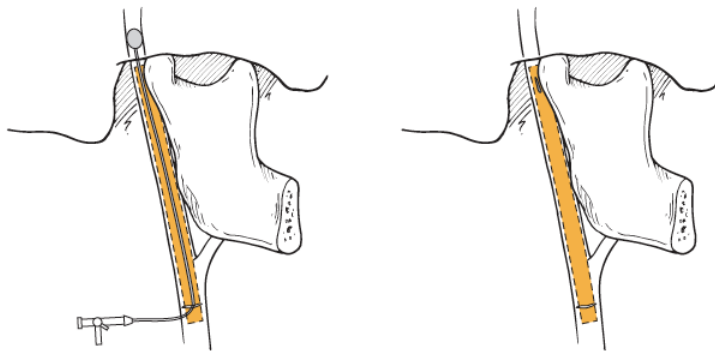


FIGURE 121-9. No. 4 Fogarty catheter method with shunt placement in an attempt to control bleeding of the internal carotid artery near the skull base. (Modified from Perry M: Injuries of the carotid and vertebral arteries. In Bongard FS, Wilson SE, Perry MO, eds: *Vascular injuries in surgical practice*, Norwalk, CT, 1991, Appleton & Lange.)

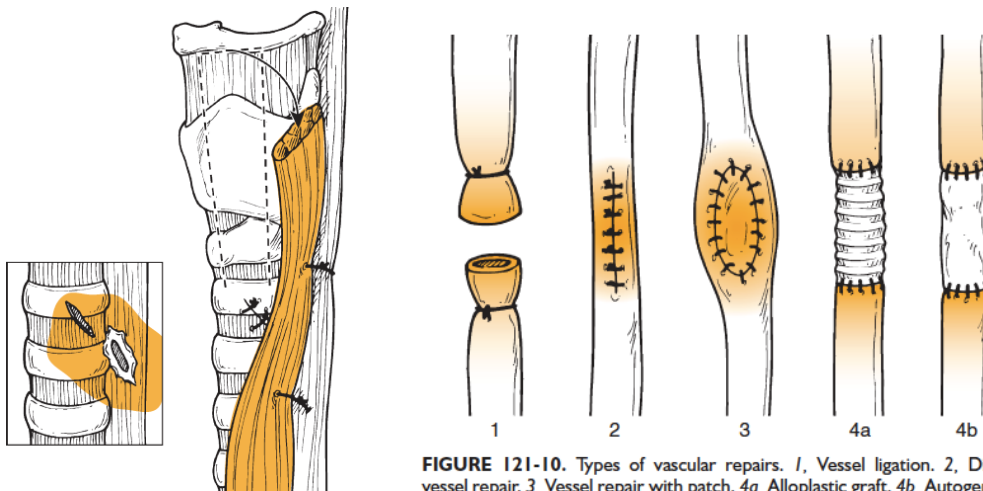


FIGURE 121-10. Types of vascular repairs. 1, Vessel ligation. 2, Direct vessel repair. 3, Vessel repair with patch. 4a, Alloplastic graft. 4b, Autogenous graft. (From Dichtel WJ, Miller RH, Feliciano DV, et al: Lateral mandibulotomy: a technique of exposure for penetrating injuries of the internal carotid artery at the base of the skull. *Laryngoscope* 1984;94:1142.)

FIGURE 121-12. Interposition of strap muscle between esophageal and tracheal repair. (From Miller RH: *The surgical atlas of airway and facial trauma*, Philadelphia, 1983, WB Saunders.)

Blunt Neck Trauma

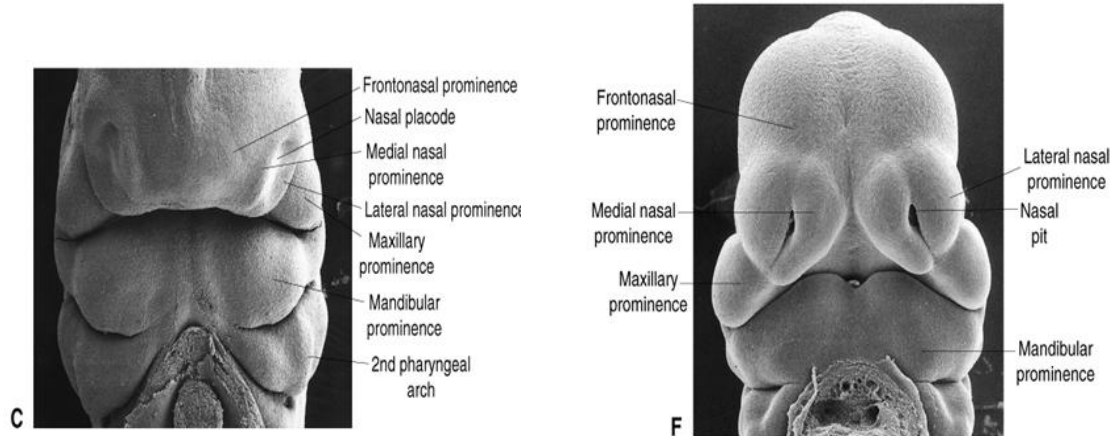
- The laryngotracheal airway and cervical spine are the most clinically susceptible to injury.
- Vascular injuries are potentially devastating but are uncommon overall, occurring in 0.08–1.5 percent of blunt neck trauma
- May need anticoagulant after trauma.
- Despite the widespread use of advanced safety mechanisms, such as shoulder harness seatbelts and airbags, **motor vehicle collisions** remain the most common etiology for blunt neck trauma
- Other mechanisms include blunt object impact sustained in assault, and sports injuries, crush injuries, and hanging or clothesline trauma.
- Presenting Signs and Symptoms as in penetrating neck trauma
- In blunt neck trauma, symptoms can present with a delayed onset and can be easily underdiagnosed.
- **Initial Diagnostic Airway Evaluation**
 - Initial diagnostic airway evaluation with flexible laryngoscopy is helpful since significant edema may occur during first 12–24 hours.
 - CT may be considered for surgical planning in symptomatic patients or in asymptomatic patients with suspected laryngeal injury.
 - **Securing the airway is advocated in the setting of acute airway symptoms, such as stridor or respiratory distress, prior to considering imaging.**
- **Hemodynamic Instability or Signs of Vascular Injury**
 - bruit, expanding/pulsating hematoma, hemorrhage, or loss of pulse
 - **Warrant surgical exploration.**
- **Hemodynamically Stable Patients Showing Risk Factors**
 - Hemodynamically stable patients should undergo initial diagnostic imaging with CTA if at-risk factors are present, including:
 - severe cervical injury
 - anoxic brain injury from hanging
 - closed head injury with diffuse axonal injury
 - midface or complex mandibular fractures
 - marked neck soft tissue swelling injury
 - high-risk cervical spine fractures
 - basilar skull fractures involving the carotid canal
- **Cervical Spine Injury Assessment**
 - After clinical examination, cervical spine injury assessment should include initial lateral and anteroposterior plain x-ray films if possible.

Rhinology:

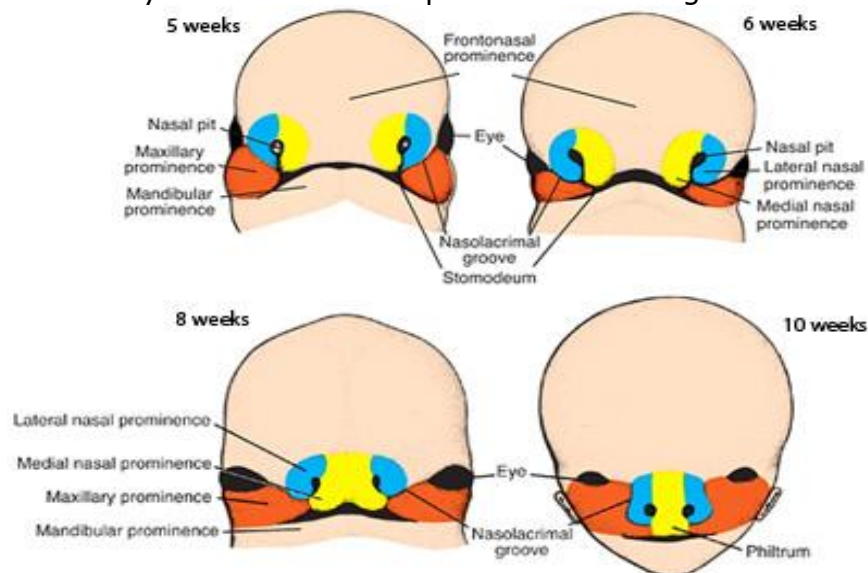
- 1- Embryology of Nose and PNS**
- 2- Anatomy of Nose and PNS**
- 3- Physiology of Nose and PNS**
- 4- Evaluation of Nasal obstruction**
- 5- Nasal Septum and Its Diseases**
- 6- Epistaxis**
- 7- Allergic and Non-Allergic Rhinitis**
- 8- Anti-Histamine**
- 9- Intranasal Corticosteroids (INCs)**
- 10- Rhinosinusitis**
- 11- Fungal Rhinosinusitis**
- 12- Complications of Rhinosinusitis**
- 13- Endoscopic sinus surgery and its complications**
- 14- Frontal Sinus Surgery**
- 15- External Sinus Surgery**
- 16- SinoNasal Neoplasms**
- 17- CSF Rhinorrhea**
- 18- Facial Trauma**
- 19- Granulomatous and Systemic Diseases Affecting the Nose**

Embryology of Nose and Paranasal Sinuses:

- The neural crest cells are the precursors of the nose.
- **During 5th week**, The frontonasal prominence differentiates inferiorly into two projections known as Nasal Placodes.
- The nasal placodes invaginate to form nasal pits.
- They create a ridge of tissue that surrounds each pit and forms the lateral and medial nasal prominences.
- The stomodeum (primitive mouth) is surrounded by mandibular and maxillary prominences bilaterally which are derivatives of 1st branchial arch.
- The maxillary prominences increase in size and grow medially.



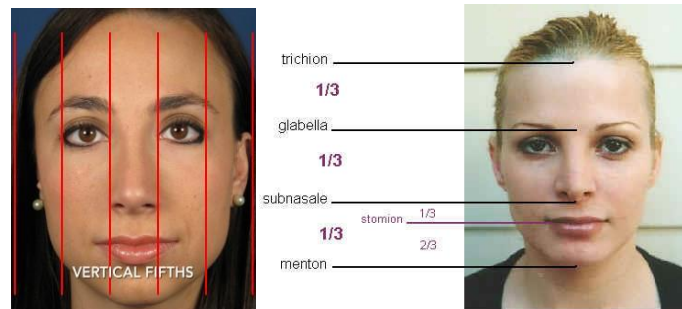
- Fusion of medial nasal prominence with maxillary process forms the upper maxilla and philtrum of upper lip.
- The maxillary and lateral nasal prominences are separated by the nasolacrimal groove which undergo canalization to form the nasolacrimal duct.
- The maxillary prominences enlarge to form the cheeks and maxillae.
- The maxillary and lateral nasal prominences merge with each other.



- Nose is formed from **5 facial prominences** :
 - 1- The frontal prominence gives rise to the bridge
 - 2- The merged medial nasal prominences provide the crest and tip
 - 3- The lateral nasal prominences form the sides (alae).
- **Septum** arises from the posterior midline growth of the frontonasal process and midline extensions from maxillary process.
- Descending septum merges with fused palate to form 2 nasal passageways.
- **During 5–8th week**, three medially directed projections arise from mucous membrane of the lateral wall of the nose.
- This serves as the beginning of the development of paranasal sinuses. Between these projections small lateral diverticula to form the meati of the nose.
- The medial projections arising from the lateral wall of the nose forms the following structures:
 1. **Anterior projection:** Forms "aggr nasi"
 2. **Inferior (maxilloturbinate) projection:** Forms the inferior turbinate and maxillary sinus.
 3. **Superior projection (ethmoidoturbinate):** Forms the superior turbinate, middle turbinate, ethmoidal air cells and their corresponding drainage channels.
- Paranasal air sinuses develop as diverticula of the lateral nasal wall and extend into the maxilla, ethmoid, frontal, and sphenoid bones. They reach their maximum size during puberty and contribute to the definitive shape of the face.
- **Order to reach FULL development E → M → S → F**

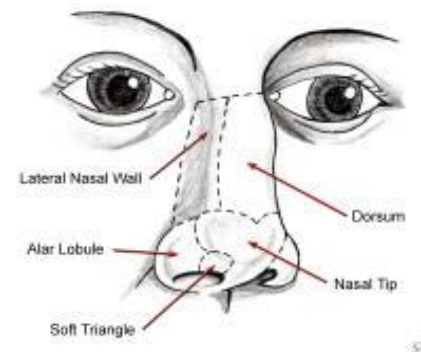
Anatomy of External Nose:

- Pyramidal in shape with its root up and base directed downwards.
- Consist of **Osteocartilaginous framework** covered by muscles and skin.
- Nose cartilage is composed of **Hyaline cartilage**.
- Nose occupies:
 1. Central horizontal 1/3 of the face (from Glabella to Subnasale).
 2. Vertical 1/5 of the face (between the medial canthi).

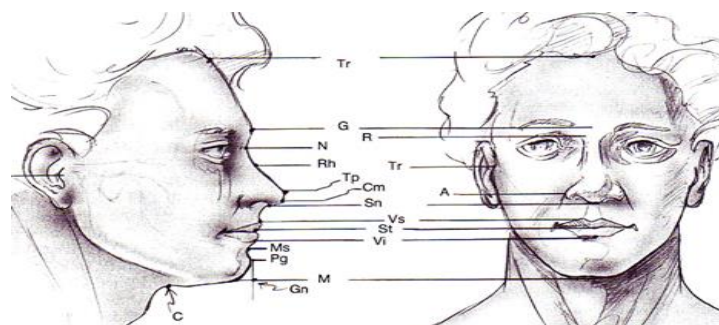


- Divided into 6 Subunits:

1. Nasal dorsum
2. Sidewalls
3. Tip
4. Columella
5. Ala/sill
6. Soft triangles



- **Glabella (G):** Most prominent point in the mid-sagittal plane of forehead.
- **Radix (R):** Root of the nose.
- **Nasion (N):** Depression at the root of the nose (Nasofrontal suture).
- **Rhinion (Rh):** Junction of bony and cartilaginous nasal dorsum (The point of maximal prominence at nasal dorsum).
- **Tip-defining point (Tp):** Anterior most projection of the nasal tip.
- **Columellar point (Cm):** Anterior most soft-tissue point of columella.
- **Subnasale (Sn):** Junction of columella and upper lip at the nasal base.
- **Alar crease (A):** Most posterior aspect of the nose.

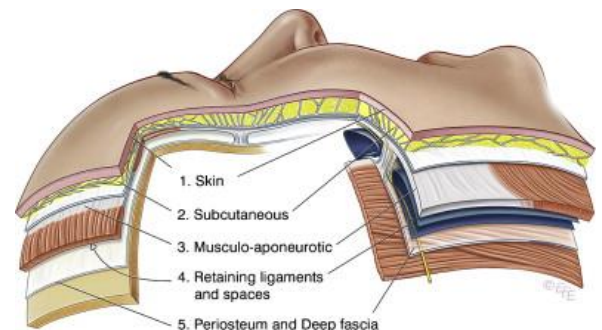


- **Skin:**
- Thickest at Nasion.
- Thinnest at Rhinion.
- Thicker again at tip with Numerous Sebaceous Glands.
- Very thin again at caudal-most aspect of alar margins and columella.
- Relatively mobile skin in the upper part and more adherent in the lower part.

- **Subcutaneous layers:**

1. Superficial Fatty Layer: directly connected to Dermis
2. Fibromuscular Layer: comprises Nasal SMAS (Superficial Muscular Aponeurotic System)
3. Deep Fatty Layer: contains Neurovascular system
4. Periosteum or Perichondrium

- Osteocartilaginous framework of nose is covered by muscles which bring about movements of the nasal tip, ala and the overlying skin.



- **Motor nerve supply:**

- Facial nerve. (CN-VII)

- **Sensory nerve supply:**

- Ophthalmic Nerve (V1):
 1. Supratrochlear Nerve → Nasal Dorsum
 2. Infratrochlear Nerve → Nasal Dorsum
 3. External Nasal branch of Anterior Ethmoid Nerve → Nasal Tip
- Maxillary Nerve (V2):
 1. Infraorbital Nerve → Malar, Lateral Nose and Subnasal region.

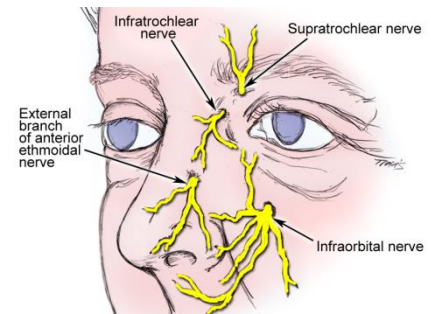
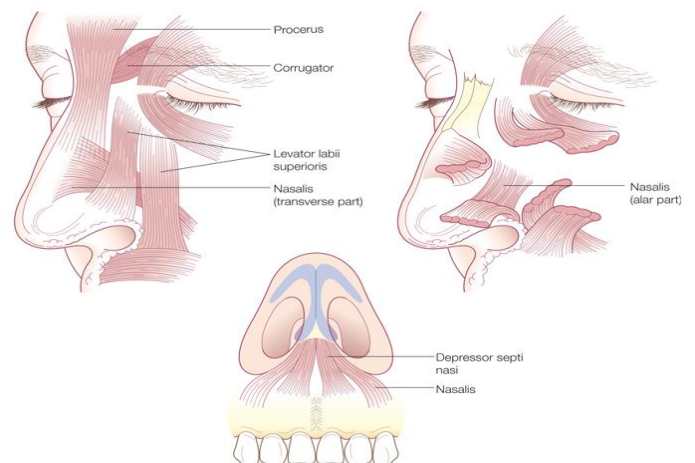


TABLE 170.1
INVESTING NASAL MUSCULATURE

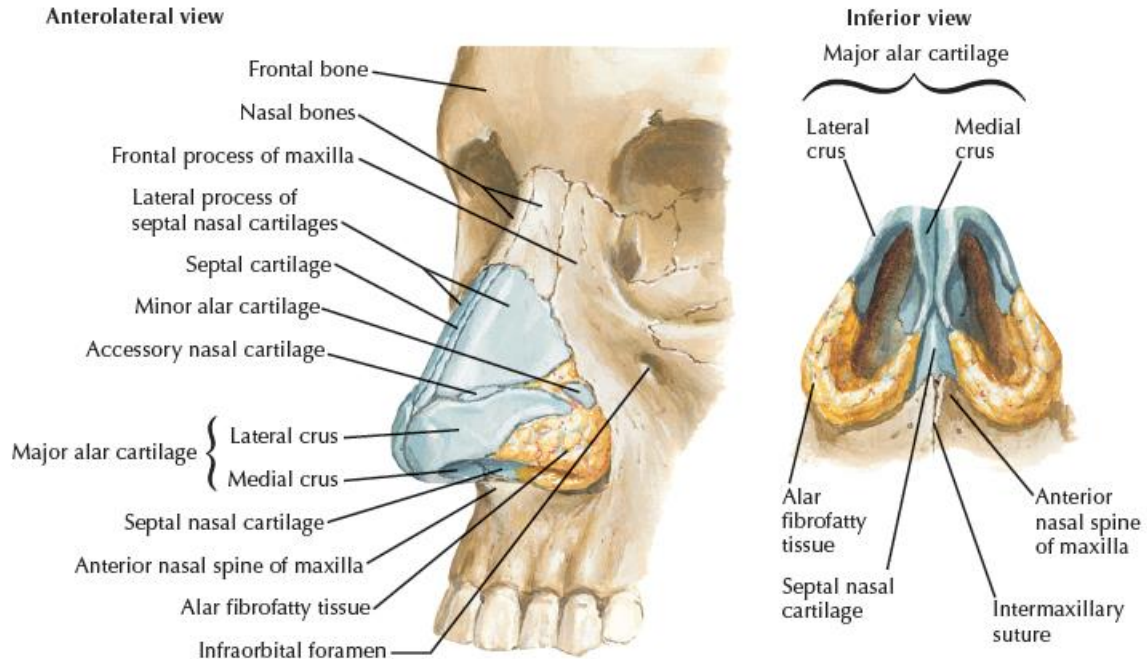
Elevator muscles	
Procerus	
Levator labii-superioris alaeque nasi	
Anomalous nasi	
Depressor muscles	
Alar nasalis	
Depressor septi nasi	
Compressor muscles	
Transverse nasalis	
Compressor narium minor	
Minor dilator muscles	
Dilator naris anterior	



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- **Osteocartilaginous Framework:**

1. Upper third = Osseous vault
2. Middle third = Upper cartilaginous vault
3. Lower third = Lower cartilaginous vault



- **Osseous vault:**

Upper 1/3 and provides principal nasal structural support.

- Composed of:

1. Nasal bones.
2. Nasal process of Frontal bone.
3. Frontal processes of Maxilla.

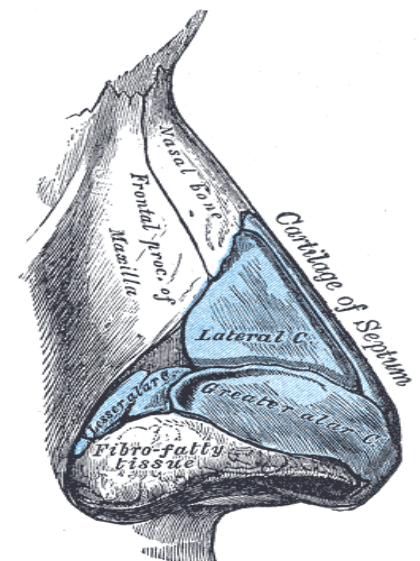
- **Upper cartilaginous vault:**

- Mid 1/3 (Mid-vault).

- Composed of:

1. Paired Upper Lateral Cartilages (**ULC**).
2. Cartilaginous septum fused in midline to ULC.

- The lower free edge of upper lateral cartilage is seen intranasally as Limen Nasai or Internal Nasal Valve.



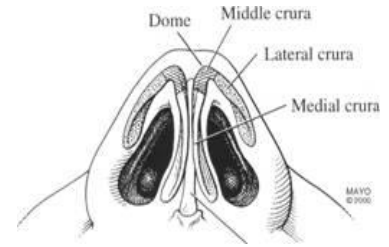
- **Lower cartilaginous vault:**

- Lower 1/3.

- Composed of:

1. Paired Lower Lateral Cartilages (**LLC**) (Greater Alar Cartilages)
2. Lesser Alar (Sesamoid) Cartilages.

- Lower Lateral Cartilages (**LLC**) is critical in defining Nasal tip and Nostril shape
- **Composed of:**
 1. Medial crus: connected to caudal septum and provides support to columella
 2. Middle crus (Dome): bridges between medial and lateral crura
 3. Lateral crus: forms Ala.



- Lesser Alar (sesamoid) cartilages are two or more in number lie above and lateral to alar cartilages and connected with one another and with the adjoining bones by perichondrium and periosteum.
- Most of the free margin of nostril is formed of Fibrofatty tissue, not Alar cartilage.

- **Nose Tip Support:**

- **MAJOR:**

1. Medial and Lateral crura of LLC.
2. Attachment of Medial crus to caudal border of Quadrangular cartilage.
3. Attachment of Caudal border of ULC to Cephalic border of LLC.

- **MINOR:**

1. Ligamentous sling spanning Domes of LLC.
2. Dorsal portion of cartilaginous nasal septum.
3. Sesamoid complex extending the support of lateral crura to the pyriform aperture.
4. Attachment of LLC cartilage to the overlying skin and musculature
5. Nasal spine
6. Membranous portion of nasal septum

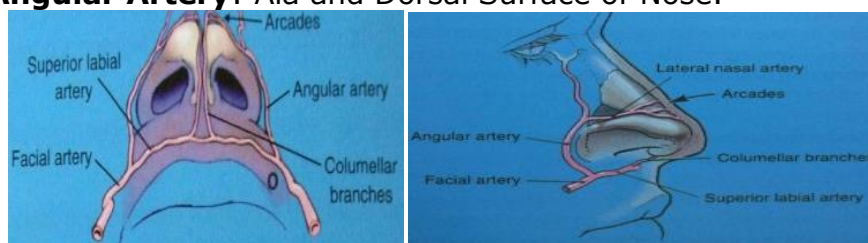
- **Arterial supply of External Nose:**

- Internal Carotid Artery → **Ophthalmic Artery:**

1. **Dorsal Nasal Artery:** Bridge of Nose.

- External Carotid Artery → **Facial Artery:**

1. **Superior Labial Artery:** Sill, Columella and Anterior Septum.
2. **Angular Artery:** Ala and Dorsal Surface of Nose.



- Venous drainage mainly through **Angular vein** → Anterior Facial Vein → Internal jugular vein.

Anatomy of Nasal Cavity and Paranasal Sinuses:

- Divided into Right and Left nasal cavities by Nasal Septum.
- Communicates with the exterior through Nostril and with the Nasopharynx through Choana.

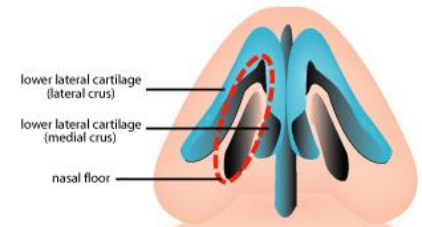
○ **External Nasal Valve: (Nasal Vestibule)**

- Anterior and inferior part of nasal cavity (Nostril).
- Lined by Skin and contains Sebaceous glands, Hair follicles and Vibrissae.

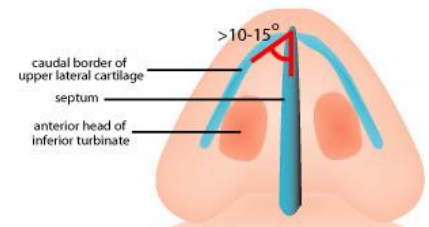
- Formed by:

1. Lateral Crus of Lower Lateral Cartilage (LLC) .
2. Columella
3. Nasal sill

The External Valve



The Internal Valve



- **Internal Nasal Valve: (Limen Nasi)**

- Narrowest part of Nasal Airway.
- Cross-section area is **0.73 cm²**.
- Forms an angle of **10-15 degree**.
- Bordered by:
 1. Septum
 2. Anterior edge of Inferior Turbinate.
 3. Caudal edge of Upper Lateral Cartilage (ULC).

- **Lining Membranes:**

- **Vestibule:**

- Skin → Keratinized Stratified Squamous Epithelium.

- **Lower 2/3 of Nasal Cavity + Paranasal Sinuses:**

- Respiratory mucosa → Pseudostratified Ciliated Columnar Epithelium.

- **Upper 1/3 of Nasal Cavity:**

- Olfactory Neuroepithelium.

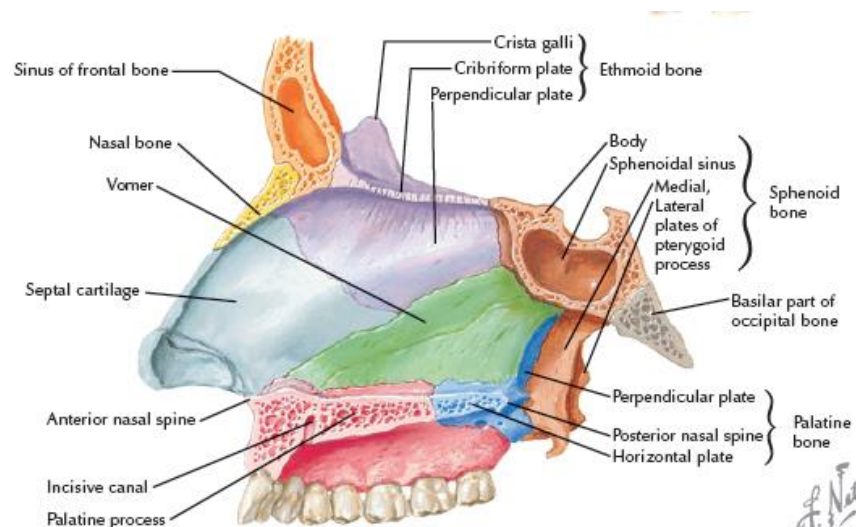
- **Borders of Nasal Cavity:**

- Roof:

1. Nasal bones
2. Cribriform plate of Ethmoid.
3. Body of Sphenoid.

- Floor:

1. Palatine process of Maxilla
2. Horizontal Plate of Palatine bone.



- **Medial wall of Nasal Cavity:**

- Composed of:

1. **Columella:**

- Formed of Medial Crura of LLC united together by Fibrous tissue and covered on either side by Skin.

2. **Membranous Septum:**

- Double layer of skin with no bony or cartilaginous support between Columella and Quadrangular cartilage.

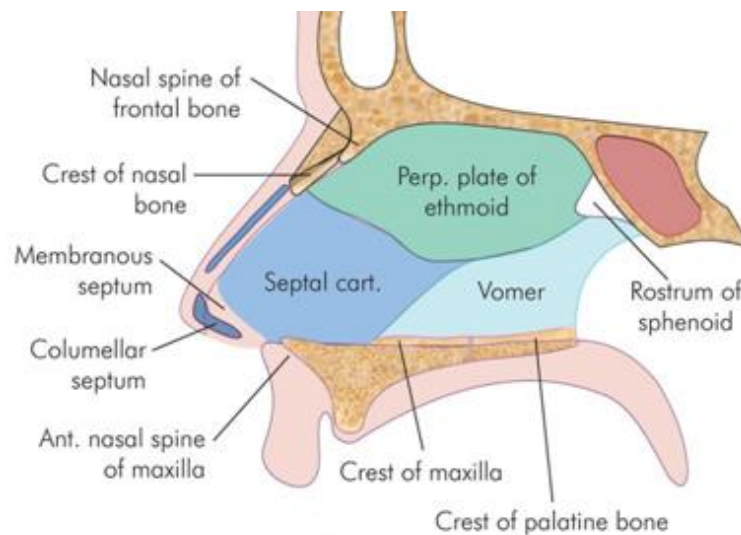
3. **Septum Proper:**

- Principal constituents:

1. Perpendicular Plate of Ethmoid.
2. Vomer Bone.
3. Septal (Quadrangular) cartilage.

- Minor constituents:

1. Anterior Nasal spine of Maxilla.
2. Maxillary crest.
3. Palatine bone crest.
4. Rostrum of sphenoid.
5. Nasal spine of frontal bone.
6. Crest of nasal bones.



- Superior border of Septal (Quadrangular) Cartilage runs from Nasal bones to Nasal tip and functions to support Dorsum of Cartilaginous part of Nose.

- Nasal growth centers:

- Located between Quadrangular cartilage and Vomer.
- Affected in trauma or during pediatric septoplasty causing:
 1. Short nose
 2. Saddle nose deformity
 3. Drop of tip

- **Lateral wall of Nasal Cavity:**
- Formed by:
 1. **Ethmoid Bone:**
 - Superior Turbinate.
 - Middle Turbinate.
 - Uncinate Process.
 2. **Inferior Turbinate Bone**
 3. **Maxilla:**
 - Frontal Process.
 4. **Palatine Bone:**
 - Perpendicular plate.
 5. **Lacrimal Bone.**
 6. **Nasal Bones.**

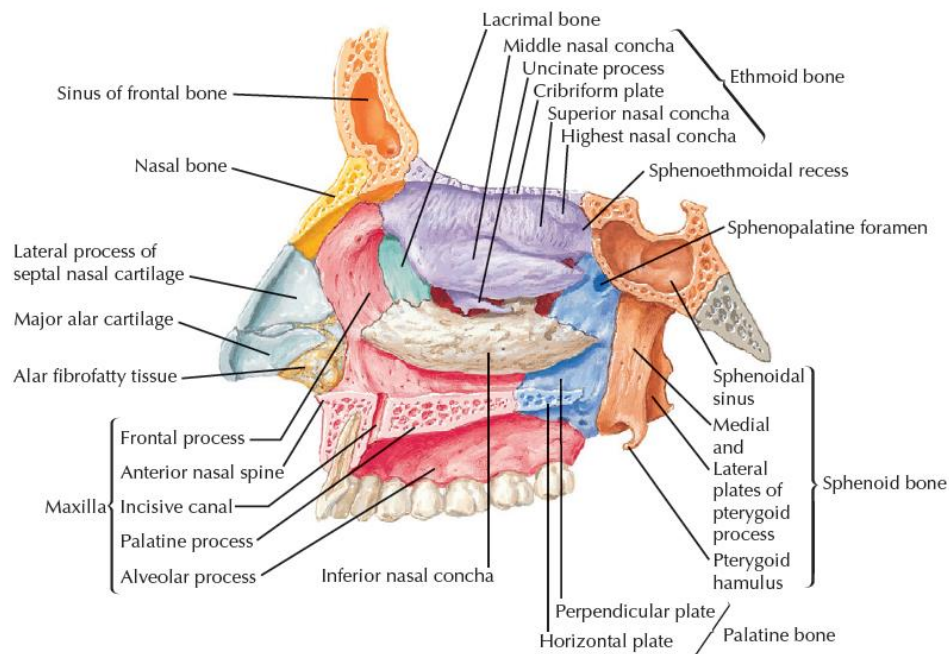
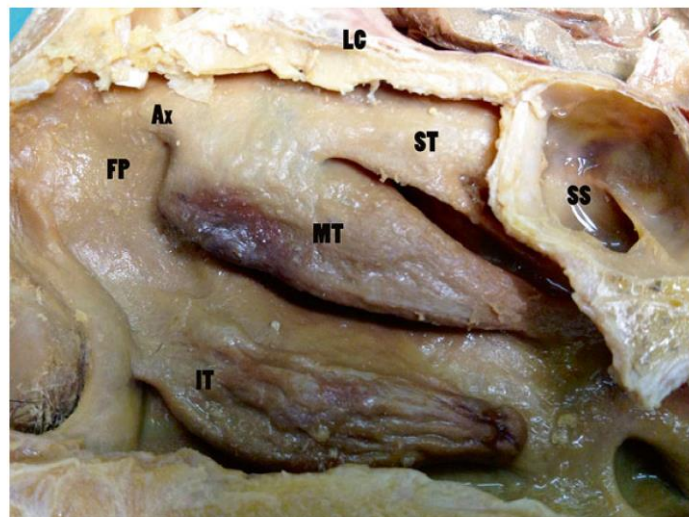


Fig. 2.11 Lateral wall of the nasal cavity. *LC* lamina cribosa, *Ax* axilla, *FP* frontal process, *ST* superior turbinate, *MT* middle turbinate, *IT* inferior turbinate, *SS* sphenoid sinus



- Lateral wall contains three meatuses underneath three turbinates:
 - Turbinates are bony projections covered by Erectile mucosa serves to increase the interior surface area.
- **Inferior Turbinate:**
- A separate bone (Not part of ethmoid bone).
- Articulates with inferior margin of the maxillary hiatus via its maxillary process.
- Inferior meatus:
 - is the area of the lateral wall of the nose covered medially by the inferior turbinate
 - Drains:
 1. Nasolacrimal Duct (Hasner's valve).

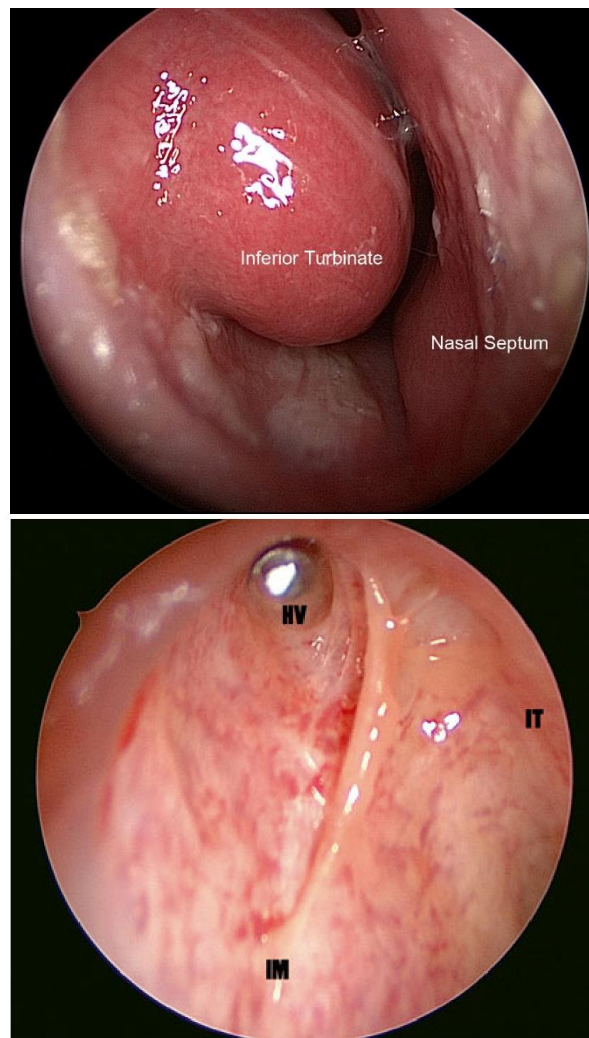
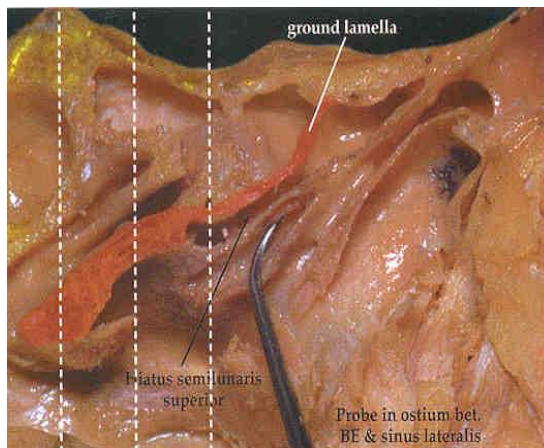


Fig. 2.6 Endoscopic view of the inferior meatus. 45° angled endoscope. *IM* inferior meatus, *HV* Hasner's valve, *IT* inferior turbinate

- **Middle Turbinate:**
- Part of Ethmoid bone.
- Attached to Lateral wall by a bony lamella called **Ground or Basal Lamella (S-shaped Manner)**.

Basal Lamella of Middle Turbinate			
Part	Site	Plane	Attachment
1st	Anterior	Sagittal (Vertical)	Skull Base
2nd	Middle	Frontal (Oblique)	Lamina Papyracea
3rd	Posterior	Horizontal	Perpendicular Plate of Palatine Bone



- The most anterior part of the middle turbinate fuses with the inferior part of agger nasi to form the "axilla".
- Middle meatus:
 - o The area of the lateral wall of the nasal cavity covered medially by the middle turbinate.
 - o Drains:
 1. Maxillary sinus.
 2. Anterior Ethmoid sinus.
 3. Frontal Sinus.

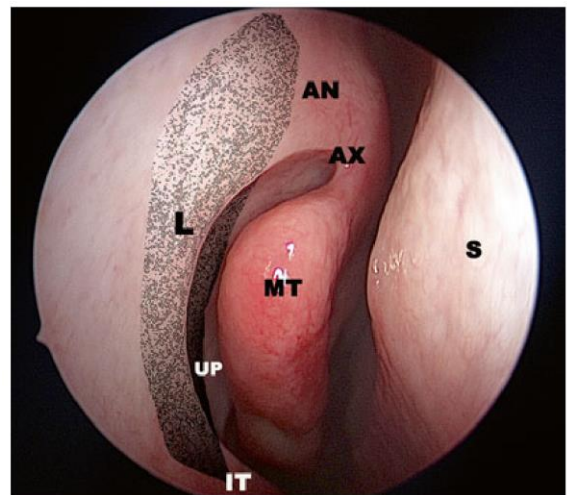


Fig.2.9 Endoscopic view of the right nasal cavity. *L* projection of the lacrimal system, *UP* uncinate process, *MT* middle turbinate, *IT* inferior turbinate, *S* septum, *AN* agger nasi, *Ax* axilla

- **Superior Turbinate:**
- Part of ethmoid bone.
- Located above the middle turbinate and bearing olfactory epithelium on its medial surface.
- There may also be a supreme turbinate.
- Superior meatus:
 - o The area of the lateral wall of the nose covered medially by the superior turbinate.
 - o Drains:
 1. Posterior ethmoid sinuses.
- **Sphenoethmoidal Recess:**
- Located in front of the anterior wall of the sphenoid and medial to the superior turbinate.
- Drains:
 - o Sphenoid sinus.

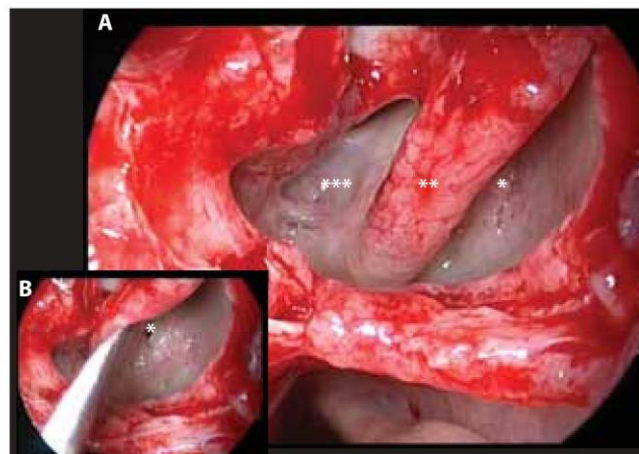
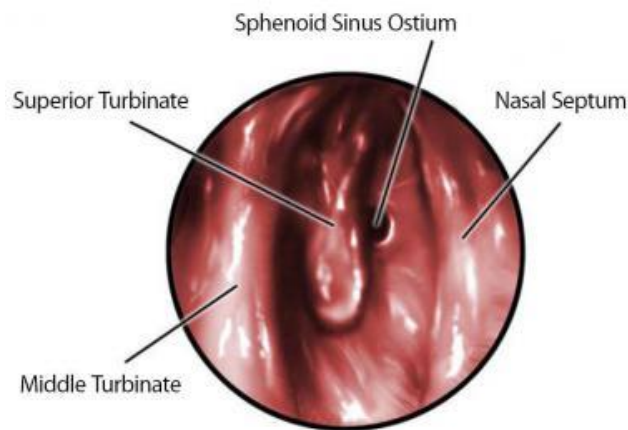
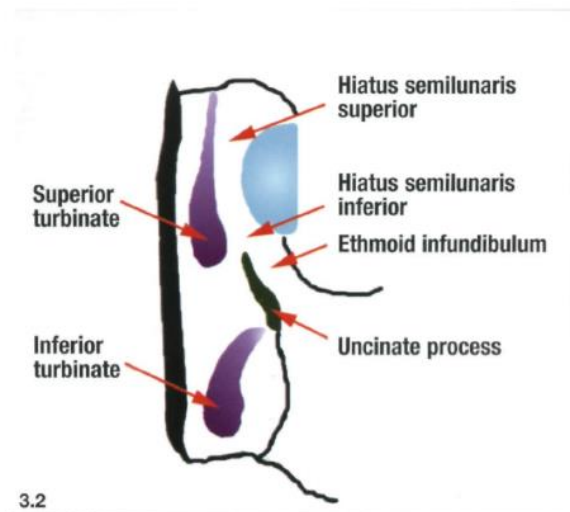
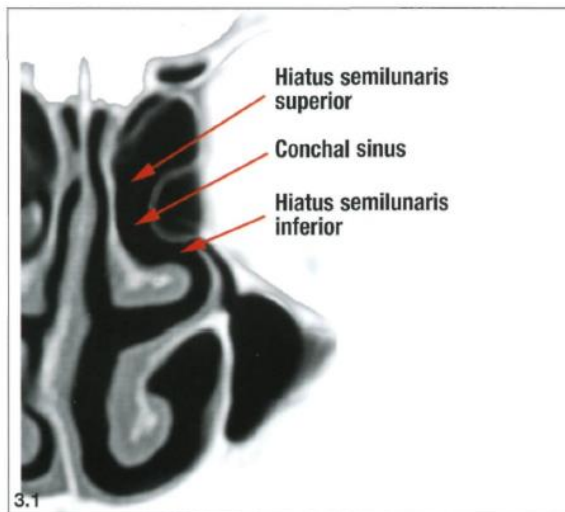
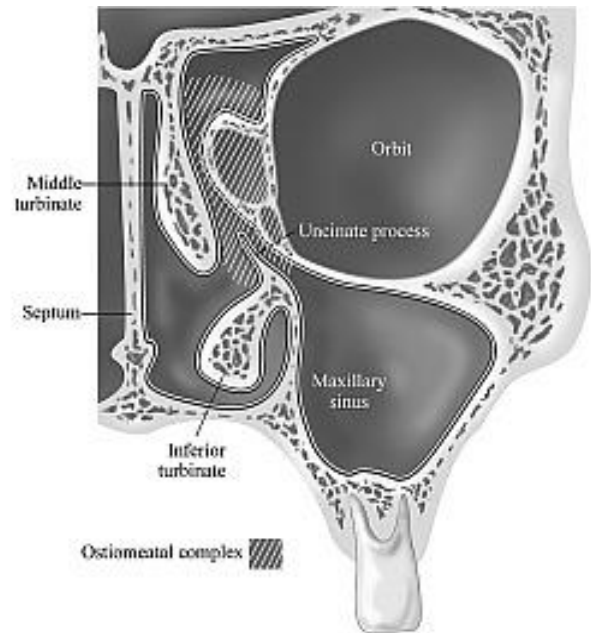


Figure 40. A) Sphenoethmoidal recess (*), superior turbinate (**) and posterior ethmoid (***). B) Sphenoid sinus ostium (*). Right side.

- **Osteomeatal Complex:**
- Narrow anatomical region in Anterior Ethmoid, Lateral to Middle Turbinate.
- Consist of:
 1. **Bony structures:**
 - Middle Turbinate.
 - Uncinate Process.
 - Bulla Ethmoidalis.
 2. **Air spaces:**
 - Frontal Recess.
 - Ethmoidal Infundibulum.
 - Middle Meatus.
 3. **Ostia:**
 - Anterior Ethmoid Ostium.
 - Maxillary Sinus Ostium.
 - Frontal Sinus Ostium.



Figures 3.1, 3.2
Localization of hiatus semilunaris inferior and the ethmoid infundibulum (coronal cross-section)

- **Uncinate Process (UP):**
- Sickle-like thin bony part of Ethmoid bone.
- Located Medial to Ethmoidal Infundibulum, Lateral to Middle Turbinate.
- Runs in the sagittal plane from Antero-superior to Postero-inferior direction.
- It has a concave free posterior margin that lies parallel to the anterior surface of the ethmoidal bulla.
- Hiatus Semilunaris Inferior lies immediately behind the free edge.
- Attachments:
 - Posteroinferiorly it attaches to the perpendicular process of the palatine bone and the ethmoidal process of the inferior turbinate.
 - Anteriorly it is attached to the lacrimal bone and in the sagittal plane, may have a "common" attachment to the medial surface of the agger nasi cell and the middle turbinate.
 - Variable Superior Attachments (60% most commonly to Lamina Papyracea).
- Surgical note:
 - It is important to check the CT scan for the distance from the uncinate process to the medial wall of the orbit to evaluate the width of the ethmoidal infundibulum.

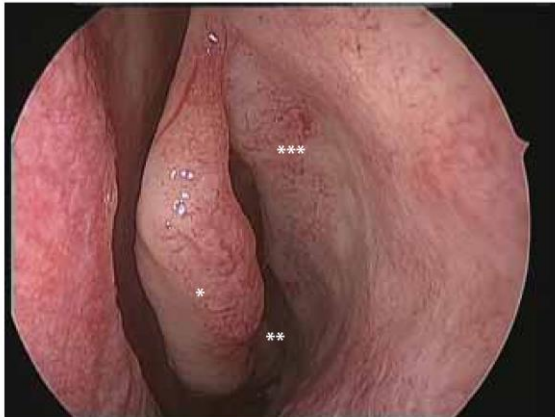


Figure 9. Left middle turbinate (*), middle meatus (**) and uncinate process (***).



Figure 5. Right everted uncinate process (*), middle turbinate (**) and nasal septum (***).

- **Fontanelles:**
- Thin boneless segments of lateral nasal wall above the Inferior Turbinate.
- They are closed with mucosa, connective tissue and in continuity with the maxillary periosteum.
- Uncinate Process divides Fontanelles into Anterior and Posterior fontanelle.
- Results in Accessory ostium of Maxillary Sinus, in 5% of the normal population and up to 25% of patients with chronic rhinosinusitis.
- Surgical note:
 - o Natural ostium of the maxillary sinus lies between the anterior and posterior fontanelles of the maxillary sinus and cannot usually be seen with a 0 degree endoscope without removing the uncinate process mainly due to its oblique orientation in the sagittal plane.
 - o If an ostium is seen, it is most likely an accessory ostium (in the absence of previous sinus surgery).

Figure: Endoscopic view of the right natural maxillary ostium anteriorly (arrowheads), as well as an accessory ostium in the posterior fontanelle (black arrow), using a 30-degree endoscope.

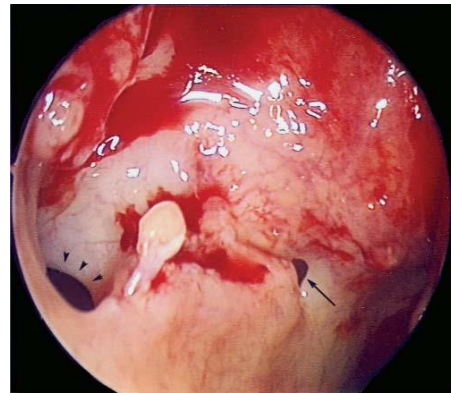


Figure 14. Accessory ostium in the anterior fontanelle (*) and paradoxically bent middle turbinate (**).

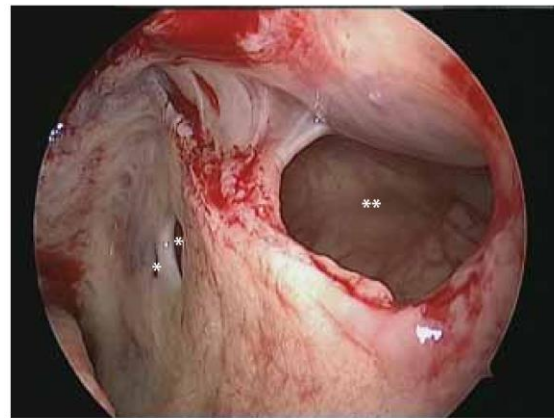


Figure 15. Accessory ostia in the posterior fontanelle (*) and left maxillary sinus ostium (**).

- **Agger Nasi:**
- Most Anterior ethmoid cell.
- Has a variable degree of pneumatization in up to 90% of patients.
- It is the first pneumatization seen on sagittal and coronal CT, posterior to the lacrimal bone and anterior to the free edge of the uncinate process.
- Seen on intranasal examination as a small prominence on the lateral nasal wall:
 - Anterior and superior to the attachment of the middle turbinate.
 - Anterior to Upper margin of Nasolacrimal duct.
 - Posterior wall of Agger nasi cells forms Anterior wall of Frontal recess.
- Major dimensions are correlated with Frontal sinus diseases and Lacrimation.

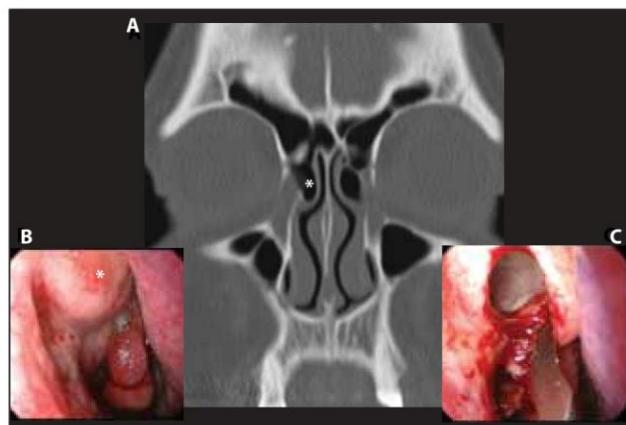
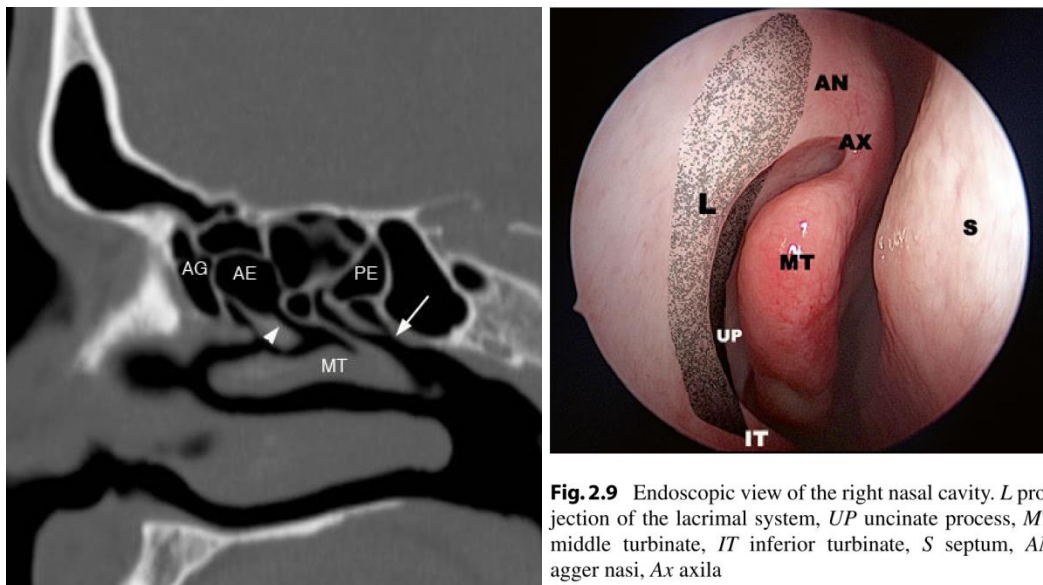


Figure 8. A) The agger nasi (*) is the most anterior part of the ethmoid, and may be seen on intranasal examination as a small prominence on the lateral nasal wall just anterior to the attachment of the middle turbinate. B) Agger nasi cell prior to resection. C) Agger nasi cell post opening.

Ethmoidal Infundibulum:

- Funnel shaped space.
- Channels drainage of Maxillary, Anterior Ethmoid and Frontal sinuses into Middle Meatus.
- Limited Medially by:
 - o Uncinate Process.
 - o Frontal Process of Maxilla.
- Limited Laterally by:
 - o Lamina Papyracea.
- Limited posteriorly by:
 - o Anterior face of ethmoidal bulla.
- Opens into the middle meatus via Hiatus Semilunaris Inferior.
- Natural ostium of Maxillary sinus is situated in Posterior Lower part of Infundibulum.
- Relationship to frontal recess varies with Uncinate Process attachments:
 - o **Terminal recess** of the ethmoidal infundibulum is formed if the superior attachment of the uncinate process is onto the lamina papyracea or the base of an agger nasi cell, thus forming a blind end to the ethmoidal infundibulum superiorly.
- **Hiatus Semilunaris Inferior:**
- Crescent-shaped gap between Posterior free edge of Uncinate Process and Anterior wall of Bulla Ethmoidalis.
- Communication between Middle meatus and Infundibulum.
- **Hiatus Semilunaris Superior:**
- Cleft-like communication between Bulla Ethmoidalis and skull base.
- Opens into middle meatus.

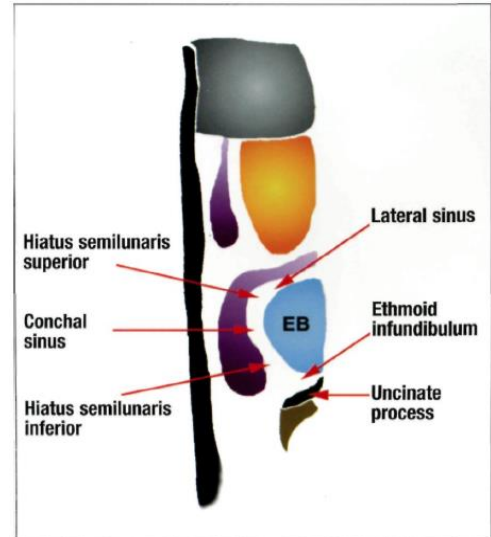
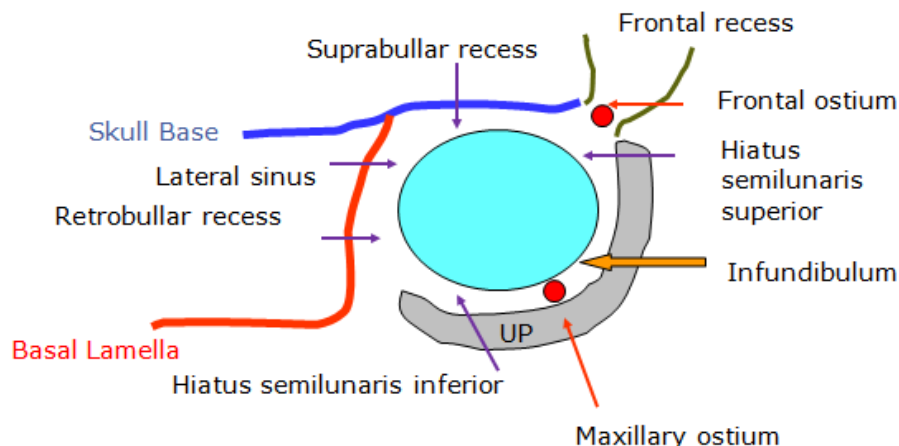


Figure 2.1
Localization of the lateral sinus (axial cross-section)
EB ethmoid bulla
UP uncinate process

Sagittal Section



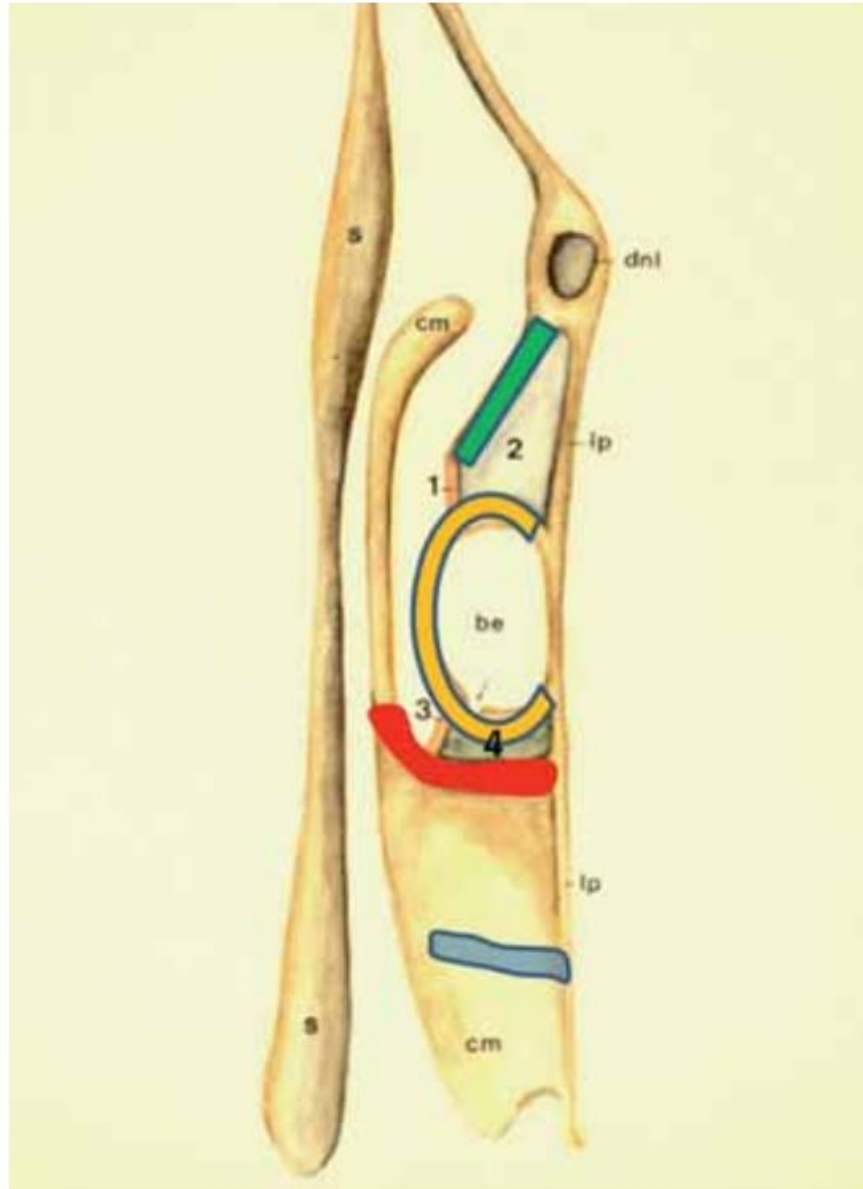


Figure 19. Schematic drawing in the axial plane through the frontal portion of the basal lamella of the middle turbinate (red). Green : Uncinate process; Yellow : Ethmoidal bulla; Blue : Basal lamella of superior turbinate. s = nasal septum, cm = concha media / middle turbinate, dnl = nasolacrimal duct, lp = lamina papyracea. 1 = hiatus semilunaris (inferior), 2 = ethmoidal infundibulum, 3 = hiatus semilunaris superior, 4 = retrobullar recess. be = ethmoidal bulla.

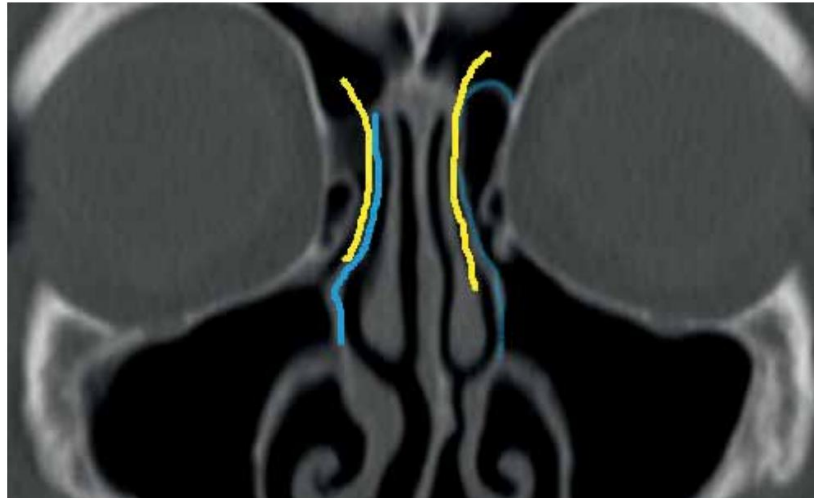


Figure 21. If the uncinate process (blue line, right side) attaches to the skull base, the infundibulum will be continuous with the frontal recess superiorly (yellow line). If the uncinate process (blue line, left side) attaches to the lamina papyracea, the infundibulum will end blindly in the terminal recess. The maxillary sinus opens into the ethmoidal infundibulum, the frontal drainage pathway (yellow line) is medial to the uncinate process.

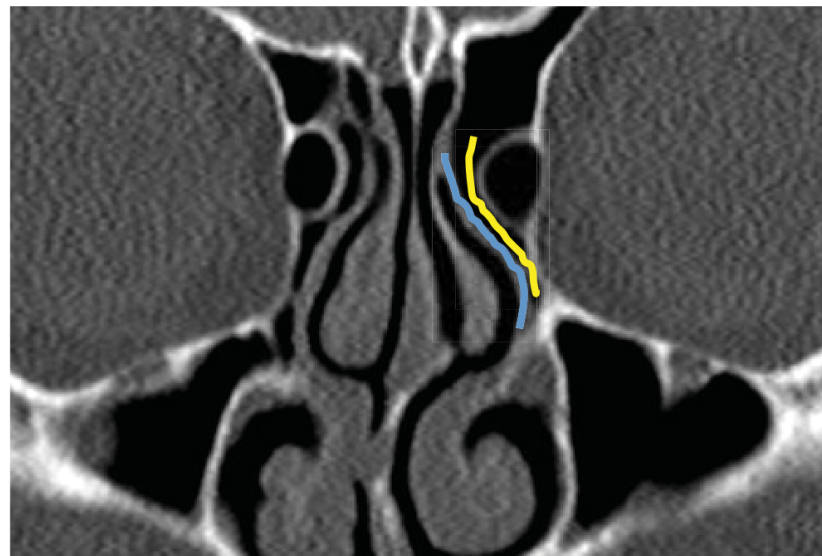


Figure 22. If the uncinate process (blue line) attaches to the middle turbinate, the infundibulum will be continuous with the frontal recess superiorly (yellow line), the frontal drainage pathway thus being lateral to the uncinate process (as on right side in Figure 21).

- **Bulla Ethmoidalis:**
- **Most consistent landmark in sinus surgery.**
- Largest Anterior Ethmoidal Air cell.
- Situated within Middle meatus
- Posterior to Uncinate Process
- Anterior to Basal Lamella of Middle Turbinate.
- The anterior face of the bulla forms the posterior border of:
 - o Inferior semilunar hiatus
 - o Ethmoidal infundibulum
 - o Frontal recess.
- Opens into the superior semilunar hiatus or retrobullar recess (70%).



Figure 16. Enlarged ethmoidal bulla (*).

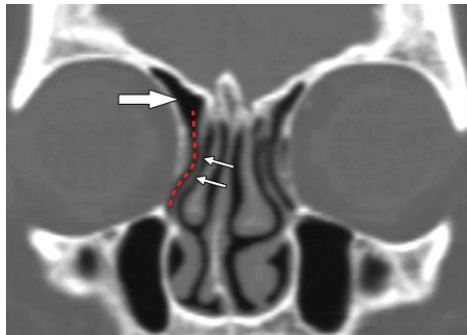
- If the ethmoidal bulla reaches the ethmoidal roof:
 - o It forms the posterior border of the frontal recess.
- If the ethmoidal bulla does not reach the ethmoidal roof:
 - o An air-filled space called **Supra-bullar recess** is present between the superior aspect of the bulla and the ethmoidal roof.
 - Bordered inferiorly by roof of ethmoidal bulla.
 - Bordered medially by middle turbinate.
 - Bordered laterally by lamina papyracea.
 - Bordered superiorly by roof of the ethmoid.
 - Found in 70% of cadavers.
 - Laterally it may give rise to an air-containing cleft extending above the orbit, known as a supraorbital recess.



Figure 20. A suprabullar recess (*) may give rise to an air-containing cleft extending above the orbit. This is a supraorbital recess (**), formerly known as supraorbital cell; ethmoidal bulla (***)).

- If the posterior wall of ethmoidal bulla is separate from the basal lamella of the middle turbinate:
 - An air-filled space called **Retro-bullar recess** is present.
 - Bordered medially by middle turbinate.
 - Bordered laterally by lamina papyracea.
 - It opens medially into the middle meatus via the superior semilunar hiatus.
 - Found in 90% of cadavers.
- **Lateral Sinus:**
 - Formed by Supra-bullar + Retro-bullar Recesses.
 - This term has been abandoned.
- **Conchal Sinus:**
 - Space between Middle turbinate and Medial wall of Bulla Ethmoidalis.
- Surgical note:
 - If the bulla is poorly or non-pneumatized, the medial wall of the orbit is potentially at risk.
 - It is also important that the surgeon appreciates the proximity of the skull base when the bulla is pneumatized superiorly.

- **Frontal Recess:**
- Most Anterior Superior aspect of Anterior Ethmoid Sinus that forms connections with Frontal Sinus.
- Hourglass shape, Narrow waist at Frontal ostium.
- Boundaries:
 - Anteriorly: Agger Nasi.
 - Laterally: Orbit.
 - Medially: Middle Turbinate.
 - When Ethmoid Bulla lamella reaches Skull base, it forms Posterior border of Frontal Recess.
 - When it doesn't reach Skull base, Supra-Bullar Recess communicates directly with Frontal Recess.



- Patency of Frontal Recess depends on:
 1. Superior attachment of Uncinate Process.
 2. Agger Nasi cell.
 3. Ethmoid bulla.
 4. Presence or Absence of Supra-bullar cell.
 5. Presence or Absence of Frontal bullar cell (Posterior).
 6. Presence or Absence of Frontal cells (Anterior).
 7. Presence or Absence of Supraorbital cell.
 8. Presence or Absence of Inter-frontal sinus septal cell.
- Frontal beak (Nasofrontal process):
 - Thick bone underlying the nasion comprising the nasal process of the frontal bone medially, the frontal process of the maxilla laterally with a potential contribution from the nasal bone infero-anteriorly.
- Surgical note:
 - Frontal recess is best studied in all three planes on CT, but especially the sagittal views.
 - On endoscopic examination, the access to the frontal sinus is medial to the attachment of the uncinate process in the majority of cases.

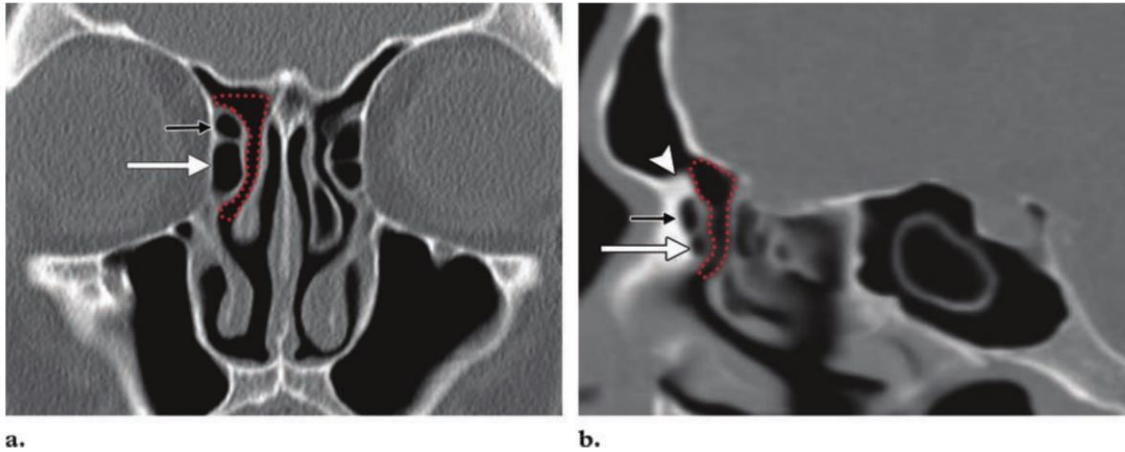


Figure 1. Normal frontal recess anatomy. Coronal (**a**) and sagittal (**b**) CT images show the right frontal recess (dotted red line), which is bounded anteriorly and laterally by an agger nasi cell (white arrow) and a type 1 frontal cell (black arrow), medially by the middle turbinate, and posteriorly by the ethmoid bulla and bulla lamella. The nasofrontal process (arrowhead in **b**) forms the floor of the frontal sinus and demarcates the level of the frontal sinus ostium.

Types of Frontal Recess Cells		
Cell Type	Description	Best Planes for Viewing
Frontal cell		
Type 1	Single cell above the agger nasi cell; it does not extend into the frontal sinus, and its posterior wall is a free partition in the frontal recess	Coronal, sagittal
Type 2	A tier of two or more cells above the agger nasi cell; its posterior wall is a free partition in the frontal recess	Coronal, sagittal
Type 3	Single large cell above the agger nasi cell; it extends into the true frontal sinus, and its posterior wall is a free partition in the frontal sinus or recess	Coronal, sagittal
Type 4	Isolated cell in the frontal sinus; its anterior or inferior margin is the anterior table or floor of the frontal sinus, and its posterior wall is a free partition in the frontal sinus	Coronal, sagittal
Supraorbital ethmoid cell	A cell extending over the orbit from the frontal recess; its posterior wall is the skull base, and it may mimic a septated frontal sinus	Axial, coronal
Frontal bullar cell	A cell above the ethmoid bulla pneumatizing into the frontal sinus; its superior wall is the skull base, and its anterior wall extends into the frontal sinus	Sagittal
Suprabullar cell	A cell above the ethmoid bulla; its superior wall is the skull base, and its anterior wall does not extend into the frontal sinus	Sagittal
Inter-frontal sinus septal cell	A pneumatized frontal sinus septum; it is associated with a pneumatized crista galli	Axial, coronal

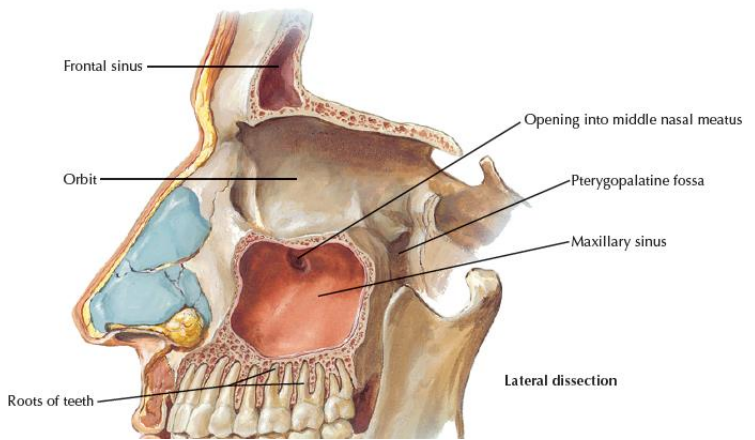
- **Maxillary Sinus:**

- **1st** sinus to develop in-utero.
- Present at birth.
- Biphasic Growth:

1. Birth - 3 years.
2. 7 - 18 years old.

- Largest Paranasal sinus.
- Adult volume 15 ml
- Pyramidal shape occupies body of maxilla.
- Borders:

- o **Anterior wall:** Facial surface of Maxilla.
- o **Posterior wall:** Infratemporal and Pterygoplatine fossae.
- o **Medial wall:** Uncinate process, Fontanelles, Inferior Turbinate
- o **Lateral wall:** Zygomatic Process of Maxilla.
- o **Roof:** Orbital Floor, Infraorbital nerve and vessels.
- o **Floor:** Alveolar and Palatine Processes of Maxilla.



- Roots of All Molars are in close relation to the floor.
 - o Roots of **1st and 2nd Molar** are Dehiscent into the sinus (2%).
- Infraorbital Nerve (**V2**) crosses Orbital floor to exit to Anterior portion of Maxilla via the infraorbital Foramen.
 - o Infraorbital nerve is dehiscent into the sinus (15%)

- Ostium:

- o Situated high up in Medial wall.
- o Opens in Posterior part of Ethmoid Infundibulum into Middle Meatus.
- o It is orientated slightly off set from the parasagittal plane facing posteriorly and is usually around 5mm in diameter.
- o **Accessory ostium** present in 25% mainly in Posterior nasal Fontanelle, Postero-inferior to Natural ostia.

- Blood supply:

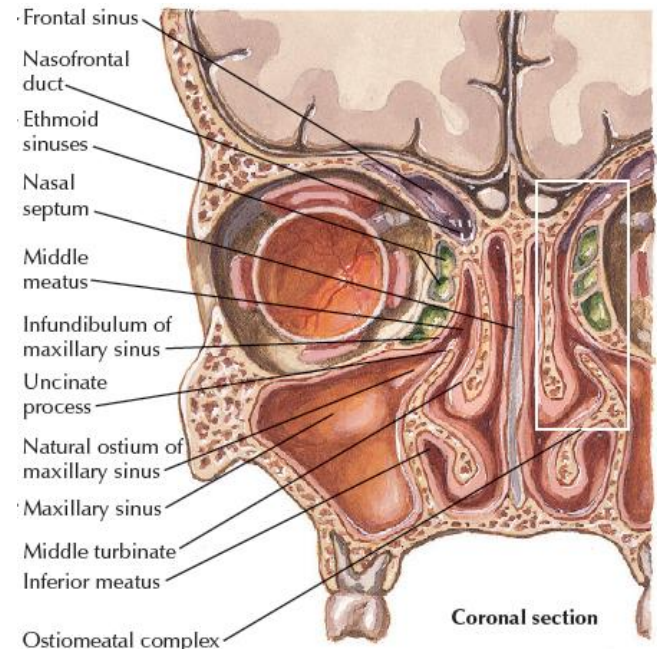
1. Maxillary Artery + Facial Artery
2. Maxillary vein

- Innervation:

1. Infraorbital nerve (**V2**).

- Radiology:

1. 1st Radiologic evidence at age of 4-5 months.
2. Occipito-mental view → **Water's view**.



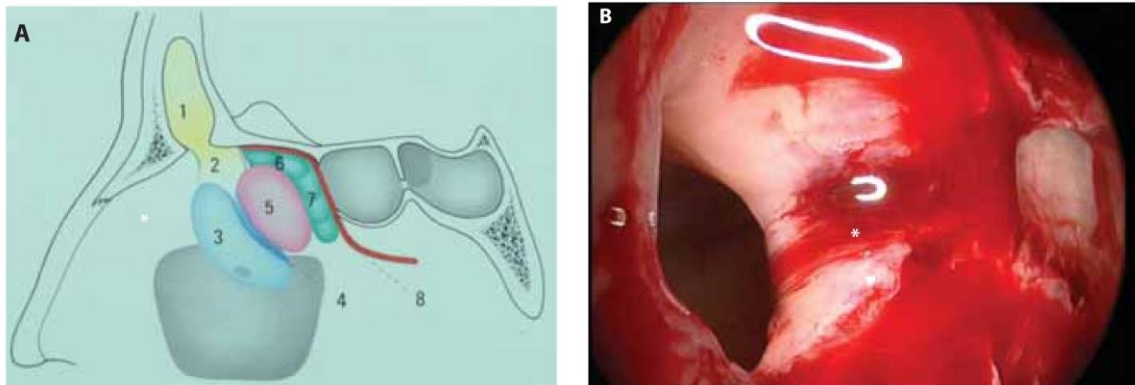


Figure 10. A) Schematic, simplified drawing of structures of the middle meatus after removal of the middle turbinate. 1 = frontal sinus, 2 = frontal recess, 3 = uncinete process over ethmoidal infundibulum, 4 = hiatus semilunaris, 5 = ethmoidal bulla, 6 = suprabullar recess, 7 = retrobullar recess, 8 = basal lamella of middle turbinate. B) Right maxillary sinus ostium (untouched) and transport of secretion over posterior margin (*).

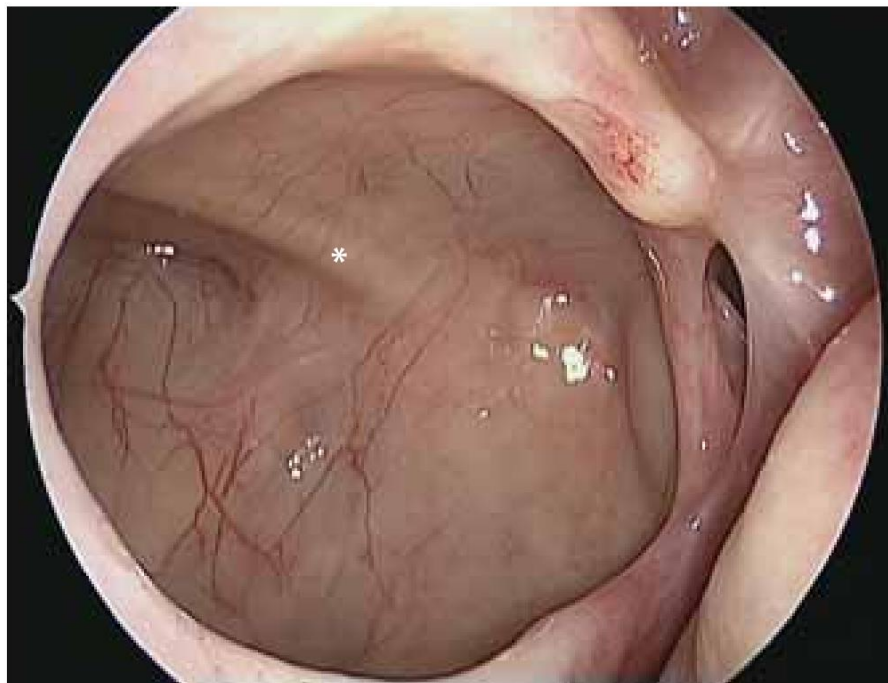
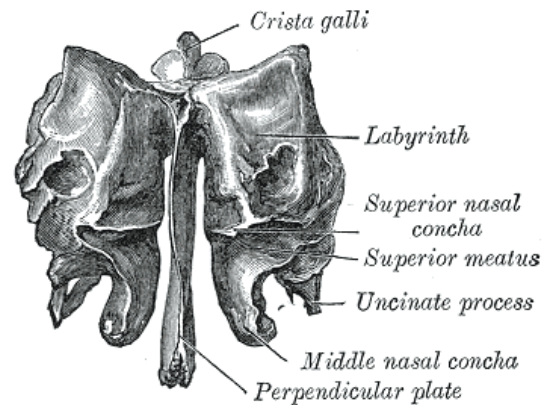


Figure 11. Infraorbital nerve seen through a large middle meatal antrostomy.

- **Ethmoid Bone:**

- Composed of:

1. Perpendicular Plate.
2. Crista galli.
3. Cribriform plate.
4. Bilateral Labyrinths (containing air cells, superior and middle turbinates).



- **Crista Galli:**

- Strong process formed by Upper part of Perpendicular plate above Cribriform plate.
- Falx cerebri attaches to its posterior border, whereas its anterior border is joined to the frontal bone by 2 small alae, completing the margins of the foramen caecum.
- Crista galli is pneumatized in 13% of the patients.

- **Horizontal Cribriform Plate:**

- Perforated by Olfactory nerve fibers, Dura, and Ethmoid vessels and nerves.
- Lies at a lower level than the roof of adjacent Ethmoid labyrinth.
- Slope downward as it passes posteriorly.
- Divided into 3 parts:

1. **Horizontal Medial Portion:**

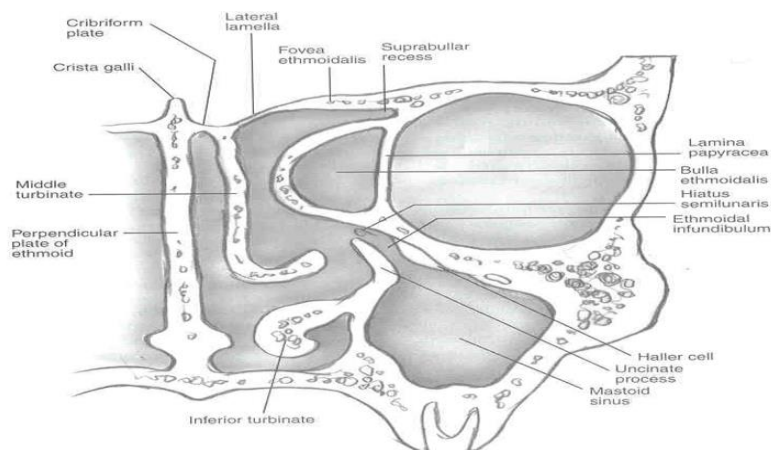
- o Thick.
- o Contains 20 or more foramina for olfactory fibers.

2. **Lateral Lamella:**

- o **Thinnest portion of skull base.**
- o Arises from horizontal segment at an angle to join Orbital Plate of Frontal bone.
- o The angle at which it rises determines the height of Olfactory groove/Fossa (**Keros Classification**).
- o Ethmoidal Arteries enter Olfactory Fossa through an openings in Lateral lamella.

3. **Fovea Ethmoidalis:**

- o Thick Lateral Ethmoid Roof.



- **Olfactory cleft:**
- Part of the superior nasal cavity where the majority of the olfactory epithelium is present.
- Bounded superiorly by the cribriform plate, medially by the superior nasal septum, laterally by the superior part of the medial aspect of the middle turbinate and superior turbinate.



Figure 31. The olfactory cleft (*)

- **Olfactory Fossa:**
- Contains the olfactory bulbs and tracts.
- Bounded inferiorly by the cribriform plate, laterally by the lateral lamella of the cribriform plate and medially by the crista galli.
- Differences in the depths of the olfactory fossa (**Keros Classification**) between right and left side are present in 11% of men compared to 2% of women.

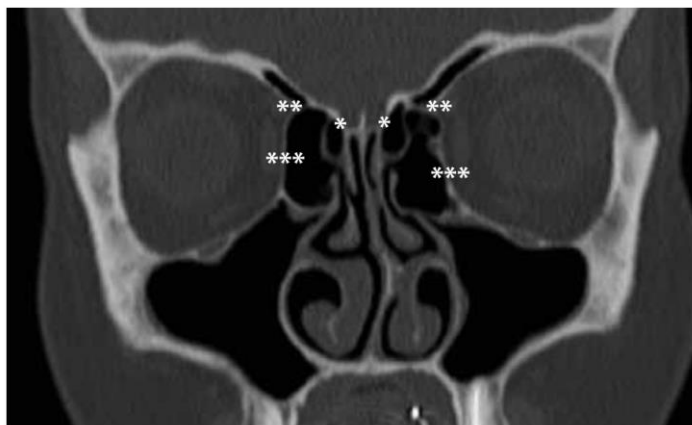


Figure 33. In this case, the lateral lamella (*) is longer, creating a moderately deep fossa (4-7mm)(49%). Anterior ethmoidal artery (**). Lamina papyracea (***).

- **Ethmoid Sinus:**
- **2nd** sinus to develop in-utero.
- Present at birth (3-4 Anterior Ethmoid cells).
- **Most developed sinus at birth.**
- Thin-walled air cavities occupies lateral masses of ethmoid bone.
- Basal lamella of Middle turbinate divides it into Anterior and Posterior group.
- Adult size: 10-15 air cells.
- Adult volume: 2-3 ml.
- **Formed from 5 Basal Lamella: (Relatively constant landmarks)**
 1. Basal Lamella of Uncinate Process.
 2. Basal Lamella of Ethmoid Bulla.
 3. Basal Lamella of Middle Turbinate.
 4. Basal Lamella of Superior Turbinate.
 5. Anterior wall of Sphenoid Sinus.

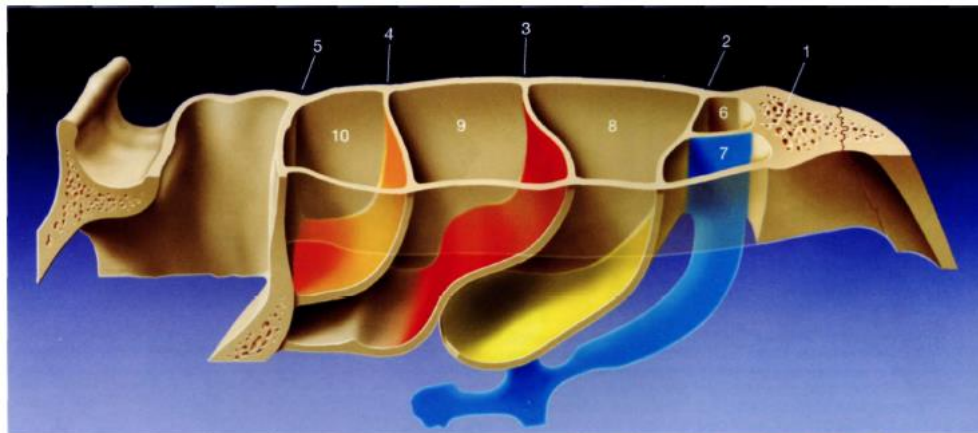
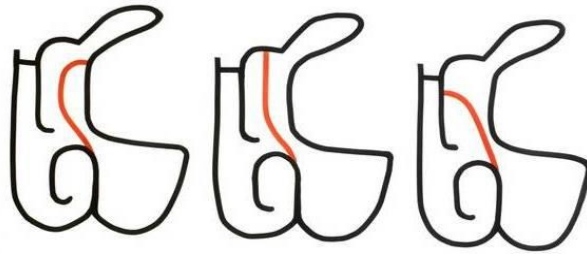


Fig. 1
Diagram of the basal lamellae of the ethmoid bone (after Terrier 1991).

1 Basal lamella of the uncinat process	5 Supreme turbinate (usually absent), anterior wall of the sphenoid sinus
2 Basal lamella of the ethmoid bulla	6 Space in continuity with the ethmoid infundibulum (variable)
3 Basal lamella of the middle turbinate	7 Anterior middle meatus
4 Basal lamella of the superior turbinate	8 Ethmoid bulla
	9 and 10 Posterior ethmoid

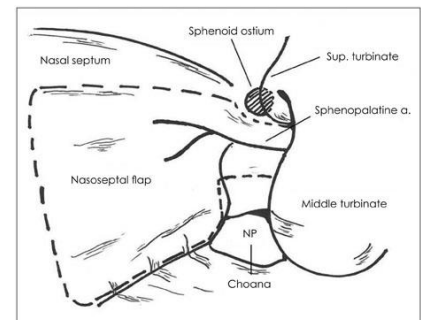
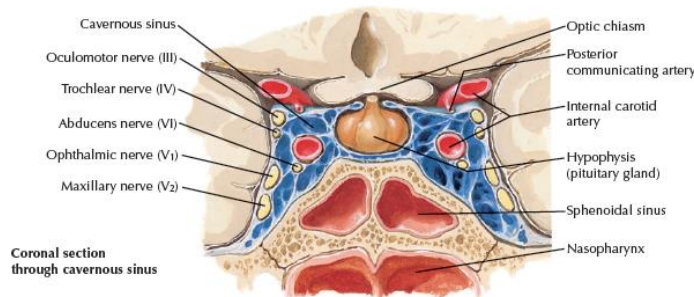
- Roof is formed by:
 - o Lateral lamella and Fovea Ethmoidalis medially.
 - o Orbital plate of the frontal bone laterally.
- Separated from Orbit Laterally by Lamina papyracea.
- Optic nerve forms close relation with Posterior Ethmoid cells.
- Anterior cells Drains into → Infundibulum into Middle Meatus.
- Posterior cells Drains into → Superior Meatus.
- Blood supply:
 1. Anterior Ethmoid + Posterior Ethmoid Artery.
 2. Maxillary + Ethmoid veins (**Cavernous sinus**).
- Innervation:
 1. Anterior Ethmoid + Posterior Ethmoid nerves. (**V1**)
- Radiology:
 1. 1st Radiologic evidence at age of 1 year.
 2. Lateral view.
 3. Occipito-Frontal → **Caldwell view**.

- **Frontal Sinus:**
- Not present at birth.
- Invade frontal bone at age of **4 years**.
- Between outer and inner table of Frontal bone.
- Ratio of table thickness between outer and inner table is 2:1
- Adult volume: 2-3 ml at age of 12-12 years.
- Underdeveloped in **5%**
- Usually 2 Asymmetric Frontal sinuses separated by thin septum.
- Ostium Located Postero-medially in the floor.
- Drainage depends on Superior attachment of Uncinate Process:
 1. Attaches to Lamina papyracea (**Most common**):
 - Drains into Middle Meatus medial to Ethmoidal Infundibulum (between uncinate process and middle turbinate)
 2. Attaches to Ethmoid Roof (Skull base):
 - Drains into Ethmoidal Infundibulum.
 3. Attaches to Middle Turbinate:
 - Drains into Ethmoidal infundibulum.



- Relations:
 - Anterior wall: Skin over forehead
 - Posterior wall: Meninges and Frontal lobe.
 - Floor: Orbit.
- Blood supply:
 - **Supraorbital + Supratrochlear artery**
 - **Ophthalmic (Cavernous sinus) + Supraorbital (Anterior Facial) veins.**
- Innervation:
 - Supraorbital + Supratrochlear nerves (**V1**)
- Radiology:
 - 1st Radiologic evidence at age of 6 year.
 - Lateral view.
 - Occipito-Frontal → **Caldwell view**.

- **Sphenoid Sinus:**
 - Not present at birth.
 - Occupies body of Sphenoid.
 - Usually 2 Asymmetric sinuses separated by thin septum.
 - Reach Sella turcica by age of 7 years.
 - Adult size at 12-18 years
 - Adult volume: 0.5-8 ml.
- Ostium:
 - o Situated in Upper part of face of sphenoid at the level of inferior one third of the superior turbinate and along a horizontal plane through floor of the orbit.
 - o Located medial to posterior end of superior turbinate in 80% and laterally in 20%.
 - o Drains into Sphenoethmoidal Recess.
- Anatomical Relationships of Sphenoid Ostium:
 - o 30 degree from Nasal Cavity floor. (Most reliable)
 - o Adjacent to Posterior border of nasal septum.
 - o 6-8 cm Posterior to Anterior nasal spine.
 - o 1.5 cm Above Choanal floor
 - o 1/3 up from Choana to Skull base.
- Relations:
 - o **Roof:** Pituitary gland, Optic chiasm.
 - o **Floor:** Nasopharynx, Pterygoid canal.
 - o **Medial:** other sphenoid bone
 - o **Lateral wall:** Cavernous sinus, Internal Carotid Artery, CN- II, III, IV, V1, V2, and VI



- Anterior wall of sphenoid (Face of sphenoid) is often thin and is crossed inferiorly by the posterior nasal artery (septal branch of the sphenopalatine artery).
- Blood supply:
 - o Sphenopalatine Artery.
 - o Maxillary vein.
- Innervation:
 - o Sphenopalatine nerve.
- Radiology:
 - o 1st Radiologic evidence at age of 6 year.
 - o Lateral view.
 - o Submento-vertex → **Basal view.**

- **Prominences inside sphenoid sinus:**

- Lateral wall:
 - **Optic nerve:**
 - Present in 40% of patients with dehiscence in 6%.
 - **Internal carotid artery:**
 - Present in 65% of patients with dehiscence in 25%.
 - **Maxillary nerve (V2).**
- Floor:
 - Vidian nerve of the pterygoid canal.

- **Recesses inside sphenoid sinus:**

- Lateral Recess:
 - Results from pneumatisation of pterygoid process.
- Optico-carotid recess (OCR):
 - Lies on posterolateral wall of sphenoid sinus between optic nerve and internal carotid artery.
 - Results from pneumatisation of anterior clinoid process.

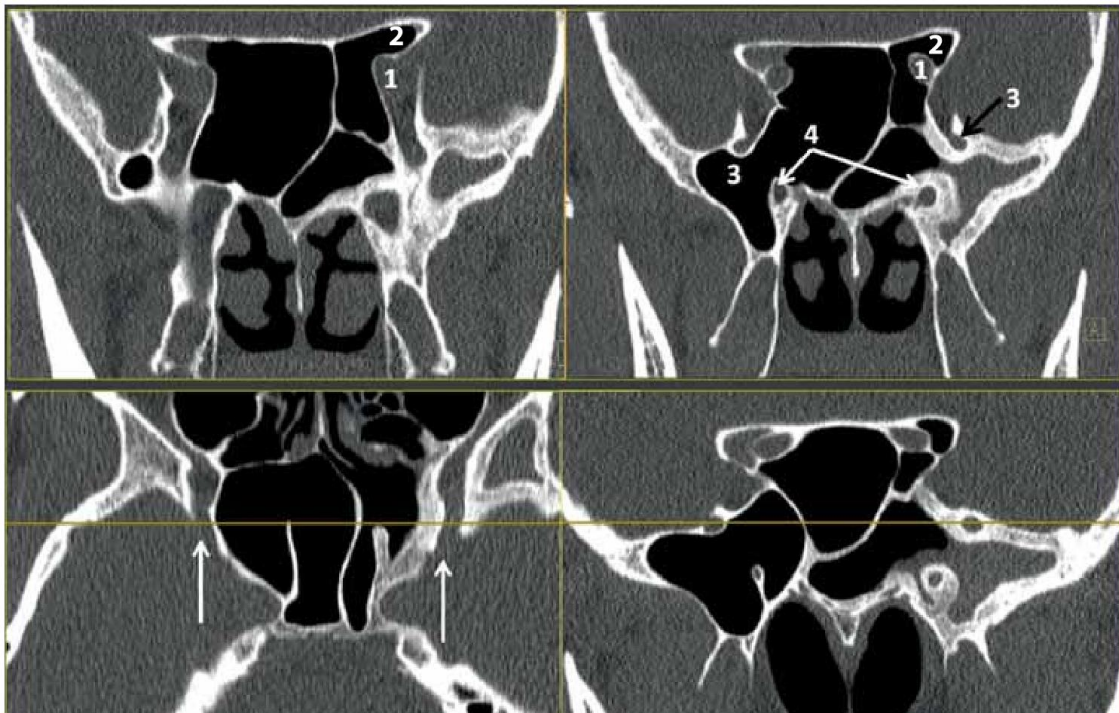


Figure 43. Complex sphenoid anatomy with extensive pneumatisation. Clockwise: 1 = (bulge of) optic nerve, 2 = pneumatized clinoid process. Please note, that in contrast to an optico-carotid recess, here pneumatisation towards the anterior clinoid goes superior to the optic nerve, 3 = foramen rotundum, 4 = Pterygoid (Vidian) nerve. When the axial CT level is placed through the foramen rotundum bilaterally, the corresponding canals can be seen on either side (arrows). Note "crab eye" appearance of (Vidian) nerve in pterygoid canal. There is bone thickening following long standing chronic sphenoiditis on the left side.

Figure. An endoscopic picture showing both the sphenoid sinus cavities. CP, carotid protuberance; CR, clival recess; IS, intersphenoid septum; mOCR, medial opticocarotid recess; OCR, lateral opticocarotid recess; OP, optic protuberance; PS, planum sphenoidale; SF, sellar floor

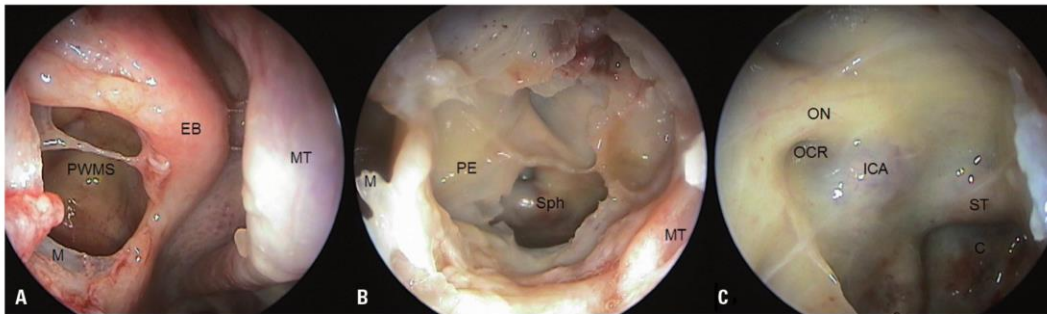
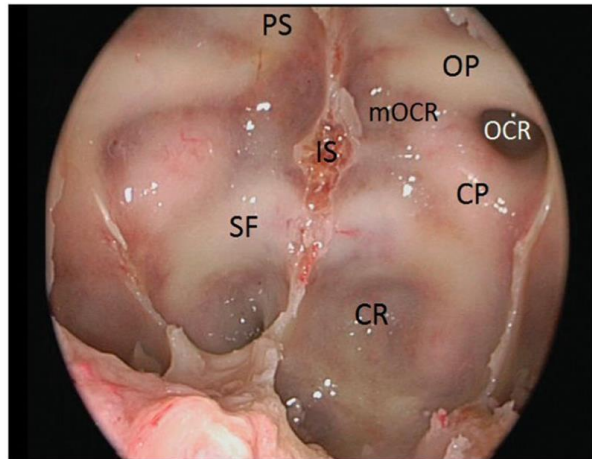


Fig 1. [A] Endoscopic view of the right nasal cavity with 0° telescope after the removal of the uncinate process and opening of the ethmoidal bulla (EB). The maxillary antrostomy (M), posterior wall of the maxillary sinus (PWMS) and middle turbinate can be observed. [B] Transthemoidal approach to the sphenoid sinus (Sph), labeled the maxillary antrostomy (M), middle turbinate (MT) and posterior ethmoid (PE). [C] Structures on the posterolateral wall in the sphenoid sinus: optic nerve (ON), prominence of the internal carotid artery (ICA), optic-carotid recess (OCR), sella turcica (ST) and clivus (C).

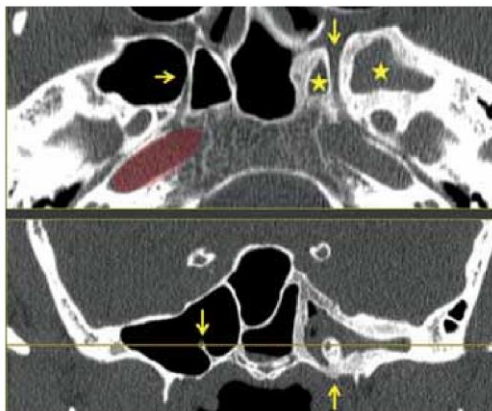


Figure 44. Axial cut through the level of the pterygoid canals with nerve and artery (arrows). Note the relationship to the horizontal carotid (shaded red on the right), just where this curves up into its vertical paraclival segment. Asteriks: Opacified lateral recess of sphenoid on the left.



Figure 46. Coronal CT cut through sphenoid sinuses. Internal carotid artery (*), optic nerve (**) and optico-carotid recess (***).

- **Canals associated with sphenoid sinus:**

- Pterygoid (Vidian) canal:
 - Runs anteriorly from foramen lacerum through the sphenoid to open into pterygopalatine fossa.
 - Transmits nerve of the pterygoid canal composed of the great petrosal nerve and the deep petrosal nerve together with autonomic fibers.
 - Its position relative to sphenoid sinus is dependent on the pneumatization of the sinus, may be encased in the basisphenoid bone, partially protruding into the sinus floor or occasionally exposed within the sinus cavity and connected to the floor by a bony stalk.
- Palatovaginal canal:
 - Bony canal located in the floor sphenoid sinus, medial to pterygoid (vidian) canal.
 - Contains the pharyngeal branch of the maxillary nerve and the pharyngeal branches of the maxillary artery.
 - Results in excessive bleeding from floor of the sphenoid sinus during endoscopic skull base surgeries.
- Vomerovaginal canal:
 - Small, inconsistent canal.
 - Lies medial to the palatovaginal canal, and lead into the anterior end of the palatovaginal canal.
 - When present, it may contain a branch of the sphenopalatine artery.
- Lateral craniopharyngeal canal (Sternberg's canal):
 - Congenital bony defect in the lateral wall of sphenoid sinus.
 - Results from failure of fusion of the greater wing of the sphenoid and the presphenoid.
 - Located in the posterior part of the lateral sphenoid sinus wall, lateral to the maxillary nerve (V2).
 - Present in young children but only 4% of adults and is associated with extensive sphenoidal pneumatization.
 - Presence of this canal in combination with a raised intracranial pressure may lead to the extrusion of intracranial contents and/or 'spontaneous' CSF rhinorrhea.

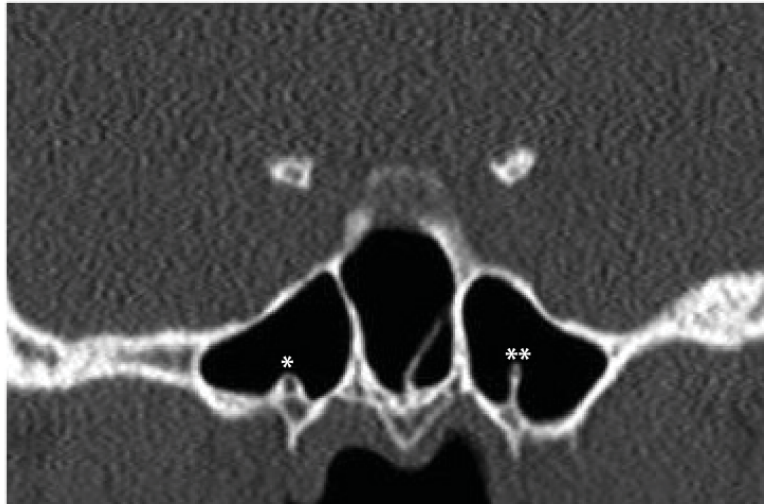


Figure 48. The pterygoid canal (*) runs anteriorly from the foramen lacerum through the sphenoid to open into the pterygopalatine fossa. The nerve may be encased in the basisphenoid bone (*), partially protruding into the sinus floor or occasionally exposed within the sinus cavity and connected to the floor by a bony mesentery (**).

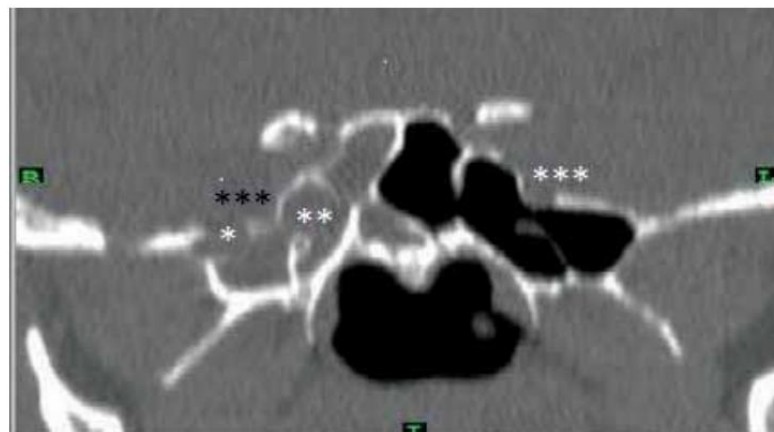


Figure 50. Lateral craniopharyngeal canal (formerly Sternberg's canal) is a congenital bony defect (*) in the lateral wall of the sphenoid sinus (**). This canal is located in the posterior part of the lateral sphenoid sinus wall, lateral to the maxillary nerve (V2) (***). Large meningo-encephalocele protruding through defect into right sphenoid sinus.

- **Nasal cavity Blood Supply:**
- Nose is richly supplied by both External (mostly) and Internal Carotid systems, both on Septum and Lateral walls.

- Internal Carotid Artery:

1. Ophthalmic Artery :

- ### 1. Anterior Ethmoid Artery:

Supply Anterior part of Lateral wall and septum.

- ## 2. Posterior Ethmoid Artery:

Supply part of lateral wall over Superior turbinate and Postero-superior portion of septum.

- External Carotid Artery:

1. Maxillary Artery :

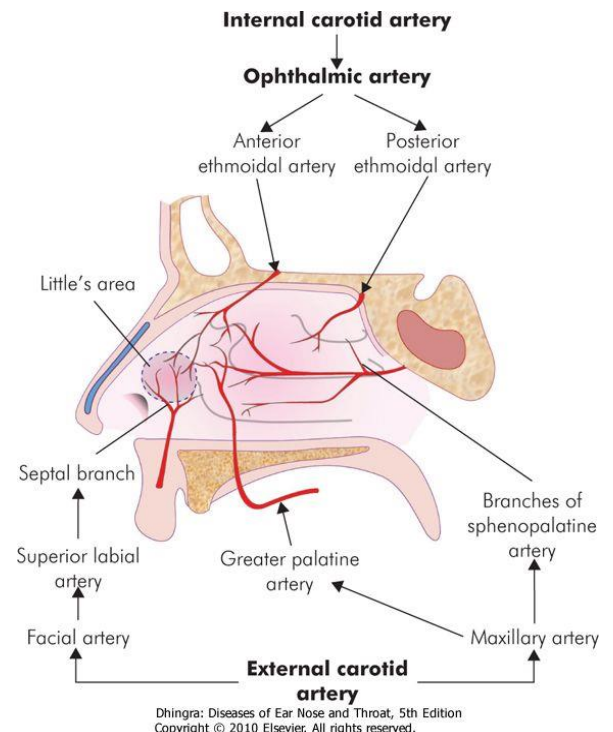
- 1. Sphenopalatine Artery:** Pass through Sphenopalatine foramen
Posterior to Middle turbinate.

- 2. Greater Palatine Artery:**
Branch of Descending Palatine Artery, pass through Incisive canal.

2. Facial Artery :

- ### 1. Superior Labial Artery:

Major blood supply to Anterior part of Nasal septum and Ala.



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- **Little's Area:**

- Most common site for Epistaxis (90%).
- Situated at Anterior Inferior part of Nasal Septum.
- Located 1.5 cm Posterior to Mucocutaneous junction above Vestibule.
- Susceptible to bleeding due to Fragile mucosa and Tight Adherence to underlying mucosa affording little resistance to mechanical stress.
- Exposed frequently to Drying effect of inspiratory current and to finger nail trauma.

- **Kiesselbach's Plexus formed by Anasotmosis of:**

1. Anterior Ethmoidal Artery (**ICA**)
2. Septal branch of Superior Labial Artery (**ECA**)
3. Septal branch of Sphenopalatine Artery (**ECA**)
4. Greater Palatine Artery (**ECA**)

- **Woodruff's Area:**

- Common source of Posterior Nasal bleeds.
- Situated under Posterior end of Inferior Turbinate.

- **Contributing Arteries:**

1. Sphenopalatine Artery (ECA)
2. Ascending Pharyngeal Artery (ECA)

- **Anterior Ethmoid Artery (AEA):**
- Large and singular.
- Originates from Ophthalmic Artery in Orbit.
- Passes between superior oblique and medial rectus muscles, penetrates lamina papyracea through the anterior ethmoidal foramen into the anterior ethmoidal cells.
- Crosses anterior ethmoidal complex from posterolateral to anteromedial at the junction of ethmoid roof and posterior border of Frontal recess.
- Passes either at level of skull base (Most common) or as much as 5mm below this level in a mucous membrane mesentery or a thin bony lamella (dehiscent in 40%).
 - o In presence of supraorbital recess, the artery is very likely to be exposed in its posterior margin.
- The artery then enters the anterior cranial fossa through the lateral lamella of the cribriform plate, which is the weakest portion of skull base.
- Eventually supplying Cribiform Plate and Anterior septum.
- CT landmarks for identifying the location of AEA:
 1. Sulcus of anterior ethmoidal artery (nipple or pyramidal sign):
 - The only well defined corticated break in the anterior lamina papyracea.
 2. The plane at the posterior globe and last 0.5 cm of crista galli.
 3. The coronal plane where the superior oblique and the medial rectus muscle are at their largest diameters.

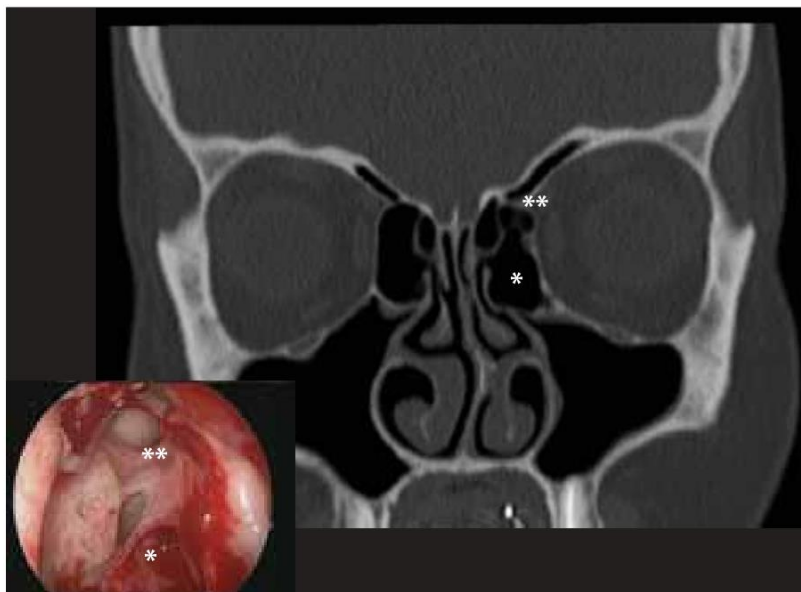


Figure 17. Ethmoidal bulla (*) and relationship to anterior ethmoidal artery (**).

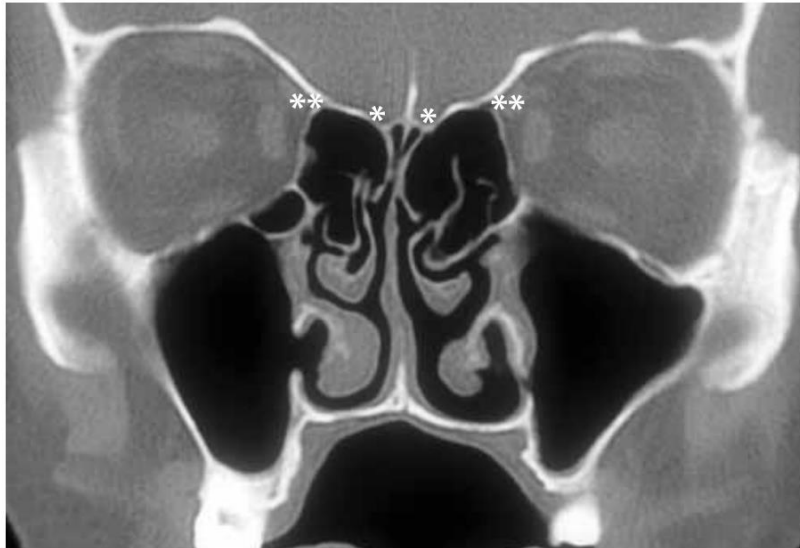


Figure 32. The lateral lamella (*) of the cribriform plate is one of the thinnest parts of the skull base. In this case, the lateral lamella is very short rendering the olfactory fossa almost flat (1-3mm) (30%). Anterior ethmoidal artery (**). Note previous inferior meatal antrostomies.



Figure 2. Anterior ethmoidal sulcus - bony sulcus (tip of arrows) on the lateral walls of the olfactory fossae, corresponding to the anterior ethmoidal sulci.

- **Posterior Ethmoid Artery:**
- Smaller and branched.
- Originates from Ophthalmic Artery in Orbit.
- Passes through the posterior ethmoidal canal into the anterior cranial fossa.
- Courses through Posterior Ethmoid cells within skull base, in front of the most superior aspect of face of sphenoid.
- Less vulnerable during surgery as it is almost never found below the level of the skull base.
- Supplies Posterior Ethmoid Sinuses, part of Superior and Middle Turbinates and small amount of Posterior septum.
- It's position is associated with position of Optic nerve near orbital vertex.

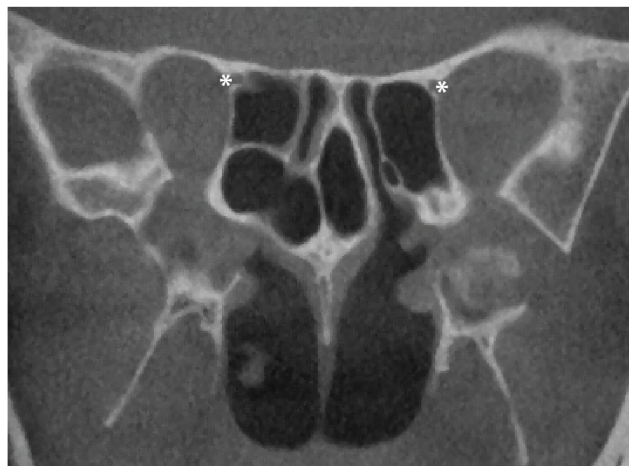
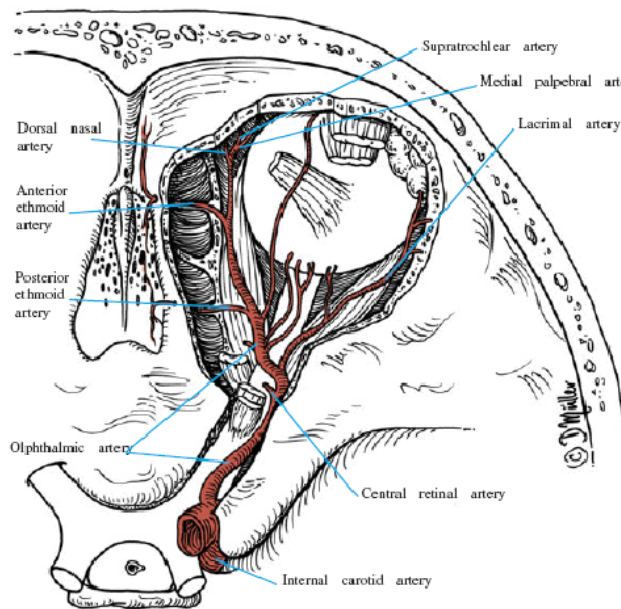


Figure 37. The posterior ethmoidal artery usually crosses within the ethmoidal roof, in front of the most superior aspect of the anterior wall of the sphenoid sinus. In 25-50%, the corticated sulcus of this artery (*) is identifiable on the coronal CT examination.

- **Venous system:**

- Anterior Ethmoid vein + Posterior Ethmoid vein → Ophthalmic vein → **Cavernous sinus**
- Sphenopalatine vein → 1- Maxillary vein → Internal Jugular vein
→ 2- Cavernous sinus
- Greater palatine vein → 1- Posterior Facial vein → External jugular vein
→ 2- Cavernous sinus

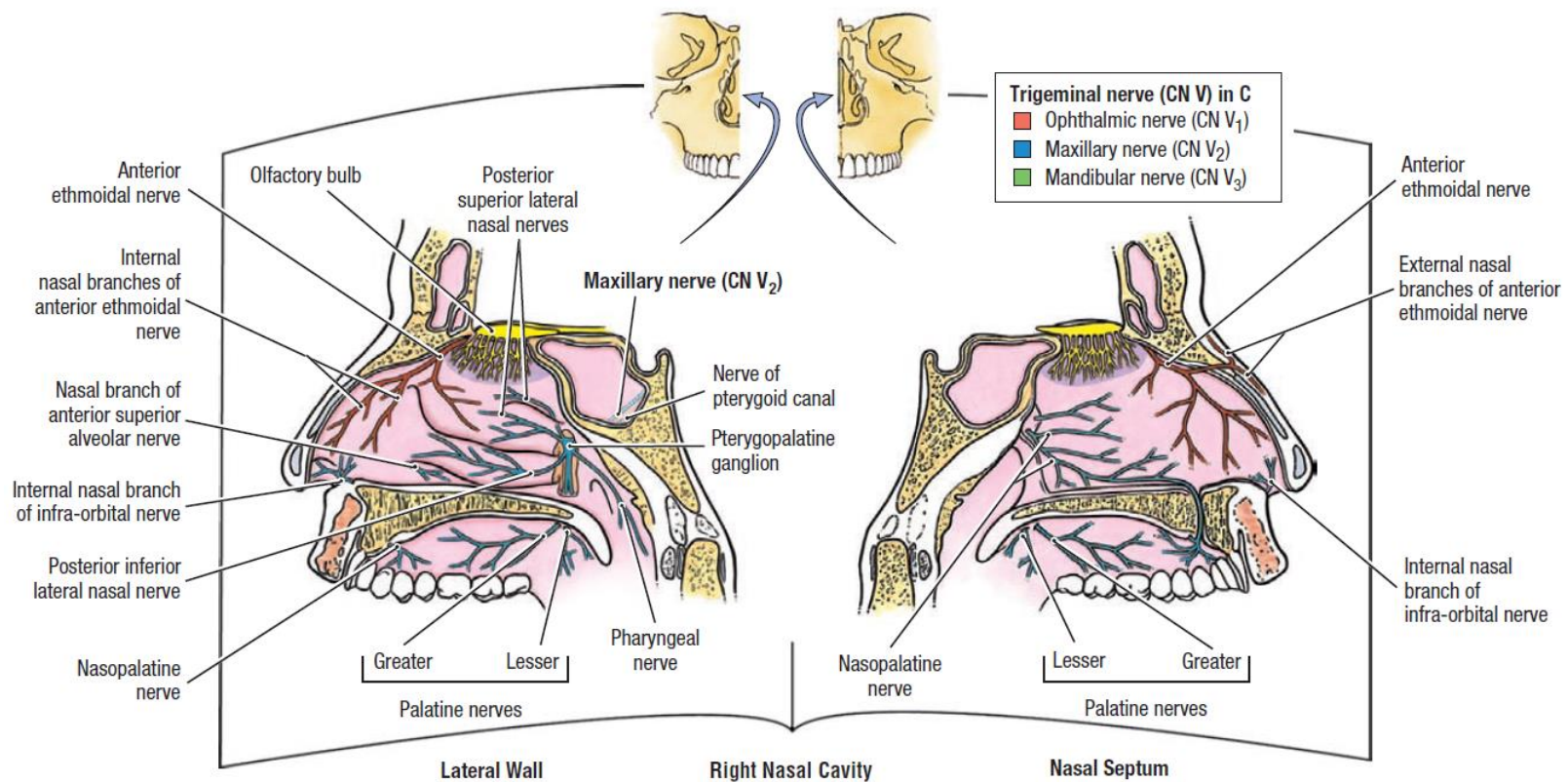
- **Retrocolumellar vein:**

- Runs vertically downwards just behind the columella.
- Crosses floor of nose and joins venous plexus on Lateral nasal wall.
- Common site of venous bleeding in young people.

- **Nerve Supply:**

- General Sensation:

1. Anterior Ethmoid Nerve (**V1**) → Antero-superior Nasal cavity.
2. Posterior Ethmoid Nerve (**V1**) → Posterior Nasal cavity.
3. Sphenopalatine Nerve (**V2**) → Postero-inferior Nasal cavity.
4. Infra-orbital Nerve (**V2**) → Vestibule



- **Special Sensation:**
- **Olfactory nerves (CN-I)**
 - Carry sense of smell.
 - Supply Olfactory Region of nose.
 - Central filaments of the olfactory cells arranged into 12-20 nerves.
 - Pass through Cribriform plate and end in Olfactory bulb.
 - Carry sheaths of dura, arachnoid and pia with them into the nose.
 - Injury to these nerves can open CSF space leading to CSF rhinorrhoea or meningitis.
- **Autonomic Supply:**
- **Parasympathetic Fibers:**
 - From **Greater Superficial Petrosal Nerve (CN-VII)** travel in Nerve of Pterygoid Canal (Vidian nerve).
 - Relay in Sphenopalatine Ganglion before reaching Nasal cavity.
 - Supply Nasal glands and Control Nasal Secretion.
 - Supply Blood vessels of Nose and cause Vasodilation.
- **Sympathetic Fibers:**
 - From T1-2, pass through Superior Cervical Ganglion.
 - Travel in **Deep Petrosal Nerve** and join Parasympathetic fibers of Greater Petrosal Nerve to form Nerve of Pterygoid Canal (Vidian nerve).
 - Reach Nasal cavity without relay in Sphenopalatine Ganglion.
 - Stimulation causes vasoconstriction.
 - Excessive rhinorrhoea in cases of Vasomotor and Allergic Rhinitis can be controlled surgically by Section of Vidian Nerve.
- **Lymphatic Drainage**
- External Nose + Anterior Nasal Cavity → Submandibular LN.
- Posterior Nasal cavity → Retropharyngeal LN → Upper Jugular LN.

Normal Anatomical Variations:

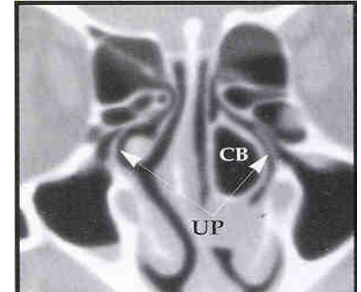
- NASAL SEPTUM:

1. **Nasal Septum Deviation (70%)**
2. **Nasal Septum Spur (25%)**
3. **Nasal Septum Pneumatization (2%)**

- MIDDLE TURBINATES:

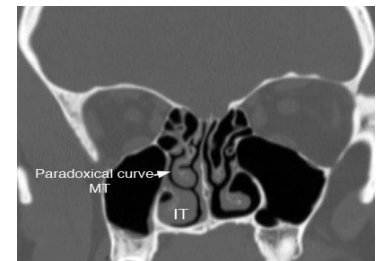
1. Concha Bullosa: (25%)

- Pneumatization of Middle Turbinate by Extension of Ethmoid sinus cells.
- Causes Middle Meatus or Infundibulum Obstruction.
- Related to deviation of Nasal septum to Contralateral side.
- **Excision of Lateral wall** relieves the obstruction.



2. Paradoxical Turbinate: (7%)

- Convexity of the middle turbinate is directed towards Medial wall of Maxillary sinus.



- UNCINATE PROCESS

- Variations of the uncinate process include:
 - o Medialized.
 - o Everted (paradoxical).
 - o Aerated (uncinate bulla).
 - o Lateralized, concave uncinate (narrow the infundibulum leading to an atelectatic infundibulum).
- Variations in Superior attachment of Uncinate Process are classified according to Criteria developed by Landsberg & Friedman to:
 1. **Type 1: Into Lamina Papyracea (50%)**
 2. **Type 2: Into Posterior wall of Agger Nasi cell (20%)**
 3. **Type 3: Into Lamina Papyracea and Junction of Middle Turbinate with Cribriform plate (15%)**
 4. **Type 4: Into junction of Middle Turbinate with Cribriform plate (10%)**
 5. **Type 5: Into Skull base (4%)**
 6. **Type 6: Into Middle Turbinate (2%)**

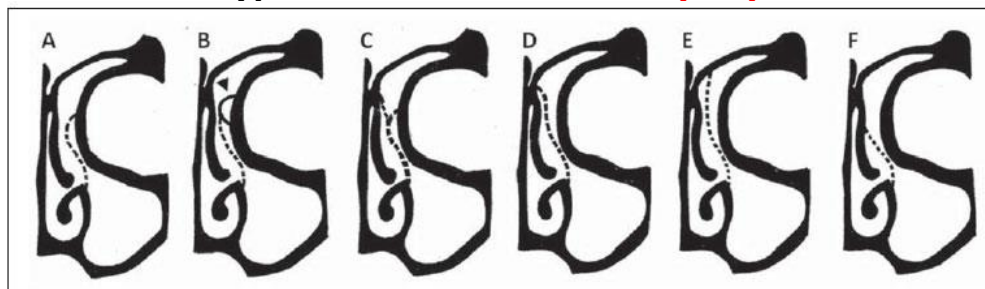


Figure 5. Landsberg & Friedman classification of superior uncinate process insertion. **A:** Type 1 (insertion into the lamina papyracea). **B:** Type 2 (insertion into the posterior wall of agger nasi cell). **C:** Type 3 (insertion into the lamina papyracea and junction of the middle turbinate with the cribriform plate). **D:** Type 4 (insertion into junction of the middle turbinate with the cribriform plate). **E:** Type 5 (insertion into the skull base). **F:** Type 6 (insertion into the middle turbinate).

- **ETHMOIDAL BULLA:**

1. **Tours Lateralis: (8%)**

- Poorly or completely Non-pneumatized Ethmoidal Bulla.

2. **Tours Ethmoidalis:**

- Persisting and Non-pneumatized second Basal Lamella.

- **CRIBRIFORM PLATE:**

- Depth of Olfactory fossa is determined by Height of Lateral lamella of Cribriiform plate.
- **Keros Classification of Olfactory Fossa:**
 - **Type 1:** Depth of 1-3 mm **(25%)**
 - **Type 2:** Depth of 4-7 mm **(70%)**
 - **Type 3:** Depth of 8-16 mm **(0.5%)**
 - **Type 4:** Depth of >16 mm or Asymmetric.
- The higher the Keros grade, the greater the chance of injury of the cribriform plate and olfactory fossa, with consequential risk for iatrogenic cerebrospinal fluid fistula and olfactory impairment.

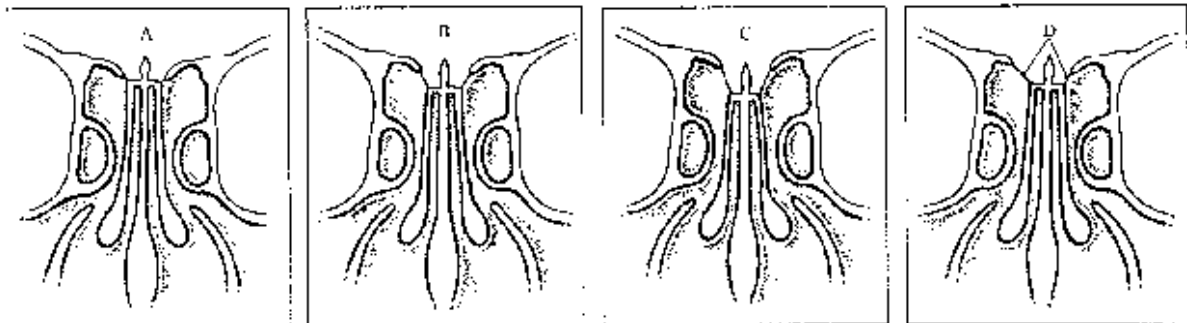


Fig 6. Configurations of ethmoid roof. Three skull base formations are A) type 1, in which olfactory fossa is 1 to 3 mm deep, B) type 2, in which it is 4 to 7 mm deep, and C) type 3, in which it is 8 to 16 mm deep. D) Asymmetric skull base is anatomic variant that should be identified with preoperative computed tomography to avoid skull base injury. Note thin lateral lamella of cribriform plate.

ETHMOID CELLS

1. **Haller cells: (15%)**

- Infraorbital Ethmoid cells.
- Located Anteriorly to Ethmoid bulla, along Orbital floor.
- Adjacent to Natural ostium of Maxillary sinus.
- May cause Mucociliary drainage obstruction.

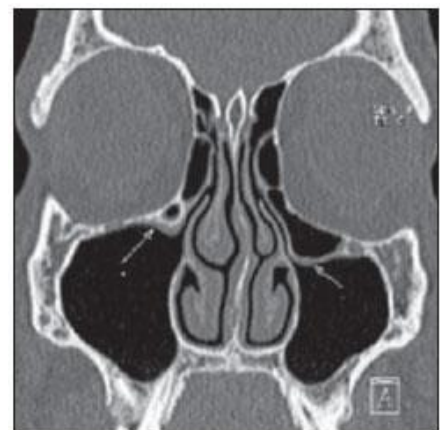


Figure 8. Haller cell. Bilateral infraorbital ethmoid cells, larger at right.

2. Onodi cells: (10%)

- Lateral and Posterior pneumatization of Most Posterior Ethmoid cells.
- Located Supero-lateral to Sphenoid sinus.
- Commonly mistaken as sphenoid cells.
- Optic Nerve & Internal Carotid Artery may be exposed within or lie immediately Lateral to Onodi cells.
- During FESS, attempts to localize Sphenoid Sinus via instrumentation through Posterior most Ethmoidal air cells can lead to Optic nerve, and even, Internal Carotid Artery injury.

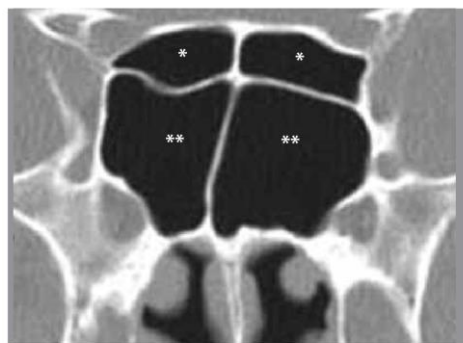
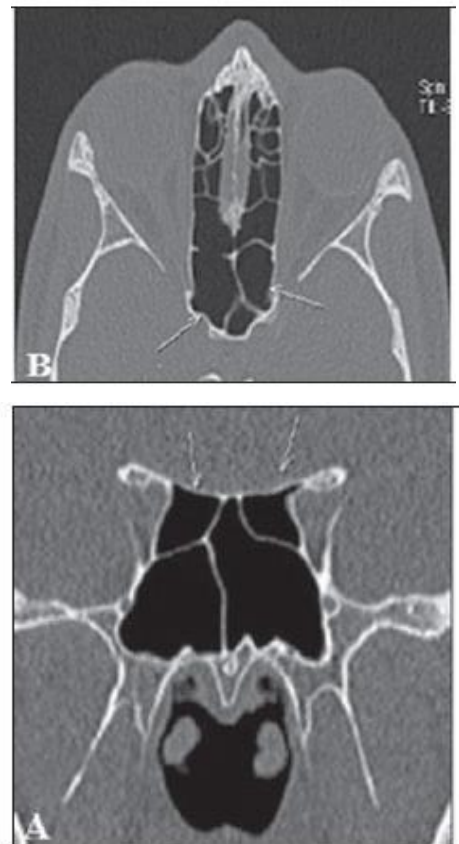
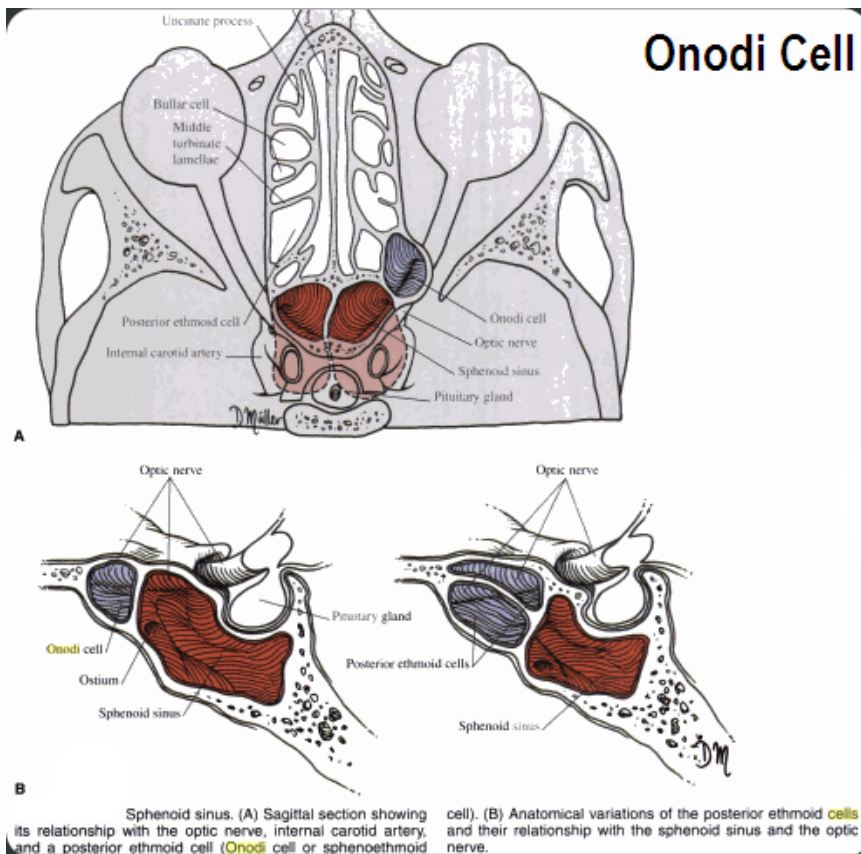


Figure 55. Sphenothmoidal (Onodi) cell (*) and sphenoid sinus (**).

3. **Bulla Frontalis: (20%)**

- Also called **Frontal Cells**
- Anterior Ethmoid cell invades Frontal Recess or Sinus.

1. **Type I (37%):**

- Single Frontal Recess cell Above Agger Nasi cell

2. **Type II (20%):**

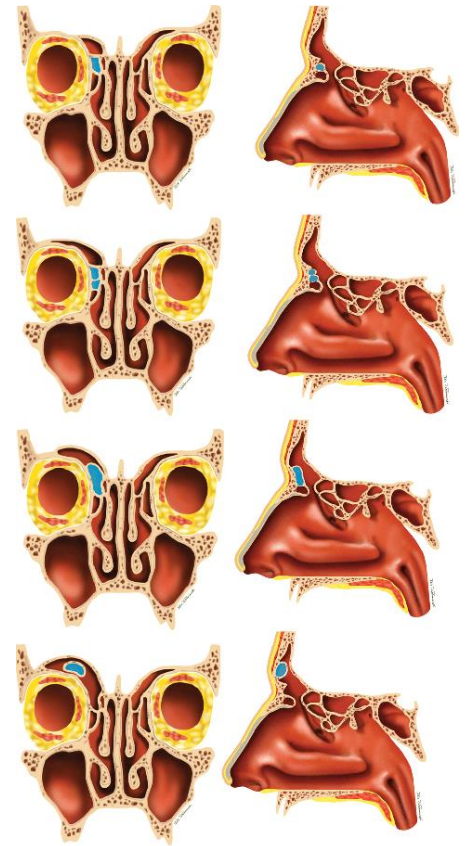
- Two or more cells in Frontal Recess above Agger nasi cell

3. **Type III (10%):**

- Single Massive cell above Agger nasi and Pneumatizing into Frontal sinus

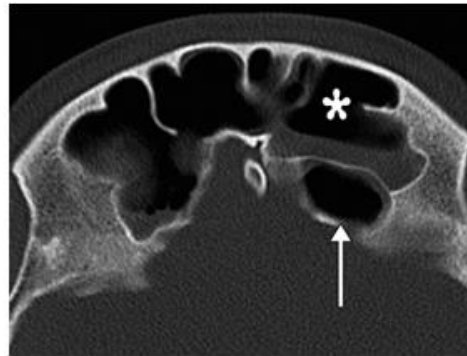
4. **Type IV (5%):**

- Single isolated cell within Frontal sinus.



4. **Supraorbital Ethmoidal cells: (15%)**

- Pneumatization of Orbital Plate of Frontal bone by Ethmoid cells.



- SPHENOID SINUS:

- Types of Pneumatization (Hamberger Classification):
 - 1. Sellar (85%):**
 - Pneumatization extending beyond Sella Turcica.
 - 2. Presellar (10%):**
 - Pneumatization extends upto Anterior part of Sella Turcica.
 - More common in children
 - 3. Conchal (3%):**
 - Limited pneumatization not extending to Sella Turcia.
 - More common in children

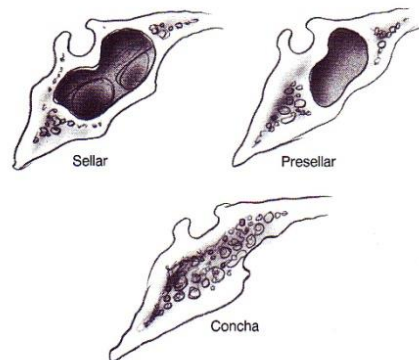


Figure 26.5 The Hamberger classification of sphenoidal pneumatization.

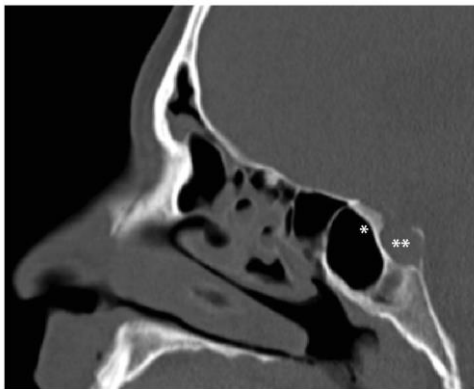


Figure 41. Sphenoid sinus: A pre-sellar sinus extends as far as the anterior bony wall (*) of the pituitary fossa (**).



Figure 42. Sphenoid sinus (*) that extends posterior to the pituitary fossa (**). Clivus (***).

Physiology of Nose:

- Functions of Nose:

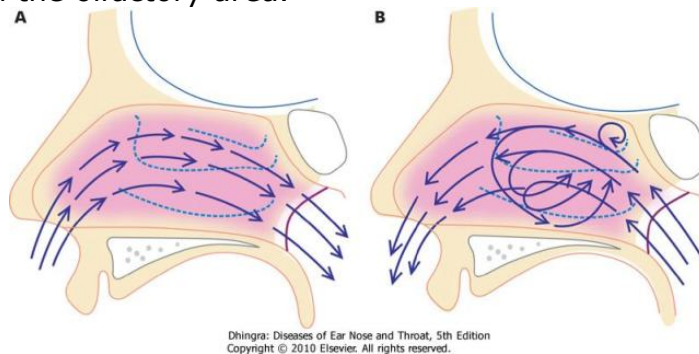
1. Respiration.
2. Air conditioning.
3. Protection of lower airway.
4. Vocal resonance.
5. Olfaction.

RESPIRATION:

- Nose is the natural pathway for breathing.
- Mouth breathing is an acquired act through learning.
- Nose permits breathing and eating to go on simultaneously.

- During quiet Inspiration:

- Inspiratory air current passes through middle part of nose between Turbinates and Nasal septum.
- Very little air passes through Inferior Meatus or Olfactory Region of nose.
- Weak odorous substances have to be sniffed before they can reach the olfactory area.



- During Expiration:

- Expiratory air current follows same course as during inspiration.
- However, Entire air current isn't expelled directly through Nares.
- Friction offered at Internal Nasal Valve (Limen Nasi) converts it into eddies under cover of Inferior and Middle Turbinates and this ventilates Sinuses through Ostia.

- Nasal Cycle:

- Found in 80% of population.
- Nasal airflow is regulated by volume of venous sinusoids in nasal erectile tissue located mainly in Inferior turbinate.
- Nasal mucosa undergoes rhythmic cyclical congestion and decongestion.
- When one Nasal chamber is working, total nasal respiration equal to that of both nasal chambers, is carried out by it.
- Nasal cycle varies every 2½-4 hours.
- Total nasal resistance remains constant throughout the cycle.

- **Regulation of Nasal microvasculature:**
- **Sympathetic:**
- Provides vasoconstrictor tone to Arteries and Capacitance veins.
 1. Norepinephrine (Mainly)
 2. Neuropeptide Y (Weak)
 3. Avian pancreatic polypeptide APP
- **Parasympathetic:**
- Controls Secretions and provides Vasodilatation.
 1. Acetylcholine (Mainly)
 2. Vasoactive intestinal peptide VIP
 3. Peptide histamine isoleucine PHI
- **Airflow Resistance:**
- Nose accounts for 50% of Total Airway Resistance.
- **Areas of Airway Resistance within Nose:**
 1. **Nasal Vestibule:**
 - Provide 1/3 of resistance.
 - Susceptible to collapse from -ve pressure during inspiration.
 - Facial muscles attached to Nasal vestibule contract during inspiration to prevent collapse
 2. **Internal Nasal Valve:**
 - Narrowest part with Cross-sectional area of 0.73 cm²
 - Highest resistance.
 - Susceptible to congestion from septal and inferior turbinate venous sinusoids.
 3. **Turbinated Nasal cavity:**
 - Minimal resistance compared to others
- **Nasal Airflow Controlling factors:**
 1. Autonomic Regulation.
 2. Nasal cycle.
 3. Exercise (Epinephrine release causes decongestion)
 4. Nitric oxide
 5. Head and body position (altered venous pressure)
 6. Sex hormones (pregnancy, puberty, and menstruation lead to increased nasal obstruction)
- **Rhinomanometry**
- Measure Nasal Resistance (pressure and airflow).
- Normal value of Nasal Resistance in Adults **0.25 Pa/cm³/s.**
- For a patient to be symptomatic, Total Nasal Resistance should be **>0.3 Pa/cm³/s.**

- **Anterior Rhinomanometry:**
- Measures Nasopharyngeal pressure and flow through **Trans-nasal** probe into Nasopharynx from one nostril at a time while closing other nostril.
- **Limitations:**
 1. Septal perforations.
 2. Incomplete occlusion.
 3. Floppy septum.
 4. Nasal cycle.
 5. Non-physiologic to do one side at a time.

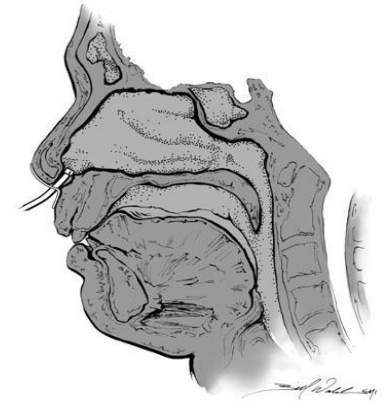


Illustration by William E. Walsh, CMI; Northwestern University Medical Student and Certified Medical Illustrator

- **Posterior Rhinomanometry:**
- Measures Nasopharyngeal pressure and flow with a pressure detector placed **Trans-orally** to Nasopharynx with Bilateral nasal flow.
- **To differentiate between Mucosal Hypertrophy and structural defects causing Nasal obstruction Using Rinometry:**
 - o Nose should be decongested.
 - o **If Less than 35%** improvement in Resistance → Structural defect.

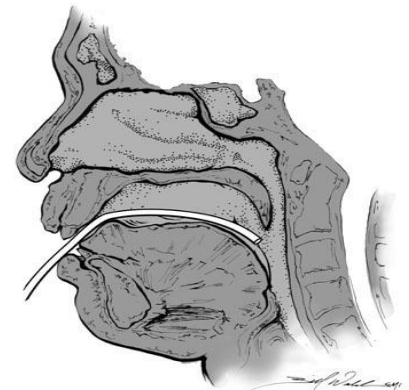


Illustration by William E. Walsh, CMI; Northwestern University Medical Student and Certified Medical Illustrator

- **Acoustic Rhinometry**
- Indirectly measure Nasal Cross-sectional area.
- Acoustic pulses enter Nasal passage through a nose piece, impact nasal structures, and are reflected back to a microphone.

AIR-CONDITIONING OF INSPIRED AIR:

- Nose is Air-conditioner for lungs.
- Filters and purifies inspired air and adjusts its temperature and humidity.
- **Temperature control:**
- Regulated by large surface of nasal mucosa.
- Middle and Inferior Turbinates and adjacent parts of Septum are highly vascular with cavernous venous spaces or sinusoids which control blood flow, and size of Turbinates.
- This also makes an efficient "Radiator" mechanism to warm up the cold inspired air to near body temperature (**37°C**).

- Humidification:
- Nasal mucous membrane adjusts the relative humidity of the inspired air to 85% or more.
- Water is provided by nasal mucous membrane which is rich in mucous and serous secreting glands.
- About 1000 ml of water is evaporated from the surface of nasal mucosa in 24 hours.

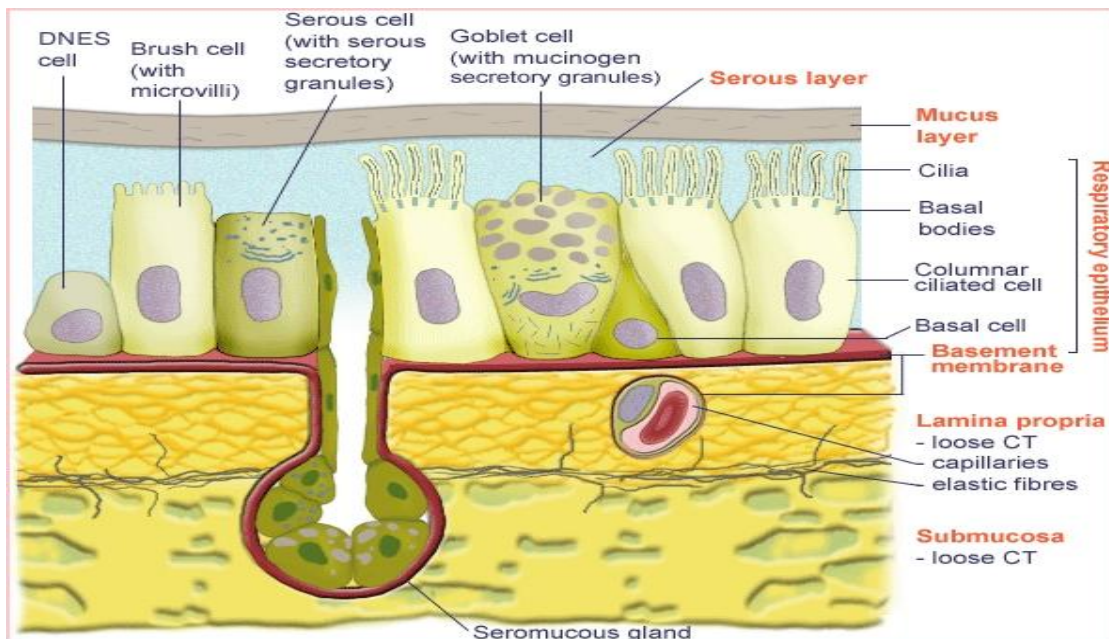
PROTECTION OF LOWER AIRWAY:

1- Filtration and Purification:

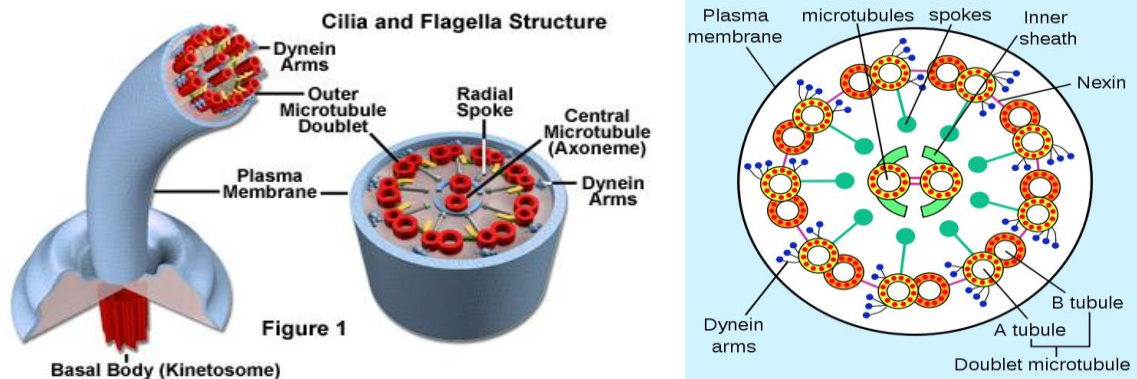
- **Nasal vibrissae** filters particles up to **3 μm** .
- **Nasal mucus** traps particles as fine as **0.5 μm** .
- Particles **< 0.5 μm** pass through Nose into Lower Airways

2- Mucociliary Mechanism:

- Nasal mucosa is rich in Goblet cells and Seromucinous glands.
- Mucous blanket is a continuous sheet spread over Normal mucosa
- Consists of:
 - 1. Superficial Mucus layer:**
 - Secreted by Goblet cells.
 - Floats on Serous layer.
 - Propelled by tips of the beating cilia.
 - 2. Deep Serous layer:**
 - Secreted by Seromucinous glands.
 - 3. Cilia.**



- **Cilia:**
- Hair-like motile structures.
- Function to move a cell or to help transport fluid or materials past them.
- Each cilium has an array of longitudinal microtubules arranged as 9 doublets formed in outer circle around a central pair (**9+2 pattern**).



- Radial spokes connect Outer Microtubular doublets with a Central sheath of protein around the central tubules.
- Cross-section of the cilia reveals Inner and Outer Dynein arms, which are attached each microtubule doublet.
- Dynein, a type of ATPase, provides energy for microtubule sliding resulting in ciliary bending.
- Defects in Ciliary component (inner and/or outer dynein arms, central apparatus, radial spokes) cause abnormal ciliary movements.
- Cilia are constantly beating within Serous layer and moving the mucous blanket in only one direction towards Nasopharynx to be swallowed.
- Moves at a speed of **1 cm/min.**
- Complete sheet of mucus is cleared into Nasopharynx every **10-20 minutes.**
- Factor Affecting Ciliary movements:
 1. Drying
 2. Drugs (Adrenaline)
 3. Excessive heat or cold (Temp < 18C)
 4. Alteration in Neutral PH of Nasal secretions.
 5. Ciliary Structural Abnormality (Dynein arm defect)
 6. Smoking
 7. Infections
 8. Noxious fumes like sulphur dioxide and carbon dioxide.

- Saccharin test:
- Measures mucociliary transit time.
- Saccharin pellet is placed in Anterior nasal cavity, dissolves, transported to Nasopharynx and Oropharynx where the sweet taste is detected.
- Normal transit time is <20 mins

Major composition of Nasal Mucus:

1. 95% Water.
2. 3% Glycoprotein
3. 2% Salts
4. IgA
5. Lysozyme
6. Lactoferrin

3- Enzymes and immunomodulators:

- Nasal secretions contain Lysozyme, Glycoproteins, IgA, IgE, Lactoferrin and interferon which provide immunity against URTI.

4- Sneezing:

- Protective reflex induced by Allergens, Ammonia, Viral infections, Exercise and other irritants which stimulate Trigeminal Afferent (CN-V)
- Complex Efferent input from Vagus Nerve (CN-X) result in:
 1. Slow inspiratory phase.
 2. Glottic and velopharyngeal closure
 3. Increase subglottic pressure
 4. Sudden glottic opening result in sneezing

VOCAL RESONANCE:

- Nose forms a resonating chamber for certain consonants in speech.
- In phonating nasal consonants (**M/N/NG**), sound passes through Nasopharyngeal isthmus and is emitted through Nose.
- When nose (or Nasopharynx) is blocked, speech becomes Denasal, i.e. M/N/NG are uttered as B/D/G respectively.

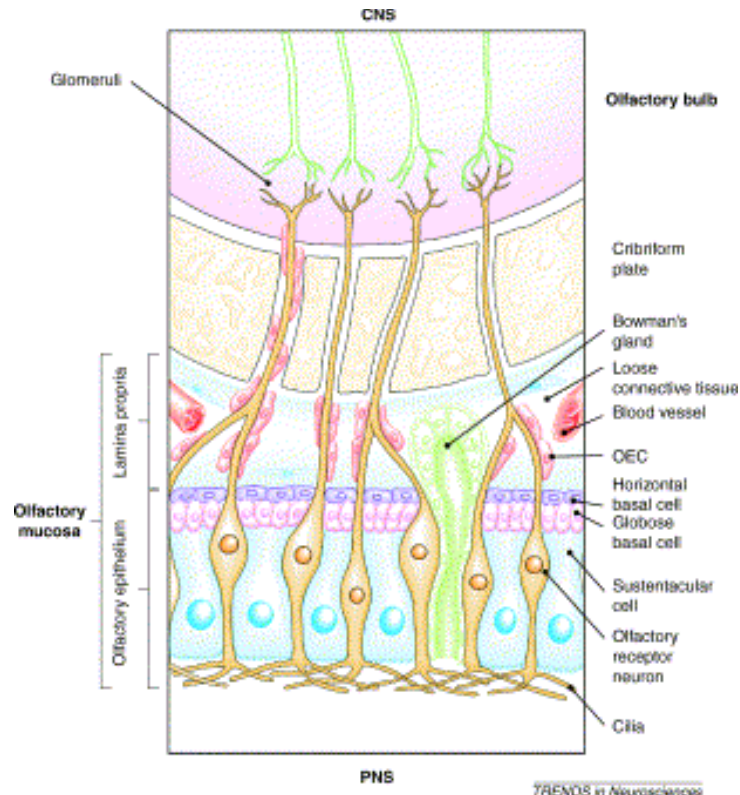
NASAL REFLEXES:

- Several reflexes are initiated in Nasal mucosa.
- **Nasopulmonary Reflex:**
 - Also known as Nasobronchial Reflex / Nasolaryngeal Reflex.
 - Ipsilateral reflex mediated by Trigeminal nerve (CN-V) and Vagus Nerve (CN-X).
 - Nasal stimulation cause reduction of breathing and even cessation (apnoea) due to constriction of bronchioles and laryngeal inlet.
 - More predominant in elderly.

- Demonstrated by increased levels of CO₂ in blood following packing of nasal cavities.
- Care should be taken while packing Nasal cavities in elderly.
- Nasal obstruction leads to increased pulmonary resistance and is reversed when nasal obstruction is surgically treated.
- Pulmonary hypertension or Cor-pulmonale can develop in children with longstanding nasal obstruction due to tonsil and adenoid hypertrophy and can be reversed after removal of the tonsils and adenoids.

OLFACTION:

- Less developed in human.
- Recognition and learning come after age of 2 years.
- Required Turbulent Airflow from Anterior Nares or Choanae.
- Pungent odors (vinegar, ammonia) are never used to test sense of smell as they stimulate fibers of Trigeminal nerve (CN-V) and cause irritation in Nose rather than stimulate Olfactory receptors.
- **Olfactory Epithelium:**
- Pseudostratified Columnar Epithelium in Roof of Nose (Cribiform plate; Upper septum; Superior and Part of Middle Turbinate)
- **Olfactory Epithelial Cell Types:**
 1. **Bipolar Olfactory Receptor Cells:**
 - Ciliated (Long, No Dyein Arms, 7 transmembrane receptors)
 - 6 millions.
 - Each one expresses a Single Odorant Gene.
 - 400 different genotypes.
 2. **Microvillar Cells:**
 - Unclear role.
 3. **Sustentacular Cells:**
 - Supporting cells.
 - Insulate receptor cells from each other.
 - Microvilli instead of cilia.
 4. **Bowman's Gland cells:**
 - Mucous Secretion.
 5. **Horizontal Basal Cells (Dark):**
 - In basement membrane
 6. **Globose Basal Cells (Light):**
 - In basement membrane



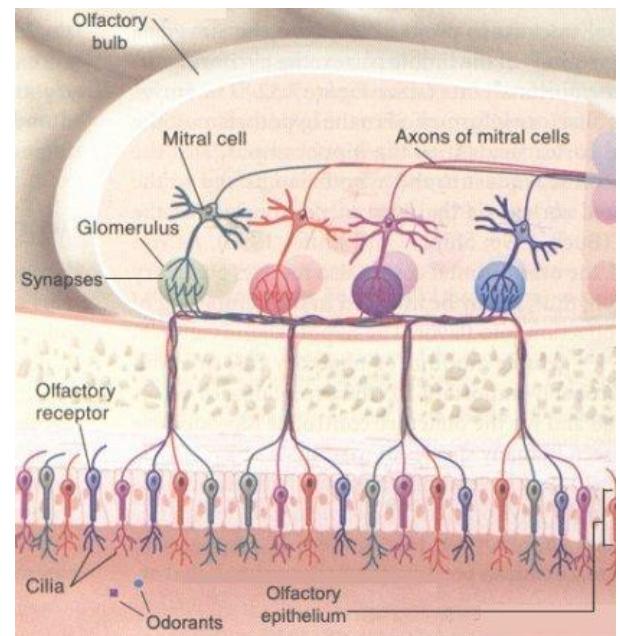
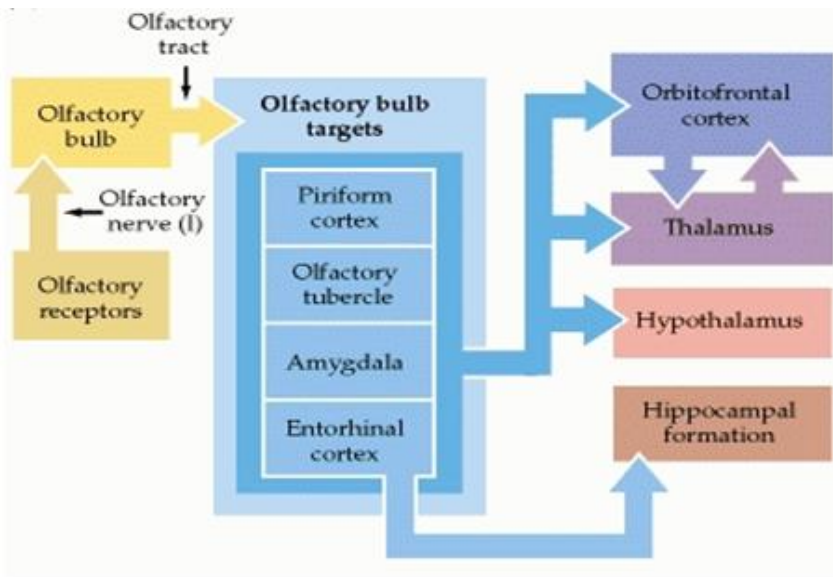
- **Physiology of Olfactory System**
- Odorants have to be water soluble, or bound to odorant binding protein before binding to the receptors.
- Threshold of perception is lower than identification, a smell is sensed before it is recognized.
- Olfactory responses show marked adaptation and thresholds increase with exposure.
- **Intranasal Chemosensory Systems:**
 - 1. Olfactory Nerve (CN-I):**
 - Major role.
 - Found in Olfactory Epithelium in Roof of Nose.
 - Mediates Smell and Flavor.
 - 2. Trigeminal Nerve (CN-V):**
 - Found throughout Nasal Mucosa.
 - Mediate Somato-Sensory Sensation (burn, irritation, cooling, tickling)
 - Induces Reflexive responses (Mucous Secretion, halting inhalation to prevent injury)
 - Noxious stimuli
 - 3. Nervus Terminalis (CN-0):**
 - Unclear role in humans.
 - Loose plexus of nerve fibres present in all mammals.
 - Distinguished by Presence of Ganglia.
 - Related to Reproduction and Pheromone detection.

- **Olfactory Mechanism:**

- Odorant enters Olfactory Cleft.
- → Dissolves in mucus.
- → Odorant binding protein OBP concentrate the solubilised odorant.
- → Binds to Olfactory receptor at Sensory Cilia.
- → Stimulate G-protein (cAMP-dependant) cascade for depolarization.
- → Synaptic connections from secondary neurons before entering brain.
- → Dentate and semilunate gyri.

- **Olfactory Pathways:**

- Olfactory receptor cells at cribriform plate (**1st Order Neuron**)
- → Olfactory Nerve (**CN-I**)
- → Ipsilateral Olfactory Bulb Mitral cells (**2nd Order Neuron**)
- → Axons project through Olfactory Tract.
- → Olfactory cortex in Medial Temporal Lobe.



- Each Olfactory Bulb Mitral cell is contacted by at least 1000 Olfactory Nerve Fibers.
- Each Olfactory Receptor Cell detects a single type of odorant.
- Olfactory epithelial cells are capable of Regeneration.
- Around 40 Olfactory Nerve Bundles cross Cribriform plate.

- **Olfactory Disorders:**
- Impair quality of life, pose safety risks and interfere with one's profession.
- **Anosmia:**
 - o No sense of smell.
 - o Trauma, Congenital.
- **Hyposmia (Microsmia):**
 - o Decreased Sense of Smell.
 - o Smokers, Postmenopausal, Elderly.
- **Hyperosmia:**
 - o Heightened sense of smell.
 - o Sensitive to odors.
 - o Hunger, Cystic Fibrosis, Addison's disease.
- **Presbyosmia:**
 - o Age related smell loss.
- **Dysosmia:**
 - o Distorted smell perception.
 - o During degeneration and recovery.
- **Parosmia (Cacosmia):**
 - o Change in quality of olfactory cue
 - o Intracranial tumors should be excluded.
- **Phantosmia:**
 - o Perception of odors that are not present
 - o Olfactory hallucinations.
- **Effect of Aging in Smell:**
 - o Age is the biggest factor causing olfactory decline.
 - o Starts to drop after age of 65 and worsen after 80 years.
 - o **Causes of Presbyosmia:**
 1. Cumulative Damage to Olfactory Receptors (viral, etc.)
 2. Ossification/Closure of Cribiform foramina.
 3. Development of Early pathology (Parkinson; Alzheimer)

- **Common Causes of Anosmia and Hyposmia:**

1. Obstructive Nasal and Paranasal Disease:

- Most common cause.
- Compromise Airflow to Olfactory Bulb.
- Treat the underlying cause.

2. URTI:

- 2nd Most common cause.
- May cause Parosmia.
- Viral-induced Neuronal injury, Epithelial damage or obstruction.
- No effective management
- Observation.

3. Head trauma:

- 3rd Most common cause.
- Shearing force injure olfactory neurons axons at cribriform plate
- More common in occipital injuries.
- No effective management.
- Observation.

- **Olfactory-Related Syndromes:**

1. Kallmann Syndrome:

- Hypogonadotropic hypogonadism.
- Anosmia secondary to incomplete olfactory bulb and stalk, hypothalamus or olfactory epithelium.
- SNHL
- Cleft palate.

2. Familial Anosmia

- AD with variable penetrance.
- Associated with Baldness and vascular headaches.

3. Foster-Kennedy Syndrome:

- Ipsilateral Anosmia or Hyposmia.
- Ipsilateral Optic Nerve Atrophy.
- Ipsilateral Central Scotoma.
- Contralateral Papilledema.
- Caused by Meningioma of Olfactory groove.

- **Olfaction Tests:**

1. Smell Identification Test (UPSIT):

- Fast and Reliable.

2. Electrophysiologic tests :

- For research tools only

3. Odor event-related potentials (OERPs)

4. Electro-olfactogram (EOG)

Physiology of Paranasal Sinuses:

Functions of Paranasal Sinuses:

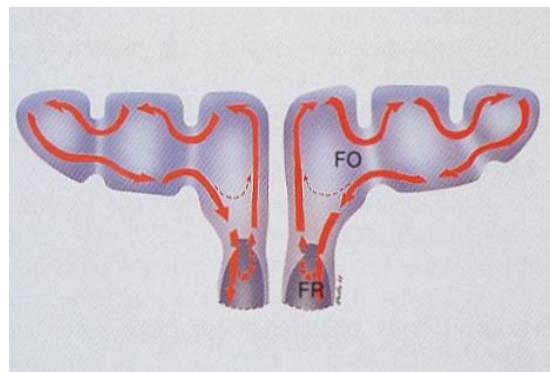
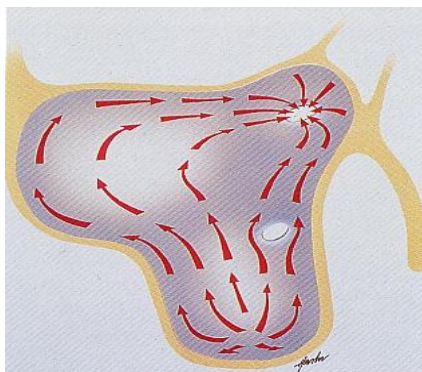
1. Air-conditioning and Humidification.
2. Increase Surface area for Olfaction.
3. Provide Resonance to voice.
4. Thermal insulators.
5. Lighten Skull bones.
6. Trauma Protection.
7. Contribute to Facial growth.

Ventilation of Sinuses:

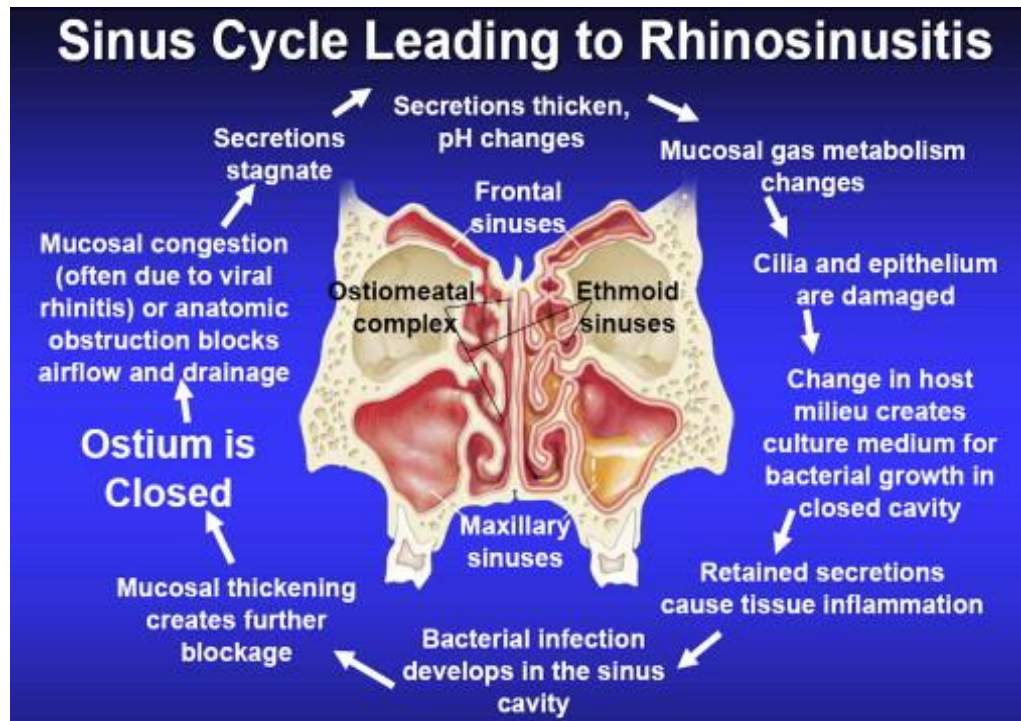
- Takes place through their ostia.
- During Inspiration, Air current causes Negative pressure in Nose.
During Expiration, Positive pressure is created in Nose and this sets up eddies which ventilate Sinuses.
- Ventilation of sinuses is Paradoxical; they are Emptied of Air during Inspiration and filled with Air during Expiration.
- Reverse of what takes place in lungs which fill during inspiration and empty during expiration.
- PO₂ is lower in sinuses than in Nose.
- PO₂ is lowest in Frontal Sinuses.
- If ostium Blocked → Oxygen tension drops further → Ciliary motion impaired → Stasis of secretions.

Mucociliary Clearance:

- Mucus travels to Ostium in a Spiral manner.
- Cilia are more marked near Ostia to help in drainage.
- Cilia propel mucus into Meatuses then Nasopharynx.
- Sinuses essentially Sterile as bacteria are promptly taken out.
- Mucociliary clearance of Maxillary and Sphenoid sinuses is towards Natural Ostium.
- Mucociliary Clearance of Frontal and Ethmoid sinuses is Downwards and Aided by Gravity.
- Mucus in Frontal sinus drains toward Ostium only from Lateral side, mucus Medial to Ostium must course Superiorly to join Lateral flow toward Ostium.



- **Nitric Oxide (NO):**
- Produced in Paranasal Sinuses.
- Functions as:
 1. Bronchodilator.
 2. Vasodilator leading to increased Congestion.
 3. Contributes to plasma Protein Extravasation during Allergic Reaction
 4. Increased Mucociliary Clearance and Motility.
 5. Bacteriostatic.
 6. Immune modulation.
- **Physiology of Normal Sinuses is maintained by:**
 1. Normal sinus secretion.
 2. Properly functioning cilia.
 3. Patent sinus ostia.



Evaluation of Nasal Obstruction:

- Nasal obstruction is a symptom and not a diagnosis.
- Defined as discomfort manifested as a feeling of insufficient airflow through the nose.
- Evaluation of nasal obstruction has both subjective and objective measures.
- In the meantime, clinical assessment including nasal endoscopy remains the cornerstone of diagnosis.

History:

- Character of Nasal Obstruction:

- Onset
- Duration
- Constant vs. intermittent
- Unilateral (tumors, normal nasal cycle) vs. bilateral obstruction

- Associated Symptoms:

- Mouth breathing
- Snoring
- Anosmia/hyposmia/taste disturbances
- Tearing (Nasolacrimal duct obstruction or Allergy)
- Nasal Secretions:
 - Purulent or thick (infectious)
 - Watery and clear (vasomotor rhinitis, allergy)
 - Salty and clear (CSF leak)
- Allergic symptoms:
 - Sneezing
 - Itchy and watery eyes
 - Clear rhinorrhea
- Sinusitis symptoms:
 - Facial pain
 - Headaches
 - Postnasal drip
- Acute infection symptoms:
 - Fevers
 - Malaise
 - Purulent or odorous nasal discharge

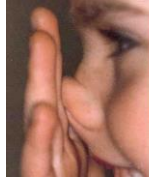
- Contributing Factors:

- Potential toxin and allergen exposure
- Known drug allergies
- Medications use.
- History of immunodeficiency
- History of asthma,
- History of rhinosinusitis
- History of facial trauma or surgery

Physical Examination:

- General Appearance:

- Midface deformities (Adenoid face) as a result from chronic mouth breathing.
- Allergic shiners or allergic salute
- Externa nasal deformity.



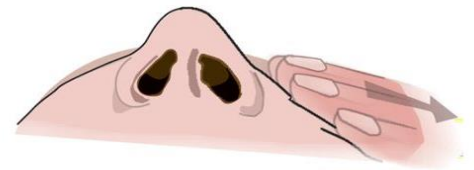
- External Nose:

- Observe for nasal bridge narrowing by previous osteotomies.
- Observe for severe tip ptosis.
 - Tilting the tip superiorly will improve the obstruction.
- Observe for collapsing of lateral cartilages during inspiration indicates valve incompetence.

- Internal Nose:

- Internal nasal valve should be examined initially by simply lifting the nasal tip superiorly without using nasal speculum.

- **Cottle Maneuver** tests nasal valve integrity by retracting the cheek laterally, pulling the upper lateral cartilage away from the septum and widening the internal nasal valve angle.



- If nasal obstruction relieved, this suggests the cause of obstruction is related to nasal valve area (Dorsal septal deviation, lack of upper lateral cartilage integrity).
- Not always reliable as it cant specify level of the obstrctu in nasal valve (Internal valve Vs. External valve)
- **Modified Cottle Maneuver** is more specific test to identify level of obstruction.
- Thin instrument (cotton swab) placed at level of External and Internal nasal vavle to enable direct observation of the nasal valve area as it widened.

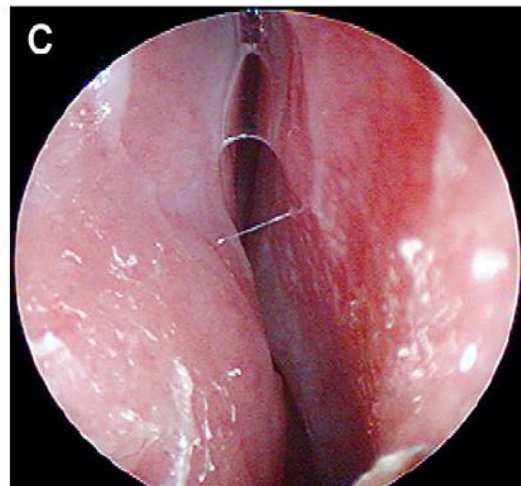
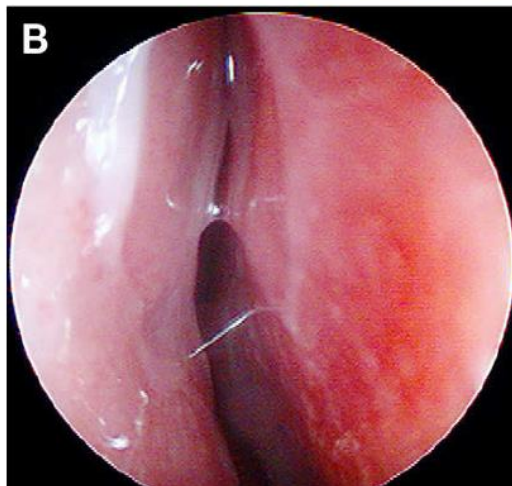
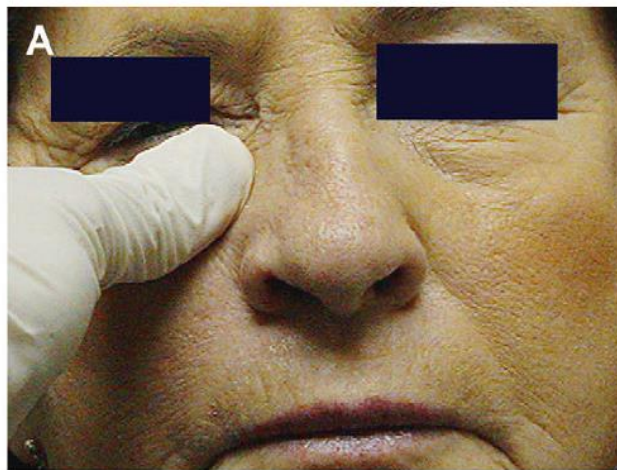


Fig. 2. (A) Performance of the Cottle maneuver. View of the right nasal valve before (B) and during (C) the Cottle maneuver.

- Complete examination of nasal cavity is best accomplished with diagnostic nasal endoscopy to evaluate for :
 - Quality of nasal mucosa
 - Septal deviations
 - Turbinates (hypertrophic, scarring, choncha bullosa)
 - Ostiomeatal complex obstruction
 - Nasal polyposis
 - Nasal masses
 - Foreign bodies
 - Choanal opening
 - Nasopharynx

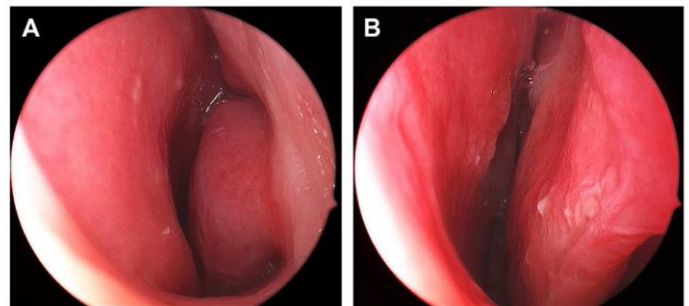
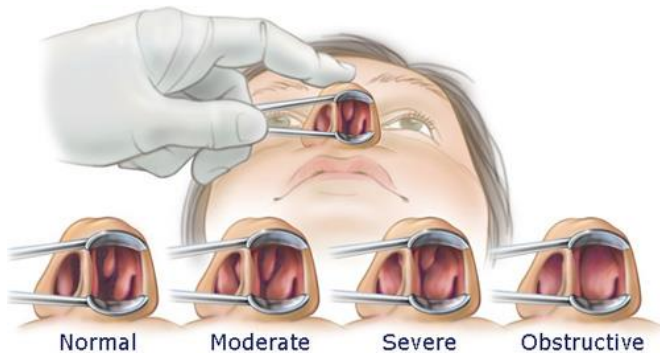


Fig. 3. The inferior turbinate before (A) and after (B) decongestion with oxymetazoline.

- Nasal endoscopy is done before and after decongestion:
 - If nasal obstruction improves with decongestion alone, this suggests a mucosal inflammatory disorder of inferior turbinates.
 - No response suggests the etiology of nasal obstruction is of a rigid, structural nature such as nasal valve obstruction, septal deviation, bony hypertrophy of inferior turbinate or some mucosal inflammatory disorders such as rhinitis medicamentosa and diffuse nasal polyposis.

**TABLE
28.1**

**RIGID SINONASAL ENDOSCOPY
3-PASS TECHNIQUE**

Endoscopic Pass	Structures Visualized
1st Pass Medial to the inferior turbinate (along floor of nose)	Inferior Turbinate Inferior aspect of Middle turbinate Nasopharynx Eustachian tube torus
2nd Pass Medial to the middle turbinate	Septal relationship with middle turbinate (TS1-4) Superior turbinate Sphenoethmoidal recess / sphenoid os
3rd Pass Medial to the middle turbinate (within the middle meatus)	Lateral portion of middle turbinate Accessory fontanelles

Radiological Imaging:

- Required to assist with diagnosis if the source of obstruction is not evident by patient history and physical examination.
- **CT Scan:**
 - Gold standard imaging modality for inflammatory diseases of the paranasal sinuses.
 - Essential for preoperative planning.
 - Evaluates structural/bony abnormalities (DNS, nasal bone fractures, choanal atresia, sinus disease, concha bullosa).
 - Coronal plane is preferred:
 - Provides excellent detail of the OMC
 - Significant neck extension during image acquisition makes this position difficult.
 - Current CT sinus protocols:
 - Non-contrasted thin axial slice thicknesses <3 mm (ideally 1 mm is required for accurate image-guidance compatibility)
 - High-resolution multiplanar reconstruction in coronal and sagittal planes.
 - Contrast is needed in the following cases:
 1. Sinonasal neoplasm
 2. Complicated CRS
 3. Intracranial and orbital extension
 - New multidetector CT scanners allow for acquisition of up to 64 slices with one rotation of the tube.
 - Significantly reduces acquisition time and minimizes motion artifact.
 - Radiation dose from High-resolution CT scan of sinuses yield a dose of 0.96 mSv.
 - To optimize bony detail while preserving soft tissue definition, viewing CT images on a width-level ratio of 2,000/200 is recommended.
 - Findings on CT scan:
 - Bony contours is assessed for:
 - Expansion (chronic disease process)
 - Thickening (osteitis from chronic inflammation)
 - Erosion (more aggressive, acute pathology),
 - Mucosal thickening is assessed for:
 - Laterality
 - Extent of sinuses involved.
 - Characteristics of sinus opacification:
 - Heterogeneity indicates fungal rhinosinusitis

**TABLE
28.2****COMMON INDICATIONS
FOR SINUS CT****Sinonasal Disease Indication**

Acute Rhinosinusitis	Suspected complication Severe illness Failure to improve with medical management Clinical deterioration on medical management Immunocompromised state
Chronic Rhinosinusitis	Failure of medical management Preoperative planning
Neoplasm	Defining extent of lesion (staging)
Trauma	Suspected frontal sinus fracture Suspected cerebrospinal fluid leak

**TABLE
28.3****INFORMATION OBTAINED FROM
THE DIFFERENT PLANES OF
SINUS CT****Sinus CT Plane Helpful Information**

Coronal	Septal deviation OMC Uncinate attachment type (A,B,C) Middle turbinate variations Lamina papyracea anatomy Ethmoid anatomy Skull base anatomy/lateral lamella length Anterior ethmoid artery anatomy Sphenoid anatomy/sphenoethmoid cell (Onodi) Identify optic nerve and carotid artery dehiscence.
Sagittal	Frontal recess anatomy Slope of the skull base Sphenoid pneumatization pattern
Axial	Sphenoid anatomy/attachment of superior turbinate

**TABLE
28.12****RADIOLOGIC RADIATION
EXPOSURE COMPARISONS (34)**

CT Studies	Radiation Dose (mSv)
In-office Sinus CT	0.2
Abdomen and Pelvis CT	15
Chest CT (low dose)	1.5
Conventional Sinus CT	0.96
Mammogram	0.40
Chest x-ray PA and Lateral	0.10
Dental intraoral x-ray	0.005

- MRI:

- Evaluates nasal tumors, complicated rhinosinusitis, intracranial and intra-orbital involvement.

**TABLE
28.4****COMPARISON OF COMMON SINONASAL RADIOLOGIC
MODALITIES**

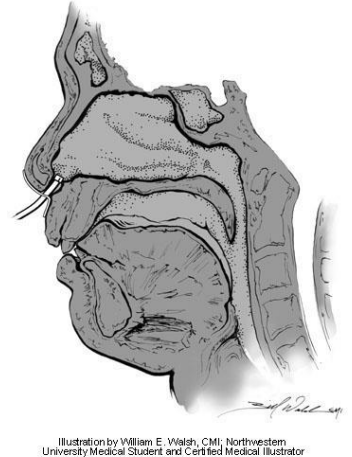
Radiologic Modality	Advantages	Disadvantages
Plain films	Fast acquisition time Low cost	Poor anatomic detail Limited clinical value
CT	Excellent bony detail Multiplanar reconstruction Surgical planning	Radiation exposure Limited soft tissue differentiation
MRI	Fast acquisition time Detailed soft tissue definition Differentiate secretions from soft tissue Lack of radiation exposure Multiplanar reconstruction	Poor bony definition Long acquisition times in confined space Expensive Contraindicated with certain metal implants

Objective evaluation of nasal airway patency:

- Objective tests aim to quantify nasal patency.
- Can be used pre- and post-decongestion in an attempt to predict the efficacy of certain medical and surgical therapies.
- Commonly used in the research setting, but clinical use is not widespread due to:
 - o Expense
 - o Availability of equipment
 - o Variability of operator
 - o Inconsistencies correlation between subjective and objective measures)
- Objective tests include:
 1. Rhinomanometry (RM):
 - Used as screening tool for nasal airway obstruction.
 2. Acoustic Rhinometry (AR):
 - Used for identification of the site of obstruction.
- **Rhinomanometry:**
 - o Simultaneously measures Transnasal pressure (Pa) and Nasal airflow (cm³/s) during normal inspiration and expiration through the nose, which is used to calculate Nasal resistance (Pa/cm³/s)
 - o Resistance from each side of the nose can be compared with each other and with total nasal resistance, to identify how each nasal passage is contributing to the patient's complaint.
 - o More sensitive and specific for patients with functional nasal obstruction (such as rhinitis).
 - o Method:
 - During passive method, the patient holds their breath while air is pumped into the nasal cavity at a fixed rate.
 - During active method (Most common method), patients own nasal respiration is used as the source of airflow.
 - Nasal Airflow is measured using Facemask.
 - Rhinomanometry is performed with and without decongestion.
 - For a patient to be symptomatic, Total Nasal Resistance should be **> 0.3 Pa/cm³/s.**
 - After using decongestion:
 - Improvement >35% in total nasal resistance indicates mucosal obstruction.
 - Improvement <35% in total nasal resistance indicates structural obstruction.
 - o Trans- nasal pressure can be measured using different methods:
 1. Anterior
 2. Posterior
 3. Postnasal

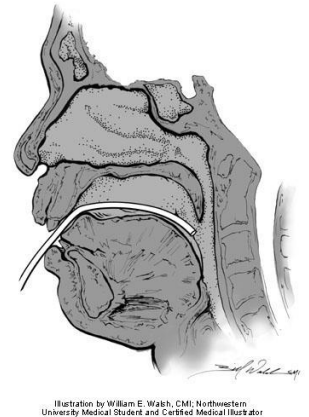
○ Anterior Rhinomanometry:

- Most commonly used method due to technical ease and improved patient comfort.
- Pressure detector is placed into opening of a single nostril not being tested to measure pressure gradient between nostril and nasopharynx of tested side.
- Pressure-flow relationship during **quiet breathing** is measured independently for both nasal cavities.
- Facemask is fitted over nose and connected to a pneumotograph to measure flow through the side to be tested.
- Total resistance can then be calculated from one nostril at a time while closing other nostril.
- Advantages:
 1. Easy.
 2. Patient comfort.
- Limitations:
 1. Inaccurate measurement with septal perforations.
 2. Incomplete occlusion.
 3. Non-physiologic to do one side at a time.



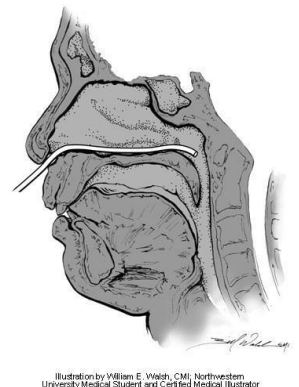
○ Posterior Rhinomanometry:

- Pressure detector is placed within the oral cavity or oropharynx.
- Total resistance can be calculated from both nasal cavity at the same time.
- Advantages:
 1. Only method which can accurately assess the contribution of adenoid hypertrophy to nasal airway obstruction.
- Limitations:
 1. Technically difficult for the patient to tolerate.



○ Postnasal Rhinomanometry:

- Pressure detector is passed through one nostril into the nasopharynx.
- Limitations:
 1. Technically difficult for the patient to tolerate.



- **Acoustic Rhinometry:**

- Simple, noninvasive and relatively cheap.
- Measures nasal airway cross-sectional area and volume.
- Most common method for measurements of nasal cavity geometry.
- More sensitive and specific when evaluating nasal airway obstruction secondary to structural abnormalities.
- Indications:
 - Quantifies nasal obstruction.
 - Anatomic assessment and structure of the nasal airway.
 - Localize areas of constriction.
 - Monitors results of medical or surgical treatment.
- Consists of:
 - sound source
 - wave tube
 - Microphone
 - Amplifier
 - Digital convertor
 - Computer
- Method:
 - Sound waves are transmitted into nasal cavity through a nose piece, impact nasal structures and then reflected back to a microphone and converted into digital impulses, which are then constructed on a rhinogram.
 - Rhinogram provides a two-dimensional anatomic assessment of the nasal airway.
 - Two separate recordings are performed, before and after nasal decongestant, to determine the mucosal and structural components.
 - Each notch on the rhinogram represents a different anatomic constriction in the nasal cavity.

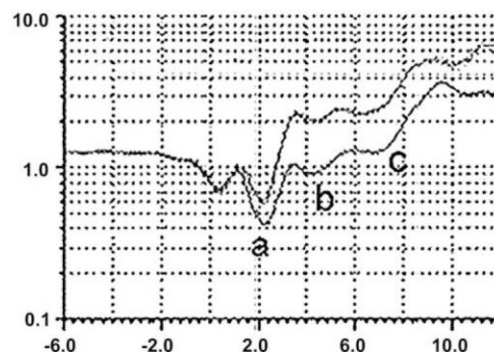
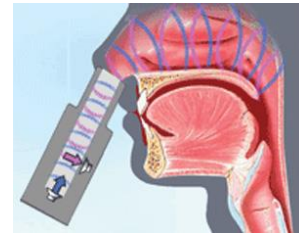


Fig. 6. Acoustic rhinogram before (*lower*) and after (*upper*) nasal decongestion. The x-axis reflects the distance from the nostril and the y-axis is the cross-sectional area of the nasal airway. Note the increase in cross-sectional area after decongestion, most pronounced at notch (b) and (c). Notch (a) represents the MCA at the nasal valve. Notch (b) represents cross-sectional area at the anterior portions of the inferior turbinate and middle turbinate. Notch (c) reflects the area of the middle/posterior end of the middle turbinate.

- **1st Notch (CSA1):** Nasal valve area.
- **2nd Notch (CSA2):** Anterior head of inferior and middle turbinates.
- **3rd Notch (CSA3):** Mid-posterior aspect of middle turbinate.
- Dual-mode Acoustic Rhinometry:
 - Variation of the standard technique whereby the nasal valve cross sectional area (CSA) is measured during apnea and during inspiration.
 - Indicated mainly for diagnosis of nasal valve collapse.
 - Normal patient should have an Inspiratory: Apnea CSA ratio equal to 1
 - Patient with nasal valve collapse have ratios < 1 due to inspiration-related narrowing:
 - Low inspiratory CSA only= Collapse



Rhinomanometry	Acoustic Rhinometry
Advantages	
More functional test	Rapid (10s each nostril)
Can be done on both nostrils simultaneously.	Minimally invasive
	Identify exact site of obstruction
Disadvantages	
Not widely available	Not widely available
Operator dependent	Operator dependent
Takes time (20–30 minutes)	Inaccurate measurement beyond narrow apertures (ie, the nasal valve)
Can't identify site of obstruction.	Less accurate in posterior nasal cavity

- **Factors affecting results of objective tests:**
 - Reducing nasal resistance:
 - Nasal cycle:
 - Exercise
 - African ethnicity
 - Increasing nasal resistance:
 - Nasal cycle
 - Supine position
 - Aspirin
 - Smoking
 - Pregnancy
 - Puberty
 - Menstruation
 - Caucasians ethnicity

- **Causes of inconsistent objective tests (No nasal airway restrictions) in patients with nasal obstruction:**
 1. Atrophic rhinitis:
 - Cause altered nasal sensation, which misinterpret as obstruction to airflow.
 2. Edema or inflammation around sensory nerve endings in nasal mucosa:
 - Create a sensation of nasal obstruction by adversely affecting the function of the sensory receptors.
 3. Poor pulmonary function:
 - Cause a patient to complain of nasal dyspnea.
 4. Cardiopulmonary insufficiency:
 - Can't tolerate any nasal resistance.
 5. Psychogenic causes.

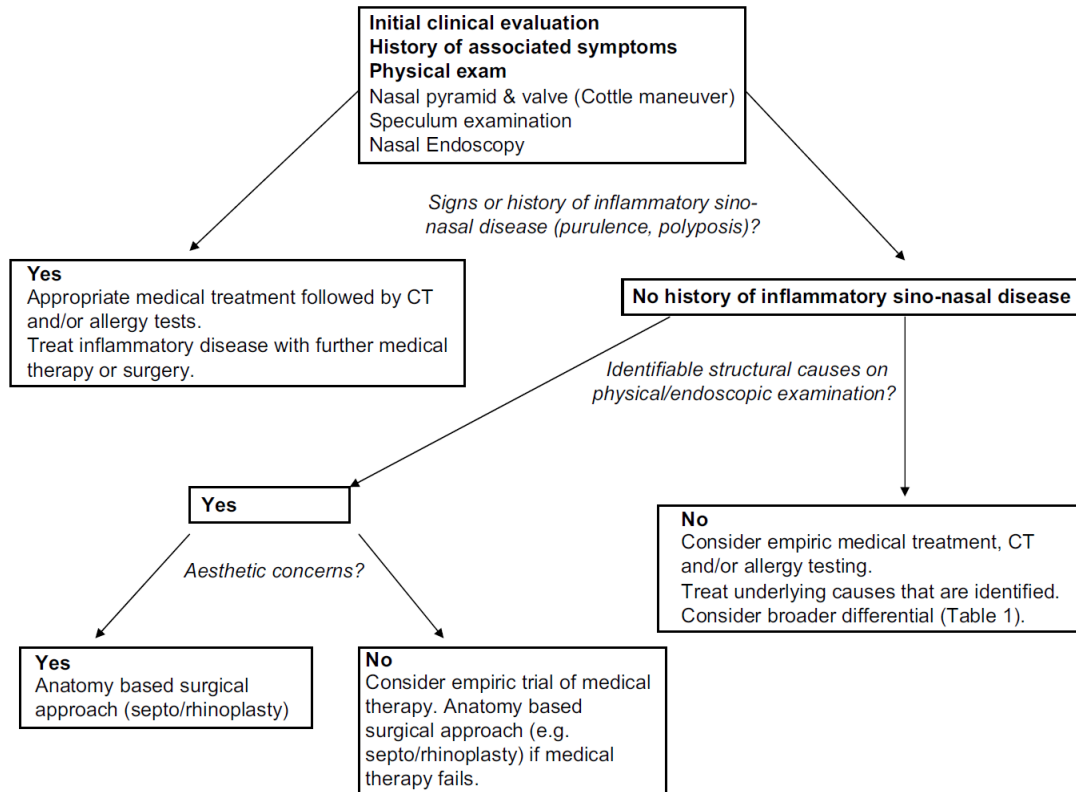


Fig. 5. Algorithm for differential diagnosis of nasal airway obstruction.

- **Differential Diagnosis of Nasal obstruction:**

- Etiology of nasal obstruction is polyfactorial.
- Differential diagnosis of nasal obstruction is broad, including physiologic and anatomic pathology.
- Patients may have a combination of these factors contributing to the symptom of nasal obstruction.

Neoplasms

Benign

Juvenile nasopharyngeal angiofibroma (JNA)
Hemangioma
Dermoid
Papilloma
Neurofibroma
Nasal osteoma
Benign salivary gland tumor
Rhinophyma

Malignant

Esthesioneuroblastoma
Malignant salivary gland neoplasm
Nasopharyngeal carcinoma
Basal cell carcinoma
Adenocarcinoma
Lymphoma
Mucosal melanoma
Squamous cell carcinoma
Sarcoma
Verrucous carcinoma
Metastatic lesion

Inflammatory

Rhinosinusitis
Nasal polyposis
Samter's Triad/Aspirin Sensitivity Triad
Inferior turbinate hypertrophy
Rhinitis
Allergic rhinitis (seasonal or perennial)
Non-allergic rhinitis (NAR)
Non-allergic rhinitis with eosinophilia (NARES)
Infectious rhinitis (bacterial, viral, fungal)
Vasomotor rhinitis
Atrophic rhinitis
Rhinitis medicamentosa

Infectious

Syphilis
Human Immune Deficiency Virus
Nasal vestibulitis

Congenital/Anatomic

Choanal atresia
Adenoid hypertrophy
Naso-septal deviation
Nasal tip ptosis
Internal/external nasal valve incompetence
Septal perforation
Concha bullosa
Cystic Fibrosis
Ciliary Dysmotility

Trauma

Synechiae
Facial nerve paralysis
Overaggressive osteotomies
Post rhinoplasty nasal valve narrowing
Empty nose syndrome (complete turbinate resection)
Cocaine abuse
Septal perforations

Medication

Anti-thyroid medications
Birth control pills
Estrogen replacements
Hypertensive medications
Calcium channel blockers
Beta blockers

Systemic

Wegener's granulomatosis
Sarcoidosis
Midline lethal granuloma
Rhinoscleroma
Histiocytosis X
Tuberculosis

Neurogenic

Encephalocele
Glioma
CSF leak

Other

Nasal foreign body
Hypothyroidism
Pregnancy
Obesity

- **Anatomic Causes:**

○ **Nasal valve collapse:**

- On inspiration, high velocity of air passing through the nasal valve will decrease the intraluminal pressure and will create a vacuum effect on upper lateral cartilages, ultimately causing collapse of the upper lateral cartilage.
- The resiliency of the upper and lower lateral cartilages counteract Bernoulli forces during deep inspiration and prevent internal and external nasal valve collapse.
- Airway collapse of the external nasal valve is prevented by activation of the dilator naris muscles during inspiration, whereas positive pressure is the driving force for nasal vestibule dilation during expiration.
- Causes:
 - Previous trauma, especially **previous rhinoplasty**, is the most common cause of a weakened nasal valve.
 - Loss of facial musculature tone due to aging (weaken the fibroareolar tissues of nasal sidewalls) or facial paralysis (nonfunctional dilator naris muscle).

○ **Septal Deviation:**

- Not every abnormality of the septum requires correction.
 - Anterior deviations in nasal valve region are more likely to cause nasal obstruction than posterior deviations.
- DNS is associated with significantly longer mucociliary clearance times than normal controls.
 - Normalization of mucociliary clearance has been observed after septoplasty.

○ **Concha Bullosa:**

- Pneumatization of the middle turbinate
- Common anatomic variant (25% of population).
- Most concha bullosae are small and asymptomatic.
- Large concha bullosa or massive bilateral concha bullosa is thought to contribute to nasal obstructive symptoms
- Concha bullosa is a significant etiology of nasal middle meatal obstructive syndrome
 - Symptoms include headaches, impaired nasal breathing and anosmia.
- Septal deviation and middle turbinate concha bullosa often occur concurrently in 80% of patients.
 - Unilateral concha bullosa and contralateral septal deviation.

▪ Types of CB:

1. Lamellar (45%):

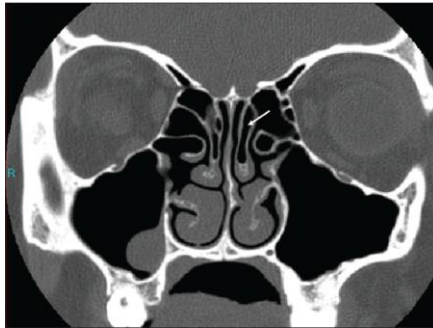
- Pneumatisation of the vertical lamella of the concha.

2. Bulbous (30%):

- Pneumatisation of inferior bulbous segment.

3. Extensive (15%):

- Pneumatisation of both vertical lamella and inferior bulbous segment.



- Patients with fixed anatomic obstructions may experience intermittent symptoms secondary to the nasal cycle and other autonomic phenomena.
- Basal cycle is often unnoticed unless other pathology exists, such as septal deviation, allergic rhinitis, and so forth.

- **Sinonasal Inflammatory Disease:**

○ **Allergic Rhinitis:**

- Most common allergic condition in the world.
- Nasal congestion is one of its most prominent symptom.
- Duration of persistent allergic rhinitis may lead to progressive worsening of nasal airflow determined by rhinomanometry.
- Strong association between asthma and allergic rhinitis:
 - 80% incidence of allergic rhinitis in patients with asthma.
 - Allergic rhinitis should be managed empirically in any patient with asthma and nasal obstruction.
- Allergic rhinitis patients are more sensitive to decreases in cross-sectional nasal area.
- Positional nasal obstruction is significantly higher in patients with rhinitis symptoms.
 - Nasal obstruction increased in:
 - **Supine position:**
 - High nasal resistance due to nasal mucosal reaction to venous changes that alter local blood flow, secondary to compression of the neck veins or hydrostatic pressures.
 - **Lateral decubitus position:**
 - Dependent inferior turbinate is engorged and the turbinate in the superior position is constricted.

- **Traumatic Causes:**

- Accidental trauma and sinonasal surgery may result in complications such as septal perforations, adhesions, nasal stenosis and empty nose syndrome.
- Precipitate in nasal obstruction by:
 - Physical blockage of airflow
 - Induction of sinusitis
 - Impaired sensation of airflow.

- **Medical and Hormonal Causes:**

- **Chronic intranasal decongestant use:**
 - Risk of developing Rhinitis medicamentosa with rebound nasal congestion.
 - Typically occurs 5-7 days after intranasal medication.
 - Microscopically:
 - Loss and destruction of ciliated epithelial cells results in the disruption of mucociliary clearance and an increase in vascular permeability resulting in interstitial edema.
 - Examples:
 - Sympathomimetic amines (ephedrine/phenylephrine).
 - Imidazoline derivatives (oxymetazoline and xylometazoline).
- **Systemic medications:**
 - Antihypertensive:
 - Reserpine, Hydralazine, Guanethidine, Methyldopa, and Prazosin.
 - Beta-blockers:
 - Propranolol and Nadolol.
 - Antidepressants and Antipsychotics:
 - Thioridazine, Amitriptyline and Perphenazine.
 -
 -
 - can also cause congestion.
 - Develops Rhinitis medicamentosa with rebound nasal congestion.
- **Hypothyroidism:**
 - Occurs in 40-60% of patients in a hypothyroid state.
 - Antithyroid medications can mimic these symptoms.
 - Increase in nasal congestion and secretions occur secondary to vascular dilation of the nasal mucosa.
- **Rhinitis of Pregnancy:**
 - Rhinopathia gravidarum.
 - Occurs in 5-30% of pregnant women.
 - Most prevalent during the first trimester.
 - Isolated pregnancy rhinitis should resolve after the gestational period.
 - Assumed etiology is a combination of generalized increases in interstitial fluid volume and direct effect of estrogen on the nasal mucosa, which causes increased vascularity and mucosal edema.

- **Neoplastic Causes:**

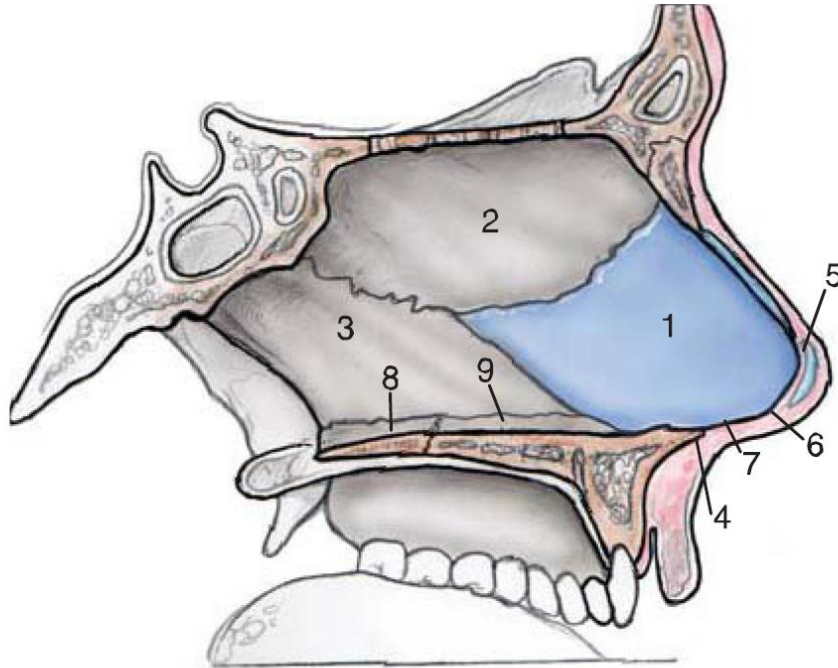
- Nasal obstruction may be associated with other nonspecific symptoms such as unilateral epistaxis or anosmia.
- Squamous cell carcinoma is the most common malignant tumor of the sinonasal tract.
- The most common benign tumors include osteomas and inverting papillomas.
- Juvenile nasopharyngeal angiofibroma must be considered in young patients with epistaxis.
- Pyogenic granuloma is a reparative vascular lesion that presents with finding of a large obstructing lesion along the anterior nasal septum, especially in a pregnant patient.

- **Atypical and Idiopathic Causes:**

- Atypical inflammatory disorders including Wegener's granulomatosis, tuberculosis, sarcoidosis, rhinoscleroma, and rhinosporidiosis may present with nasal lesions, friable mucosa, or crusting, and hence, symptoms of obstruction.
- Cocaine abuse must be ruled out in patients with such findings.
- An important diagnosis to consider in the differential is extranodal NK/T-cell lymphoma, which may initially manifest as an intranasal purple granulomatous mass with bleeding.
- A finding of nasopharyngeal lymphoid hypertrophy in an adult should be further investigated and an HIV test should be recommended.
- Prevalence of nasopharyngeal lymphoid hypertrophy in patients with early stages of HIV infection is 50-90%.
- This hypertrophy and nasal obstruction may improve as the patient's immunocompromised state declines.

Nasal Septum and Its Diseases:

- Nasal septum is a central support structure for the nose.
- When significantly deformed, septum may cause dysfunction and cosmetic deformity.
- Often, patient provides a history of trauma to the nose.
- However, many times, there is no clear history of an inciting event.
 - o Birth trauma or microfractures occurring early in life that led to asymmetric growth of the septal cartilage.



- | | |
|--|--|
| 1. Quadrangular cartilage | 6. Middle septal angle |
| 2. Perpendicular plate of Ethmoid Bone | 7. Posterior septal angle |
| 3. Vomer | 8. Maxillary crest palatine component |
| 4. Nasal Spine | 9. Maxillary crest maxillary component |
| 5. Anterior septal angle | |

Figure 42.1 Septal anatomy in sagittal view

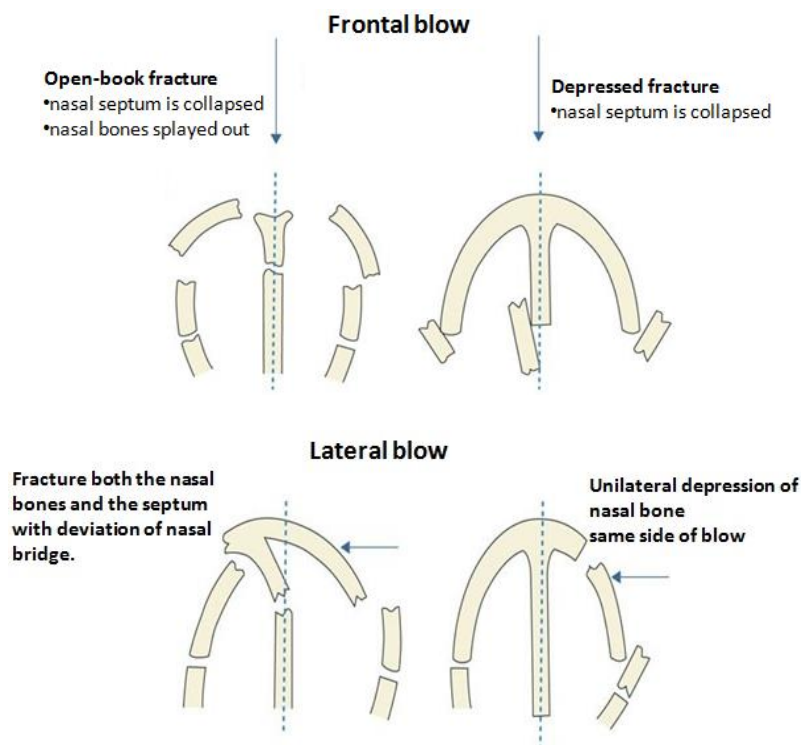
Deviated Nasal Septum (DNS):

- Most common cause of nasal obstruction.
- DNS can involve any age and sex.
- Males are affected more than females
- Deviation may involve only the cartilage, bone or both the cartilage and bone
- Evaluation of DNS causing nasal obstruction depends mainly on physical examination and possibly imaging (CT scan)
 - o Degree of septal deviation has little correlation with subjective ratings of nasal obstruction.

Causes of DNS:

1. Trauma:

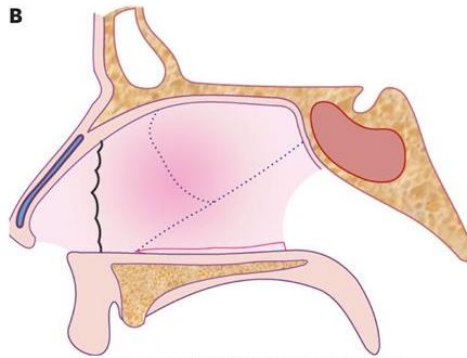
- o Resulted from:
 - **Direct Nasal Trauma:**
 - Most commonly occur in childhood.
 - Frontal blow may cause buckling, twisting, fractures and duplication of nasal septum.
 - Lateral blow may cause displacement of septal cartilage from the vomerine groove and maxillary crest.
 - **Birth Trauma:**
 - During difficult labour, nose is pressed during its passage through the birth canal as they result in septal deviation later in life.
 - Abnormal intrauterine posture may result in compression forces acting on the nose and upper jaws resulting in deviated nasal septum.



○ Types of septal fractures after trauma:

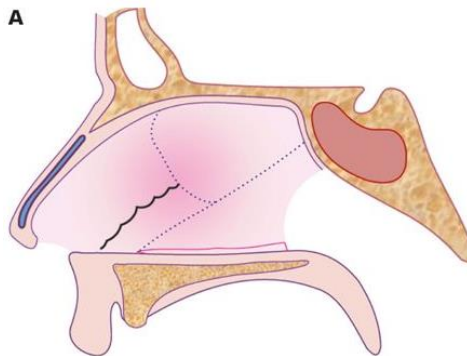
1. Vertical Chevallet Fracture:

- Resulted from frontal blow.
- Runs vertically from the anterior nasal spine upwards to the junction of bony and cartilaginous dorsum of nose.



2. Horizontal Jarjaway Fracture:

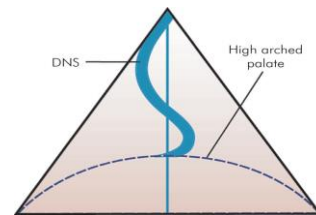
- Results from blows from lateral blow.
- Runs horizontally backwards from above the anterior nasal spine till just above the junction of septal cartilage with the vomer.



- Acute nasal injuries with mucosal tears cause profuse Epistaxis while those with intact mucosa result in septal hematoma which, if not drained early, will cause absorption of the septal cartilage and saddle nose deformity.
- Early recognition and treatment of septal hematomas is essential.

2. Developmental Error:

- Nasal septum begins as a downward growth of frontal prominence which fused with two halves of the developing palate (primary and secondary) in the midline.
- During the primary and secondary dentition, further development takes place in the palate, which descends and widens to accommodate the teeth
- When the nasal septum grows faster in certain individuals than the palate then the nasal septum starts to buckle under pressure.
- DNS may be seen in cases of cleft lip and palate and in those with dental abnormalities.
- In mouth breathers (adenoid hypertrophy), palate is often highly arched and the septum is deviated.



- Types of DNS:

1. Anterior (Caudal) dislocation:

- Septal cartilage may be dislocated into one of the nasal chambers.
- This is better appreciated by looking at the base of nose when patient's head is tilted backwards.

2. C-shaped deformity:

- Septum is deviated in a simple curve to one side.
- Nasal chamber on the concave side of the nasal septum will be wider and may show compensatory hypertrophy of turbinates and concha bullosa.

3. S-shaped deformity:

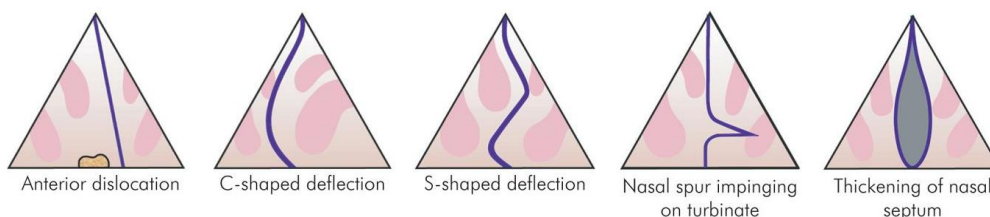
- Septum may show a S-shaped curve either in vertical or anteroposterior plane and cause bilateral nasal obstruction.

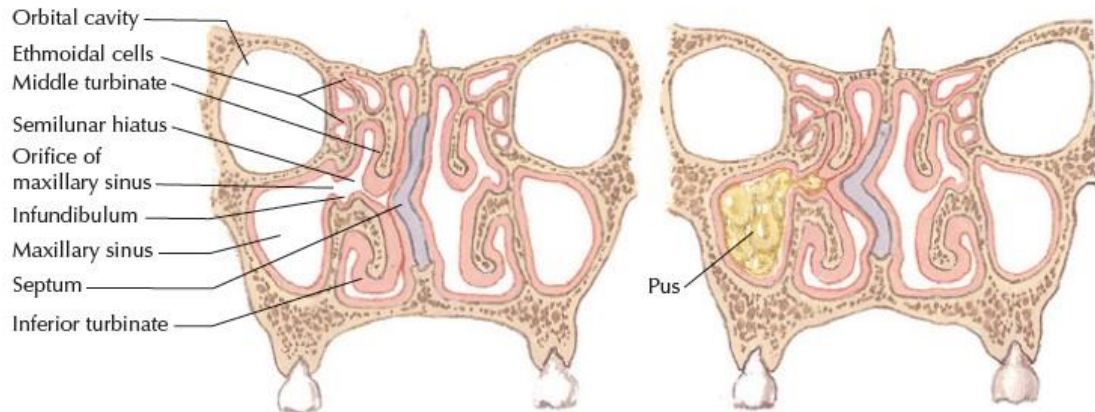
4. Spur:

- A spur is a shelf-like projection resulted from vertical compression forces and found at junction of the vomer below, with the septal cartilage and /or ethmoid bone above.
- A spur may press on the lateral wall and gives rise to headache.
- It may also predispose to repeated epistaxis from the vessels stretched on its convex surface.

5. Thickening

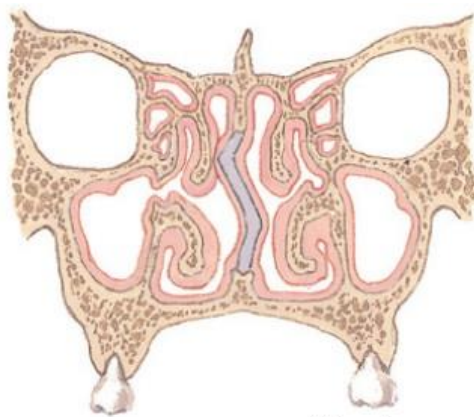
- It may be due to organized hematoma (fibrosis) or over-riding of dislocated septal fragments.



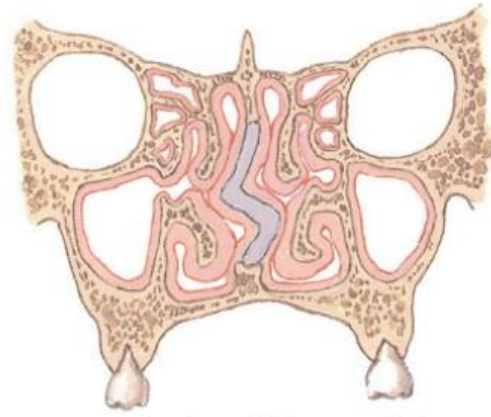


A. Septal bulge occluding airway unilaterally

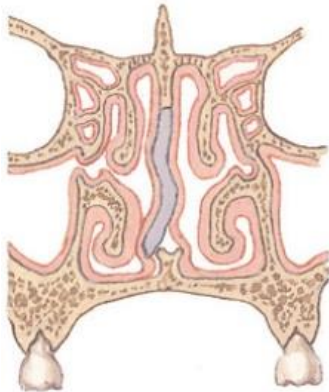
B. Septum occluding semilunar hiatus and obstructing outflow from maxillary sinus with resultant infection



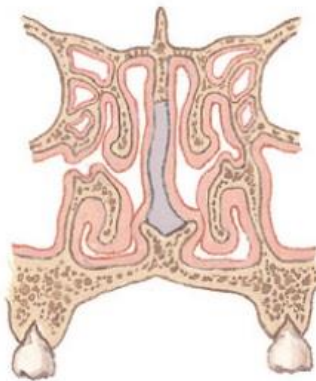
C. Septum impinging on middle turbinate and causing pain via maxillary division of trigeminal nerve



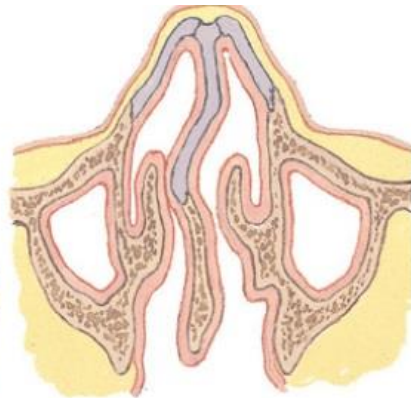
D. S-shaped septal deformity occluding airway bilaterally



E. Septum dislocated from maxillary crest



F. Excessive "wings" on maxillary crest



G. Anteroposterior S-shaped septal bulge (horizontal section)

- **Classification of DNS severity:**

- **Cottle's Classification:**

1. Simple DNS:

- Mild DNS not causing nasal obstruction.
- Most common type.
- No intervention is needed

2. Obstructive DNS:

- Severe DNS that may touch the lateral wall.
- Turbinates shrink away from the septum on vasoconstriction and nasal airway improves.
- Nasal obstruction might improve with medical management targeting the turbinate mucosa.

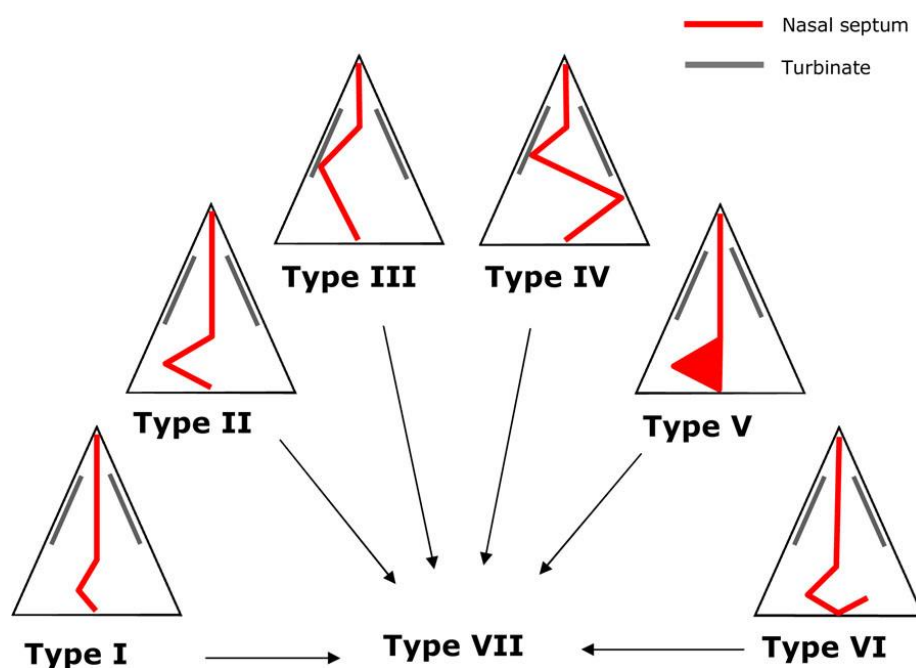
3. Impactive DNS:

- Marked angulation of septum with a spur which lies in contact with lateral nasal wall.
- No improvement of nasal airway of shrinking of the turbinate on vasoconstriction.
- Surgery is indicated in these patients



- Mladina's Classification:

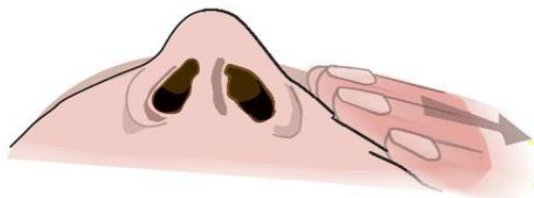
- **Type I:**
 - Mild anterior vertical ridge at the level of nasal valve area that doesn't cause nasal obstruction
- **Type II:**
 - Severe anterior vertical ridge at the level of nasal valve area that cause significant nasal obstruction
- **Type III:**
 - Posterior high vertical deviation at the level of middle turbinate.
- **Type IV:**
 - 'S'-shaped, posterior high vertical deviation at the level of middle turbinate on one side and anterior vertical deviation at the level of nasal valve area on the other side.
- **Type V:**
 - Horizontal septal crest on one side and straight septum on the other side.
- **Type VI:**
 - Horizontal septal crest on one side and sulcus on the other side.
- **Type VII:**
 - Combination of previously described septal deformity type.



Clinical Features of DNS:

1. Nasal obstruction

- Depending on the type of septal deformity, obstruction may be unilateral or bilateral.
- Respiratory currents pass through middle part of nasal cavity, therefore, **high septal deviation cause nasal obstruction more than lower ones.**
- When examining a case of nasal obstruction, one should ascertain the site of obstruction in the nose. It could be
 - **Vestibular:** (caudal septal dislocation, synechiae or stenosis),
 - **Nasal valve:** (synechiae, usually post-rhinoplasty),
 - **Attic:** (along the upper part of nasal septum due to high septal deviation)
 - **Turbinal:** (hypertrophic turbinates or concha bullosa);
 - **Choanal:** (choanal atresia or a choanal polyp.)
- **Cottle Test:**
 - Used to diagnose nasal obstruction due to abnormality of the **Nasal valve.**
 - Cheek is drawn laterally while the patient breathes quietly.
 - If the nasal airway improves on the test side, the test is positive, and indicates abnormality of the vestibular component of nasal valve.
 - Not always reliable.
 - Can't specify level of the obstruction in nasal valve (Internal valve Vs. External valve)



- **Modified Cottle Test:**
 - More specific test.
 - Thin instrument (cotton swab) placed at level of External and Internal nasal valve.
 - Accurate method to identify level of obstruction.

2. Sinusitis:

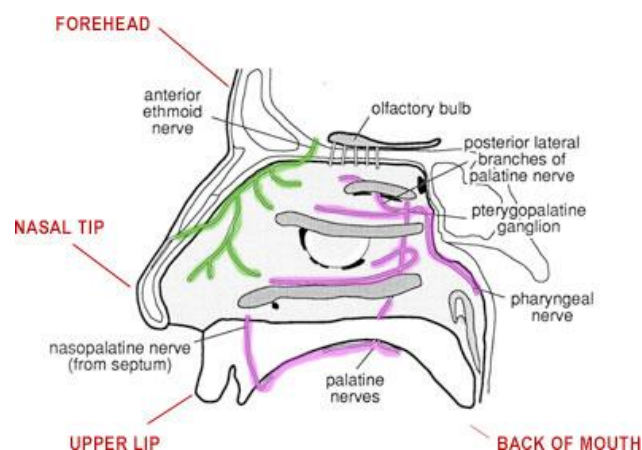
- Deviated septum may obstruct sinus ostia resulting in poor ventilation of the sinuses.
- cause to predispose or perpetuate sinus infections.

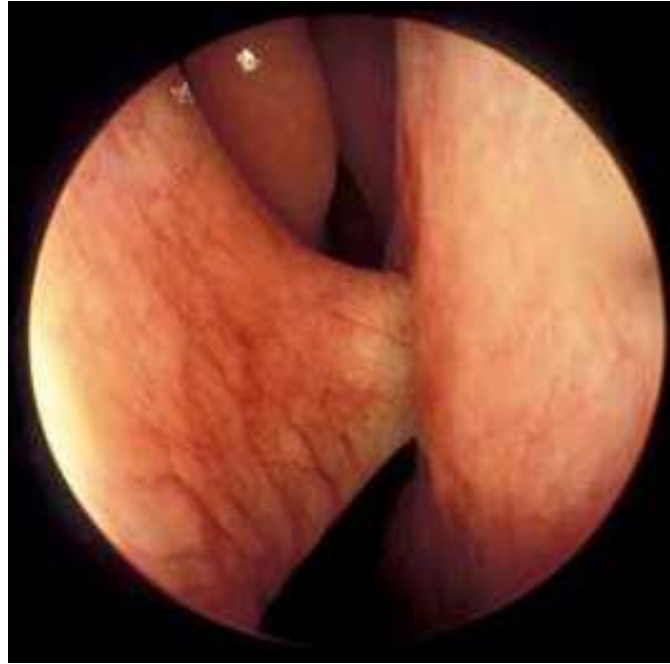
3. Epistaxis:

- Mucosa over the deviated part of septum is exposed to the drying effects of air currents leading to formation of crusts which when removed, cause bleeding.
- Bleeding may also occur from vessels over a septal spur.

4. Contact Point Headache (**Sluder's Neuralgia**):

- Also known as Anterior ethmoid neuralgia, Pterygopalatine ganglion neuralgia, or Sphenopalatine ganglion neuralgia.
- Deviated septum, especially a spur, may press on the lateral wall of nose compressing either **Anterior Ethmoid Nerve** (Branch of Ophthalmic Nerve V1) or one of the branches of **Sphenopalatine ganglion** causing release of substance P (SP) which mediates pain impulses.
- Clinical picture:
 - o Gives rise to pressure headache and facial pain.
 - o Usually the pain started after URTI.
 - o Pain localized to a single area on the face on just ONE side.
 - o Pain classically described localized to the cheek and/or between nose and eye.
 - o Pain can also be localized to upper teeth and roof of mouth.
 - o Pain described as sharp or shooting; less commonly as pressure (like someone driving a high-heel into the face)
 - o May be associated with light and noise sensitivity; not uncommon for these patients to be erroneously diagnosed as migraines without aura (MWOA).
 - o Decongestants seem to work the best to resolve headache, but only temporarily.
- Diagnosis:
 - o Clinical improvement of headache after insertion of nasal pledges soaked with either topical decongestant or local anesthetic agent.
 - o Endoscopic visualization of mucosal contact points.
 - o CT scan showing mucosal contact points.
- Treatment:
 - o Septal or turbinate surgery is considered depends on the location of mucosal contact point to correct it.
- Outcome:
 - o 80% of patients showed improvement post-op.
 - o 20% of patients showed no improvement post-op.
 - o The best post-op outcome was found in the group of patients who met all the 3 criteria for the diagnosis preoperatively with 100% response (60% cure, 40% improvement).





5. Anosmia and Hyposmia:

- Failure of the inspired air to reach the olfactory region may result in total or partial loss of sense of smell.

6. External Deformity: (As the septum goes, so goes the nose)

- Septal deformities may be associated with deviation:
 - o Cartilaginous.
 - o Both the bony and cartilaginous dorsum of nose
 - o Deformities of the nasal tip or columella.

7. Middle Ear infections:

- DNS also predisposes to middle ear infection.

Treatment of DNS:

- Asymptomatic DNS require no treatment.
- Medical management targeting nasal mucosa is typically attempted first with topical nasal steroids, antihistamines, and decongestants as tolerated.
- If the patient fails medical therapy, a surgical intervention to correct the underlying septal deformity is considered.
- **Septal surgery is usually done after the age of 17 so as not to interfere with the growth of nasal skeleton.**
 - o If a child has severe septal deviation causing marked nasal obstruction, Conservative septal surgery (Limited septoplasty) can be performed to provide a good airway.

1. Submucous Resection (SMR):

- Aggressive form of septal surgery in which most of the quadrangular cartilage and the deviated bony parts are removed leaving only the L-shaped strut for structural support.
- SMR leaves a large defect in cartilage and osseous portions of the septum which have been associated with increased risk for complications compared to septoplasty.
- SMR should not be performed in children because it may affect growth.
- Killian's incision (1cm above caudal border of septal cartilage) is ideal for posterior deviations.
- Hemitransfixation incision (at caudal edge of cartilagenous septum in the mucocutaneous junction) is ideal for anterior deviations.

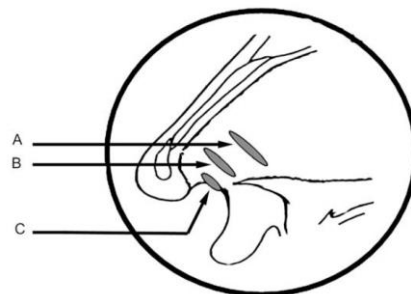


Fig. 3. Schematic of incisions used during septal surgery: A, Killian incision; B, hemitransfixion; C, external rhinoplasty incision.

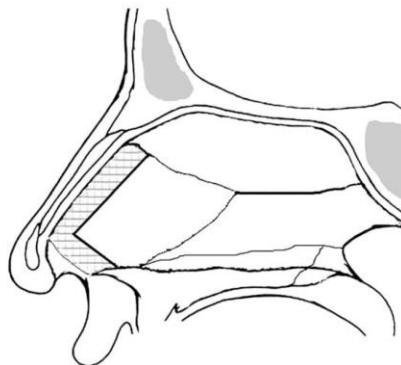


Fig. 2. Diagram of the SMR procedure. Shading demonstrates the area of L-shaped strut cartilage preserved.

2. Septoplasty:

- Septoplasty has now almost replaced SMR operation.
- Technically similar to SMR, but with conservative approach:
 - o Much of the septal framework is preserved.
 - o Only the most deviated parts are removed then corrected and repositioned.
- **Indications of Septoplasty:**
 1. Symptomatic DNS not improving with medical management.
 2. Recurrent Epistaxis.
 3. Cosmetic for caudal dislocation.
 4. For harvesting Cartilage in Septorhinoplasty.
 5. Anatomic obstruction that hinders endoscopic sinus procedures
 6. An approach during trans-septal trans-sphenoidal skull base procedures.
- **Complications of Septal Surgery:**
 1. Persistence of deviation:
 - Most common complication.
 - Associated mainly with severe deviations.
 2. Bleeding and septal hematoma formation:
 - Prevented by packing, nasal septal stents and quilting stitches.
 3. Septal Perforation:
 - Prevented by persevering both mucosal flaps.
 - Postoperative hematoma and tight quilting sutures can lead to vascular compromise of mucosa resulting in perforation.
 4. External nasal deformity
 - Can cause depression of nasal bridge and retraction of columella.
 - Resulted from excessive resection of dorsal aspect of the septal cartilage.
 - Maintenance of L-strut during surgery decrease the risk for this major complication.
 5. Dental anesthesia
 - From injury to nasopalatine nerve while removing the maxillary crest.
 6. Anosmia
 7. Infection and septal abscess formation.
 8. Toxic Shock Syndrome.
 9. CSF leak.
- **Outcome of septoplasty:**
 - o 90% of patients will have post-op improvement.
 - o Anterior deviation better post-op improvement than posterior obstruction.
 - o Presence of concomitant (AR,HIT) decrease post-op improvement and should be treated.

- **Methods of Septoplasty:**
 - Conventional Septoplasty
 - Pediatric Limited Septoplasty
 - Endoscopic Septoplasty
 - Open Septoplasty

Conventional Septoplasty:

- **Steps of Surgery:**

1. Injection of LA:

- Nose is decongested with oxymetazoline (0.05%) soaked pledges bilaterally.
- Sub-perichondrial injection of 1% lidocaine with 1/100,000 epinephrine to act as hemostatic agent and aid for hydrodissection.
- Injection is done bilaterally.
- From anterior-to-posterior direction.
- Mucosa should blanch as the injection proceeds.
- Injection should extend posterior to the deviation.

2. Mucosal incision:

- Hemitransfixion incision is done at the caudal edge of cartilagenous septum in the Mucocutaneous junction on only one side of the septum.
- Done from the side of flap elevation Except if there is caudal dislocation then always open from the side of caudal dislocation.
- The incision should extend through the mucosa and perichondrium but spare the cartilage itself.

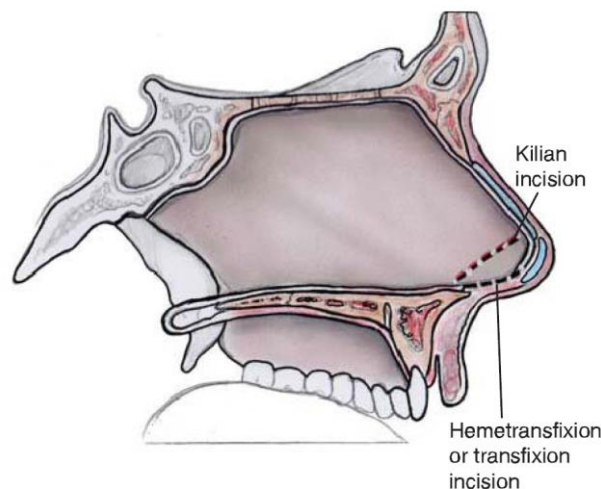


Figure 42.2 Hemitransfixion incision.

3. Elevation of Mucoperichondrial and Mucoperiosteal flaps:

- Always try to do unilateral flap elevation.
 - Usually, start elevation from the concave side or the same side of spur.
- Initially use a sharp elevator (Cottle elevator) to break the fibrous attachments of perichondrium to underlying cartilage then use blunt elevator (Freer elevator) to continue the dissection posteriorly.
- Start with elevating the mucoperichondrial flap (superior tunnel) till reaching the bony septum then elevate the mucoperiosteal flap (inferior tunnel) over the bony septum and inferiorly onto the maxillary crest and connect both tunnels together.
 - Either do the inferior tunnel over maxillary crest starting from posterior to anterior (easier) or from anterior to posterior (Need sharp dissection with knife or iris scissor at the nasal spine).
- Nasal speculum is inserted carefully into subperichondrial space to provide adequate visualization of the dissection.
- Care must be taken during flap elevation along the deviation or over any sharp septal spurs, because the mucosa tears easily at these sites.
- Broad flap elevation is done initially over and under the septal deformity with Freer elevator to provide some laxity to the flap then encroach on the spur carefully with a Cottle elevator to avoid tearing the mucosa.

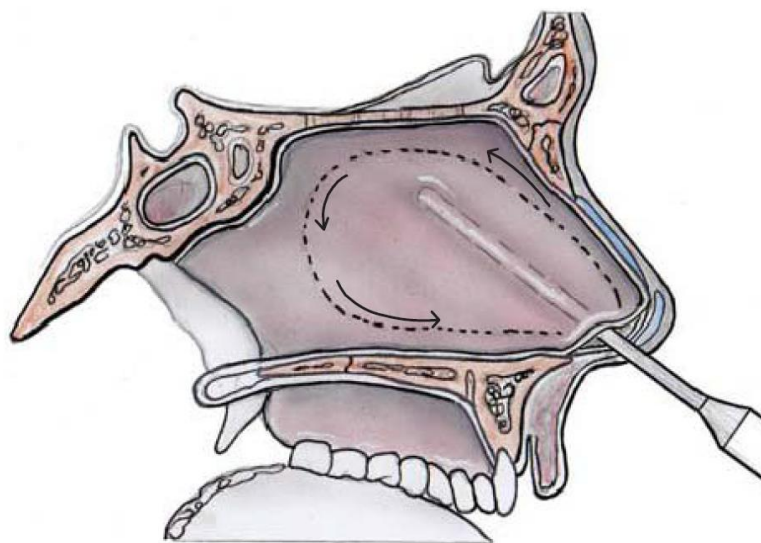
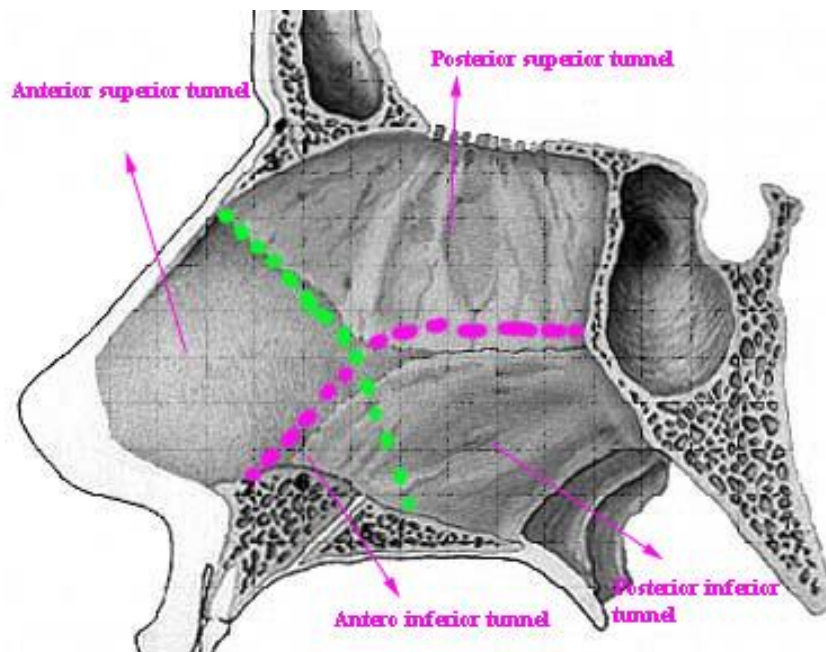


Figure 42.3 Elevation of mucoperichondrial flap.



4. Resection of the deviated septum:

- Once ipsilateral flap is elevated, Cottle elevator can be used to incise septum anterior to deviation and the contralateral flap can then be elevated.
- Cottle elevator is used to incise the septal cartilage inferior and superior to the deviation
- Grasp this segment with Tilley Henkel forceps and fracture it from its remaining posterior attachment using a rotational movement about an anterior-posterior axis.
 - Avoid pulling movement because of risk of fracturing the cribriform plate and CSF leak.
- Must leave 1cm inverted L-shaped cartilaginous strut dorsally and caudally for nasal support and preventing saddle deformity or columellar collapse with tip ptosis.
- If there is high cartilaginous deviation, do cross hatching and scoring to make this part weaker then try to push it centrally.
- True-cut forceps or Jansen-Middleton forceps (Double action) can be used to remove deviated bony septum.
- Always crush the resected portions of cartilage or bone septum and put it back between the flaps.

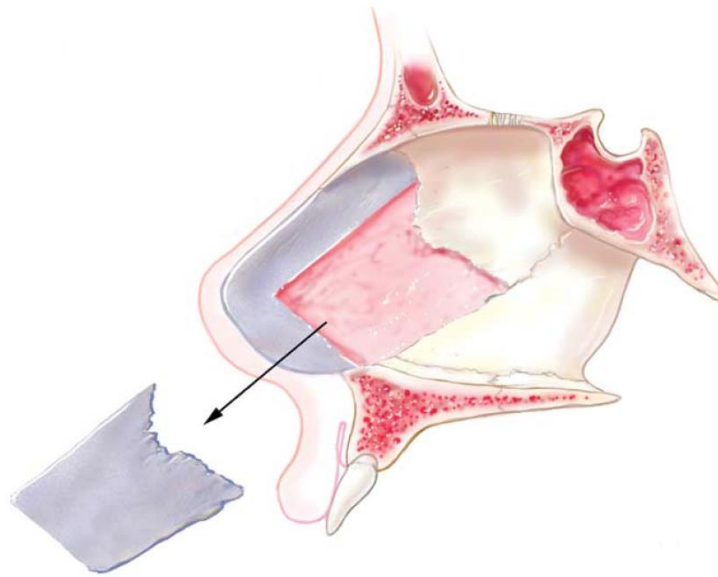
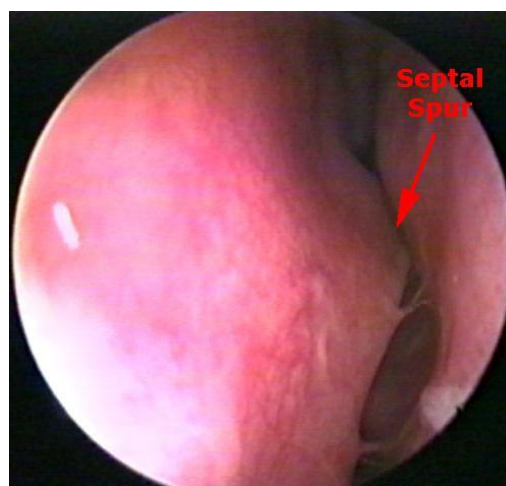
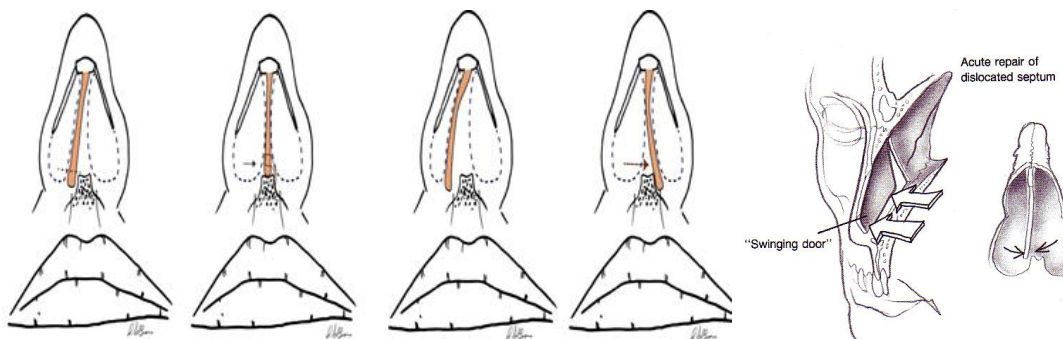


Figure 4. Maintain a generous L-strut.

- If there is cartilaginous spur in one side only:
 - Better to elevate from the same side in mild spur with low risk of tearing the flap.
 - If there is severe spur with high risk of tear then elevate from the other side and resect the cartilage corresponding to site of spur and push the cartilage to elevated side (to prevent other flap from tearing).
- If there is bony maxillary crest spur:
 - If it is difficult to elevate the flap from same side, elevate the opposite side then separate the cartilage inferiorly from the crest and then push the cartilage to the side of spur.
 - Take Chisel and fracture the crest then take Tilley Henkel forceps and grasp the crest and twisted to side of spur to prevent tearing of opposite flap and then remove the crest.

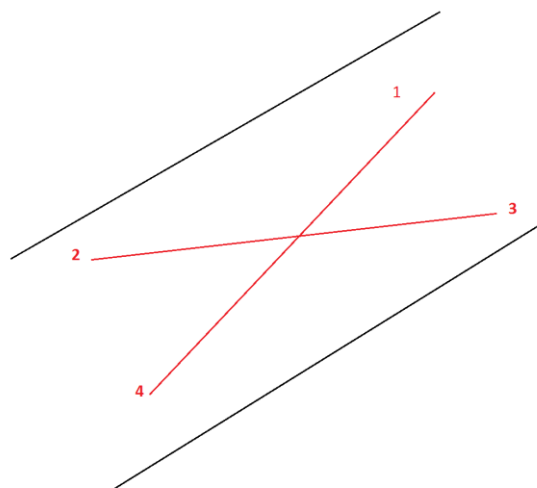


- If there is caudal dislocation:
 - Utilize the “**Swinging door procedure**” to correct the caudal deviation while preserving the cartilage for tip support.
 - Hemitransfixation incision is preformed at the caudal edge of the septum.
 - Caudal septum is exposed by bilateral subperichondrial flap elevation.
 - Skin hooks are used to help retract the flap as it is elevated initially.
 - Flaps are then elevated widely with superior and inferior tunnels’ on both sides of the inferior septum over and under the maxillary crest.
 - Cartilaginous septum is mobilized from three areas with Cottle elevator (Posterior bony attachments, inferior maxillary crest, anterior nasal spine) leaving only its superior attachment intact.
 - Segment of cartilage along the floor of the nose is resected creating a gap along the floor then then reposition of cartilaginous septum with the caudal end to more midline position.
 - Other method is done by not resecting the inferior segment of cartilage along the floor of the nose, and only reposition the cartilaginous septum to other side of nasal spine.
 - Fix it to nasal spine with absorbable sutures.
 - Create a small pocket in a retrograde fashion in the columella between the medial crura into which caudal septum put in a midline position.
 - If the caudal end is long, trim few millimeters of it and excise part of the ipsilateral mucosa.
 - Suture the caudal septum using septocolumellar sutures.



5. Repositioning of the flaps:

- Once septal deformity is resected, flaps are placed back to their anatomic positions and the nasal cavities are examined.
- If there is any large tear in the mucosa, it can be sutured with absorbable sutures.
- Continuous quilting sutures (through and through) with absorbable sutures done to re-approximate the mucoperichondrial flaps to prevent flappy septum and hematoma collection.
 - Starting from one side supero-posteriorly then going through the cartilage to the other side supero-posteriorly then going from the same side supero-anteriorly to exit to the other side supero-anteriorly then go from infero-posteriorly to exit from other side infero-posteriorly then go from same side infero-anteriorly to exit from other side infero-anteriorly and make the surgical note.
 - X-shape from one side and two parallel horizontal lines from the other side.
- Mucosal incision is closed using an absorbable sutures.
- A unilateral mucosal incision can be made in the ventral portion of the septum to allow for drainage of blood, preventing postoperative septal hematoma.
- Nasal packing in form of silastic splints is placed on each side of the septum to compress the flaps together and secured in place using Prolene horizontal mattress suture through the membranous septum.
- Splints are removed in the clinic 2 to 5 days after surgery.
- Antibiotic with adequate coverage for Staphylococcus aureus infection (toxic shock syndrome secondary to nasal packing) should be instituted.



Endoscopic Septoplasty:

- Developed as a result of limited nasal access during endoscopic sinus surgery due to septal spurs or deviations.
- It gives better visualization to the superior and posterior portions of nasal septum.
- Steps of Surgery:
 1. Injection of the nasal septum with local anesthetic with vasoconstrictive properties under endoscopic guidance.
 2. Incision is made either over the spur or just anterior to the deviation.
 3. Mucoperichondrial-mucoperiosteal flap is raised above and below this area with suction Freer elevator.
 4. Deviated cartilage or bone is then resected as described earlier with endoscopic guidance.
 5. Mucosal flaps usually do not need to be closed unless the incision is large.

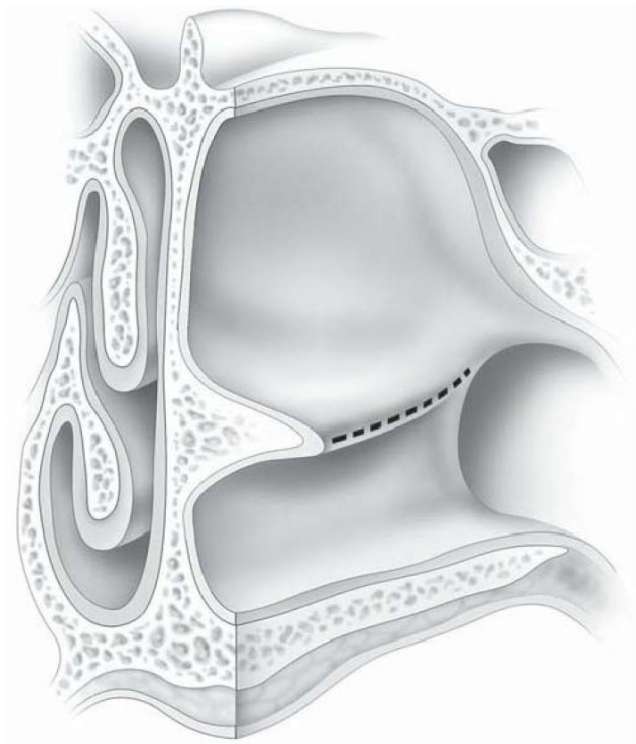


Figure 42.4 Endoscopic septoplasty: Mucoperichondrial flaps are elevated above and below the septal spur.

Open Septoplasty:

- Indications of open septoplasty:
 1. Severe anterior DNS located within the anterior 2 cm of the caudal septum.
 2. DNS associated with external nasal deformity.
- Two methods of open septoplasty:
 1. **Open Septorhinoplasty:**
 - Approached through a rhinoplasty incision, which is made at the midcolumnella.
 - Corrects the septal deformity and external nasal deformity without en bloc removal of cartilaginous septum.
 2. **Extracorporeal Septoplasty:**
 - This technique involves separation of cartilaginous septum from the upper lateral cartilages, perpendicular plate of the ethmoid bone, vomer, and maxillary crest, followed by en bloc removal.
 - The appropriate dimensions of the desired dorsal and caudal L-strut is then measured and carved from the straight segments of the previously removed quadrangular cartilage.
 - The strut is then re-implanted between the mucoperichondrial flaps and sutured into appropriate position.
 - Any disruption of this point results in notching or saddling of the dorsum.

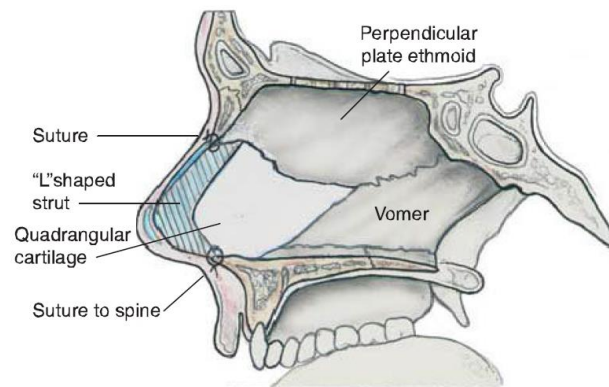
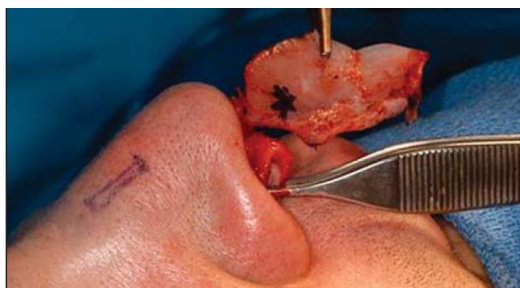
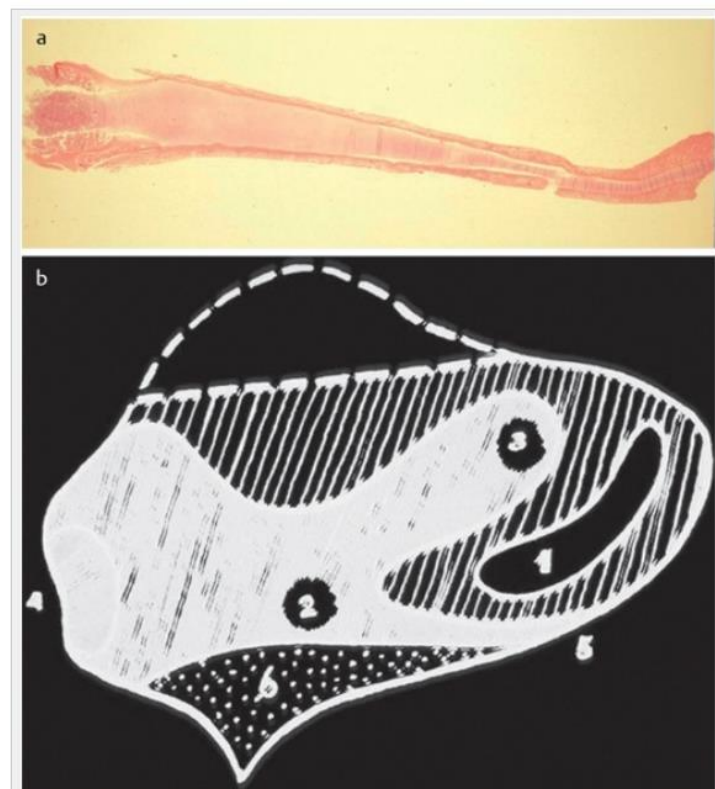


Figure 42.5 Explanation and reimplantation of L-shaped strut during extracorporeal septoplasty.



Pediatric Septoplasty:

- Nasal septum is considered important for midface growth.
- Dimensions and anatomical features changes with increasing age.
- In young children, cartilaginous septum is based on the sphenoid.
- 2 Major growth centers found in the nasal septum:
 - 1. Spheno-dorsal zone:**
 - Area of thick cartilage extends from sphenoid in antero- superior direction to the nasal dorsum.
 - Vertical growth in the sphenodorsal zone results in increased length and height of the nasal dorsum.
 - 2. Spheno-spinal zone:**
 - Area of thick cartilage extends from sphenoid in antero- superior direction nasal spine.
 - Sagittal growth in sphenospatial zone contributes to anterior projection of the nose and maxilla.
- Injury to either of these growth centers (violation of bony cartilaginous junction) may lead to a nasal and midface deformities (short nose, droppy tip, saddle nose deformity).



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Figure 5: (a) Microscopic section through the neonatal septum from the sphenoid (left) to columella; caudal end is dislocated (artifact); the diameter of the septal cartilage diminishes from sphenoid to columella by appr. a factor of 9. (Haematoxylin-azophloxin); (b) Schematic representation of thinner and thicker parts of the neonatal septum. (1) ventrocentral area of thin cartilage, (2) spheno-spinal and (3) spheno-dorsal zone of thick cartilage, (4) sphenoid, (5) anterior nasal spine, (6) vomer anlage. The interrupted line delineates the most superior part of the septum and cartilaginous crista Galli; (2) and (3) represent the growth zones which contribute to the length of nasal dorsum and the size of premaxilla/maxilla.

- Growth of the nasal septum occurs in two phases:
 - Cartilaginous septum grows and reaches adult size by age of 2 years old.
 - Later growth of the total septum is due to development of perpendicular plate by ongoing ossification of septal cartilage at septo-ethmoidal junction.

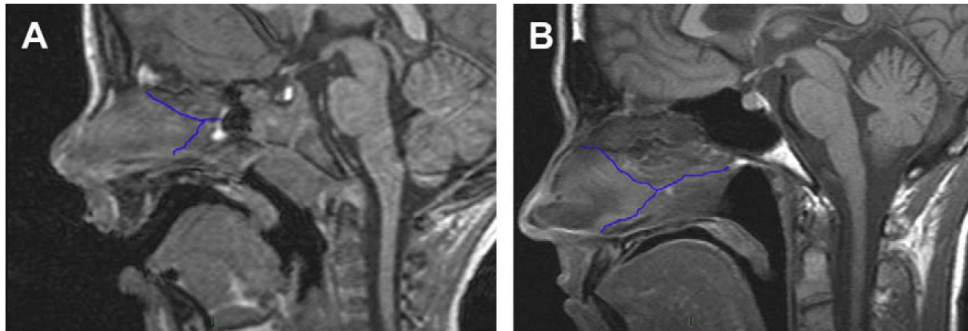


Fig. 3. Sagittal MRIs of a 3-year-old boy (A) and a 27-year-old man (B). The blue lines mark the osteochondral suture lines that delineate the different parts of the nasal septum. Note the predominance of cartilage present in the younger patient.

- The main concern in pediatric septoplasty is the potential for altering or stunting the growth of the nose or midface.
- However, not performing surgery on children affected by nasal septal deviation can lead to dental malocclusion, facial abnormalities and respiratory morbidity.
- Animal studies results:
 - Resection of large amounts of septal cartilage without preservation of the mucoperichondrium in young rabbits caused significant underdevelopment of the maxilla and saddling of the nasal dorsum.
 - However, Submucous resection of cartilage with preservation of a mucoperichondrial flap in young pups did not result in any growth disturbances.
- Human studies results:
 - Most of the studies were on children with a strong indication to undergo septal manipulation at a young age.
 - Septoplasty in children is advocated without serious long term provided doing the following
 - Mucoperichondrium must be left intact.
 - No wide cartilaginous resections must be made.
 - Area of contact between the septum, the vomer, and the perpendicular lamina of the ethmoid must be preserved.
 - Resected cartilage must be repositioned.

- Deviated nasal septum alone is rarely significant enough to be the sole cause of obstructive breathing.
- Nasal endoscopy is indicated in evaluation of child with snoring and should be done before considering septoplasty to rule out all other factors contributing to the nasal obstruction.

Table 2

Absolute and relative indications for pediatric septoplasty [3].

Absolute indications

- Septal abscess
- Septal hematoma
- Severe deformity secondary to acute nasal fracture
- Dermoid cyst
- Cleft lip nose

Relative indications

- Severely deviated septum causing significant nasal airway obstruction
-

- Timing of surgery:
 - Children with mild nasal obstruction can wait for surgery until nasal growth process is complete.
 - Age 16 years for boys.
 - Age 14 years for girls.
 - Children with severe nasal obstruction due to anterior septal deviation can underwent septoplasty as young as 6 years of age.
- **Methods of correcting septal deviation in children:**
 - Closed Septal Repositioning
 - Limited Septoplasty

Closed Septal Repositioning:

- Provide lasting results in pediatric population when correctly performed due to the relative dominance of cartilage in pediatric septum compared to adult, which is more amenable to closed repositioning.
- Less-invasive procedure and minimizes the risk of disrupting mucoperichondrium and cartilage of the developing nasal septum.
- Steps of Surgery:
 - Flat elevator (Asch or Walsham forceps) is inserted into nasal cavity that has been narrowed by septal displacement and the septum is reduced.
 - If the procedure is being performed for an acute septal fracture, one must convert a greenstick fracture to a full fracture before attempting closed repositioning.
 - If the procedure is unsuccessful, proceed to a surgical septoplasty.

Pediatric Limited Septoplasty:

- Steps of Surgery:

1. Targeted endoscopic resection of spurs without flap elevation:

- Indicated in acutely deviated spur of septal cartilage abutting causing nasal airway obstruction.
- Mucoperichondrium can be resected using microdebrider without elevating a flap from the anterior septum.
- This targeted approach will leave only a small area of exposed cartilage at base of the spur, which mucosalizes quickly.
- Great care must be taken to preserve intact the mucoperichondrial covering of the opposite side to ensure that a total perforation is not created.

2. Targeted spur resection

- Indicated in broad septal spurs in which targeted endoscopic resection without flap elevation would leave an unacceptable amount of exposed cartilage, placing the septum at higher risk for perforation.
- Starts with limited conservative elevation of submucoperichondrial flap through either a Killian or Hemitransfixion incision on ipsilateral side of the spur.
- A Freer or Cottle elevator is then used to make a transcartilaginous incision just anterior to the base of the spur.
- Small contralateral mucoperichondrial flap is elevated opposite the spur.
- Resect only the cartilage or bone causing the spur.
- This method minimizes the amount of flap elevation and disruption of blood supply to cartilaginous septum.

3. Cartilage weakening without resection:

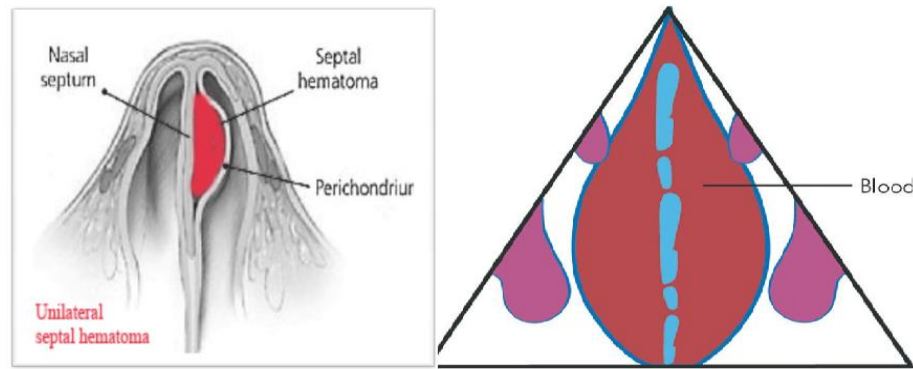
- Consider making incisions on convex side of cartilaginous septal deviation, allowing the septum to swing back to the midline.
- This only requires elevation of a mucoperichondrial flap on convex side of the septal deviation.

4. Cartilage Resection:

- Done only if the more conservative techniques mentioned above do not alleviate the anatomic pathology causing the septal deviation.
- Submucoperichondrial resection of deviated quadrilateral cartilage is done in a similar fashion to that performed in adult.
- Re-implantat the resected cartilage or bony portions after appropriate manipulations to straighten the cartilage or trim excess length that causes bowing. This also applies for bony portions

❖ **Septal Hematoma:**

- Collection of blood under perichondrium or periosteum of the nasal septum.
- **Most common in children.**
- **Results from:**
 - o Nasal Trauma.
 - o Septal surgery.
 - o Spontaneous in bleeding disorders.
- It causes elevation of the mucosa off the cartilaginous septum causing loss of the cartilaginous vascular supply.
 - o **Avascular cartilage can remain viable only for 3 days after compromise of the perichondrium.**
- If the mucosa remain intact, will result in the formation of hematoma.
- If the trauma is severe enough to fracture the septal cartilage, the blood will seep to the opposite side causing bilateral septal hematoma.

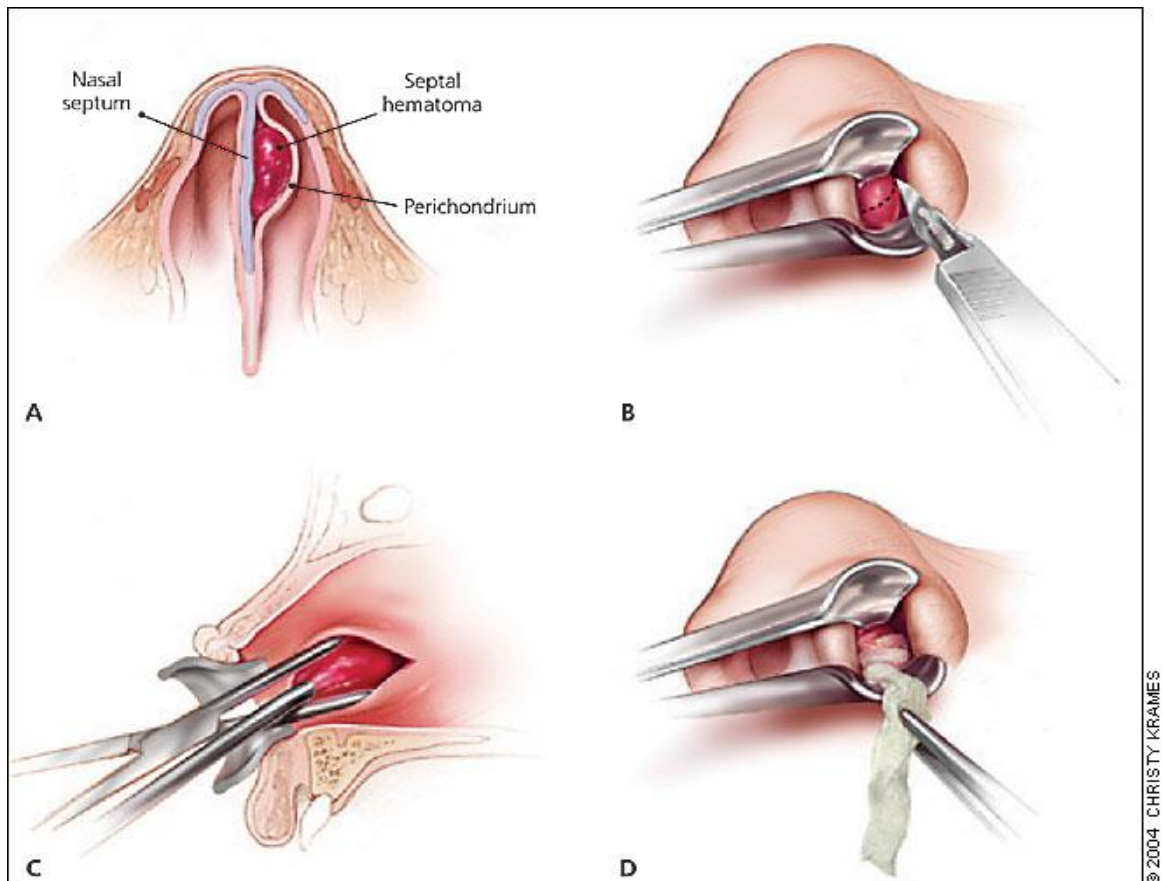


- **Clinical Features:**
- Symptoms usually appear within the first 24-72 hours.
- Bilateral nasal obstruction is the (**commonest**)
- Frontal headache
- Sense of pressure over the nasal bridge.
- Examination reveals **smooth rounded swelling of the septum** in both the nasal fossae.
- **Palpation** may show the mass to be **soft and fluctuant**.
- Sign of trauma.



- **Treatment:**

1. Early surgical drainage to prevent complications.
 - Small Haematomas can be **Aspirated** with a wide bore sterile needle.
 - Large Haematomas are **Incised and Drained** by a small antero-posterior incision parallel to the nasal floor with excision of a small piece of mucosa from the edge of incision for better drainage.
2. Nasal packing on both sides after evacuation to prevent Reaccumulation.
3. Systemic antibiotics to prevent **septal abscess**.



- **Complications of Septal Heamatoma:**

1. Septal thickening due to organization of Septal hematoma into Fibrous tissue.
2. Septal Abscess secondary to infection.
3. Septal Necrosis and Depression of nasal dorsum (Saddle nose deformity).
4. Cavernous sinous thrombosis.

❖ **Septal Abscess:**

- Mostly, results from **secondary infection of septal haematoma**.
- Occasionally, it follows **furuncle** of the nose or upper lip.
- It may also follow acute infection such as **typhoid or measles**.

- **Clinical Features:**
 - Bilateral nasal obstruction.
 - **Pain and tenderness** over the bridge of nose.
 - Patient may also complain of **fever with chills**
 - **Frontal headache**.
 - Skin over the nose may be **red and swollen**.
 - Internal examination of nose reveals:
 - Smooth bilateral swelling of the nasal septum .
 - Fluctuation swelling.
 - Septal mucosa is often congested.
 - Submandibular lymph nodes may also be enlarged and tender.

- **Treatment:**
 - I&D as early as possible.
 - Incision is made in the most dependent part of the abscess and a piece of septal mucosa excised.
 - Pus and necrosed pieces of cartilage are removed by suction.
 - Incision may require to be **reopened daily for 2-3 days** to drain any pus or to remove any necrosed pieces of cartilage.
 - Systemic antibiotics are continued at least for a period of **10 days**

- **Complications:**
 - **Depression of the cartilaginous dorsum** (Necrosis of septal cartilage)
 - **Septal perforation** due to Necrosis of septal flaps.
 - Meningitis
 - cavernous sinus thrombosis

❖ **Perforation of Nasal Septum:**

- Through-and-through defect in any portion of septum with no overlying mucoperichondrium or mucoperiosteum on either side.
- Provides direct communication between the right and left nasal cavities.
- Nasal septal perforations may be stratified based on size:
 - Small perforations: **≤ 0.5 cm**
 - Medium perforations: **0.5-2 cm**
 - Large perforations: **> 2 cm**

- **Traumatic Causes:**

- Post septal surgery.
 - Most common cause (>50%).
 - Mainly if bilateral opposing tear in the flap.
- Post bilateral opposing cauterization of Septum for Epistaxis.
- Habitual nose picking.
- Nasal intubation.
- Septal Hematoma.
- Rhinolith or Neglected foreign body

- **Inflammatory Causes:**

- **Lupus:**
 - Cartilaginous Perforation.
- **Wegener's granuloma:**
 - Cartilaginous and Bony Perforation.
- Sarcoidosis
- Churg-Strauss syndrome

- **Infectious Causes:**

- Septal Abscess.
- Invasive Fungal infection.
- **Tuberculosis and Leprosy:**
 - Cartilaginous Perforation.
- **Syphilis:**
 - Bony Perforation.

- **Neoplastic Causes:**

- Lymphoma.

- **Inhalants:**

- Prolonged use of Steroid sprays.
- Prolonged use of Vasoconstrictive sprays.
- Cocaine addicts.
- Sulfuric acid fumes
- Glass dust
- Mercurials
- Phosphorus

- **Pathophysiology:**

- Diminished blood supply can lead to cartilaginous and mucosal necrosis.
- After perforation occurs, mucosal edges epithelialize, preventing closure of the defect.
- Symptoms arise from altered nasal laminar airflow and may be severely disturbing to the patient.
- Some patients may be completely asymptomatic.

- **Clinical Features:**

- Symptoms tend to be related to size and location of the perforation.
- **Most symptomatic perforations are Large and Anterior.**
- Posterior perforations tend to be less symptomatic because of humidification from nasal mucosa and turbinates.
 - Small perforations cause whistling sound during inspiration or expiration.
 - Larger perforations develop crusts which obstruct the nose.
 - Epistaxis can result from picking at or removing crusted, dried mucous secretions.
 - Persistence of dry nasal crust, coupled with chronic manipulation, can lead to progressive enlargement of the defect.
 - Progression of an enlarging anterior septal perforation will eventually cause deterioration of the dorsal and caudal septal support of the nose.

- **Investigations:**

1. Laboratory evaluation:

- Elevated (p-ANCA): **Churg-Strauss syndrome.**
- Elevated (c-ANCA): **Wegener's granulomatosis.**
- Elevated (ESR).
- Elevated Rheumatoid factor (RF).
- Elevated (ACE) and serum calcium: **Sarcoidosis.**

2. Biopsy of Posterior edge of the perforation

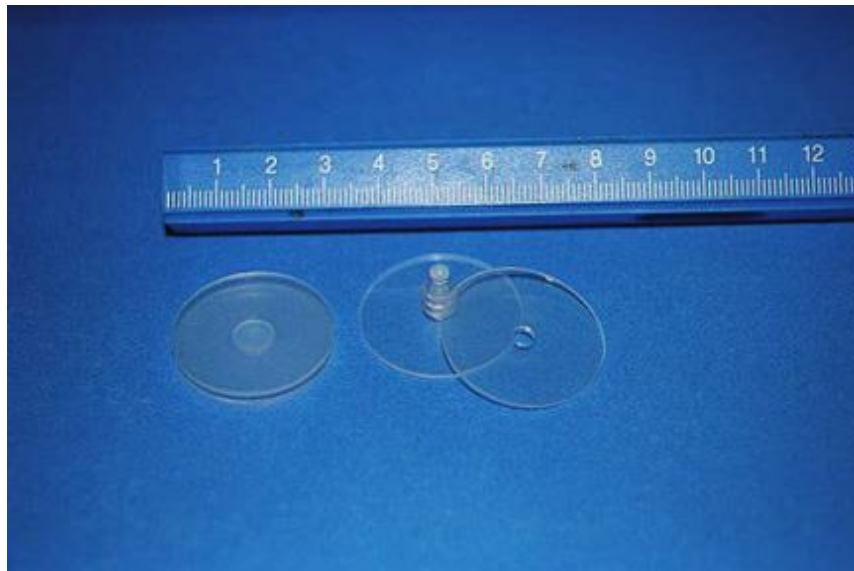
- Send for pathology as well as acid-fast bacilli and fungal cultures.
- Biopsies of the superior margin of the perforation should be avoided because they contribute to the vertical diameter of the defect.

3. CT sinuses to look for co-existing sinus disease and serve as an aid in preoperative sizing of the perforation.



Treatment:

- No treatment is necessary if Asymptomatic
- Always be find out the cause before treatment of perforation.
- **Nasal Hygiene:**
 - Avoid digital manipulations.
 - Routine nasal irrigation with saline or regular humidification to reduce crusts.
 - Antibiotic ointment or any petroleum-based ointment to prevent the drying and hardening of crusted material.
- **Nasal Septal Prosthesis:**
 - Achieves Temporary closure of the perforation.
 - Office based.
 - Available in various sizes.
 - Remain in place for one year or more.
 - Duration is very dependent on patient's diligence with good nasal hygiene and proper care of the prosthesis.
 - Indications for device removal:
 1. Need to size-up the prosthesis
 2. Relief of chronic discomfort from the button
 3. To enable ongoing cleaning and maintenance.
 - Septal buttons complications:
 - Increase frequency of Epistaxis.
 - Allow crusted material to collect around the flanges.
 - Intranasal pain
 - Enlargement of the defect.



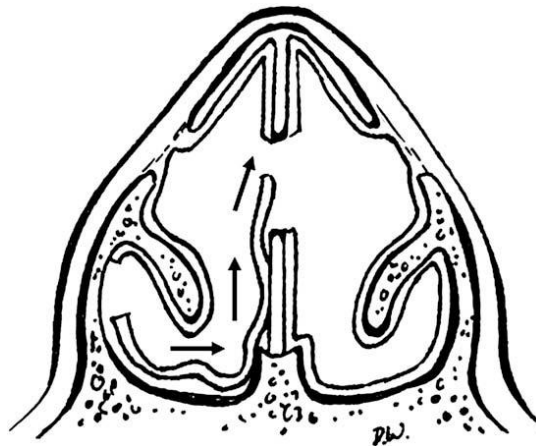
- **Surgical Repair:**

1. Approximation of septal mucosa with interpositional graft:

- **Used for Small defects < 5mm.**
- Endonasal approach.
- Choices for interpositional graft material include bone, cartilage, periosteum and acellular dermal allograft (Alloderm).
- Postoperative care with use of silastic splints for 2 to 3 weeks followed by careful nasal hygiene.

2. Sliding or Rotating Mucoperichondrial Flap:

- **Used for Medium defects 0.5-2 cm.**
- Using Nasal floor mucosa.
- Areas of exposed nasal floor are left to re-mucosalize.
- Postoperative care with use of silastic splints for 2 to 3 weeks followed by careful nasal hygiene.



3. Inferior Turbinate Flap:

- **Used for Large defects > 2 cm.**
- Difficult.
- other flaps can be used like Tunneled sublabial mucosal flap, Facial artery musculomucosal flap (FAMM), and Radial forearm-facial free flap.

- **Contraindications of Septal perforation repair:**

- 1.** Current Cocaine users.
- 2.** Active Granulomatous disease.
- 3.** Malignancy.

- **Capillary Haemangioma:**
- Arises from the caudal septum at Little's Area (Kiesselbach's plexus).
- Soft, dark red, pedunculated or sessile tumor.
- Smooth but may become ulcerated and present with recurrent Epistaxis and nasal obstruction.
- **Treatment:**
 - o Local excision with a cuff of surrounding mucoperichondrium.
 - o May consider preoperative embolization.

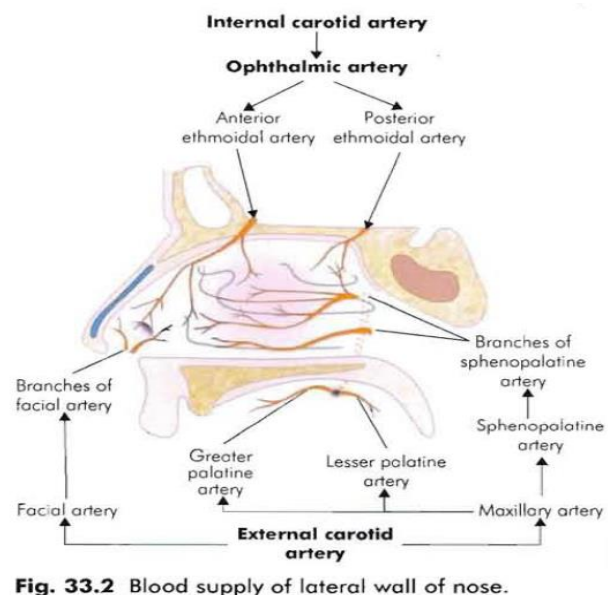
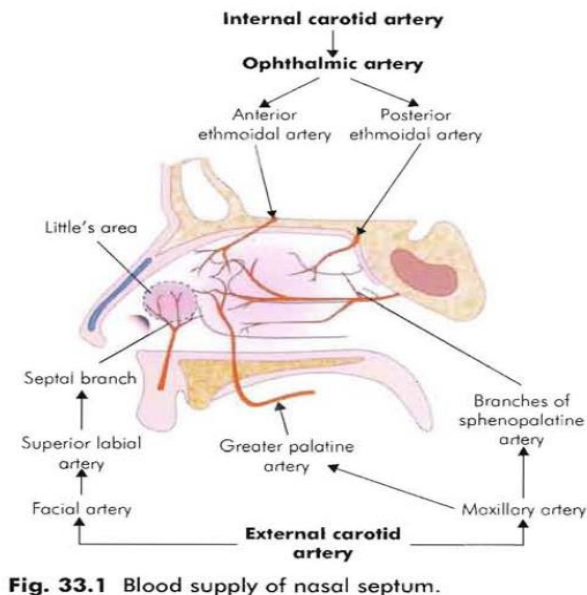


- **Lobular Capillary Hemangioma:**
- Known previously as **Pyogenic Granulomas**.
 - o Misnomer.
 - o Thought to be caused by bacterial infection which is wrong.
- Friable polypoid lesion.
- Usually located on Nasal septum.
- May be secondary to **Trauma**.
- Commonly in female patients during pregnancy usually in 3rd decade of life.
- Presents with Epistaxis and unilateral obstruction.
- **Treatment:**
 - o Most resolves spontaneously.
 - o Surgical Excision.



Epistaxis

- 10% of population experience an episode of epistaxis each year.
- 10% of those will see a physician.
- 1% of those seeking medical care will need a specialist.
- Increased incidence in **childhood** followed by a peak incidence in **6th decade**.
- Epistaxis in children under 2 years old is rare and should raise the suspicion of an underlying illness or child abuse.
- Younger individuals overwhelmingly present with minor bleeding derived from the anterior septum.
- Older patients are more likely to present with a severe acute bleed.
- Males > females.
- Winter worse than summer.



Blood Supply of the Nose:

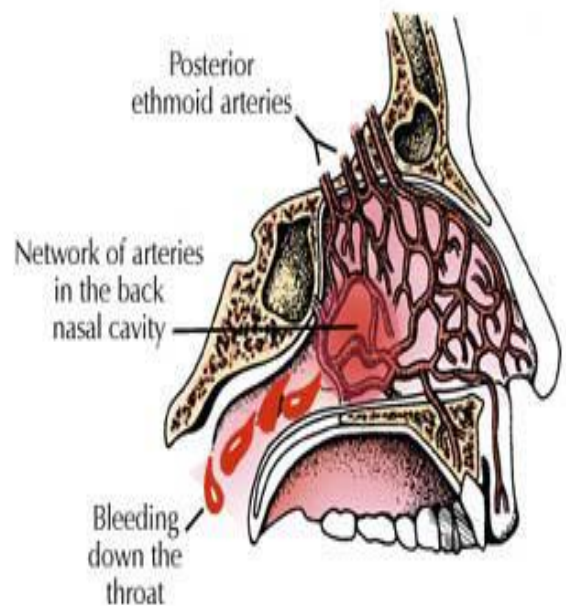
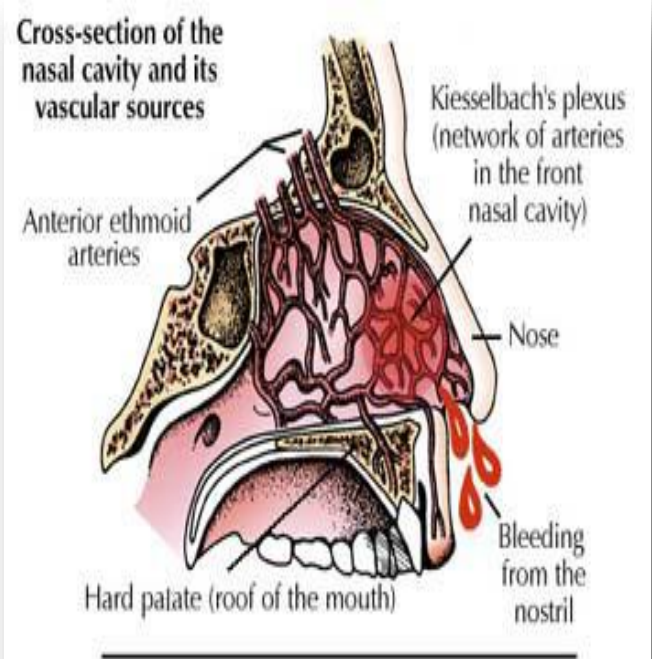
- **External Carotid System:**
 - 1. Sphenopalatine artery** (branch of maxillary artery) gives:
 - A. Lateral posterior nasal artery.
 - B. Septal posterior nasal artery:
 - Cross the anterior face of sphenoid along the septum.
 - May be injured during sphenoid surgery.
 - 2. Greater palatine artery** (branch of maxillary artery)
 - 3. Superior labial artery** (branch of facial artery).
- **Internal Carotid System:**
 - 1. Anterior ethmoidal artery**
 - 2. Posterior ethmoidal artery**
 - Both are branches of ophthalmic artery.
 - Each one divides into a medial branch (Little's area & septum) and a lateral branch (superior & middle turbinate).

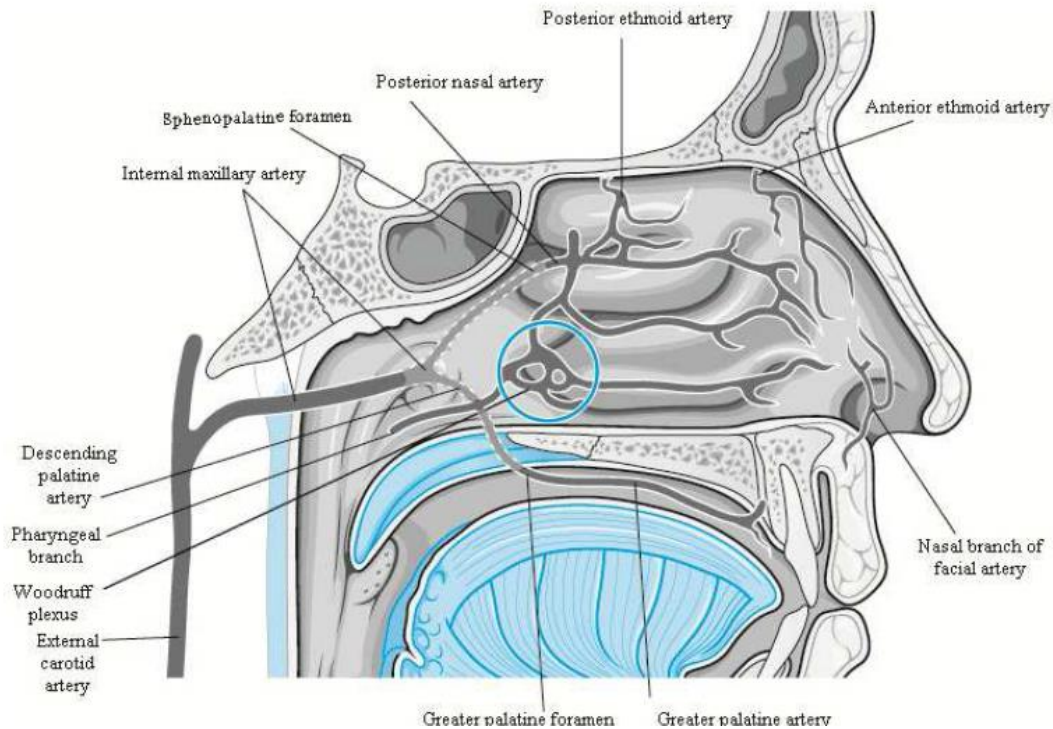
Kesselbach's Plexus/Little's Area:

- Situated in the anterior inferior part of nasal septum, just above the **vestibule**.
- Four arteries anastomose here:
 1. **Anterior Ethmoid (Opth)**
 2. **Superior Labial (Facial)**
 3. **Sphenopalatine (IMAX)**
 4. **Greater Palatine (IMAX)**
- Vasculature runs just under mucosa (**not squamous**).
- This area is exposed to the drying effect of inspiratory current and to finger nail trauma.
- It is the usual site for epistaxis in **children and young adults**.

Woodruff's Plexus (Nasopharyngeal Plexus):

- Vascular area situated under the posterior end of inferior turbinate.
- Site of anastomosis of internal maxillary artery branches:
 1. **Pharyngeal (IMAX)**
 2. **Sphenopalatine (IMAX)**
- Posterior epistaxis may occur in this area.





Retrocolumellar vein:

- This vein runs vertically downwards just behind the columella, crosses the floor of nose and joins venous plexus on the lateral nasal wall.
- This is a common site of venous bleeding in young people.

Classification of Epistaxis

○ Bleeding may be classified as:

1. Anterior Epistaxis
2. Posterior Epistaxis

○ Bleeding may also be classified as:

1. Primary (Idiopathic): Accounts for the majority of cases.
2. Secondary: Results from known local or systemic factors.

Anterior Epistaxis	Posterior Epistaxis
○ Blood flows out from the front of nose with the patient in sitting position. ○ More common ○ Mostly from Little's area or anterior part of lateral wall. ○ Mainly occurs in children or young adults. ○ Mostly caused by trauma. ○ Usually mild, can be easily controlled by local pressure or anterior pack.	○ Mainly the blood flows back into the throat. ○ Patient may swallow it and later have a "coffee-colored" vomitus. ○ This may erroneously be diagnosed as hematemesis. ○ Less common ○ Mostly from Woodruff's Plexus. ○ Often difficult to localize the bleeding point. ○ After 40 years of age. ○ Spontaneous; often due to hypertension or from invasive trauma. ○ Usually severe and requires hospitalization and posterior pack (postnasal pack). ○ Most of the time (95%) posterior epistaxis results from poorly packed anterior epistaxis.

Causes of Epistaxis		
Riyadh et al. Notes		
Secondary		Primary
Local	Systemic	Idiopathic
<u>Nose:</u> <u>Trauma:</u> - Blunt, digital, devices, violent sneeze. <u>Surgery:</u> - Sinus surgery, septoplasty, turbinate surgery, endoscopic skull base. <u>Mucosal Dryness:</u> - Colder weather and low humidity (winter seasons). <u>Anatomic deformities:</u> - Septal deviation, spur, septal perforation. - Occurs due to the drying effects of turbulent airflow. <u>Sinonasal tumors:</u> - Benign, vascular, malignant. <u>Foreign bodies</u> <u>Inflammatory and granulomatous disease.</u> <u>Topical medications:</u> - Topical nasal steroid spray. <u>Atmospheric changes:</u> - High altitudes, sudden decompression (Caisson's disease). <u>Nasopharynx:</u> - Adenoiditis - Juvenile angiofibroma - Malignant tumors	<u>Cardiovascular system:</u> - Hypertension, arteriosclerosis, mitral stenosis, pregnancy (hypertension and hormonal). <u>Disorders of blood and blood vessels:</u> - Aplastic anemia, leukemia, thrombocytopenic and vascular purpura, hemophilia, Christmas disease, scurvy, vitamin K deficiency, Hereditary hemorrhagic telangiectasia. <u>Liver disease:</u> - Hepatic cirrhosis (deficiency of factor II, VII, IX & X Protein C and S). <u>Kidney disease:</u> - Chronic nephritis (uremia). <u>Drugs:</u> - Salicylates (Asprin), anticoagulant therapy, Alcohol abuse. <u>Mediastinal compression:</u> - Tumors of mediastinum (raises venous pressure in the nose). <u>Acute general infection:</u> - Influenza, measles, chickenpox, whooping cough, rheumatic fever, infectious mononucleosis, typhoid, pneumonia, malaria, dengue fever. <u>Vicarious menstruation:</u> - Epistaxis occurring at the time of menstruation.	Most common cause

Etiology and Age

- **Children:** Foreign body, Nose picking, Coagulation disease.
- **Adults:** Trauma, Idiopathic.
- **Old age:** Hypertension, Aspirin and warfarin, Alcohol abuse, Tumors.

Site of Epistaxis:

1. Little's area:

- In 90% cases of epistaxis, bleeding occurs from this site.

2. Above the level of middle turbinate:

- Bleeding from above the middle turbinate and corresponding area on the septum is often from the anterior and posterior ethmoidal vessels (internal carotid system).

3. Below the level of middle turbinate:

- Results from the branches of sphenopalatine artery.
- It may be hidden, lying lateral to middle or inferior turbinate and may require infrastructure of these turbinates for localization of the bleeding site and placement of packing to control it.

4. Posterior part of nasal cavity:

- Blood will flow directly into the pharynx.

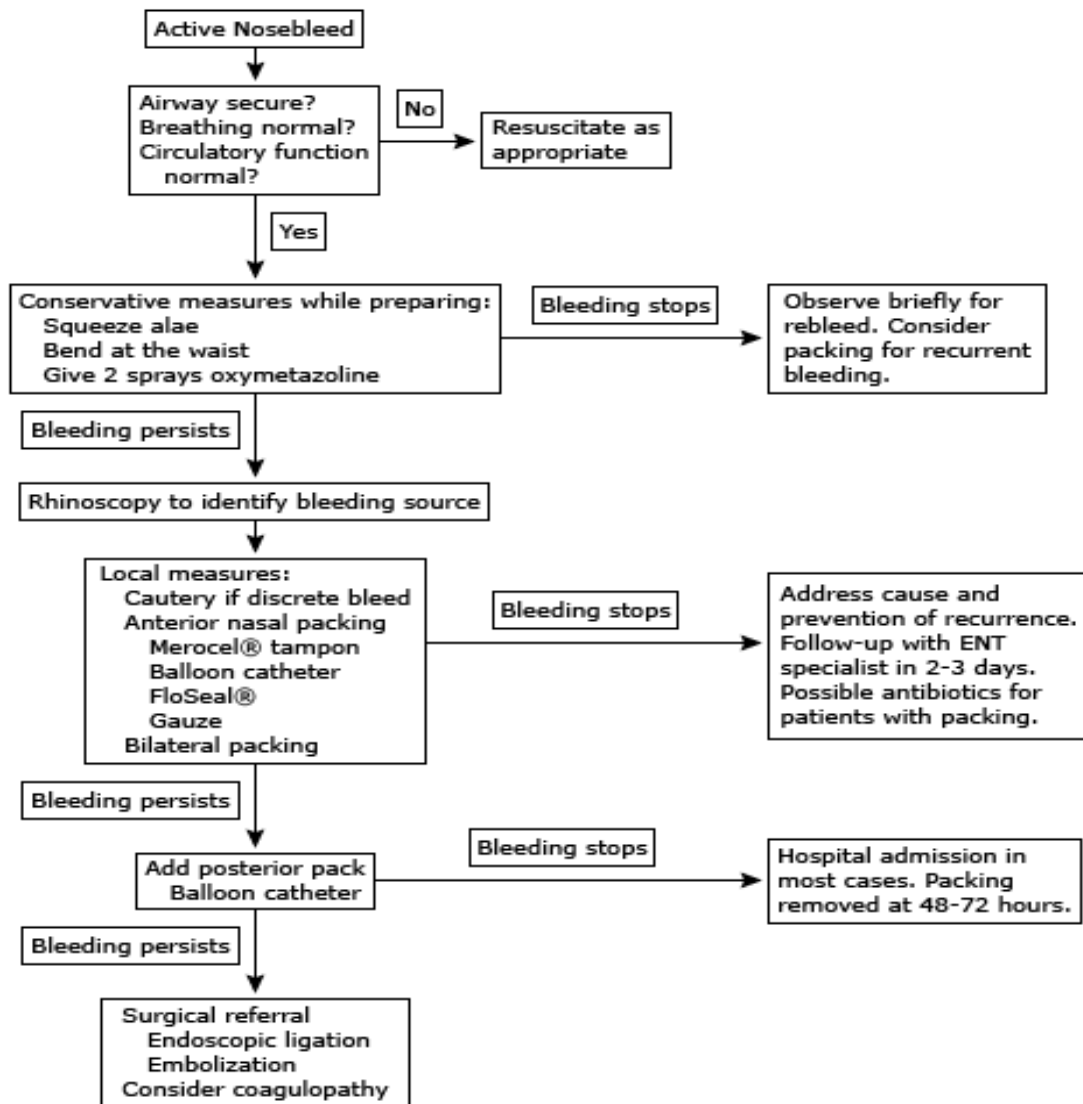
5. Diffuse:

- From septum and lateral nasal wall.
- Often seen in general systemic disorders and blood dyscrasias.

6. Nasopharynx.

Management of Epistaxis:

Non-surgical	Surgical
1. Topical decongestion. 2. Cauterization: • Silver nitrate, electrocautery. 3. Packing: • Absorbable vs. non-absorbable • Anterior, posterior, combined, balloon packs. 4. Greater palatine canal injection. 5. Radiologic Embolization of internal maxillary & facial arteries.	1. Sphenopalatine artery ligation: • Trans-antral vs. Endoscopic. 2. Ethmoid artery ligation: • Lynch frontoethmoidectomy approach vs. Endoscopic. 3. Maxillary artery ligation: • Trans-antral vs. Endoscopic. 4. External carotid artery ligation.



- **First Aid:**

- ABCs and V/S (Needs IV?).
- Patient should sit up with body tilted forward to allow blood to be spit out and not swallowed & mouth breathing (**Trotter's method**).
- Initial attempt to stop bleeding by applying continuous pressure to nasal alae for 5-10 minutes.
- Cold compresses should be applied to the nose to cause reflex vasoconstriction.
- Apply vasoconstrictive agent (Oxymetazoline) and if necessarily Local anesthetic agent (Lidocaine).
- Perform systematic evaluation of the patient prior to controlling the bleeding (May not be possible for heavy bleeding).

- **History:**
 - Characterize Epistaxis:
 - Mode of onset.
 - Amount of epistaxis
 - Duration of epistaxis.
 - Intermittent versus continuous bleeding.
 - Side of bleeding.
 - Epistaxis History:
 - Previous episodes.
 - Hospitalizations.
 - Packing.
 - Medical History:
 - History of known bleeding tendency in patient or family.
 - History of known medical disease (Hypertension, Leukemia, Mitral valve disease, Cirrhosis, Nephritis).
 - Drug History:
 - ASA and Anticoagulants.
 - Excessive Nasal steroid and vasoconstrictive agents.
 - Social History:
 - Cocaine Abuse.
 - Alcoholism.
 - Smoking.
 - Toxin Exposure:
 - Ammonia.
 - Sulfuric acid
 - Phosphorus
 - Other Contributing Factors:
 - Previous Nasal surgery.
 - Recent trauma.
 - Dry environment.
 - High altitude living.
 - Home CPAP ventilators or oxygen.
 - AR, Sinusitis and URTI.
- **Physical Exam:**
 - Acquire adequate lighting (Head light), Nasal speculum, bayonet Forceps.
 - Suction clots to aid visualization.
 - Attempt to **localize** active bleeding.
 - Examine for septal perforations, mucosal lacerations, polyps Nasoseptal deformities, foreign bodies, masses, etc.
 - For Chronic or Recurrent epistaxis without an obvious bleeding source patient should undergo an Endoscopic exam.
- **Lab Tests (For severe or recurrent epistaxis):**
 - CBC.
 - Coagulation profile.
 - LFT.
 - Creatinine.
 - Cross and Match.

- **Acute Medical Management:**

- First Aid Maneuvers.
- Correct Hypovolemia (IVF or Blood transfusion).
- Control Hypertension (antihypertensive agents).
- Correct Coagulopathy (fresh frozen plasma, platelets, cryoprecipitate).

- **Cauterization**

- Types:

1. Silver nitrate
2. Chromic acid pearls
3. Electrocautery
4. Cryotherapy

- Indication:

- Minor epistaxis with single bleeding point located in easily visualized regions (Kiesselbach's plexus).

- Advantages:

- Simple, quick, minimal tissue damage, no packing required.

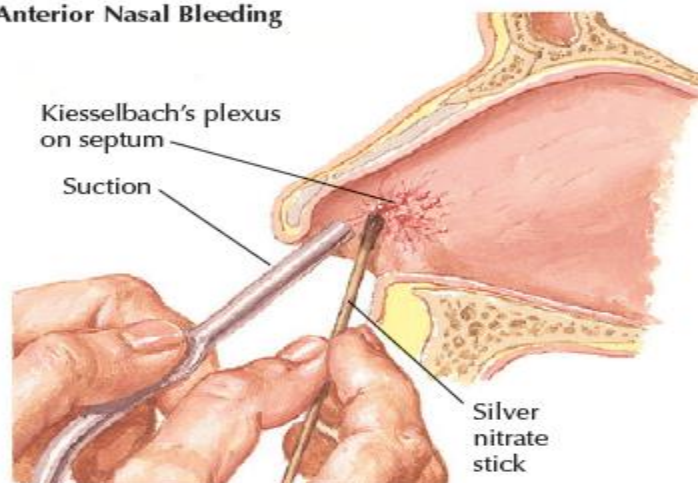
- Disadvantages:

- Allows for coagulation of superficial vessel only.
- Risk of perichondrial exposure, cartilage injury, septal perforation (**avoid cauterization both sides of the septum at similar points, do one side and then about a month later the other**)

- Endoscopic Cauterization:

- Widely used as an effective treatment method for epistaxis, as success rate is up to 90%.
- May be used for **posterior** and difficult to visualize bleeding.
- Can replace arterial ligation method in many cases.
- Very effective if the exact location of the bleeding vessel is known or the bleeding is active at the time of the procedure.
- Using a suction cautery, monopolar or bipolar diathermy.

Cauterization of Anterior Nasal Bleeding



- **Anterior Nasal Packing:**

- **Indications:**

- Active anterior epistaxis that doesn't stop with 1st aid maneuvers or cauterization.
- Diffuse bleeding (site is difficult to localize).

- **Types:**

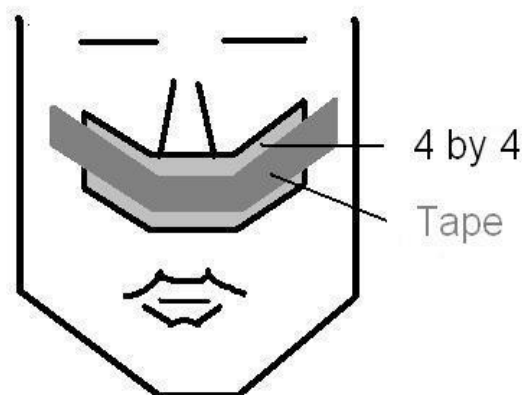
1. Non-Absorbable pack.
2. Absorbable pack.

Non-Absorbable	Absorbable
<u>Types:</u> 1. Ribbon gauze (strip-gauze) soaked with Vaseline. 2. Merocel packs (Hydroxylated polyvinyl acetate/HPVA). 3. Rapid-Rhino (Inflatable pack). ○ Pack can be removed after 24 hours if bleeding has stopped. ○ Sometimes, it has to be kept for 2-3 days to allow vessel to develop a mature thrombus; in that case, Systemic antibiotics (Anti-staph) should be given while the pack is in place to prevent Toxic shock syndrome. • <u>Advantages:</u> ○ Simple ○ Proper anterior pack controls most posterior bleeding. ○ Does not require inpatient monitoring. • <u>Disadvantages:</u> ○ Nasal obstruction. ○ Risk of pressure necrosis (nasal and septal cartilage). ○ Hypoxia ○ Sinusitis ○ Bacteremia ○ Epiphora ○ Toxic shock syndrome (S. aureus exotoxin).	<u>Types:</u> 1. Surgicel (Oxidized cellulose). 2. Floseal (Human Gelatin-thrombin-matrix). 3. Tisseel (Fibrin glue). 4. Surgifoam (Porcine gelatin sponge). 5. Nasopore (Polyurethane foam). ○ More expensive. ○ Provides less mechanical tamponade so used in mild observable bleeding point. ○ Pro-coagulant effect. ○ Useful for coagulopathies. ○ May be used after cauterization attempts. ○ Significant reduction in the rate of re-bleeding at 1 week & more patient satisfaction and comfort compared with non-absorbable nasal pack. • <u>Advantages:</u> ○ Simple and Quick. ○ Pro-coagulant effect. ○ Less damage than cautery. ○ Potential to avoid non-absorbable packing. ○ Dissolves and avoid physical trauma of removal. • <u>Disadvantages:</u> ○ Risk of re-bleed in severe bleeding.



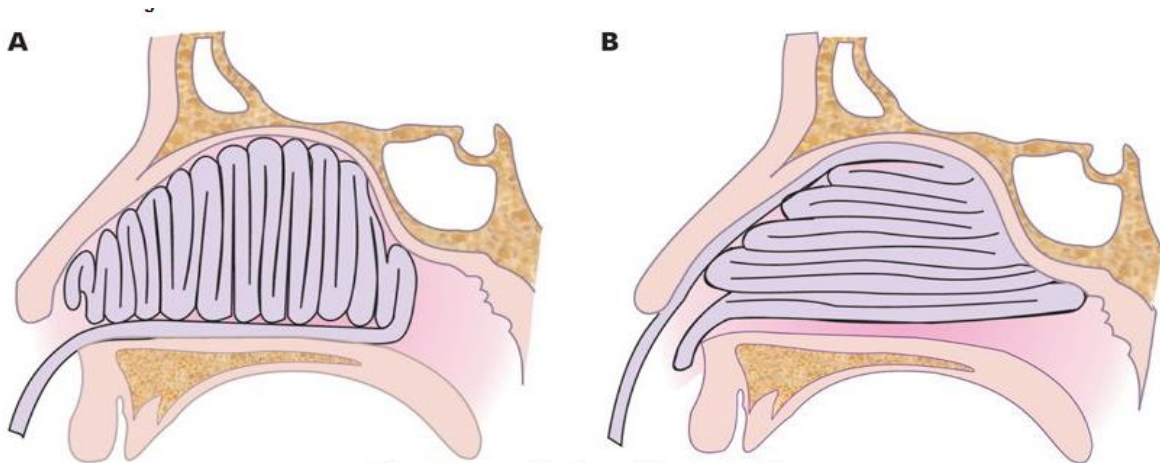
Merocel Pack Method:

- Size up the nostrils.
- Most people's nasopharynx is approximately 7-8 cm long.
 - That's why they made the Merocel that long. But you can always trim these things to custom fit the patient.
 - The trick is to not cut off too much.
- Apply a generous amount of **Fucidine** to the leading edge of the Merocel as this will make it a lot easier on the patient.
- The patient should be sitting up straight with their head against the head rest (so they don't move back).
- Gently but firmly glide the Merocel in at a perpendicular angle to the forehead.
- Try to stay slightly closer to the septum which is less sensitive than the turbinates.
- The rule with all packs, whether you use Merocel, Vaseline gauze or Posteriors, you must pack both sides.
- Once both packs are in, you can inject the non-bleeding side with Normal Saline (to expand it) if it hasn't expanded with blood.
- Always tie the merocels together, leaving about a centimeter of slack between the nose and the knot.
- If they're loose the patient will invariable pull them out "by accident"
- The final step is applying a "**moustache dressing**" by folding a 4 by 4 into thirds and tape it over their nostrils.



Vaseline Ribbon Gauze Method:

- More useful in patients which have unusual anatomy (major septal deviation), a bleeding mass or are post-op nose procedure bleeds.
 - The reason is that **you can custom fit the pack to the necessary defect**, something the Merocel will not allow you to do.
- After you've examined and anaesthetized the mucosa, few centimeters of gauze are folded upon itself and inserted by forceps along the floor, and then the whole nasal cavity is packed tightly by layering the gauze from floor to the roof and from before backwards.
- Packing can also be done in vertical layers from back to the front .
- About 1 meter gauze (2.5 cm wide in adults and 12 mm in children) is required for each nasal cavity.
- Two important things about Vaseline gauze that should be kept in mind:
 1. It really helps to see where you are layering the stuff, If you keep banging into the turbinates or septum it will bleed and hurt more.
 2. When you layer it, place the strips onto the floor of the nose and gently press each layer into the next so that the whole pack becomes one solid thing, This will ensure that it's tight enough to stop the bleeding and more importantly that it's **not too loose** at the back and starts to hang down the patient's nasopharynx, If that happens, you have to cut it in their mouth or worse, take it out and re-do the pack since they could swallow or aspirate the thing!
- Once both sides are nicely packed apply a moustache dressing.



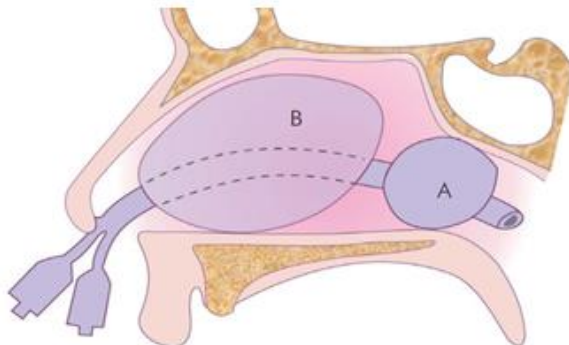
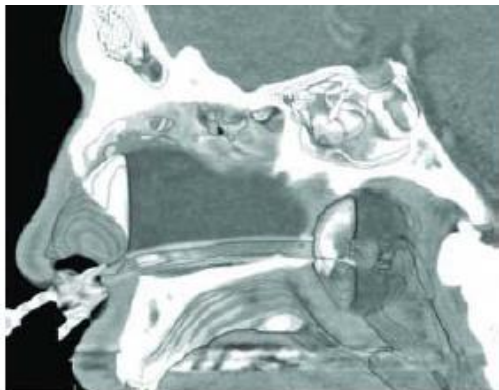
- **Posterior Nasal packing (Postnasal Pack):**
- Indications:
 - Active posterior epistaxis that doesn't stop with appropriate-sized bilateral anterior nasal packing.
- Types:
 1. Vaseline Ribbon Gauze
 2. Foley catheter
 3. Pneumatic nasal catheter (2 balloons)
- Posterior nasal packing is placed to close off the choana to prevent escape of bleeding into the nasopharynx.
- Good bilateral anterior packing (Merocel or Ribbon gauze is actually a type of posterior pack.
- Posterior packing can theoretically induce **naso-pulmonary reflex**, leading to:
 - Hypercarbia.
 - Hypoxia.
 - Decreased lung volume.
 - Bronchoconstriction.
 - Cardiac arrhythmias.
- For that reason, patients requiring posterior packs should be hospitalized for:
 - Continuous cardiopulmonary monitoring.
 - Antibiotics.
 - Oxygen supplementation if needed.
 - Mild sedation/analgesia.
 - IVF.
- Advantages:
 - Can control severe bleed in the emergency room or office (life-saving).
- Disadvantages:
 - Risk of airway compromise (requires hospital monitoring).
 - Requires patient cooperation (painful).
 - May require intubation or general anesthesia.
 - Eustachian tube dysfunction (hearing loss).
 - Other risks similar to anterior nasal packing.

Foley's Catheter Posterior Nasal Packing:

- Very quick & potentially lifesaving way to pack the nasopharynx.
- Prepare:
 - Two Foley's size 12-14 F (test the balloons).
 - Prefilled water syringes.
 - Plastic clamps and some 2 by 2 gauzes.
- Place one in each nostril.
- Inflate the balloons about 10 cc and pull them out until there is moderate resistance (don't pull so hard).
- At this point you can do an **anterior pack** with Vaseline gauze or Merocel.
- Secure the Foley's with clamps.
- Cut the non-clamped portion off.
- Make sure you pad the skin contact points (avoid necrosis).

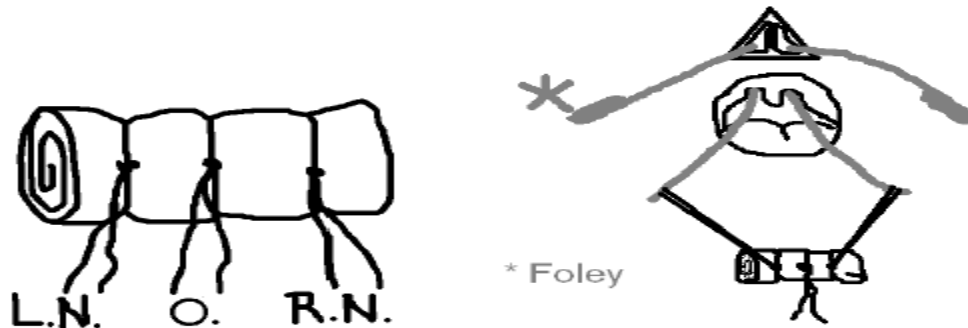
Pneumatic nasal catheter:

- Catheter with two balloons.
- One placed in the nasopharynx and the other in the nasal cavity.
- Designed for easier placement of a posterior pack.
- Provides less trauma and is simple to adjust pressure.



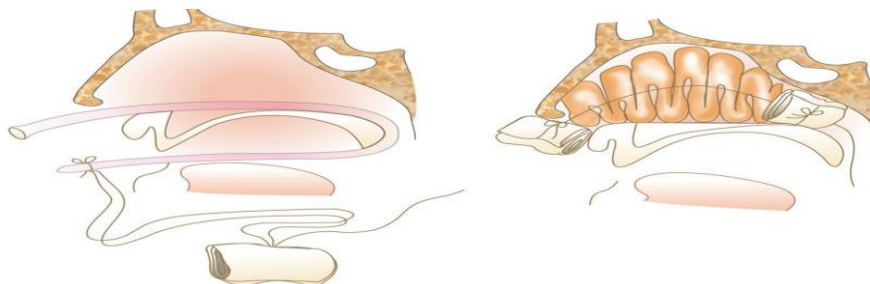
Vaseline Ribbon Gauze Posterior Packing:

- **Prepare:**
 - Single 4 by 4 folded into thirds.
 - 2 small Foley's catheters.



(**LN:** Left Nostril, **O.:** Oral/Mouth, **RN:** Right Nostril)

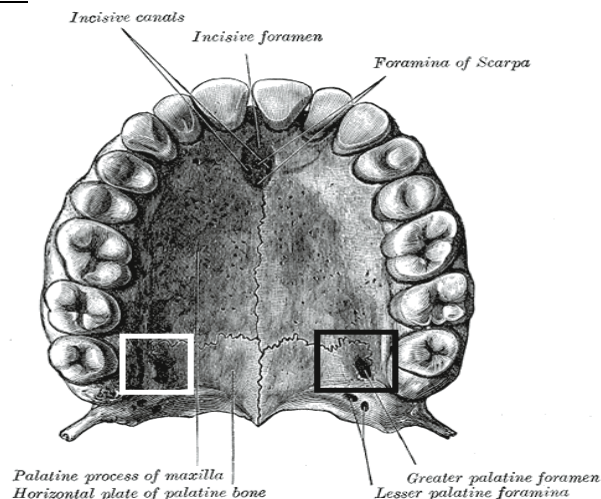
- Spray the nose and oropharynx with xylocaine spray, insert the cotton/gauze with local and Otrivin.
- Insert a foley into the left nostril and grab the end of it inside the mouth with a bayonet forceps.
- Pull it out just enough so you can tie the „LN“ suture to its end.
- Do the same thing on the right side.
- **When both the sutures are tied on, moisten the pack with Fucidine:**
 - This makes it easier to insert and prevents infection that often happens within 24-48 hours.
- Then with your dominant hand holding both foleys out of the nose and the other hand holding the pack at your index and middle finger tips, pull the foleys out while simultaneously pushing the pack behind the uvula/soft palate.
- It should be snug and you shouldn't see any of the pack in the back of the mouth.
- Now you need to tie the 2 silk sutures to the columella (middle of the nostrils).
- **Make sure you put a rolled „2 by 2“ under the knot to prevent necrosis of the suture into the skin.**
- Remember that the 3rd or middle silk suture has to come out of the patient's mouth and you need to tape it securely onto their cheek.
- This is the one you'll use to pull the pack out usually in 72 hours.
- Once tied, you now just pack the nose with Vaseline gauze as above.



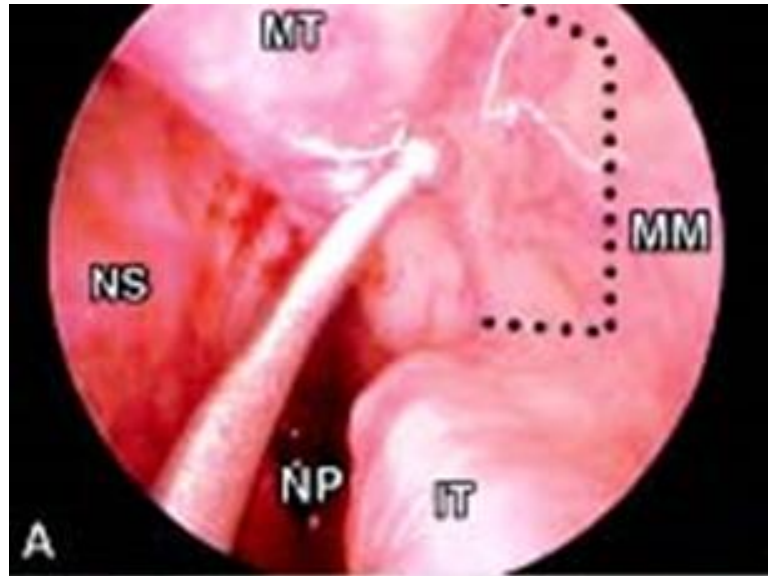
- **If a proper posterior nasal packing didn't stop the bleeding, consider the following options:**
 1. **Embolization.**
 2. **Surgery for ligation.**

SURGICAL MANAGEMENT

- Indications:
 - Uncontrolled epistaxis & failure medical management after 72 hours.
 - Identifiable bleeding site.
 - Patient requires transfusion.
 - Nasal anomaly precluding packing.
 - Patient refusal/intolerance of packing.
- Types:
 - **Sphenopalatine artery ligation:**
 - Trans-antral vs. Endoscopic.
 - **Ethmoid artery ligation:**
 - Lynch frontoethmoidectomy approach vs. Endoscopic.
 - **Internal Maxillary artery ligation:**
 - Trans-antral vs. Endoscopic.
 - **External carotid artery ligation:**
 - Trans-cervical.
- 1. **Sphenopalatine artery ligation (Endoscopic Approach):**
 - Trans-palatal injection of the greater palatine foramen is performed using local anesthetic agent to minimize the bleeding "maxillary vasospasm" after removal of the posterior pack "The landmark is the point midway between the second molar tooth and the midline of the palate".



- Make a vertical mucoperiosteal incision immediately behind the posterior fontanelle (1cm anterior to the posterior insertion of the middle turbinate to the lateral wall).
- The incision should be about 1cm in length and should extend from high up in the middle meatus (at the level of the basal lamella) to the inferior turbinate.



- Dissect submucosally in a posterior direction from the vertical incision behind the posterior fontanelle.
- The first structure to be encountered is the **Crista ethmoidalis**.
 - Part of the perpendicular process of the Palatine bone.
 - It is the most important landmark as the SPA is located just posterior to it.

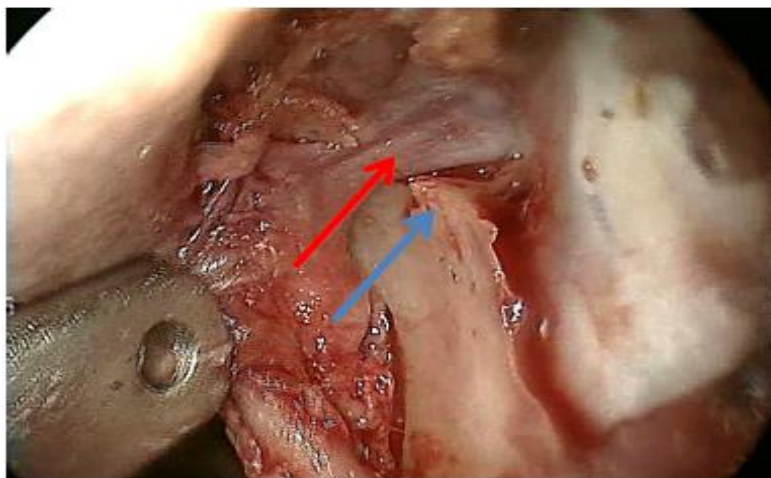


Figure 6: Blue arrow points to crista ethmoidalis; SPA is visible behind it (red arrow)

- Take care when dissecting around this area not to accidentally disrupt the SPA as this can cause significant bleeding.
- Elevate a mucoperiosteal flap to expose the SPA as it exits its foramen.
- Use a Kerrison punch or Up-cut to remove the crista ethmoidalis and to follow the SPA laterally into the pterygo-palatine fossa. Identify the main or anterior branch of the SPA.

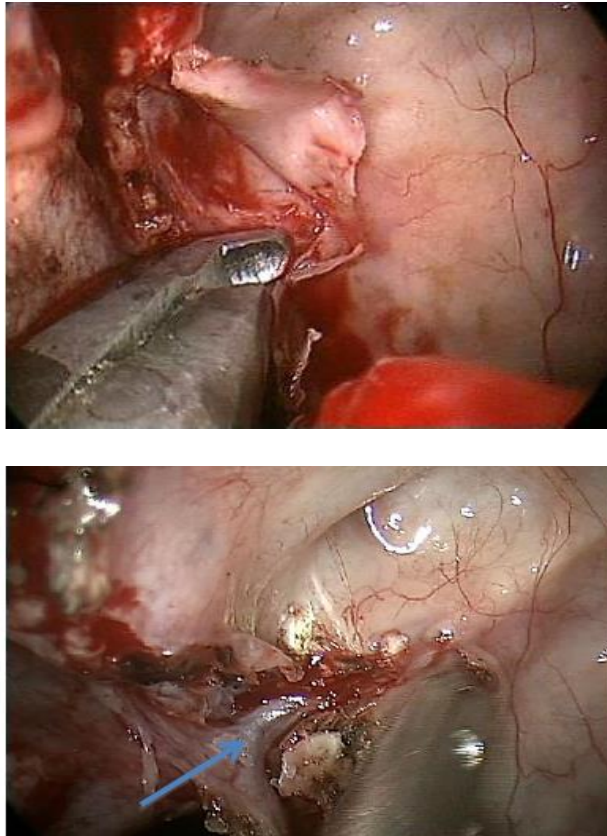


Figure 9: The most anterior or main branch of the SPA is identified (blue arrow) and followed into the pterygo-palatine fossa for a few millimetres

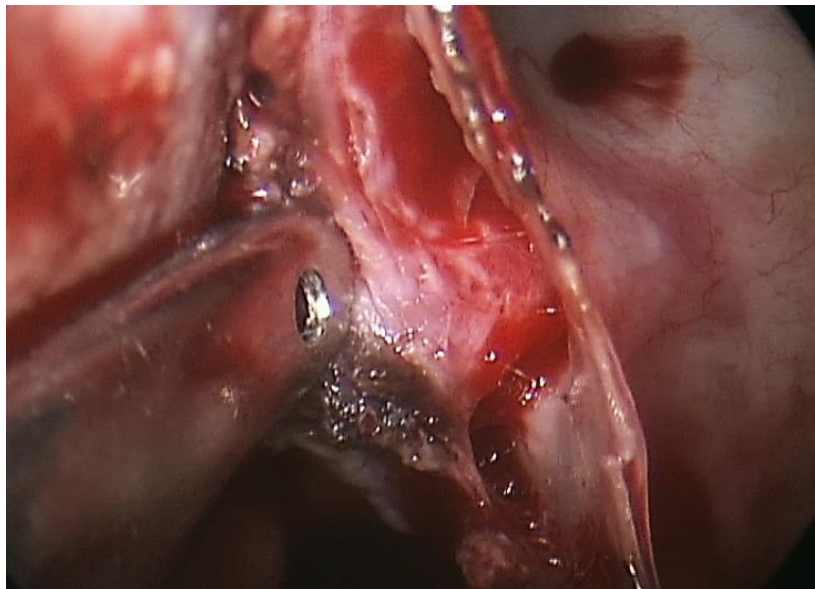
- Either cauterize the vessel with bipolar cautery or apply a ligaclip.
- The author prefers bipolar cautery as Ligaclips often are dislodged.
- Transect the artery to provide access to determine the presence of more arterial branches posteriorly, superiorly and inferiorly.



Fig. 10: Two ligaclips applied to the proximal end of the SPA. The artery was cut to find posteriorly-located branches

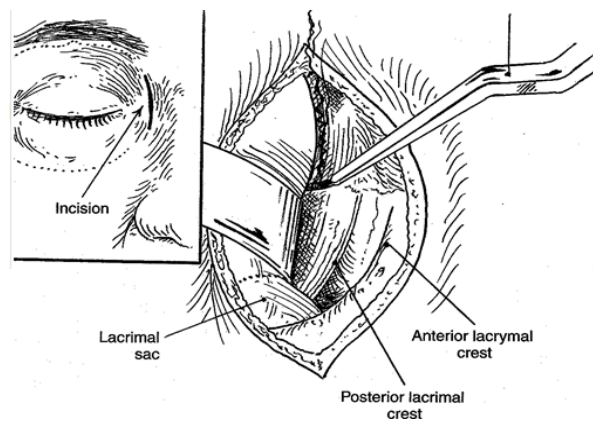
- Replace the mucosal flap and place a small piece of Surgicell over the mucosal flap; no additional nasal packing is required.
- Observe the patient in hospital over-night.

- **Alternative technique in the event that the SPA is not found:**
- It may be difficult to locate the SPA especially in the following circumstances:
 - Edematous nose secondary to nasal packing.
 - Anatomical anomalies (deviated nasal septum, large concha bullosa etc.).
 - Limited space in the nasal cavity due to a combination of the above factors and/or large turbinates.
- If the tip of the endoscope cannot be positioned in the posterior part of the middle meatus and limited space prevents one from making the mucosal incision, the following should be done:
 - Do a **standard uncinectomy**, preserving the superior part of the uncinete.
 - **Enlarge the maxillary sinus ostium posteriorly** using a through-cutting Blakes-ley forceps until the **posterior wall of the maxillary sinus is** encountered.
 - This provides sufficient access so that the mucoperiosteal flap can be raised from the level of the posterior wall of the maxillary sinus.
 - Occasionally a limited septoplasty or reduction of a concha bullosa is required to gain adequate access to the posterior middle meatus.
- 90-100% success rate with low complication rate.
- See video <http://www.sinusvideos.com/endoscopic-ligation-of-spa-sphenopalatine-artery/>

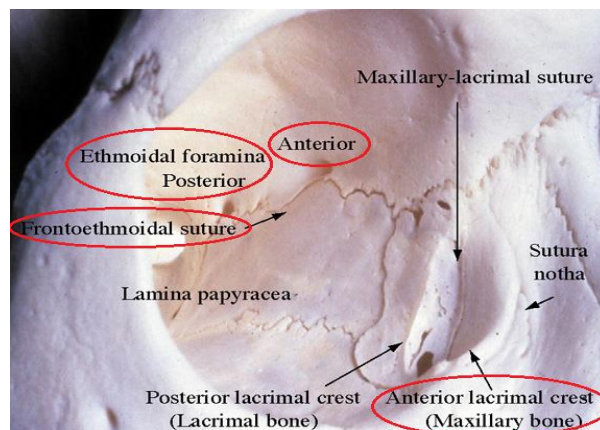


2. Ethmoid Artery Ligation:

- Anterior ethmoid artery bleeding usually cause anterosuperior bleeding above the middle turbinate.
- Epistaxis from anterior ethmoid artery is usually occurs from:
 - **Facial trauma** with fractures through the skull base that avulse or lacerate the anterior ethmoid artery can occur.
 - **Post-ESS** with injury to the AEA may not respond to packing.
- Epistaxis from posterior ethmoid artery is rare as the artery is most commonly runs in the skull base.
- Approaches for Ethmoid artery ligation:
 - **Endoscopic:**
 - Usually performed when Anterior Ethmoid Artery is injured during endoscopic sinus surgery.
 - **Lynch incision:**
 - Begins at the inferior margin of the medial aspect of the eyebrow and extends inferiorly along the lateral nose midway between the nasal dorsum and the medial canthus to the level of the medial canthus.



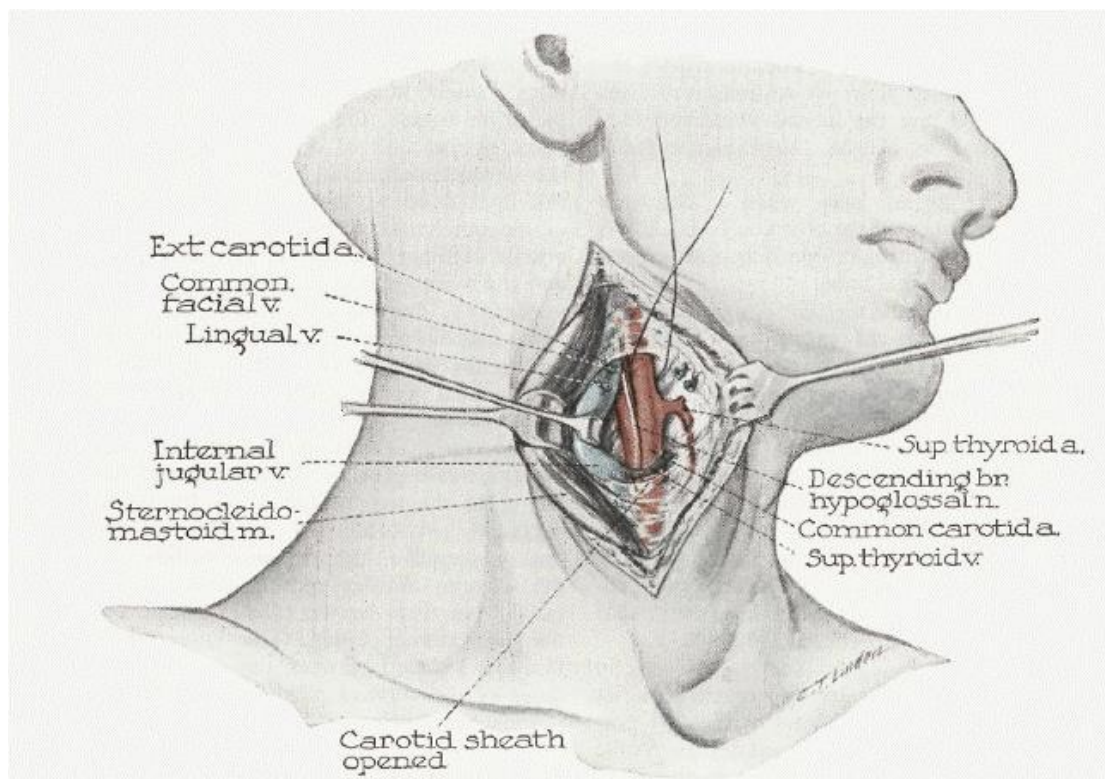
- Anterior Ethmoid artery is located **24 mm** posterior to **anterior maxillary crest** at the level of frontoethmoidal suture line.
- Posterior Ethmoid Artery is located **12 mm** posterior to Anterior Ethmoid Foramen.
- Optic Nerve is located **6 mm** posterior to Posterior Ethmoid Foramen.



- Complications:
 - 1. Epiphora:**
 - Injury to the nasolacrimal apparatus.
 - 2. Diplopia:**
 - Temporary is common and usually self-limited as edema resolves.
 - Horizontal gaze diplopia, however; may result as a consequence of trauma to the medial rectus muscle with subsequent fibrosis.
 - Blindness is a potential consequence.
 - 3. Retro-bulbar hemorrhage.**
 - 4. Optic nerve injury.**
 - 5. CSF leak:**
 - If the fovea ethmoidalis or cribriform plate is disrupted.
 - 6. Supratrochlear nerve injury:**
 - Cause anesthesia, hypesthesia, or neuralgia of the median forehead.
 - 7. Hypertrophic scar formation.**
- 3. Internal Maxillary Artery Ligation:**
 - Rarely used now as SPA ligation is available.
 - **Two branches of IMAX must be ligated in pterygopalatine fossa as well to prevent recurrent epistaxis from collaterals:**
 - 1.** Sphenopalatine artery.
 - 2.** Descending palatine artery.
 - Approaches:
 - **Endoscopic trans-pterygoid:**
 - Performed by doing mega maxillary antrostomy followed by entering the pterygopalatine fossa through posterior maxillary wall.
 - IMAX is very tortuous vessel with multiple branches, easy to miss main trunks.
 - **Caldwell-Luc:**
 - Traditional Caldwell-Luc approach followed by entering the pterygopalatine fossa through posterior maxillary wall.
 - Failure rate of 10-20%.
 - Complication rate of 15-20%, including:
 - Facial paresthesia and pain infraorbital nerve
 - Dental pain and numbness
 - Hematoma
 - Oroantral fistula
 - Sinusitis
 - Blindness (rare)

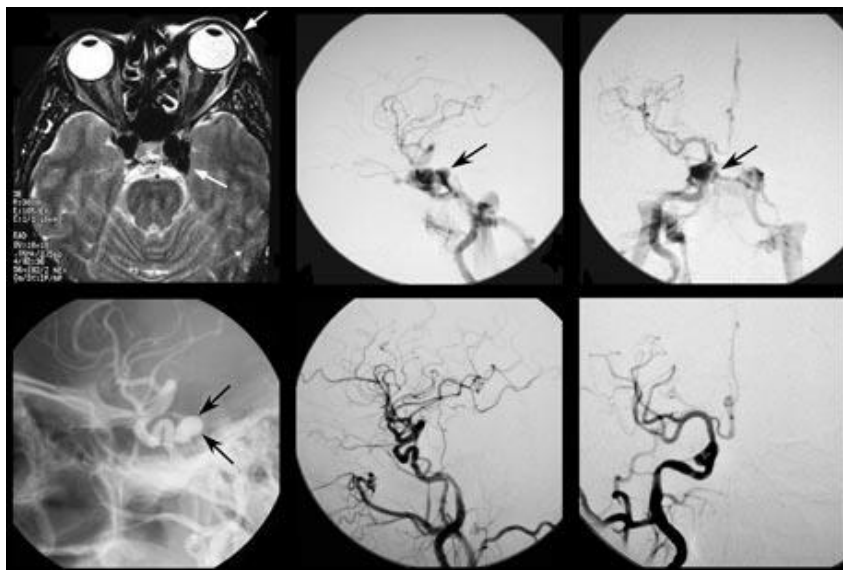
4. **External Carotid Artery Ligation:**

- Rarely used now due to the improvement in the endoscopic approaches for ligation of terminal branches and embolization.
- **Low success rate of only 55%** due to possible recurrence of collateral circulation (because of ICA system contributions).
- Indications:
 - Severe uncontrolled, life-threatening bleeding.
 - Lack of experts in endoscopic approaches or interventional radiology.
- Trans-cervical Approach:
 - Vertical incision is made along the anterior border of Sternocleidomastoid muscle between hyoid and superior border of thyroid cartilage.
 - Sub-platysmal flap elevation is performed.
 - Sternocleidomastoid muscle is retracted posteriorly to visualize internal jugular vein, which is retracted to identify the carotid arteries.
 - Since the origin of external carotid artery lies medial to the internal carotid artery, the 2 vessels may be mistaken unless it is remembered that the external carotid is the only vessel that gives off branches in the neck.
 - More than two branches should be identified to confirm external carotid artery from internal carotid artery, and then **external carotid artery is ligated above the origin of superior thyroid artery (or lingual artery).**



EMBOLIZATION:

- Indications:
 - Similar to the indications of surgical ligation plus:
 - Surgically inaccessible sites.
 - Inoperable candidates.
 - Re-bleeding after surgical arterial ligation.
- Effective only when bleeding is >0.5 ml/min.
- More than 90% success rate with low complication rate of 0.1%.
- Only able to embolize external carotid artery and its branches.
- Advantages:
 - Diagnostic (defines bleeding site) and therapeutic.
 - Can be repeated.
 - Can be done under local anesthesia.
- Disadvantages:
 - Risk of embolic event (pulmonary emboli, stroke).
 - Requires active bleeding.
 - Complications.
- Complications:
 - Minor (18-45%)
 - Major (4%)
 - Cerebral embolization with hemiplegia.
 - Blindness.
 - Ophthalmoplegia
 - Skin Necrosis.
 - Facial pain/edema.
 - Groin hematoma (most common) "catheter site "
- **Contraindications:**
 - Severe atherosclerosis.
 - Ethmoid bleed.



Osler-Weber-Rendu Syndrome (Hereditary Hemorrhagic Telangiectasia/HHT):

- Pathophysiology:
 - Autosomal dominant "chromosome 9" → defect in contractile elements (elastic and muscular layers) of vessels, results in arteriovenous malformations.
- Clinical picture:
 - Characterized by ectatic vessels of the skin, mucous membranes, and viscera.
 - Diagnosed by "Curacao criteria".

**TABLE
32.2**

**CURAÇAO CRITERIA FOR THE
CLINICAL DIAGNOSIS OF HHT**

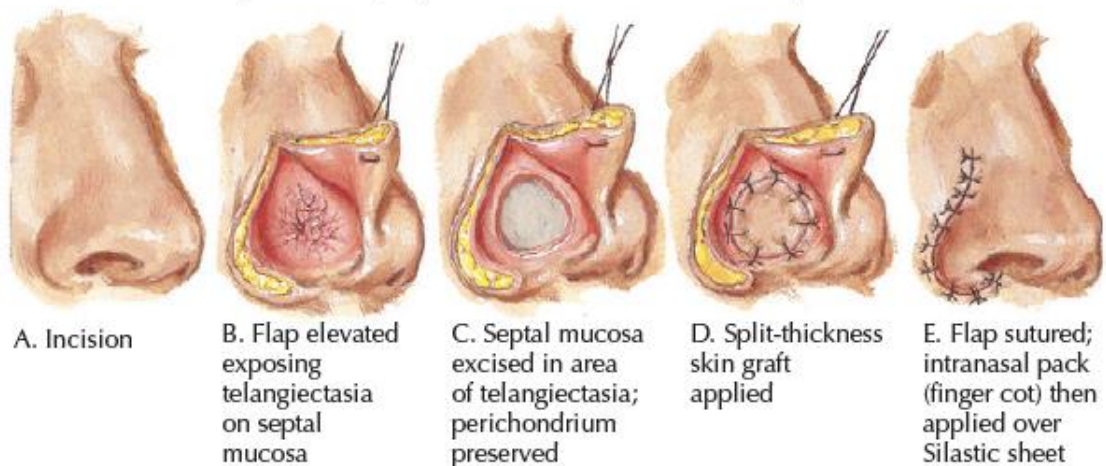
Criteria	Description
Epistaxis	Recurrent
Telangiectasia	Multiple at characteristic sites (lips, oral cavity, fingers, nose)
Visceral lesions	GI telangiectasia; pulmonary, cerebral, hepatic, spinal AVMs
Family history	A first degree relative with HHT according to these criteria
0–1 criteria	Unlikely
2 criteria	Possible or suspected
3 or more criteria	Definite



A HHT patient with telangiectasias over right anterior septum and Inferior turbinate

- Complications:
 - Morbidity and mortality due to multi organ arteriovenous malformations, and associated hemorrhages pulmonary, GI & CNS bleeds.
- Treatment
 - **Medical therapy:**
 - Episodes of epistaxis can be treated with high-dose antifibrinolytic agents (tranexamic acid).
 - **Septoplasty:**
 - Just elevation of mucoperichondrial flap and then repositioning it back helps to cause fibrosis and constrict blood vessels.
 - Also, removing any septal spur which is sometimes the cause of epistaxis.
 - **Septodermoplasty (Saunders' Dermoplasty):**
 - Done by removing the diseased mucosa from anterior $\frac{1}{2}$ septum, floor of nose, lateral wall and replace it with STSG (split thickness skin graft), cutaneous, myocutaneous, microvascular free flaps, or autografts.
 - **Radiofrequency coblation**
 - **Laser ablation (KTP)**
 - **Embolization**
 - **Young's procedure:**
 - Definitive treatment for severe disease.
 - Nostrils are sealed off completely.

Septal Dermoplasty for Recurrent Severe Anterior Epistaxis





right Internal valve following septodennoplasty with replacement of the mucosa using an allograft.

- **Preventive and Chronic Management:**
- Moisture to reduce crusting and bleeding:
 - Saline spray.
 - Petroleum ointment.
 - Emollients.
 - Humidification.
- Avoid local trauma:
 - Digital manipulation.
 - Nose blowing.
 - Excess straining.
- Antimicrobial ointment to excoriated lesions.
- Long term control of HTN.
- Educate about positioning of nasal sprays away from septum.

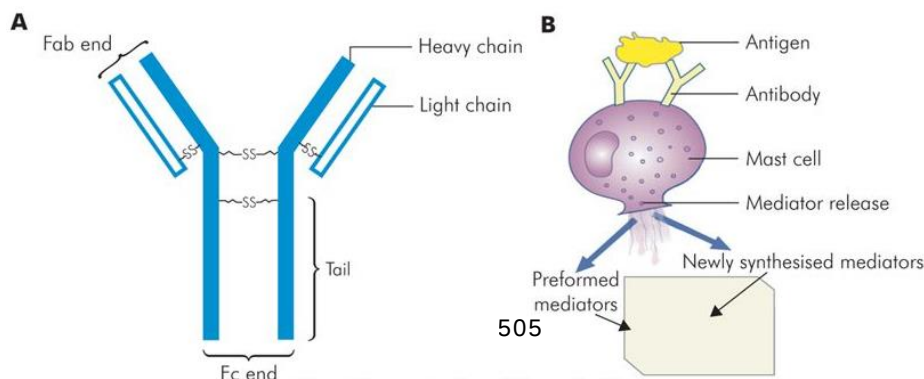
Allergic Rhinitis:

- **IgE-mediated** immunologic response of nasal mucosa to air-borne allergens.
- No age or sex predilection.
- May start in infants as young as 6 months or older people.
- Usual onset is at 12-16 years of age.
- **Etiology:**
- Inhalant allergens are often the cause.
 - o Pollen from trees and grasses, mould spores, house dust, debris from insects or house mite are common offenders.
- Food allergy is rarely an important cause.
- Genetic predisposition plays an important part.
 - o If one parent is Allergic, **20%** risk of developing allergy.
 - o If both parents are Allergic, **50%** risk of developing allergy.
- **Pathogenesis:**
- After initial antigen exposure, Antigen-presenting cells (Macrophages, HLA class II) present processed peptides to T-Helper cells (CD4) releasing Interleukins (IL-4 and IL-13).
- IL-4 and IL-13 favor B-cell transformation to sensitized Plasma cell with specific IgE production.

- Clinical phases of allergic response:

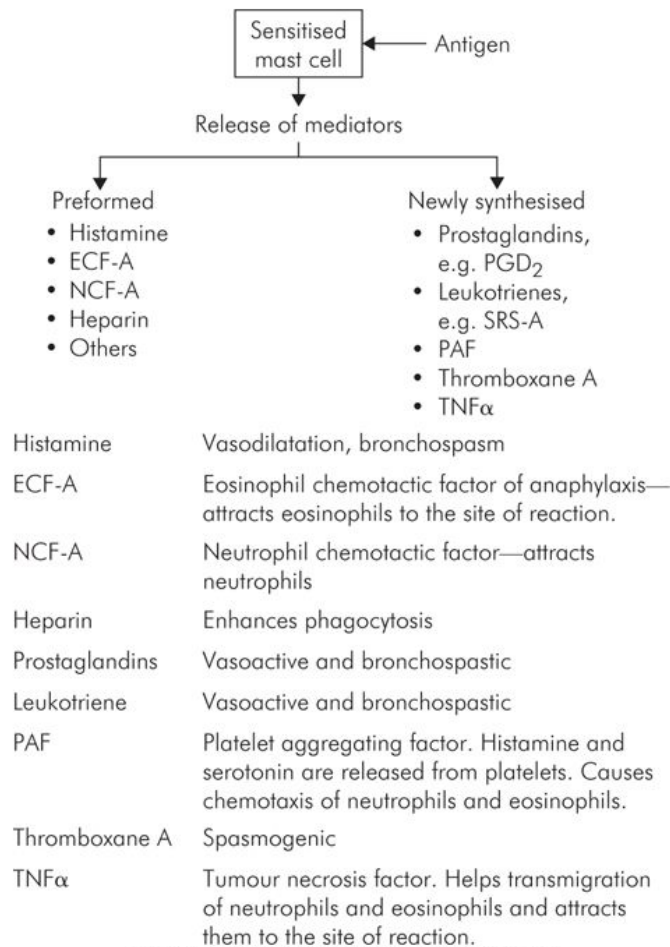
1. **Acute (Early) Phase:**

- o Type I immediate hypersensitivity.
- o Occurs immediately within 5-30 min after exposure to the specific allergen.
- o **Antigen** is attached to IgE by its **Fab End**.
- o **Mast cells** are attached to IgE by its **Fc End**.
- o Degranulation of Mast cells occur with release of several chemical mediators (Histamine, Serotonin, Proteases, Leukotrienes, Prostaglandins) responsible for symptomatology of allergic disease:
 - Vasodilation.
 - Mucosal edema.
 - Infiltration with eosinophils.
 - Excessive secretion from nasal glands.
 - Smooth muscle contraction.
- o Rechallenged allergens stimulate mast cells more quickly and require less antigen load.



2. Late (Delayed) Phase:

- Occurs 2-8 hours after exposure to allergen without additional exposure.
- Asymptomatic phase prior to late phase.
- Due to infiltration of inflammatory cells (Eosinophils, Neutrophils, Basophil, Monocytes and CD4) at site of antigen deposition causing swelling, congestion, thick secretion.
- IL-5 play an important role.
- Mast cells do not remain active.
- **Eosinophilia is the hallmark of Allergic response.**



- Two clinical types of AR:

1. Seasonal:

- < 4 days/week, or
- < 4 weeks/year.

2. Perennial:

- > 4 days/week, or
- > 4 weeks/year.

TABLE 1–4. Hypersensitivity Types

	Type	Mediators	Reaction
I:	Anaphylactic	IgE	• immediate, self-limiting • IgE mediated, stimulates mast cells and basophils which release histamine and other inflammatory mediators
II:	Cytotoxic	IgG, IgM	• IgG, IgM multivalent binding to phagocyte or complement • eg, Transfusion reactions, Goodpasture's Syndrome, Bullous Pemphigoid
III:	Immune Complex	IgG, IgM, IgA	• antibody and complement complexes cause increased blood viscosity • removed by reticulo-endothelial system • eg, Renal Deposition, Arthritis, Glomerulonephritis, Serum Sickness
IV:	Cell-mediated	T-cells	• Delayed-type Hypersensitivity reaction (T-cell mediated) • eg, Graft Rejection, Contact Dermatitis
V:	Interference with Receptor	Ig	• antibody “resembles” a ligand and thus blocks or stimulates the receptor • pathophysiology of autoimmunity (eg, Hashimoto's Thyroiditis, Myasthenia Gravis, Graves' Disease)

Table 1 – Basic characteristics of classes of immunoglobulins

Class	Structure	Properties
IgA	Dimeric and Monomeric	Found in gastrointestinal, respiratory and urogenital tract mucosa. Prevents the colonization by pathogens. Also present in saliva, tears and milk.
IgD	Monomeric	Membrane immunoglobulin. It is part of the membrane receptor of naïve B lymphocytes (BCR).
IgE	Monomeric	Involved in allergic and parasitic processes. Its interaction with basophils and mastocytes causes histamine release.
IgG	Monomeric	Main immunoglobulin of acquired immunity. It has the capacity to cross the placental barrier.
IgM	Monomeric Pentameric	It is part of the membrane receptor of naïve B lymphocytes (BCR). Form found in the serum, secreted early in acquired immune response.

- **Diagnosis of Allergic Rhinitis:**
- History:
 - Nasal SSx:
 - Sneezing, congestion, watery rhinorrhea, itching
 - Ocular SSx:
 - Redness, itching, epiphora, conjunctivitis, burning
 - Otologic SSx:
 - Aural fullness (ET dysfunction),, OME
 - Laryngeal SSx:
 - Scratchiness, dry, irritated, cough
 - Oral SSx:
 - Palatal itching, hypogeusia
 - Associated Disorders:
 - Chronic rhinosinusitis (obstruction from mucosal edema), nasal polyps, asthma, OME
- Physical exam:
 - Eyes:
 - Periorbital puffiness, darkening of skin under eyes (Allergic shiners) from venous congestion, fine creases in eyelids (Dennie's lines), conjunctival injection.
 - Ears:
 - OME, CHL
 - Nose:
 - watery rhinorrhea, congested turbinates, nasal tip transverse crease (Allergic salute) from chronic nose wiping, nasal twitching (from itching), nasal obstruction relieved after nasal decongestant.
 - Mouth:
 - Mouth breather.
 - Throat:
 - Prominent pharyngeal lymphoid tissue (Cobblestoning).
- Adjunctive Testing:
 - **Nasal Endoscopy:**
 - Evaluate for nasal polyps, osteomeatal unit obstruction, adenoid tissue.
 - **Nasal Smear:**
 - Obtained from inferior turbinate mucosa.
 - >25% eosinophils on nasal smear suggests allergy.
 - Neutrophils suggest infection.
 - **Total Serum IgE:**
 - Not always accurate or cost-effective.

- **Skin Allergy Testing:**
- Best test for AR.
- Most convenient and least expensive screening method.
- Avoid antihistamine 48-72 hours prior to testing (increases false negative).
 - **Scratch Test:**
 - Scratch skin with placement of allergen or scratch with allergen.
 - Largely been replaced for more objective and more reliable techniques.
 - **Prick Test:**
 - Series of allergens are inserted by needle into skin.
 - Positive "wheal-and-flare" reactions are compared to controls.
 - Rapid and safe test.
 - Risk of anaphylaxis.
 - Misses less sensitive allergy.
 - Grading is subjective.
 - **Intradermal Test:**
 - Similar to prick test except allergen is placed intradermally.
 - More sensitive than prick test.
 - More time consuming and painful.
 - Greater risk of anaphylaxis.
 - Subjective grading.
 - **Skin Endpoint Titration:**
 - Series of increasing concentrations of specific allergen are introduced intradermally to titrate to a positive response.
 - Useful for determining immunotherapy concentrations.
 - Highly sensitive.
 - Determines quantitative measurements.
 - Time-consuming.
- **In Vitro Allergy Testing:**
- **Indications:**
 - Equivocal skin test results
 - High risk of anaphylaxis (severe asthma, prior history).
 - Skin disorders (eczema).
 - Uncooperative patient (children and infants).
 - Failed immunotherapy (negative skin test is not an indication for in vitro allergy testing).
- **Advantages:**
 - Highly specific.
 - No risk of anaphylaxis.
 - No effect from skin condition (skin color, eczema, dermatographia) or medications (B-blockers, antihistamines, tricyclics).

- Disadvantages:
 - Less sensitive.
 - Requires up to 1–2 weeks for results.
 - More expensive.
- Radioallergosorbent Test (RAST):
 - Serum reacts with a series of known allergens.
 - Radiolabeled anti-IgE identifies specific antigen-IgE complexes.
- Enzyme-linked Immunosorbent Assay (ELISA):
 - Similar to RAST except fluorescing agents are used for markers of antigen-IgE complexes.
- Complications of allergic rhinitis
 1. Recurrent sinusitis because of obstruction to sinus ostia.
 2. Nasal polypi.
 3. Serous otitis media.
 4. Orthodontic problems and other ill-effects of prolonged mouth breathing especially in children.
 5. Bronchial asthma. Patients of nasal allergy have 4 times more risk of developing bronchial asthma.

Management:

- Anaphylaxis:
 1. ABC's: establish airway, oxygenation, and IV access
 2. Inject up to **0.3 ml of Epinephrine** intramuscularly (IM)
 3. Consider **Dopamine** for hypotension
 4. If needed repeat injection of up to 0.3 ml of epinephrine IM
 5. Add **Diphenhydramine 50 mg**, **Dexamethasone 4 mg**, and **Cimetidine 300 mg** through IV
 6. If needed repeat injection of up to 0.3 ml of epinephrine IM.
- Level 1a: Avoidance:
 - Avoidance is based on specific allergy (most successful if the antigen involved is single).
 - Dust: mattress cover, foam pillows, plastic case, low carpet or hardwood floors, frequent dusting and vacuuming, consider synthetic carpets
 - Molds: disinfect bathroom, clean furnace, dehumidify basement, clean refrigerator, avoid gardening, address potential sources of molds indoors. Pollens: air conditioning, air cleaners, keep windows closed, avoid cutting grass
 - Animals: keep out of bedroom, use special shampoos high efficiency particulate air filters may be used for all airborne allergies
 - home humidity prevents nasal dryness
 - masks may be helpful in unavoidable allergy exposure

- **Level 1b: Symptomatic Relief:**

- Nasal Saline Irrigations:
 - Removes nasal mucus and crusts.
 - Aids in mucociliary clearance.
 - Thins tenacious mucus.
- Nasal Antihistamines (H1-receptor Antagonists):
 - Not very potent.
 - Ex: Azelastine.
- First Generation Antihistamines (H1-receptor Antagonists):
 - Primarily for sneezing, itching, and rhinorrhea.
 - Side effects include sedation, dryness, confusion tolerance, and aggravation of prostatism.
 - Ex: Diphenylhydramine, Chlorpheniramine, Promethazine, Hydroxyzine.
- Second Generation Antihistamines (H1-receptor Antagonists):
 - Lipophobic (Not cross blood-brain barrier).
 - Less sedating.
 - Inhibit release of inflammatory mediators.
 - Ex: Loratadine, Astemizole, Certirizine, Fexofenadine, Terfenadine.
- Nasal Decongestant (Adrenergic Agonist):
 - Vasoconstrictive effect.
 - Rapid mucosal decongestion.
 - Side effects include central nervous system stimulation (anxiety, anorexia), cardiac arrhythmias, hypertension, seizures, insomnia, psychosis.
 - Must limit use to 3-5 days to prevent rebound congestion, tolerance and rhinitis medicamentosa.
 - Ex: Phenylamines and Oxymetazoline.
- Oral Decongestant (Adrenergic Agonist):
 - Vasoconstrictive effect.
 - Reduced potency.
 - More systemic side effects.
 - Avoid risk of rhinitis medicamentosa.
 - Ex: Phenylephrine and pseudoephedrine.

- Topical Corticosteroids:
 - Local reduction of inflammatory cells in nasal mucosa, decreased capillary permeability, reduces edema.
 - Single most effective maintenance therapy for AR.
 - Take > 1 week for maximal effect.
 - Use regularly for maximum benefit.
 - Break their use for 1-2 weeks every 2-3 months.
 - Minimal side effects (epistaxis, candidiasis, nasal dryness)
 - Decreases both acute and late phase reactions
 - < 2% go systematically.
 - Oral steroid used only for severe disease.
 - Ex: Fluticasone, Belcomethasone.
- Nasal Anticholinergics:
 - Specifically useful for rhinorrhea.
 - Also indicated in vasomotor rhinitis and viral rhinitis.
 - Ex: Ipratropium bromide.
- Oral Antileukotrienes:
 - Competitive inhibitor of leukotrienes receptors on smooth muscles throughout airway.
 - inhibits the late phase reaction decreasing eosinophil infiltration.
 - considered for combination therapy.
 - Ex: Montelukast.
- Nasal Mast Cell Stabilizers:
 - Stabilizes mast cells preventing release of mediators in acute and late phase reactions.
 - Effective only for prophylaxis.
 - Minimal side effects (sneezing, epistaxis, nasal irritation).
 - Safe to use in pregnancy (?)
 - Ex: Cromolyn
- **Level 2: Management of Complicating Factors:**
 - Must evaluate and treat potential concurrent disorders, which may mimic allergy including vasomotor rhinitis, sinusitis, and rhinitis medicamentosa before changing treatment regimens.

- **Level 3: Chronic Severe Symptoms (Corticosteroids):**

- Most potent medication for symptomatic relief of severe persistent AR.
- May be given orally, nasal aerosols, or via intratubinal injections (cause bleeding in injected side and facial flushing in 2%, blindness in 0.006%).
- Mechanism of Action: decreases inflammatory migration, blocks arachidonic metabolites, decreases vascular permeability.
- Side Effects of Oral Corticosteroids: increased gastric acid production (consider prophylactic concurrent H₂-blocker), hypertension, masks signs of infection, sodium retention, hypokalemia, posterior subcapsular cataracts, central nervous system stimulation (psychosis, seizures, insomnia), menstrual irregularities, aseptic necrosis of femoral head.

- **Level 4: Immunotherapy:**

- A method of desensitization that utilizes a controlled dosing of specific allergens to reduce the symptoms of AR and the complication of allergies.
- Indications:
 - Severe, persistent symptoms.
 - Unresponsive to maximal medical therapy.
 - Allergens that cannot be avoided (patient doesn't want long term Rx).
 - Coexisting asthma.
- Advantages: suppresses allergy
- Disadvantages: patient must be reliable for multiple injections, requires a chronic regimen (3 years), risk of worsening symptoms and anaphylactic shock.
- Contraindications:
 - Pregnancy (anaphylaxis risks hypoxia in fetus), autoimmune disorders, immunological compromised patients, B blockers (increases sensitivity to allergens), easily avoidable allergens, noncompliant patients, food allergens, Hx of previous anaphylaxis.
- Mechanism of Action: uncertain, initial small doses of allergen cause a rise in allergen-specific IgG which prevents binding of IgE, IgE may also become "exhausted"
- It is discontinued if uninterrupted treatment for 3 years shows no clinical improvement.
- Subcutaneous immunotherapy is the most common.
- **Monoclonal Antibodies:**
 - Targeting specific IgE responsible for allergic symptoms.
 - Ex: Omalizumab.

Mild Intermittent	Moderate-Severe Intermittent	Mild-Moderate Persistent	Severe Persistent
			Short course oral corticosteroid 3-7 days
Intranasal cromolyn	Intranasal corticosteroid		
Oral or local nonsedative H ₁ -antagonist			
Intranasal decongestant (<3 days) or oral decongestant			
Topical ocular cromone or antihistamine/cell stabilizer if ocular symptoms			
Allergen and irritant avoidance			
			Immunotherapy

Non-Allergic Rhinitis:

- **Viral Rhinitis (Common Cold/Coryza):**
- Caused by Rhinovirus (most common), Coronavirus, Parainfluenza virus, Adenovirus, Enterovirus, Respiratory Syncytial Virus (RSV).
- Airborne droplets.
- Incubation period is 1-4 days.
- Illness lasts for 1-2 weeks.
- **Clinical features:**
 - Burning sensation at Nose.
 - Nasal stuffiness.
 - Rhinorrhoea.
 - Watery and profuse.
 - Mucopurulent due to secondary bacterial invasion.
 - Sneezing.
 - Low grade fever.
- **Treatment:**
 - Self-limiting and resolves spontaneously after 1-2weeks.
 - Bed rest
 - Hydration.
 - Symptomatic treatment
 - Antihistamine
 - Nasal decongestants
 - Analgesics.
 - Antibiotics when secondary infection supervenes.
- **Complications:**
 - Sinusitis, pharyngitis, tonsillitis, bronchitis, pneumonia and otitis media.

- **Bacterial Rhinitis:**
 - Typically secondarily infected viral rhinitis.
 - Pneumococcus, streptococcus or staphylococcus.
 - Clinical picture is similar to viral rhinitis except thick, greenish and foul smelling rhinorrhea.
 - Treatment is similar to viral rhinitis in addition to Antibiotic regimen.
- **Irritative Rhinitis:**
 - Acute rhinitis caused by exposure to dust, smoke or irritating gases such as ammonia, formaline, acid fumes, etc.
 - May result from trauma inflicted on nasal mucosa during intranasal manipulation, e.g. removal of a foreign body.
 - Immediate reaction with sneezing, rhinorrhoea and nasal congestion.
 - Symptoms may pass off rapidly with removal of the offending agent or may persist for some days if nasal epithelium has been damaged.
 - Recovery depends on the amount of epithelial damage and infection that supervenes.
- **Hypertrophic Rhinitis:**
 - Characterized by thickening of mucosa, submucosa, seromucinous glands, periosteum and bone, mainly over Turbinates.
 - Common causes are recurrent nasal infections, chronic sinusitis, chronic irritation of nasal mucosa due to smoking, industrial irritants, prolonged use of nasal drops and vasomotor and allergic rhinitis.
- **Clinical picture:**
 - o Nasal obstruction is the predominant symptom.
 - o Thick and stick nasal discharge.
 - o Headache, heaviness of head or transient anosmia.
 - o Hypertrophy of turbinates.
 - Turbinal mucosa is thick and does not pit on pressure.
 - Little shrinkage with vasoconstrictor drugs due to presence of underlying fibrosis.
 - Maximum changes are seen in Inferior turbinate.
- **Treatment:**
 - Attempt should be made to discover the cause and remove it.
 - Nasal obstruction can be relieved by reduction in size of turbinates by:
- **Mucosal Destructive Techniques:**
 - o **Partial or total turbinectomy.**
 - High risk of Atrophic rhinitis.
 - Generally not recommended.
- **Mucosal Preserving Techniques:**
 - o **Inferior turbinate outfracturing:**
 - Easy.
 - Poor long term results.
 - o **Submucosal Diathermy(SMD)/Radiofrequency/Ablation/Coblation:**
 - Office based.
 - Poor long-term results.

- **Submucosal Resection (SMR):**
 - Removes bony while preserves turbinal mucosa for its function.
 - Excellent long-term results.
- **Compensatory Hypertrophic Rhinitis:**
- Seen in cases of marked deviation of septum to one side.
- The roomier side of nose shows compensatory hypertrophy of inferior and middle turbinates to reduce the wide space to overcome effects of drying and crusting that always attend wider nasal space.
- Hypertrophic changes in these cases are **not reversible** with correction of nasal septum and require reduction of turbinates at time of septal surgery.
- **Atrophic Rhinitis (Empty Nose Syndrome/Ozanea):**
- Chronic inflammation of nose characterized by atrophy of nasal mucosa and turbinate bones.
- Nasal cavities are roomy and full of foul-smelling crusts
- **Pathology:**
 - Ciliated columnar epithelium is replaced by stratified squamous epithelium.
 - Atrophy of seromucinous glands, venous sinusoids and nerves.
 - Obliterative endarteritis of mucosal arteries, periosteum and bone.
 - Resorption of turbinates bone causing roomy nasal chambers.
 - Paranasal sinuses are small due to their arrested development
- **Causes:**
 - Excess nasal surgery (Excessive turbinectomy).
 - Suspected genetic factors:
 - More common in East Asia, Egypt, Greece.
 - Endocrinal disturbance:
 - Starts at puberty.
 - Females more than males.
 - Cease after menopause.
 - Nutritional deficiency:
 - Deficiency of vitamin A, D or iron.
 - Chronic bacterial infections and Irritant exposure:
 - Klebsiella ozaenae, (Perez bacillus), diphtheroids, P. vulgaris, Esch. coli, Staphylococci and Streptococci.

- Clinical picture:
 - Foul smell from Nose.
 - Merciful Anosmia (Patient is unaware of foul smell).
 - Nasal obstruction due to large crusts filling the nose.
 - Epistaxis when crusts are removed.
 - Roomy Nasal cavities with atrophy of turbinates.
 - Septal perforation and dermatitis of nasal vestibule may present
 - Nose may show a saddle deformity.
 - Atrophic Pharyngitis.
 - Cough and hoarseness of voice (Atrophic laryngitis).
 - Hearing-impairment due of obstruction to ET tube and OME.
 - Small and underdeveloped paranasal sinuses with thick walls.
- Prognosis:
 - Persists for years but there is a tendency to recover spontaneously in middle age.
- Treatment:
- **Medical:**
 - Moisture:
 - Nasal irrigation and removal of crusts.
 - Warm normal saline or an alkaline solution.
 - 25% glucose in glycerine.
 - Inhibits growth of proteolytic organisms which responsible for foul smell.
 - Vitamin A and D and iron supplements.
 - Local and systemic antibiotics for secondary infections.
- **Surgical:**
 - Young's operation:
 - Both nostrils are closed completely just within nasal vestibule by raising flaps.
 - Opened after 6 months or later.
 - Mucosa may revert to normal and crusting reduced.
 - Modified Young's operation:
 - To avoid the discomfort of bilateral nasal obstruction.
 - Aims to partially close the nostrils.
 - Claimed to give the same benefit as Young's.
 - Narrowing the nasal cavities:
 - Narrowing size of nasal airway helps to relieve the symptoms.
 - Techniques:
 - Submucosal injection of teflon paste.
 - Insertion of fat, cartilage, bone or teflon strips under the mucoperiosteum of floor and lateral wall of nose and mucoperichondrium of septum.
 - Section and medial displacement of lateral wall of nose.

- **Rhinitis Sicca:**
 - Crust-forming disease
 - Patients who work in hot, dry and dusty surroundings (bakers, iron- and goldsmiths).
 - Ciliated columnar epithelium undergoes squamous metaplasia with atrophy of seromucinous glands
 - Crusts form on anterior part of septum and their removal causes ulceration and epistaxis and septal perforation
 - Treatment consists of correction of occupational surroundings and application of bland ointment or one with an antibiotic and steroid.
 - Nose pricking and forcible removal of crusts should be avoided.
 - Nasal douche, like the one used in cases of atrophic rhinitis.
- **Rhinitis Caseosa:**
 - Uncommon, usually unilateral and mostly affecting males.
 - Nose is filled with offensive purulent discharge and cheesy material.
 - Arises from chronic sinusitis with collection of inspissated cheesy material.
 - Sinus mucosa becomes granulomatous.
 - Bony walls of sinus may be destroyed, requiring differentiation from malignancy.
 - Treatment is removal of debris and granulation tissue and free drainage of the affected sinus.
 - Prognosis is good.
- **Nonallergic Rhinitis with Eosinophilia Syndrome (NARES):**
 - Nasal eosinophilia without allergy.
 - Clinical picture:
 - o Symptoms of perennial rhinitis.
 - o Eosinophils on nasal smears.
 - o **Negative allergic tests.**
 - Treatment:
 - o Symptomatic relief similar to allergic rhinitis.
- **Vasomotor Rhinitis (VMR/Hyperreflexive Rhinopathy):**
 - Non-allergic rhinitis but clinically simulating allergic rhinitis.
 - Persists throughout the year.
 - All tests of nasal allergy are negative.
 - Pathophysiology:
 - o **Overactivity of parasympathetic system** which causes vasodilation and engorgement of nasal mucosa venous sinusoids and excessive secretion from nasal glands.
 - o Nasal mucosa is also hyperreactive and responds to several non-specific stimuli, e.g. change in temperature, humidity, blasts of air, small amounts of dust or smoke.

- Clinical picture (Similar to AR):
 - Morning profuse and watery rhinorrhoea.
 - Paroxysmal sneezing.
 - Alternating nasal obstruction
 - Postnasal drip.
- Diagnosis of exclusion, Negative allergy work-up.
- Treatment:
- **Medical:**
 - Avoidance of physical factors which provoke symptoms, e.g. sudden change in temperature, humidity, blasts of air or dust.
 - Anticholinergic Nasal sprays (ipratropium bromide).
 - Corticosteroid nasal sprays.
 - Hypertonic saline nasal sprays
 - Short course of oral and topical decongestants or Antihistamines
- **Surgical (For refractory cases):**
 - Nasal obstruction can be relieved by measures which reduce the size of nasal turbinates.
 - Other associated causes of nasal obstruction, e.g. polyp, deviated nasal septum, should also be corrected.
 - Excessive rhinorrhoea, not corrected by medical therapy and bothersome to the patient, can be relieved by sectioning the parasympathetic secretomotor fibres to nose (**Vidian Neurectomy**).
- **Rhinitis Medicamentosa:**
- Semi-ischemic state secondary to prolonged use of any topical nasal decongestants.
- Results in rebound congestion from:
 - Decreased vasomotor tone.
 - Increased parasympathetic activity.
 - Increased vascular permeability.
 - Decreased ciliary activity.
- Irreversible if vagal tone becomes atonic.
- Avoided by limiting topical decongestants to 3–5 days.
- Clinical picture:
 - Mucosal edema.
 - Nasal obstruction.
 - Dryness.
 - Irritation.
- Treatment:
 - Discontinue topical decongestants.
 - Aggressive saline irrigation.
 - Oral decongestants.
 - Nasal steroid spray.
 - Short-term oral corticosteroids.
 - Avoided by limiting topical decongestants to 3–5 days.

- **Drug-induced Rhinitis:**
- Antihypertensive drugs such as Reserpine, Guanethidine, Methyl dopa and **Propranolol** are sympathetic blocking agents and cause nasal stuffiness.
- Anticholinesterase drugs, e.g. **neostigmine**, used in the treatment of myasthenia gravis, have acetylcholine like action and cause nasal obstruction.
- **Contraceptive pills** also cause nasal obstruction because of oestrogens.

- **Rhinitis of Pregnancy:**
- Most common hormonal rhinitis.
- **Pathophysiology:**
 - o Multifactorial (cholinergic effects from increased estrogen may contribute).
- Symptoms manifests near end of 1st trimester and resolves after delivery.
- **Avoid skin testing (risk of anaphylaxis).**
- Use RAST testing and nasal cytology.
- **Treatment:**
 - o Refractory to most regimens.
 - o Conservative management (nasal saline irrigations, avoidance of allergens).
 - o **Avoid steroids, decongestants and antihistamine (may place fetus at risk).**

- **Honeymoon Rhinitis:**
- Follows sexual excitement leading to nasal stuffiness.

- **Emotional Rhinitis:**
- Nose may react to several emotional stimuli.
- Psychological states like anxiety, tension, hostility, humiliation, resentment and grief are all known to cause rhinitis.
- Treatment is proper counselling for psychological adjustment.
- Imipramine, which has both antidepressant and anticholinergic effects has been found useful.

- **Rhinitis due to Hypothyroidism**
- Hypothyroidism leads to hypoactivity of sympathetic system with predominance of parasympathetic activity causing nasal stuffiness and 'colds'.
- Replacement of thyroid hormone relieves the condition.

- **Gustatory Rhinitis:**
- Spicy and pungent food may in some people produce rhinorrhoea, nasal stuffiness, lacrimation, sweating and even flushing of face.
- Cholinergic response to stimulation of sensory receptors on the palate.
- Spicy food, particularly the red pepper, contains capsaicin which is known to stimulate sensory nerves.
- Can be relieved by anticholinergic (ipratropium bromide) nasal spray a few minutes before meals.

- **Non air-flow Rhinitis**
- Seen in patients of laryngectomy and tracheostomy.
- Nose is not used for air flow and the turbinates become swollen due to loss of vasomotor control.
- Similar changes are also seen in nasopharyngeal obstruction due to choanal atresia or adenoidal hyperplasia.

Nasal Steroids	Budesonide: - Rhinochort. - Class B for pregnancy Mometasone: - Nasonex - Class C for pregnancy - Used for Children above 2 years. Fluticasone Propionate: - Flixonase - Class C for pregnancy Fluticasone Furoate: - Avamys - Class C for pregnancy	Inhibit phospholipase A2 that controls the release of Arachidonic acid (Precursor to Prostaglandins and Leukotrienes).
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Anti-Histamine:

- Block histamine receptors in sensory nerve endings and blood vessels.
- Types of histamine receptors:
 - o **Histamine H1-receptor:**
 - Found in mast cells, smooth muscle, and endothelium in the body.
 - Histamine, acting on H1-receptors, produces pruritus, vasodilation, hypotension, flushing, headache, tachycardia, bronchoconstriction, increase in vascular permeability, potentiation of pain, and more.
 - o **Histamine H2-receptor:**
 - Found in upper GIT, primarily in the stomach.
- H1-Antihistamines:
 - o Inhibit the activity of the H1 receptor.
 - o Clinically used in the treatment of histamine-mediated allergic conditions.
 - o Types:
 - **First-generation (non-selective, sedating):**
 - Inexpensive and widely available.
 - Highly cross the blood-brain barrier (sedative).
 - Non-selective:
 - o H1-blocker (peripheral and central)
 - o Anti-cholinergic
 - Examples:
 - o **Diphenhydramine (Benadryl, Diphen):**
 - Pregnancy Category B.
 - 25-50 mg Q4 in adults.
 - 5 mg/kg/day in pediatrics > 12 years.
 - **Second-generation (selective, less-sedating)**
 - Newer drugs that are much more selective for peripheral H1 receptors.
 - Cross the blood-brain barrier to a much lower degree than the first-generation antihistamines:
 - o Less-sedative unless used in high doses.
 - Examples:
 - o **Cetirizine (Zyrtec):**
 - Pregnancy Category B.
 - 5-10 mg daily in adults.
 - 2.5 mg daily in pediatrics.
 - o **Loratadine (Claritin):**
 - Pregnancy Category B.
 - 10 mg daily in adults.
 - 5 mg daily in pediatrics.
 - o **Azelastine (Nasal)**

- **Third-generation (selective, non-sedating)**
 - Active metabolites of second-generation antihistamines.
 - Little evidence for any advantage compared to second-generation antihistamines.
 - Does not cross blood-brain barrier (non-sedative).
 - Examples:
 - **Levocetirizine (Xyzal):**
 - Pregnancy Category B.
 - 5mg daily in adults.
 - 2.5 mg daily in pediatrics.
 - **Desloratadine (Clarinx):**
 - **Pregnancy Category C.**
 - 5 mg daily in adults.
 - 2.5 mg daily in pediatrics.

Intranasal Corticosteroids (INCs):

- Efficacy and safety of intranasal corticosteroids (INCs) are well established for the management of allergic rhinitis, rhinosinusitis, and nasal polyps.
- Concerns about systemic absorption and adverse effects (AEs) remains among some patients, caregivers, and health care providers.
- **Side effects of systemic corticosteroids:**
 - Growth inhibition induced by hypothalamus-pituitary-adrenal (HPA) axis suppression.
 - Decreased bone mineral density
 - Myopathy
 - Cataract
 - Glaucoma
 - Hypertension
 - Hyperglycemia
- **Factors leading to the misconception that consistent use of INCs will cause systemic effects:**
 - Infrequent reports of systemic AEs with some of older INCs.
 - Concomitant use of INCs with inhaled corticosteroids (ICS), which have greater systemic absorption than INCs, for treatment of comorbid asthma, resulting in the perception of an additive inhibitory effect on the HPA that is more pronounced than the suppression seen with ICSs alone.
 - Increasing chronic use of INCs in a broader patient population.
- **Mechanism of Action:**
 - Anti-inflammatory effects by inhibiting:
 - 1. Production of pro-inflammatory cytokines:**
 - Interleukin IL-1
 - Interleukin IL-2
 - Interferon IFN-alpha
 - Tumor necrosis factor TNF
 - 2. Synthesis of pro-inflammatory enzymes:**
 - Macrophage products
 - Collagenase
 - Elastase
 - Plasminogen activator
 - 3. Lymphocytes proliferation.**
 - 4. Delayed type hypersensitivity.**

- **INCs Generations:**
 - **First Generation:**
 - Examples:
 - **Flunisolide (FLU):**
 - Flunisolide 29mcg/spray.
 - **Triamcinolone acetonide (TTA):**
 - Nasacort 55mcg/spray.
 - **Beclomethasone dipropionate (BDP):**
 - Beconase 42mcg/spray.
 - **Second Generation:**
 - Overall improvement relative to first generation.
 - Varies regarding potency and systemic bioactivity.
 - No detectable effects on measures of HPA axis function at recommended doses.
 - Examples:
 - **Fluticasone furoate (FF):**
 - Avamys 27.5mcg/spray.
 - Most recently introduced INCs.
 - **Mometasone furoate (MF):**
 - Nasonex 50mcg/spray.
 - One of the most potent to date INCs with almost undetectable systemic availability.
 - **Fluticasone propionate (FP):**
 - Flixonase 50mcg/spray.
 - One of the most potent to date INCs with almost undetectable systemic availability.
 - **Budesonide (BUD):**
 - Rhinocort Aqua 32mcg/spray.
 - **Ciclesonide:**
 - Omnaris 50mcg/spray
- **Pharmacokinetic Differences:**
 - Systemic bioavailability is the amount of drug that reaches the systemic circulation.
 - Reflects the sum of nasal and intestinal absorption, as well as clearance by first-pass hepatic metabolism.
 - The main determinant of systemic bioavailability of INCs is direct absorption from the nose, where there is no first-pass inactivation.
 - Second-generation INC agents have low systemic bioavailability compared with older INCs which minimize the risk of systemic adverse events.

- Delivery device of INCs:
 - Delivery of aqueous suspensions by pump sprays provides better drug deposition at the ciliated mucous membrane when compared with pressurized aerosol.
 - After intranasal administration, part of the drug is available for absorption at the nasal mucosa, the rest is cleared by nasociliary clearance into the throat.

Table 3. Recommended Technique for Using Topical Intranasal Corticosteroid Sprays [51]

1. Hold head in a neutral, upright position
2. Clear nose of any thick or excessive mucus, if present, by gently blowing the nose
3. Insert spray nozzle into the nostril
4. Direct the spray laterally or to the side, away from the middle of the nose (septum) and toward the outer portion of the eye or the top of the ear on that side. (If possible, use the right hand to spray the left nostril and left hand to spray the right nostril, to direct the spray away from the septum)
5. Activate the device as recommended by the manufacturer, and use the number of sprays recommended by the doctor
6. Gently breathe in or sniff during the spraying
7. Breathe out through the nose

(Adapted from Benninger MS et al. *Techniques of intranasal steroid use. Otolaryngol Head Neck Surg.* 2004;130:5-24.)

- Absorption of INCs:
 - 1. Topical absorption:**
 - Determines therapeutic efficacy.
 - Occurs at the target site (Nose).
 - 2. Systemic absorption:**
 - Determines undesired systemic effects.
 - Occurs in 2 sites:
 - GI absorption of swallowed part of INS which underwent first-pass metabolism in the liver
 - Direct blood absorption at nasal mucosa.
- Increased lipophilicity of INCs leads to less systemic absorption and less side effect due to:
 - Greater deposition of corticosteroid in respiratory mucosa.
 - Greater binding affinity to the glucocorticoid receptor.
 - Slower release from respiratory mucosa.
 - Less amount of corticosteroid absorbed in blood.
 - Less amount of corticosteroid get swallowed.
 - Poor absorption from GI.
 - Extensive first-pass metabolism in the liver.

- Rank order of lipophilicity of some INCs (highest to lowest):
 1. Mometasone furoate (MF).
 2. Fluticasone propionate (FP).
 3. Beclomethasone dipropionate (BDP).
 4. Budesonide (BUD).
 5. Triamcinolone acetonide (TTA).
 6. Flunisolide.

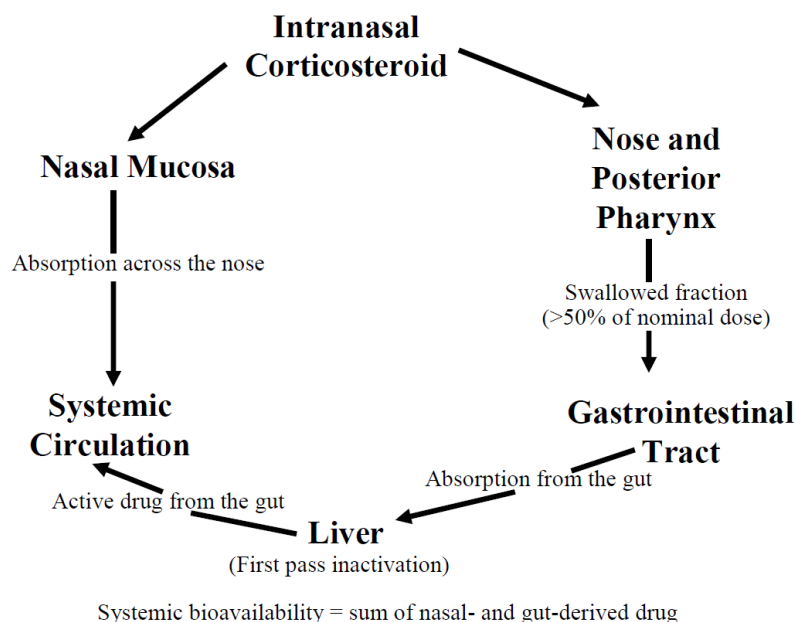


Table 1. Estimated Absolute Bioavailability of Intranasal Corticosteroids [1,9,12,14]

Corticosteroid	Systemic Bioavailability
Dexamethasone (oral)	76%
Flunisolide	49%
Triamcinolone acetonide	46%
Beclomethasone dipropionate	44%
Budesonide	34%
Fluticasone propionate ^a	<1%
Fluticasone furoate	0.5%
Mometasone furoate ^a	<0.1%
Ciclesonide aqueous	Below lower limits of assay quantification

^aA pharmacokinetic study of mometasone furoate (MFNS) and fluticasone propionate (FP) administered at 2400 µg/day—utilizing a dose 3 times higher than the maximum ciclesonide dose administered in a study by Nave et al. and 12 times the recommended dose of MFNS and FP—yielded barely detectable serum levels of each agent and estimated bioavailability of 0.42% and 0.46% for FP and MFNS, respectively [10,13,14].

- Elimination:
 - Half-life is the time it takes for the plasma concentration to be reduced by 50%.
 - Most intranasal steroid products work within the first several days of use, with some producing symptomatic relief in as few as 12 hours after the first dose.
 - There is typically a 3 to 7 day period before full benefit is seen.
 - After multiple doses of a drug, the plasma concentration rises until it reaches a steady-state concentration.
 - In pediatrics, more rapid systemic elimination of drugs is present compared to adults due to the higher hepatic blood flow in children.
 - Greatest half-live value for INCs (highest to lowest):
 1. Fluticasone propionate (FP)
 2. Mometasone furoate (MF)
 3. Budesonide (BUD)
 4. Flunisolide (FLU)
 5. Triamcinolone acetone (TAA)

- **INCs in Pregnancy:**
 - Budesonide (BUD) is only INC that given FDA Pregnancy Category B:
 - Based on a review of 3 Swedish registries covering over 2000 births from 1995 to 2001 that indicated no increased risk for overall congenital malformation from the use of intranasal BUD during early pregnancy.
 - All other INCs are Category C:
 - Because data in pregnant women are lacking and animal studies have either not been performed or revealed AEs.
 - However, No data indicate an association between INCs and congenital malformations.
 - A prospective human study of INC use during pregnancy, 8 weeks of intranasal Fluticasone propionate (FP) did not have any deleterious effects on fetal growth or pregnancy outcome.

- **INCs in Children:**
 - Mometasone furoate (MF) and Fluticasone furoate (FF) have negligible systemic bioavailability and preferable in children requiring chronic therapy.
 - INCs are approved for use as low as age 2 years with Mometasone furoate (MF).

- **Local adverse effects of INCs:**
 - Occurs in 5-10% of patients.
 - Most are mild, self-limiting and resolve without discontinuing therapy, including:
 - Epistaxis
 - Throat irritation
 - Nasal dryness
 - Burning sensation
 - Severe local adverse effects are rare and can be prevented with an appropriate administration technique that helps avoid dryness, crusting and bleeding from the septum, including:
 - Nasal mucosal atrophy
 - Nasal mucosal ulceration
 - Septal perforation
 - Histological studies from long-term studies of several INCs (MF, FP, TAA, and BDP) in patients with perennial allergic rhinitis demonstrate no evidence of atrophy or deleterious pathologic changes in the nasal mucosa after 6 months to 5 years of use.
- **Systemic adverse effects of INCs:**
 - **Effects on the HPA Axis**
 - The primary action of corticosteroids on the HPA axis is a negative feedback effect caused by suppression of corticotrophin-releasing hormone and adrenocorticotrophic hormone (ACTH) levels, resulting in lower cortisol secretion.
 - Large number of short- and long-term studies in adults and children have found no significant impact on HPA axis function with the newer INCs agents (**MF, FP, FF, BUD, Ciclesonide**).
 - **Effects on Statural Growth in Children:**
 - Systemic corticosteroids are known to exert a suppressive effect on growth through several mechanisms, including:
 - Decreased release of growth hormone
 - Inhibition of insulin-like growth factor 1 activity
 - Downregulation of growth hormone receptor expression
 - Suppression of collagen synthesis and adrenal androgen production.
 - Studies have shown that newer INCs (**MF, FP, FF, BUD, Ciclesonide**) administered at recommended doses for up to 1 year are not associated with impairment of growth or final adult height.
 - Growth suppression has been reported with long-term use of some INCs (**BDP, TTA**) when recommended doses were exceeded.

- **Effects on Bone Density:**
 - Systemic corticosteroids exert their negative effect on bone metabolism by altering both calcium homeostasis (osteoblastic and osteoclastic activity) and sex hormone production.
 - Newer INCs (**MF, FP, BUD**) do not appear to be associated with reductions in bone mineral density or osteoporosis.
- **Ocular Effects:**
 - Several recent long-term studies have demonstrated no evidence of ocular changes (high IOP or cataract) with newer INCs (MF, FF).

Rhinosinusitis:

- Inflammation of nasal cavity and paranasal sinuses.
- Rhinitis and sinusitis usually coexist and are concurrent in most individuals.
- **Rhinosinusitis in Adults: (EPOS 2012)**
 - Characterized by 2 or more symptoms:
 - Nasal Obstruction or Nasal Discharge (Anterior/Posterior).
 - ± Facial Pain/Pressure
 - ± Reduction or Loss of Smell
 - And Either:
 - Endoscopic signs of:
 1. Nasal Polyps.
 2. Mucopurulent discharge from Middle Meatus.
 3. Eedema/mucosal obstruction in Middle Meatus.
 - CT changes:
 - Mucosal changes within Ostiomeatal complex or Sinuses.
- **Rhinosinusitis in Children: (EPOS 2012)**
 - Characterized by 2 or more symptoms:
 1. Nasal Obstruction or Nasal Discharge (Anterior/Posterior).
 1. ± Facial Pain/Pressure
 2. ± Cough
 - And Either:
 1. Endoscopic signs of:
 1. Nasal Polyps.
 2. Mucopurulent discharge from Middle Meatus.
 3. Eedema/mucosal obstruction in Middle Meatus.
 2. CT changes:
 1. Mucosal changes within Ostiomeatal complex or Sinuses.
- **Rhinosinusitis: (Rhinosinusitis Task force 1996)**
 - Characterized by 2 Major or 1 Major and 2 Minor symptoms:

TABLE 46-I. Rhinosinusitis Symptoms

Major	Minor
Facial pain/pressure	Headache
Facial congestion/fullness	Fever (nonacute)
Nasal obstruction/blockage	Halitosis
Nasal discharge/purulence, discolored posterior drainage	Fatigue
Hyposmia/anosmia	Dental pain
Purulence on nasal exam	Cough
Fever (acute rhinosinusitis only)	Ear pain, pressure, and/or fullness

Diagnosis of rhinosinusitis requires two major or one major and two minor symptoms.

- **Classification of Rhinosinusitis:**

- **Acute Rhinosinusitis (ARS):**
 - <4 weeks
 - Complete resolution of symptoms.
- **Recurrent Acute Rhinosinusitis:**
 - ≥4 episodes of Acute Rhinosinusitis in 1 year with resolution of symptoms in between episodes.
- **Sub-Acute Rhinosinusitis:**
 - 4-12 weeks
 - Complete resolution of symptoms.
 - Symptom free intervals if recurrent.
- **Chronic Rhinosinusitis (CRS):**
 - ≥12 weeks
 - No complete resolution of symptoms.
 - Subject to Exacerbations
- **Acute Exacerbation of Chronic Rhinosinusitis:**
 - Sudden worsening of symptoms in a patient already diagnosed with CRS.
 - Return to baseline symptoms after treatment.

**TABLE
33.2**

**TEMPORAL CLASSIFICATION
OF RHINOSINUSITIS**

Term	Duration of Symptoms
Acute rhinosinusitis	Up to 4 wk
Subacute rhinosinusitis	4–12 wk
Chronic rhinosinusitis	Longer than 12 wk
Recurrent acute rhinosinusitis	>4 acute episodes in 1 y with resolution between episodes

- **Predisposing Factors for Development of Rhinosinusitis:**

○ **Host Factors:**

1. Paranasal Sinus Anatomic Variations and Abnormality:

- OMC refers to physiologic arrangement of structures into which frontal, maxillary, and anterior ethmoid sinuses drain.
- Patency of sinus drainage pathways is crucial for adequate mucociliary function
- One liter of mucus is produced by sinonasal mucosa per day, which is cleared by mucociliary transport.
- Ostial obstruction leads to fluid accumulation and stagnation, creating a moist, hypoxemic environment ideal for growth of pathogens.
- Examples:
 - Septal deviation
 - Hypertrophic turbinates
 - Concha bullosa
 - Paradoxical middle turbinates
 - Infraorbital (Haller) cells
 - Obstructing frontal recess cells
 - Enlarged Adenoids
 - Choanal Atresia
 - Scarring from previous surgery
 - Trauma
 - Nasal FB in pediatrics

2. Allergic Conditions:

- **Allergic Rhinitis:**
 - Allergy was presumed to be a causative factor in CRS because of the eosinophilic inflammation in NPs, and the known role of allergy as a mechanism for the production of eosinophilic inflammation.
 - 50% of CRS with NP patients have allergic sensitization.
 - However, Positive skin test didn't correlates with severity of disease or responsiveness to treatment in CRS.
 - Allergy should be addressed as a comorbid condition with CRS with NP and treated appropriately.

■ **Asthma:**

- 50% of CRS with NP patients have asthma.
- 7% of asthma patients have NPs.
- In addition to clinical association of asthma and NP disease, histopathologic and molecular findings in asthma and NPs are similar.
- Asthma and CRS may be separate clinical manifestations of a single systemic inflammatory disease "The Unified Airway".
- This linkage is supported by observation that treating upper airway inflammatory disease results in reduced burden of disease in the lower airway.
- Multiple studies showed that surgical treatment of CRS improves asthma.

■ **Samter Triad:**

- Aspirin Exacerbated Respiratory Disease (AERD).
- Adult-onset triad of:
 1. CRSwNP
 2. Asthma
 3. Aspirin Intolerance
- Urticaria, angioedema and anaphylaxis are additional clinical manifestations in some individuals.
- Affected patient will have episodes of asthma attack or allergic rhinitis symptoms after the ingestion of aspirin or other NSAID.
- Pathophysiology:
 - NSAIDs trigger these reactions by COX-1 inhibition.
 - Selective COX-2 inhibitors are usually well tolerated in AERD.
 - Resulted from abnormal metabolism of arachidonic acid in the cyclooxygenase and lipoxygenase pathways which lead eosinophilic upper and lower airway inflammation.
- Diagnosis:
 - Aspirin challenge in a controlled environment is the most definitive method of diagnosis.

- Prevalence:
 - Affects 1-5% of population.
 - Affects 10-20% of Asthmatic patients.
 - Affects 20% of CRS with NP patients.
 - Affects 30-40% of Asthmatic patients with CRS with NP.
- Prognosis:
 - AERD patients have a severe form of disease that is difficult to manage and more likely to require multiple endoscopic surgeries to manage their disease.
- Management:
 - Leukotriene modifiers (Montelukast).
 - Topical or systemic corticosteroids
 - Endoscopic sinus surgery
 - Aspirin desensitization
- **Churg-Strauss Syndrome (CSS):**
 - Allergic or autoimmune reaction to an environmental agent causing granulomatous small-vessel vasculitis.
 - Criteria for Diagnosis:
 1. Asthma (97%)
 2. Paranasal sinusitis (60%)
 3. Pulmonary infiltrates (40%)
 4. Peripheral neuropathy (70%)
 5. Blood Eosinophilia count >10%
 6. Histological proof of vasculitis with extravascular eosinophils
 - Phases of presentation:
 1. Allergic Rhinitis and Asthma
 2. Eosinophilic infiltrative disease (eosinophilic pneumonia or gastroenteritis)
 3. Systemic small-vessel vasculitis with granulomatous inflammation.
 - Labs:
 - Blood Eosinophilia count >10%
 - Elevated ESR and CRP
 - Positive p-ANCA in 40% of patients.
 - Elevated serum IgE level
 - Hypergammaglobulinemia
 - Treatment:
 - Glucocorticoids alone are usually adequate for the treatment.
 - Cytotoxic drugs are necessary in <20% of patients.

3. Congenital Conditions:

- **Cystic Fibrosis (CF):**
 - Inherited disorder associated with abnormal mucociliary clearance from the sinuses.
 - Cause increase viscosity of nasal secretions which impedes adequate clearance and leads to ciliary injury, local mucosal edema, and further inflammation.
- **Primary Ciliary Dyskinesia:**
 - Inherited disorder associated with abnormal mucociliary clearance from the sinuses.
 - Cause ciliary dysmotility that predisposes patients to both acute and chronic rhinosinusitis.
 - Kartagener syndrome is the most common form of Primary Ciliary Dyskinesia and associated with a triad of:
 1. CRS
 2. Bronchiectasis
 3. Situs inversus

4. Systemic Conditions:

- **Acid Reflux Disease:**
 - Good correlation between nasopharyngeal reflux and postsurgical patients with persistent CRS.
 - Patients with persistent CRS had more reflux at nasopharynx, upper esophageal sphincter, and at distal esophagus than controls, with the greatest difference at the nasopharynx.
- **Decrease Immunity:**
 - Can predispose to severe forms of Rhinosinusitis with high risk to develop complications.
 - History of persistent or recurrent infections despite adequate antimicrobial therapy should raise the suspicion of immune deficiency as a contributing factor.
 - Examples:
 - DM
 - HIV
 - Chemotherapy.
- **Inflammatory Disorders:**
 - Examples:
 - Wegener Granulomatosis
 - Sarcoidosis.

○ **Environmental Factors:**

1. Infectious agents:

- Post-URTI
- Swimming and diving in infected water.
- Dental infections from Molar or Premolar teeth.

2. Trauma:

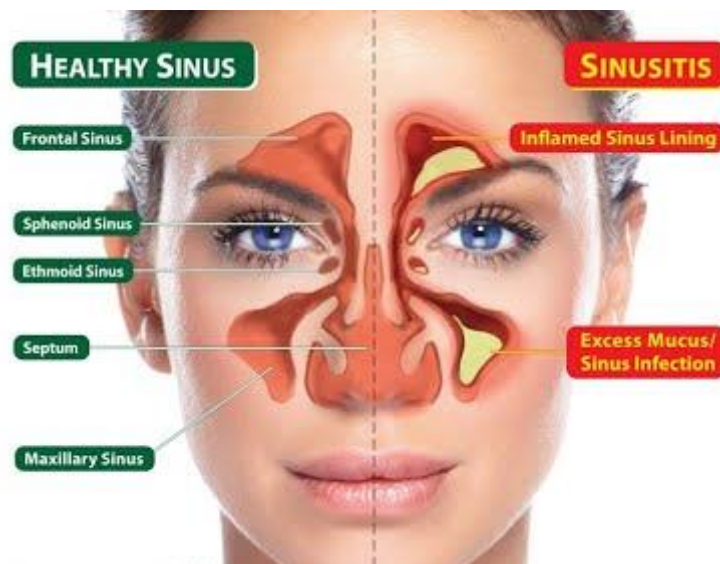
- Mucosal edema and inflammation.
- Altered bony anatomy.

3. Iatrogenic:

- Medications.
- Post-surgery.
- Nasal Packing.
- NGT.

4. Exposure to Toxins:

- Smokers and exposure to secondhand smoke is a morbid cofactors in CRS.
- Persistent smoking is significantly associated with worse symptom outcomes after endoscopic sinus surgery.
- Cause decreased ciliary beat frequency.
- Positive correlation between environmental pollution exposure and disease of upper respiratory tract in patients with pre-existing atopy.



**TABLE
33.1**

**PREDISPOSING FACTORS AND
CONTRIBUTORS TO THE
DEVELOPMENT OF RHINOSINUSITIS**

Host Factors

Congenital conditions

Examples: Cystic fibrosis
Immotile cilia syndrome

Allergic or immune conditions

Examples: Environmental allergy
Human immunodeficiency virus
Immune suppressive agents (i.e., chemotherapy)
Bone marrow transplant

Paranasal sinus anatomic abnormalities

Examples: Middle turbinate concha bullosa
Obstructing frontal recess cells
Severe nasal septal deviation

Systemic inflammatory conditions

Examples: Wegener granulomatosis
Sarcoidosis

Presence of neoplasm

Environmental Factors

Infectious agents

Examples: Viruses
Bacterial pathogens

Trauma

Examples: Resultant mucosal edema and inflammation
Altered bony anatomy

Exposure to noxious chemicals

Iatrogenic

Examples: Medications
Surgery

- **Inflammation Vs. Infection:**
- **Inflammation:**
- A cascade of processes in which mediators (leukocytes) eliminate foreign agents and repair damaged tissues.
 - **Acute inflammation:**
 - Characterized by exudation of fluid and plasma proteins from blood vessels with emigration of leukocytes (Neutrophils) to combat the offending entity.
 - Identified within minutes to hours of an insult.
 - **Sub-Acute inflammation:**
 - Interval period when overlapping patterns of inflammation are observed.
 - **Chronic inflammation:**
 - Develops if condition persists over weeks to months.
 - Hallmarks of chronic inflammation are the presence of lymphocytes, macrophages, eosinophils, and basophils, along with increased vascularity, fibrosis, and tissue necrosis.
- **Infection:**
- Diagnosed when microorganisms present in a host, directly interacting with host tissue and replicating which results in expression of disease in the host organism.
 - **Bacterial infection:**
 - Characterized by presence of ≥ 1 bacteria/ high density field ($\geq 1,000$ colony forming units/ml).

**TABLE
35.2**

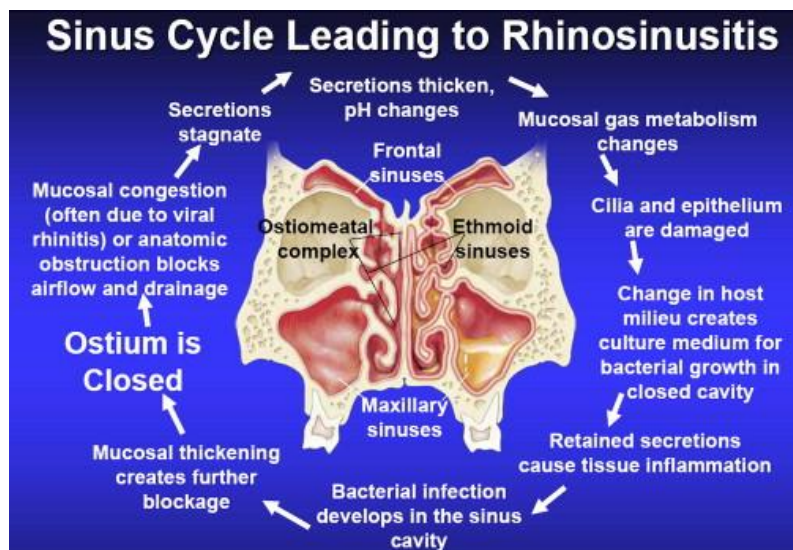
DIAGNOSTIC CRITERIA FOR SINUSITIS FROM 2007 TASKFORCE

Acute Rhinosinusitis	Up to 4wk of purulent nasal drainage (anterior, posterior, or both) accompanied by nasal obstruction, facial pain-pressure-fullness, or both: • Purulent nasal drainage is cloudy or colored, in contrast to the clear secretions that typically accompany viral upper respiratory infection, and may be reported by the patient or observed on physical examination. • Nasal obstruction may be reported by the patient as nasal obstruction, congestion, blockage, or stuffiness, or may be diagnosed by physical examination. • Facial pain-pressure-fullness may involve the anterior face, periorbital region, or manifest with headache that is localized or diffuse.
Acute Viral Rhinosinusitis	Acute Viral Rhinosinusitis (AVRS) that is caused by, or is presumed to be caused by, viral infection. A clinician should diagnose AVRS when symptoms or signs of acute rhinosinusitis are present <10 d and the symptoms are not worsening.
ABRS	Acute Viral Rhinosinusitis that is caused by, or is presumed to be caused by, bacterial infection. A clinician should diagnose ABRS when: a. Symptoms or signs of acute rhinosinusitis are present 10 d or more beyond the onset of upper respiratory symptoms, or b. Symptoms or signs of acute rhinosinusitis worsen within 10 d after an initial improvement (double worsening)
CRS	Twelve (12) weeks or longer of two or more of the following signs and symptoms: • Mucopurulent drainage (anterior, posterior, or both) • Nasal obstruction (congestion) • Facial pain-pressure-fullness • Decreased sense of smell AND inflammation is documented by one or more of the following findings: • Purulent (not clear) mucus or edema in the middle meatus or ethmoid region • Polyps in nasal cavity or the middle meatus • Radiographic imaging showing inflammation of the paranasal sinuses,
Recurrent Acute Rhinosinusitis	Four (4) or more episodes per year of ABRS without signs or symptoms of rhinosinusitis between episodes: • Each episode of ABRS should meet diagnostic criteria above

- **Acute Rhinosinusitis (ARS):**

○ Pathophysiology:

- Majority of ARS is infectious (Viral) in etiology causing Acute Viral Rhinosinusitis (AVRS) in which later on become superimposed with bacterial infection causing Acute Bacterial Rhinosinusitis (ABRS) in 2% of cases.
- → Acute inflammation of sinus mucosa.
- → Edema and hyperemia
- → Outpouring of PMN cells (Neutrophils)
- → Increased activity of serous and mucous glands
- → Exudation of fluid
- → Impaired mucociliary clearance
- → Failure of ostium to drain
- → Stasis of serous exudate
- → Superimposed secondary bacterial infection
- → Mucopurulent or purulent exudate.
- → Empyema of sinuses.
- → Destruction of mucosal lining.
- → Destruction of its bony walls.
- → Complications.



- Most common cause of Acute Rhinosinusitis is Post-viral URTI.
 - Most Acute viral URTI infections are self-limiting.
- In patients with Recurrent ARS, always consider:
 - Anatomical variations obstructing sinus drainage.
 - Odontogenic Source.
- ARS has three possible clinical courses:
 - Resolution
 - Development of complications
 - Development of Chronic Rhinosinusitis (CRS)

- **Pathogens of Acute Rhinosinusitis:**

○ **Viral (Most common)**

1. Rhinovirus (Most common)
2. Influenza virus
3. Parainfluenza virus
4. Adenovirus
5. Corona virus.
6. Respiratory Syncytial Virus (RSV)

○ **Bacterial:**

1. Streptococcus Pneumoniae **40%** (Most common)
2. Haemophilus Influenzae **35%**
3. Moraxella Catarrhalis **15%**
4. Anaerobes (Dental) **5%**
5. Staphylococcus Aureus **<5%**
6. Streptococcus Pyogenes **<5%**

○ **Fungal:**

1. Aspergillus (Most common)
2. Mucor
3. Rhizopus

- **Clinical Picture of ARS:**

○ **History:**

- Facial Pain and Tenderness:
 - Worse with straining or bending over
- Pressure Headache:
 - Forehead with frontal and maxillary sinusitis.
 - Bridge of nose, medial and deep to eye with ethmoid sinusitis.
 - Occipital headaches from sphenoid sinusitis.
- Nasal obstruction and congestion.
- Nasal discharge and postnasal drip.
- Cough and halitosis (especially in Children)
- Allergic symptoms (sneezing, watery eyes)
- Eustachian tube dysfunction,
- Hyposmia/Anosmia

○ Physical Exam:

▪ **General Examination:**

- Vital signs and temperature.
- Swelling, erythema, or edema localized over the involved cheek or periorbital edema may suggest complicated ABRS with extension into soft tissues.
- Palpation and percussion over the face and maxillary dentition may assist in localizing pain.

▪ **Eye Examination:**

- Assessment of appearance of conjunctiva
- Visual status testing
- Extraocular muscle function evaluation
- Inspection for proptosis.

▪ **Ear Examination:**

- May shows signs of AOM or OME.

▪ **Throat Examination:**

- Postnasal drip may suggest ABRS.

▪ **Nasal Examination:**

- Done before and after topical decongestion.
- Hyperemia, edema of turbinates and nasal mucosa.
- Visualize middle turbinate and inspect for pus in middle meatus.
- Nasal polyposis.

**TABLE
33.5**

**INDICATIONS FOR SINONASAL
ENDOSCOPY IN ACUTE
RHINOSINUSITIS**

Failure to improve on empiric therapy
Unilateral disease
Severe symptoms
Suspicion of complications of ABRS
Recent sinonasal surgery
Immunocompromised status



- **AVRS Vs ABRS:**

- Can't be differentiated in the first 3-4 days.
- Fever can be present in both types.
- Purulent discharge indicates presence neutrophils not bacteria, which is characteristic of acute inflammation regardless of the etiology.
- Clear rhinorrhea may be indicative of AVRS, Allergic Rhinosinusitis or Vasomotor rhinitis.

- **Diagnostic Criteria of ABRS:**

- Presence of the three cardinal symptoms of ABRS:
 1. Purulent nasal discharge or postnasal drip
 2. Nasal obstruction
 3. Facial pain, pressure, or fullness
- Duration of 10 days or longer beyond onset of URTI or worsening of ARS symptoms within 10 days after a period of initial improvement (Double-worsening).

- **Diagnostic Tests for ARS:**

- **Middle Meatus Cultures/Biopsy:**

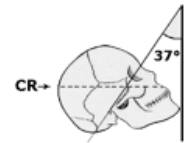
- Routine cultures or biopsy should not be done in uncomplicated ABRS.
- Blind swabs of nasal cavity are not recommended due to its contamination by normal colonizing bacteria.
- Endoscopically-directed middle meatus cultures are well correlated with cultures obtained by direct maxillary sinus aspiration.
- Cultures can be obtained with a sterile swab or utilizing suction into a sterile trap device.
- Indications of Culture in ARS:
 - Severe ARS:
 - Failure to improve with medical management
 - Clinical deterioration on medical management
 - Complicated ARS.
 - Immunocompromised patient.
- Indications of Biopsy in ARS:
 - Unilateral disease
 - Suspicious of neoplasm
 - Immunocompromised patient to rule out fungal infection.

○ **Plain X-Ray:**

- Routine imaging should not be done in patients with ARS who meet the diagnostic criteria.
- Not used anymore due to:
 - High rate of False Positives and False Negatives.
 - Not sensitive and Not specific
- Evaluates presence of:
 - Air-fluid levels
 - Mucosal thickening
 - Sinus opacification
- Four Views:

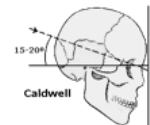
- **Occipito-Mental View:**

- Water's View.
- Best for Maxillary Sinus.



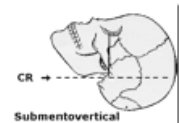
- **Occipito-Frontal View:**

- Caldwell View.
- Best for Ethmoid and Frontal Sinuses.



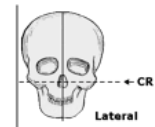
- **Submento-Vertex View:**

- Bucket-handle View.
- Best for Sphenoid and Ethmoid Sinuses.



- **Lateral View:**

- Good for All Sinuses.



○ **CT Sinuses:**

- Routine imaging should not be done in patients with ARS who meet the diagnostic criteria.
- Gold standard modality to evaluate the sinuses if needed.
- Some degree of mucosal thickening occurs in almost **30%** of asymptomatic general public.
- Lund-Mackay staging system is the most common objective staging system used to evaluate and quantitates the paranasal sinuses on CT.
- Current CT sinus protocols:
 - Non-contrasted thin axial slice thicknesses <3 mm (ideally 1 mm is required for accurate image-guidance compatibility)
 - High-resolution multiplanar reconstruction in coronal and sagittal planes.
 - Contrast is needed in the following cases:
 1. Sinonasal neoplasm
 2. Complicated Rhinosinusitis
 3. Intracranial and orbital extension.

▪ **Indications of CT in ARS:**

1. Severe ARS
 - Failure to improve with medical management
 - Clinical deterioration on medical management
2. Complicated ARS
3. Immunocompromised patient.

▪ **CT Findings in ARS:**

1. Opacification of sinuses
2. Air-fluid levels.
3. Moderate to severe mucosal thickening.
4. Intra-cranial or intra-orbital extension.

**TABLE
28.3**

**INFORMATION OBTAINED FROM
THE DIFFERENT PLANES OF
SINUS CT**

Sinus CT Plane	Helpful Information
Coronal	Septal deviation OMC Uncinate attachment type (A,B,C) Middle turbinate variations Lamina papyracea anatomy Ethmoid anatomy Skull base anatomy/lateral lamella length Anterior ethmoid artery anatomy Sphenoid anatomy/sphenoethmoid cell (Onodi) Identify optic nerve and carotid artery dehiscence.
Sagittal	Frontal recess anatomy Slope of the skull base Sphenoid pneumatization pattern
Axial	Sphenoid anatomy/attachment of superior turbinate

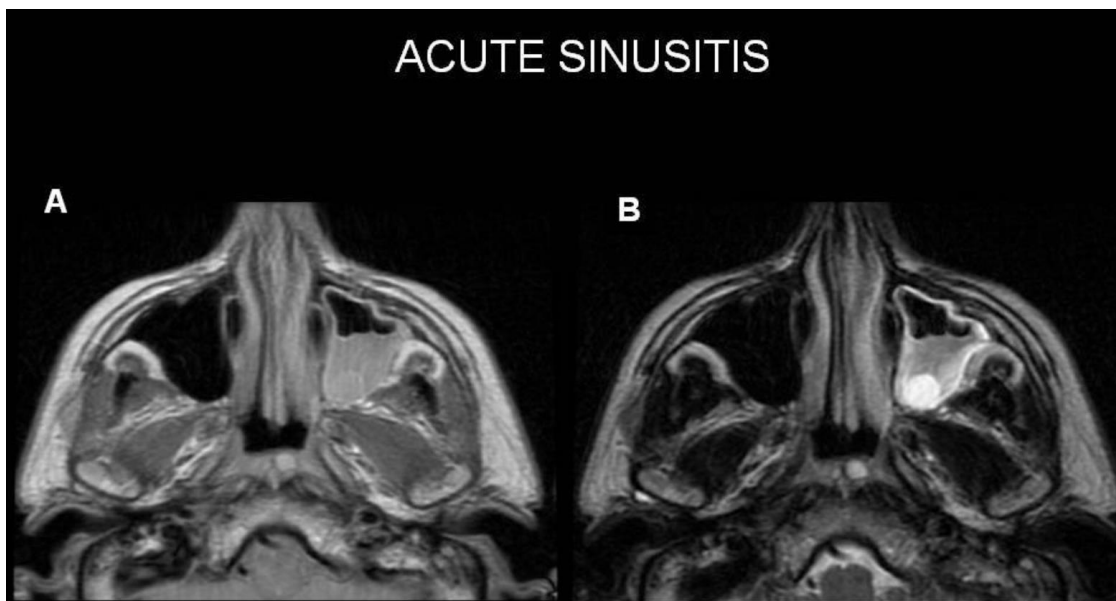
- **Evaluation of CT PNS:**

- Examine Distribution of Mucosal disease:
 - Mucosal thickening
 - Air-fluid levels suggest acute inflammatory process.
- Inspect development of sinus:
 - Symmetry.
 - Aeration of sinus cavities.
- Examine Nasal structures, Airway, and Access.
- Evaluate for underlying causes of disease:
 - OMC patency
 - Paradoxical Turbinates
 - DNS
 - Concha bullosa
- Examine for Anatomical variations and Landmarks:
 - Cribiform plate
 - Posterior ethmoidal height
 - Thickness of skull base
 - Optic nerve
 - Orbital dehiscence
 - Carotid artery)
- Correlate between CT and symptoms or Endoscopic findings.



○ **MRI Sinuses:**

- Improved soft tissue detail.
- Can delineate polyp from retained secretions.
- Poor bone resolution.
- Indications:
 - Complicated Rhinosinusitis:
 - Intracranial and Intraorbital extension.
 - Fungal Rhinosinusitis.
- Findings:
 - **T1 MRI:**
 - Mucosal thickening is HYPER-intense.
 - Fluid is HYPO-intense.
 - Mucocele is HYPER-intense (Higher protein content).
 - **T2 MRI:**
 - Mucosal thickening is HYPER-intense.
 - Fluid is HYPER-intense.
 - Tumor tissue appears HYPO-intense compared to mucosal thickening.
 - Mucocele is HYPER-intense (Higher protein content).
 - AFRS is HYPO-intense/signal void due to presence of Metallic proteinaceous material, magnesium, iron, and calcium.



**TABLE
33.6****IMAGING MODALITIES IN ACUTE RHINOSINUSITIS**

Modality	Strengths	Weaknesses	Indications in the Setting of ARS
Ultrasound	Low cost No radiation exposure	Low sensitivity	Not recommended
X-ray	Low cost Low radiation dose Portable exam	Low sensitivity Low specificity	No longer used routinely
CT	Excellent bony anatomy High sensitivity (>90%)	Higher cost Higher radiation dose Poor soft tissue differentiation Cannot distinguish AVRS from ABRS Findings do not correlate well with localizing symptoms	Suspicion for a complication of ABRS Suspicion for alternative diagnosis, such as neoplasm Presence of comorbidities that may predispose to complications of ABRS
MRI	Excellent soft tissue differentiation Excellent intracranial diagnostic capacity	Higher cost Longer scan time Poor delineation of bony anatomy Cannot distinguish AVRS from ABRS	Suspicion of neoplasm Suspicion of intracranial involvement

Note: No imaging modality is recommended in the routine diagnosis of uncomplicated ABRS.

- **Treatment of ARS:**

○ **Medical Management:**

1. Analgesic/Antipyretic (Acetaminophen):

- Decreases pain and fever.
- Recommended for AVRS and ABRS.

2. Saline Irrigations:

- Thins mucus, enhance mucociliary clearance and decrease inflammation.
- Hypertonic saline (Sinomarine) have superior anti-inflammatory effects.
- Recommended for AVRS and ABRS.

3. Topical Decongestant (Oxymetazoline):

- Reduces mucosal edema and nasal airway obstruction.
- More effective than systemic decongestant due to increase potency.
- Consider in AVRS and ABRS for 3 days duration only to avoid development of Rhinitis Medicamentosa.

4. Systemic Decongestant (Pseudoephedrine):

- Limited reduction of mucosal edema.
- Not as effective as topical decongestant

5. Non-Sedating Anti-Histamine (Loratadine):

- Dries nasal secretions.
- Consider in AVRS or ABRS with allergic component.

6. Topical Corticosteroid (Fluticasone/Mometason):

- Improved symptom control in ABRS.
- Consider in ABRS with allergic component.
- Minimal systemic absorption and carry a very low risk for systemic side-effects.
- Systemic corticosteroid (Prednisone) use was not supported in literature in AVRS or ABRS.

7. Antibiotics:

- Observation for period of 7 days without use of antibiotics should be done for adults with uncomplicated mild ABRS and assurance of follow-up.

- Indications of Antibiotics in ABRS::

1. Mild ABRS (Mild pain with Fever <38.3) with:
 - o Failure of improvement by 7 days or worsening of symptoms at any point during this period.

2. Severe ABRS (Severe pain with Fever >38.3).

- Duration of Antibiotics:

- 7-10 days in adults and 10-14 days in children.

- Factors affecting initial Antibiotics choices:

- **Patients with Penicillin-Allergy:**

1. Tetracyclines (Doxycycline)
2. Fluoroquinolones (Levofloxacin Moxifloxacin)
3. Macrolide (Clarithromycin, Azithromycin)

- **Patients during Pregnancy:**

1. Amoxicillin-clavulanate (Class B).
2. Azithromycin (Class B) for patients with Penicillin-Allergy.

- **Recent use of prior Antibiotics in the last 4-6 weeks (High risk of developing antibiotic-resistant bacteria):**

1. High-dose Amoxicillin-Clavulanate (4g/day).
2. Fluoroquinolones (Levofloxacin Moxifloxacin).

- **Having a child in daycare in the household (High risk for developing Penicillin-resistant *S. pneumoniae*):**

1. High-dose amoxicillin-clavulanate (4g/day).
2. Fluoroquinolones (Levofloxacin Moxifloxacin).

- Choices for initial empiric antibiotic therapy:
 - **Recommended (IDSA guidelines 2012):**

1. Amoxicillin-Clavulanate:

- The recommended first-line of treatment in ABRS.
- 625mg TID or 1g BID dosage.
- Addition of clavulanate to amoxicillin improves coverage for penicillin-resistant *H. influenzae* as well as *M. catarrhalis*.

2. Tetracyclines:

- Doxycycline 200mg OD for the 1st day then 100mg OD for 7 days.
- Active against all common respiratory pathogens.
- The recommended alternative for Amoxicillin in adults with penicillin allergy.
- Not recommended in children.

3. Third Generation Cephalosporin + Clindamycin:

- Combination therapy of third-generation cephalosporin (cefixime, cefpodoxime) and Clindamycin can be used for patients from geographic regions with high endemic rates of penicillin-nonsusceptible *S. pneumoniae*.

- **Not Recommended (IDSA guidelines 2012):**

- 1. Amoxicillin:**

- Not recommended currently as initial empiric therapy due to high percentage of bacterial resistance.
 - 500mg TID orally.
 - Advantages:
 - Safety, Efficacy
 - Low cost
 - Narrow microbiologic spectrum
 - Disadvantages:
 - High percentage of beta-lactamase-mediated resistance to penicillin:
 - 60% of *S. pneumoniae*
 - 30% of *H. influenza*
 - 80% *M. catarrhalis*

- 2. Cephalosporins:**

- Not currently recommended for empiric monotherapy of ABRS due to variable rates of resistance with *S. pneumoniae*.
 - Second generation:
 - Cefuroxime 500mg BID.
 - Third generation:
 - Cefpodoxime 200mg BID.
 - Cefixime 200mg BID.

- 3. Macrolides:**

- Clarithromycin 500mg BID.
 - Azithromycin 500mg OD for 1st day then 250mg BID for 4 days.
 - Alternative for Amoxicillin in patients with penicillin allergy.
 - Not recommended as initial empiric therapy due to high rates of resistance with *S. pneumoniae* (30%).

- **Failure of Medical Management:**
 - Consider a changing the antibiotics in either situation:
 - Worsening of symptoms after 48–72 hours of antibiotic treatment, or
 - Failure of improvement after 3-5 days of antibiotic treatment.
 - Patients should be re-evaluated for:
 - Confirming the diagnosis.
 - Sub-optimal antibiotic coverage.
 - Drug-resistant bacteria.
 - Non-bacterial cause.
 - Complications of ABRS.
 - Choices of Antibiotic after failure of initial empiric antibiotic therapy:
 - **Recommended (IDSA guidelines 2012):**
 - 1. High dose Amoxicillin-Clavulanate:**
 - The recommended second-line of treatment in ABRS.
 - 2g BID dosage (4g/day).
 - Indications:
 1. Failure of initial antibiotic therapy.
 2. Recent use of prior Antibiotics in the last 4-6 weeks.
 3. Recently hospitalization.
 4. Having a child in daycare in the household.
 5. From geographic regions with high endemic rates of penicillin non-susceptible *S. pneumoniae*.
 6. Immunocompromised patients.
 - 2. IV Third Generation Cephalosporins:**
 - Preferred alternative for high-dose amoxicillin-clavulanate for hospitalized patients with severe infections.
 - IV ceftriaxone and cefotaxime remain active against nearly all *S. pneumoniae*, including penicillin-resistant strains.
 - 3. Fluoroquinolones:**
 - Moxifloxacin 400mg OD.
 - Levofloxacin 500mg OD.
 - Alternatives for High dose Amoxicillin-Clavulanate in adults with penicillin allergy.
 - Levofloxacin can be used in children.

Antibiotic	Dosage	<i>Streptococcus pneumoniae</i>			<i>Haemophilus influenzae</i>	<i>Moraxella catarrhalis</i>	Anaerobic bacteria
		Sensitive	Intermediate	Resistant			
Amoxicillin	500 mg PO tid	+++	++	+	++	+	+
Clarithromycin	250-500 mg PO bid	++	++	+	++	+++	+
Azithromycin	500 mg PO first day, then						
	250 mg/d PO for 4 days	++	++	+	++	+++	+

Antibiotic	Dosage	<i>Streptococcus pneumoniae</i>			<i>Haemophilus influenzae</i>	<i>Moraxella catarrhalis</i>	Anaerobic bacteria
		Sensitive	Intermediate	Resistant			
Amoxicillin/clavulanate	500 mg PO tid	+++	++	+	+++	+++	+++
Cefuroxime	250-500 mg PO bid	+++	++	+	+++	++	++
Cefpodoxime + cefixime	200 mg PO bid	-	+++	++	+	+++	+++
	400 mg/d PO	++	-	-	+++	+++	-
Ciprofloxacin	500-750 mg PO bid	++	+	+	++	+++	+
Levofloxacin	500 mg/d PO	+++	+++	+++	+++	+++	+++
Trovafoxacin	200 mg/d PO	+++	+++	+++	+++	+++	+++
Clindamycin	300 mg PO tid	+++	+++	+++	-	-	+++
Metronidazole	500 mg PO tid	-	-	-	-	-	+++

Table 10. Antimicrobial Regimens for Acute Bacterial Rhinosinusitis in Adults
Riyadh et al. Notes

Indication	First-line (Daily Dose)	Second-line (Daily Dose)
Initial empirical therapy	● Amoxicillin-clavulanate (500 mg/125 mg PO tid, or 875 mg/125 mg PO bid)	● Amoxicillin-clavulanate (2000 mg/125 mg PO bid)
β-lactam allergy		● Doxycycline (100 mg PO bid or 200 mg PO qd) ● Doxycycline (100 mg PO bid or 200 mg PO qd) ● Levofloxacin (500 mg PO qd) ● Moxifloxacin (400 mg PO qd)
Risk for antibiotic resistance or failed initial therapy		● Amoxicillin-clavulanate (2000 mg/125 mg PO bid) ● Levofloxacin (500 mg PO qd) ● Moxifloxacin (400 mg PO qd)
Severe infection requiring hospitalization		● Ampicillin-sulbactam (1.5–3 g IV every 6 h) ● Levofloxacin (500 mg PO or IV qd) ● Moxifloxacin (400 mg PO or IV qd) ● Ceftriaxone (1–2 g IV every 12–24 h) ● Cefotaxime (2 g IV every 4–6 h)

Abbreviations: bid, twice daily; IV, intravenously; PO, orally; qd, daily; tid, 3 times a day.

**TABLE
33.7**

MEDICAL TREATMENT MODALITIES FOR ACUTE RHINOSINUSITIS

Class	Generic Example	Clinical Benefit	Recommended Use
Analgesic/antipyretic Saline irrigations	Acetaminophen	Decreases pain and fever Thins mucus, enhances mucociliary clearance, decreases inflammation	Recommended for AVRS and ABRS Recommended for AVRS and ABRS; may consider hypertonic irrigations
Topical decongestant	Oxymetazoline	Reduces mucosal edema and nasal airway obstruction	Consider in AVRS and ABRS for 3 d duration only
Systemic decongestant	Pseudoephedrine	Limited reduction of mucosal edema	Not as effective as topical decongestant
Topical corticosteroid	Fluticasone propionate	Improved symptom control in ABRS	Consider in ABRS with allergic component
Systemic corticosteroid	Prednisone	Not supported in literature	No evidence supporting use in AVRS or ABRS
Nonsedating antihistamine	Loratadine	Dries secretions	Consider in AVRS or ABRS with allergic component
Mucolytic	Guaifenesin	Thins tenacious secretions and phlegm	Improves symptom control in AVRS, limited literature support in ABRS
Antibiotic— <i>penicillins</i>	Amoxicillin ± clavulanate	Bactericidal, inhibit peptidoglycan cross-linking in cell wall synthesis	First-line for severe ABRS or mild ABRS that fails to improve with observation
Antibiotic— <i>cephalosporins</i>	Cefpodoxime proxetil, cefuroxime axetil, cefdinir	Bactericidal, inhibit peptidoglycan cross-linking in cell wall synthesis	Second most efficacious class for ABRS
Antibiotic— <i>sulfonamides and trimethoprim</i>	Trimethoprim-sulfamethoxazole	Bacteriostatic, inhibit folate synthesis, thereby preventing DNA replication and transcription	Alternative for first-line for severe ABRS or mild ABRS that fails to improve with conservative management in patients with penicillin allergy
Antibiotic— <i>tetracyclines</i>	Doxycycline	Bactericidal, inhibits protein synthesis	Good efficacy for ABRS
Antibiotic— <i>erythromycin, lincosamides, and macrolides</i>	Azithromycin, clarithromycin, erythromycin	Bactericidal, inhibits protein synthesis	Least efficacious for ABRS; consider as alternative for patients with penicillin allergy

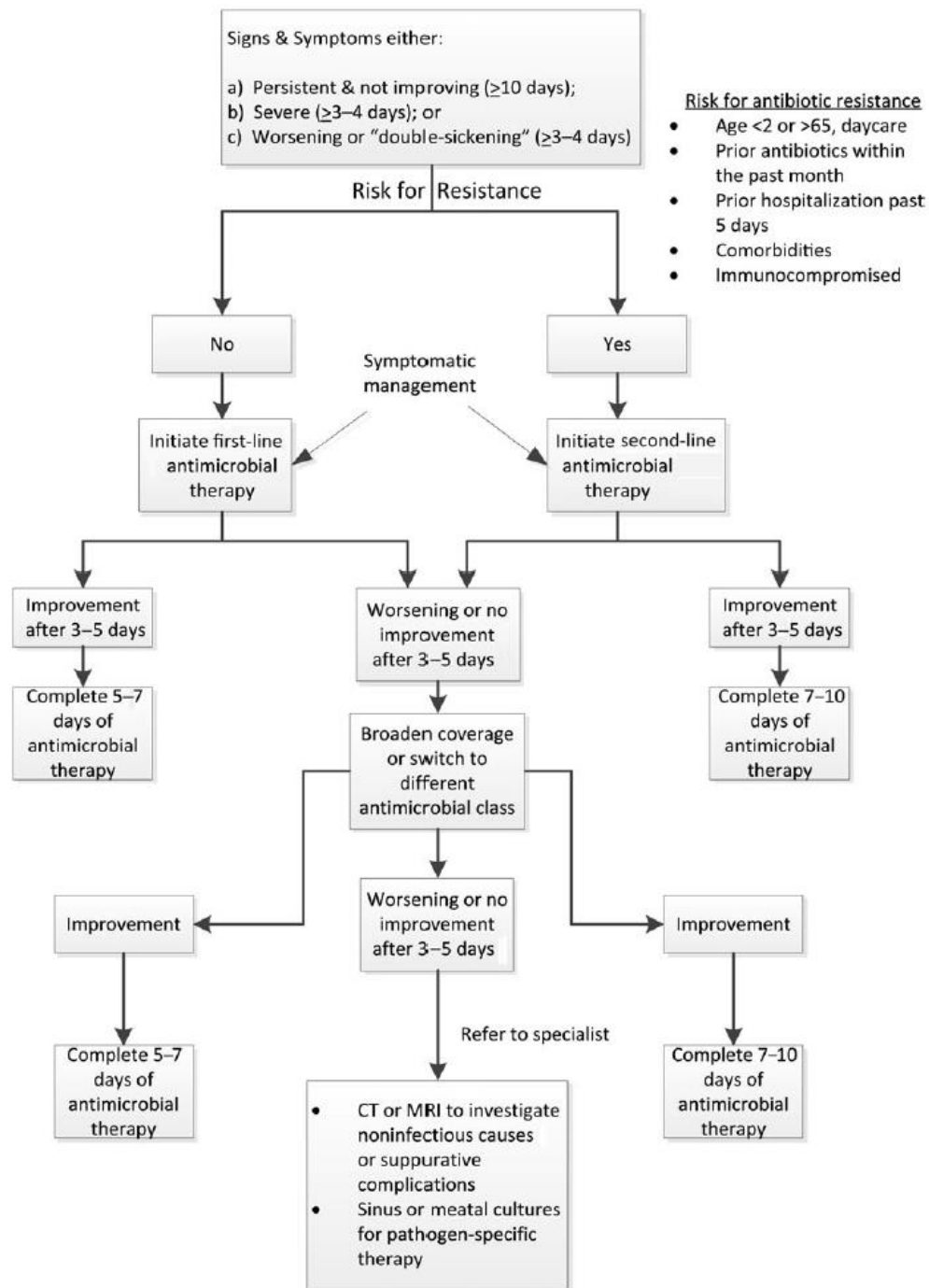


Figure 1. Algorithm for the management of acute bacterial rhinosinusitis. Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

Table 14. Indications for Referral to a Specialist

● Severe infection (high persistent fever with temperature $>39^{\circ}\text{C}$ [$>102^{\circ}\text{F}$]; orbital edema; severe headache, visual disturbance, altered mental status, meningeal signs)
● Recalcitrant infection with failure to respond to extended courses of antimicrobial therapy
● Immunocompromised host
● Multiple medical problems that might compromise response to treatment (eg, hepatic or renal impairment, hypersensitivity to antimicrobial agents, organ transplant)
● Unusual or resistant pathogens
● Fungal sinusitis or granulomatous disease
● Nosocomial infection
● Anatomic defects causing obstruction and requiring surgical intervention
● Multiple recurrent episodes of acute bacterial rhinosinusitis (ABRS) (3–4 episodes per year) suggesting chronic sinusitis
● Chronic rhinosinusitis (with or without polyps or asthma) with recurrent ABRS exacerbations
● Evaluation of immunotherapy for allergic rhinitis

- **Chronic Rhinosinusitis (CRS):**

- Chronic Rhinosinusitis is generally sub-classified as:
 - 1. CRS with Nasal Polyposis (CRSwNP):**
 - 2. CRS without Nasal Polyposis (CRSsNP).**
- Histology:
 - **CRSsNP:**
 - Fibrosis
 - Basement membrane thickening
 - Goblet cell hyperplasia
 - Subepithelial edema
 - Mononuclear cell infiltration:
 - More Neutrophilic infiltrate.
 - **CRSwNP:**
 - Intense edematous stroma with albumin deposition
 - Formation of pseudocysts
 - Subepithelial and perivascular inflammatory cell infiltration:
 - Most CRSWNPs are having eosinophilic infiltration except in Asians and patients with CR which are having more neutrophilic inflammatory infiltrate.
- Pathophysiology:
 - Dysfunctional immune response to exogenous factors at the sinonasal mucosal border leads to the mucosal inflammation, radiographic changes, and symptoms that characterize CRS.
 - CRSsNP and CRSwNP are regarded as distinct clinical entities based on diverse inflammatory and remodeling parameters.
 - **Remolding Process:**
 - Remodeling is a dynamic process in both health and disease that balances extracellular matrix (ECM) production and degradation.
 - Regulated by diverse mediators among which TGF- β protein takes a central role.
 - **CRSsNP:**
 - High concentrations of TGF- β reflecting the edema formation and lack of collagen production.
 - **CRSwNP:**
 - Low concentrations TGF- β reflecting excessive collagen deposition associated with fibrosis. in patients

■ Inflammation Process:

○ **CRSsNP:**

- Less severe inflammatory mucosal reaction.
- High TH1
- High IFN-g

○ **CRSwNP:**

- More severe inflammatory mucosal reaction.
- High TH-2 eosinophilic inflammation.
- High IL-5 and eosinophil cationic protein
 - White populations:
 - Mainly an eosinophilic inflammation.
 - 80% expressed IL-5 protein
 - More than 50% were characterized as eosinophilic infiltrate.
 - Asian populations:
 - Mainly a neutrophilic inflammation.
 - 15% expressed IL-5 protein.
 - Less than 10% were eosinophilic.

■ Microorganisms:

○ **CRSsNP:**

- Low colonization rate of *S aureus* (25%).

○ **CRSwNP:**

- High colonization rate of *S aureus* (60%) causing invasion of the epithelium and recreation of *S aureus* enterotoxins superantigens which elicit a massive inflammatory reaction:
 - Induces a significant increase in histamine levels, leukotriene numbers, and prostaglandin D2 levels, indicating mast cell activation.
 - Induces the formation of polyclonal IgE directed against multiple inhalant allergens.

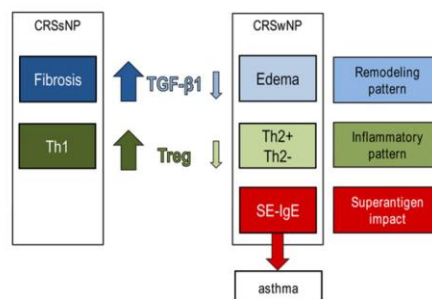


FIG 1. Phenotyping of CRS. CRSsNP and CRSwNP are regarded as distinct clinical entities based on diverse inflammatory and remodeling parameters. *Th2+*, IL-5 positive, Th2-skewed tissues; *Th2-*, IL-5 negative Th2-skewed tissues; *SE-IgE*, IgE antibodies to *Staphylococcus aureus* enterotoxins (SE). Arrows do not suggest causality.

▪ Eosinophils:

- Eosinophilic damage to sinonasal mucosa is not absolutely necessary for nasal polyposis or CRS to be present.
- Tissue eosinophilia is much higher in CRSwNP compared to CRSsNP.
- Eosinophils may be critical to polyp formation, but the relationship between CRSwNP and mucosal eosinophilia is not maintained in Asian polyps.
- Recent studies have demonstrated that tissue eosinophilia correlates with a relatively poor outcome independent of the presence or absence of polyps.
- Once present and activated, eosinophils are believed to damage the mucosa through degranulation and release of toxic mediators with resulting epithelial sloughing and tissue edema.
- The association of eosinophilia with refractory disease makes this cell a potentially important target in CRS.
- Eosinophils are steroid responsive, which explains therapeutic effects of glucocorticoids in CRS.

▪ Neutrophil:

- Role of neutrophil in CRS remains unclear.
- The highest sinus-tissue neutrophil levels are seen in Cystic Fibrosis patients.
- Degree of neutrophilic infiltrate in CRS mucosa is comparable between CRSsNP and CRSwNP as opposed to the eosinophilic infiltrate.
- Neutrophils do not appear to replace eosinophils in CRS mucosa; rather they are superimposed on the process.

▪ Mast cells:

- High levels of mast cells are seen in nasal polyps independent of atopy.
- Using anti-IgE for CRSwNP demonstrated efficacy independent of systemic allergy.

- **Nasal Polyposis (NP):**
- Nasal polyps are benign non-neoplastic masses of edematous sinonasal mucosa that arise as a result of chronic rhinosinusitis (CRS).
- No widely accepted classification system for NP disorders.
- **Factors affecting the development of NP:**
 - **Age:**
 - Nasal polyposis in childhood is very rare (0.1-0.2%) and should prompt evaluation to secure an accurate diagnosis and rule out causative conditions (CF, Kartagener syndrome).
 - Antrochoanal polyps accounts for 5% of Nasal polyps occurs more commonly in young adults.
 - AFRS is also predominantly a disease of teenagers and young adults.
 - **Genetic Predisposition:**
 - Accounts for some cases of NP disease, given the heritability of other eosinophilic inflammatory diseases such as Allergic Rhinitis and Asthma.
 - A genetic basis is well established for diseases that are known to cause NPs, such as CP and Kartagener syndrome.
 - Study found that 25% of NP patients had first degree relative with NP disease.
 - **Ethnic factor:**
 - Non-eosinophilic CRSwNP occurs more common in Asian populations
 - **Geographic factor:**
 - Allergic Fungal Rhinosinusitis (AFRS) occurs more common in warm or humid climates.

- **Pathogenesis of CRSwNP:**
 - Pathogenesis of CRSwNPs differ significantly from CRSsNP.
 - Inflammation in most CRSwNP includes an infiltrate of chronic inflammatory cells with a predominance of eosinophils, lymphocytes, and increased expression of pro-eosinophilic cytokines (IL-5) and chemokines.
 - Hypothesis for mechanisms of NPs formation:
 - **Leukotriene Hypothesis:**
 - Defects in Eicosanoid pathway is most closely associated with aspirin intolerance.
 - Increased synthesis of pro-inflammatory leukotrienes and decreased synthesis of anti-inflammatory prostaglandins (PGE2) have been proposed as a mechanism for aspirin-sensitive and aspirin-tolerant CRSwNP.
 - However, Leukotriene inhibitors have limited clinical efficacy in treatment of CRS.
 - **Staphylococcal Superantigen Hypothesis:**
 - Sinonasal cavities of CRSwNP patients are frequently colonized by Staphylococcus aureus secretes protein enterotoxin superantigens.
 - Superantigens bind to T-cell receptor and trigger cellular activation and stimulate pro-inflammatory cytokine secretion and formation of eosinophilic polyposis.
 - This hypothesis primarily addresses Caucasian CRSwNP, because superantigen effects are not seen in CRSsNP and uncommonly seen in Asian CRSwNP patients.
 - **Biofilms Hypothesis:**
 - Defect in the immune barrier might facilitate formation of Staphylococcal biofilms which permits secretion of superantigens that leads to eosinophilic polyposis.
 - **Immune Barrier Hypothesis:**
 - Defects in mechanical barrier and/or the innate immune response of sinonasal epithelium manifests as CRS.
 - Increased microbial colonization and accentuated barrier damage lead to increased stimulation of immune system with a compensatory adaptive immune response.
 - A defective immune barrier is hypothesized to predispose to CRS more broadly and can actually include the genetic forms of CRS such as those seen in CF.

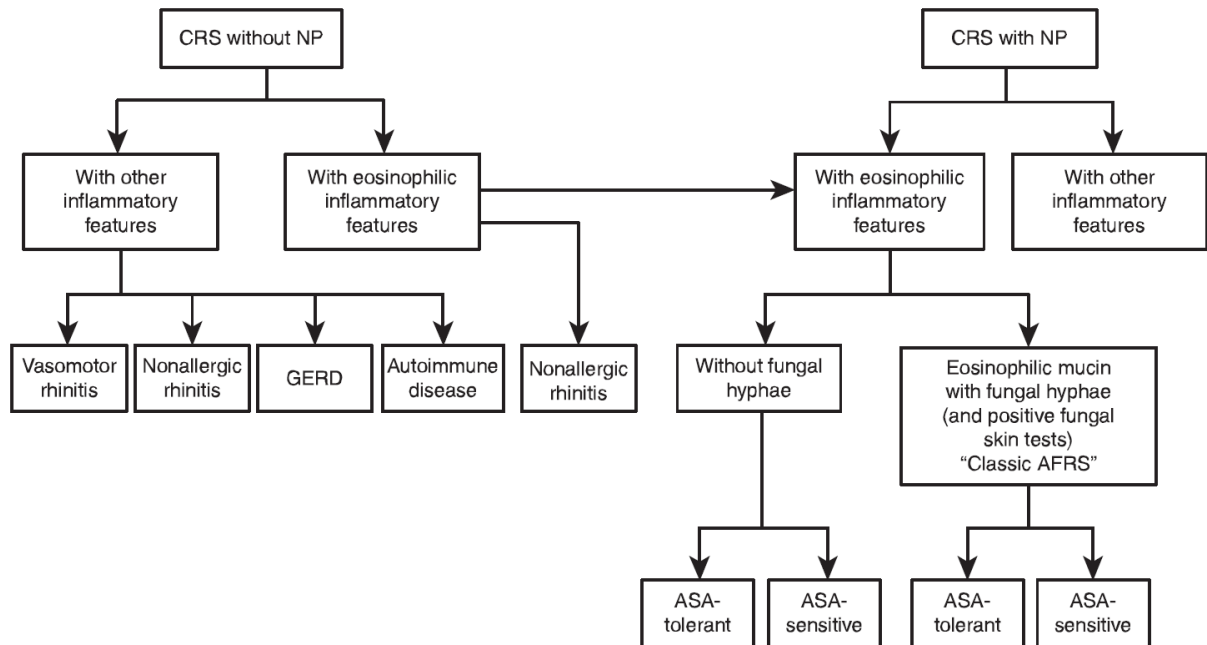


Figure 35.1 Classification for CRS.

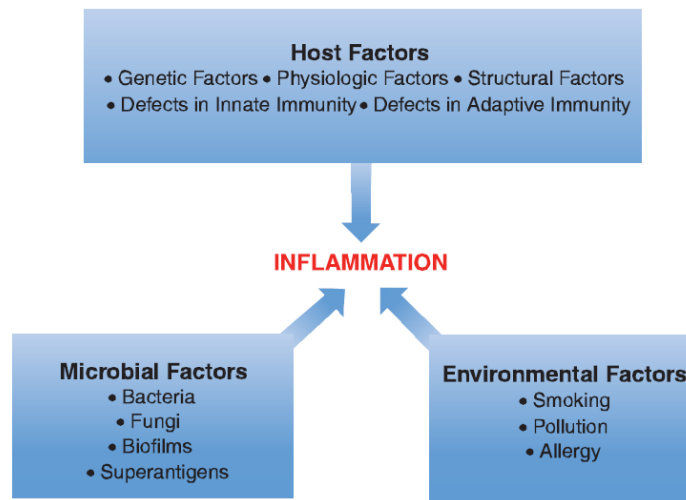


Figure 35.2 Underlying factors contributing to the pathophysiology of CRS.

- Pathogens in Chronic Rhinosinusitis:

1. Anaerobes
2. Staphylococcus Aureus
3. Streptococcus Pneumoniae
4. Haemophilus Influenzae
5. Fungal

- **Clinical Picture of CRS:**

○ History:

- Most patients will have months to years of progressively worsening sinonasal symptoms:
 - Nasal obstruction and congestion.
 - Nasal discharge and postnasal drip.
 - Allergic symptoms (sneezing, watery eyes)
 - Anosmia/Hyposmia
 - Due to obstruction of airflow into olfactory cleft or direct inflammation of olfactory cleft mucosa.
 - Facial Pain and Headache:
 - Forehead with frontal and maxillary sinusitis.
 - Bridge of nose, medial and deep to eye with ethmoid sinusitis.
 - Occipital headaches from sphenoid sinusitis.
- Subjective symptoms are poor predictor of polyp size or inflammatory disease burden in CRSwNP.
- Symptoms alone don't provide adequate warning of significant polyp recurrence after treatment of CRSwNP until nasal airway obstruction or hyposmia recur.

○ Physical Exam:

- Large NPs may be visible on anterior rhinoscopy.
- In most cases, NPs are only visible with nasal endoscopy.
- Nasal endoscopy is extremely helpful to monitor disease burden in CRSwNP.
- Long-standing cases with NP present with broadening of Nose and increased intercanthal distance.
- Polyps appearance:
 - Smooth, glistening, rounded masses.
 - Occurs in clusters.
 - Pale to yellow in color.
 - NPs may sometimes indistinguishable from tumors.
 - Painless and mobile with probing.
 - Do not bleed on touch.
- Site of origin of Nasal polyps:
 - Mainly from middle meatus lateral to Middle turbinate (Frontal, Anterior Ethmoid and Maxillary sinuses).
 - Medial to Middle turbinate (Sphenoid and Posterior Ethmoid sinuses).
 - Rarely from nasal septum or olfactory cleft.

**TABLE
34.1**

**CLASSIFICATION AND FEATURES
OF VARIOUS FORMS OF NP DISEASE**

Choanal polyp	Isolated, extends from site of origin (antrum, sphenoid, ethmoid, septum) to the posterior choana
CF	Children, neutrophilic infiltrate, sinus hypoplasia, or mucocoeles
AFRS	mucocoele formation with eosinophilic mucin, asymmetric sinus involvement, bone erosion, teens, and young adults
Eosinophilic mucin rhinosinusitis	eosinophilic mucin, symmetric sinus involvement, asthma, middle age, and older adults
AERD	asthma, history of asthma exacerbation after NSAID ingestion, symmetric sinus involvement
Churg-Strauss syndrome	asthma, peripheral eosinophilia >10%, vasculitis

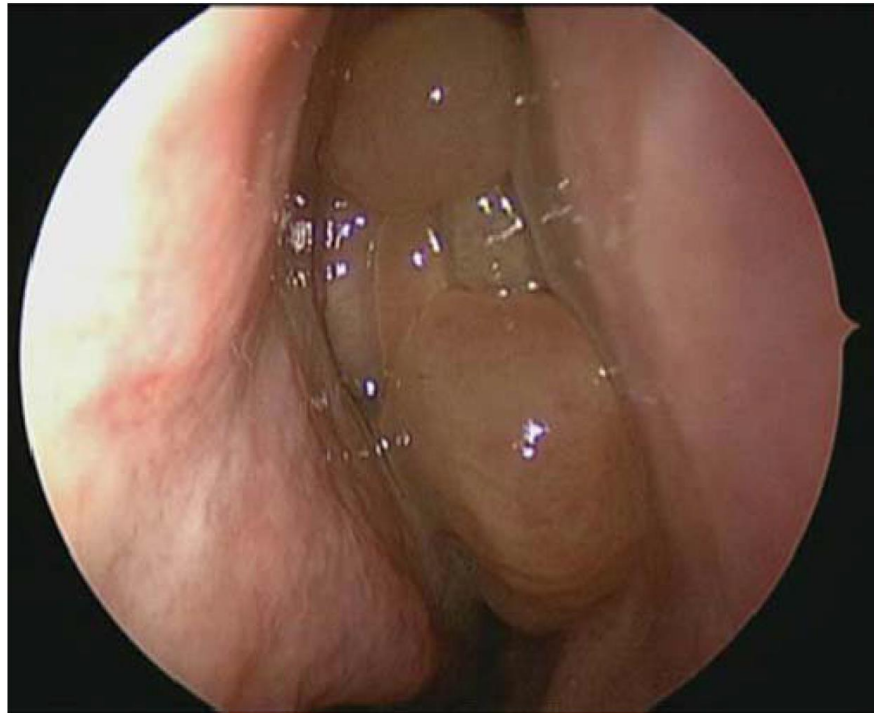


Figure 34.1 Endoscopic appearance of NPs in right nasal cavity, obscuring the middle turbinate.

- **Diagnostic Tests for CRS:**

○ **CT Sinuses:**

- Gold standard modality to evaluate the sinuses.
- Some degree of mucosal thickening occurs in almost **30%** of asymptomatic general public.
- Lund-Mackay staging system is the most common objective staging system used to evaluate and quantitates the paranasal sinuses on CT.
- Current CT sinus protocols:
 - Non-contrasted thin axial slice thicknesses <3 mm (ideally 1 mm is required for accurate image-guidance compatibility)
 - High-resolution multiplanar reconstruction in coronal and sagittal planes.
 - Contrast is needed in the following cases:
 1. Sinonasal neoplasm
 2. Complicated Rhinosinusitis
 3. Intracranial and orbital extension.
- **Indications of CT in CRS:**
 1. Failure of medical management.
 2. Complicated CRS
 3. Pre-op planning
- **CT Findings in CRS:**
 1. Mucosal Thickening
 2. Osteitic Thickening
 3. Bone Erosion
 4. Polyposis
 5. Intra-cranial or intra-orbital extension



**TABLE
28.3****INFORMATION OBTAINED FROM
THE DIFFERENT PLANES OF
SINUS CT**

Sinus CT Plane	Helpful Information
Coronal	Septal deviation OMC Uncinate attachment type (A,B,C) Middle turbinate variations Lamina papyracea anatomy Ethmoid anatomy Skull base anatomy/lateral lamella length Anterior ethmoid artery anatomy Sphenoid anatomy/sphenoethmoid cell (Onodi) Identify optic nerve and carotid artery dehiscence.
Sagittal	Frontal recess anatomy Slope of the skull base Sphenoid pneumatization pattern
Axial	Sphenoid anatomy/attachment of superior turbinate

- Evaluation of CT PNS:

- Examine Distribution of Mucosal disease:
 - Mucosal thickening
 - Air-fluid levels suggest acute inflammatory process.
- Inspect development of sinus:
 - Symmetry.
 - Aeration of sinus cavities.
- Examine Nasal structures, Airway, and Access.
- Evaluate for underlying causes of disease:
 - OMC patency
 - Paradoxical Turbinates
 - DNS
 - Concha bullosa
- Examine for Anatomical variations and Landmarks:
 - Cribriform plate
 - Posterior ethmoidal height
 - Thickness of skull base
 - Optic nerve
 - Orbital dehiscence
 - Carotid artery)
- Correlate between CT and symptoms or Endoscopic findings.

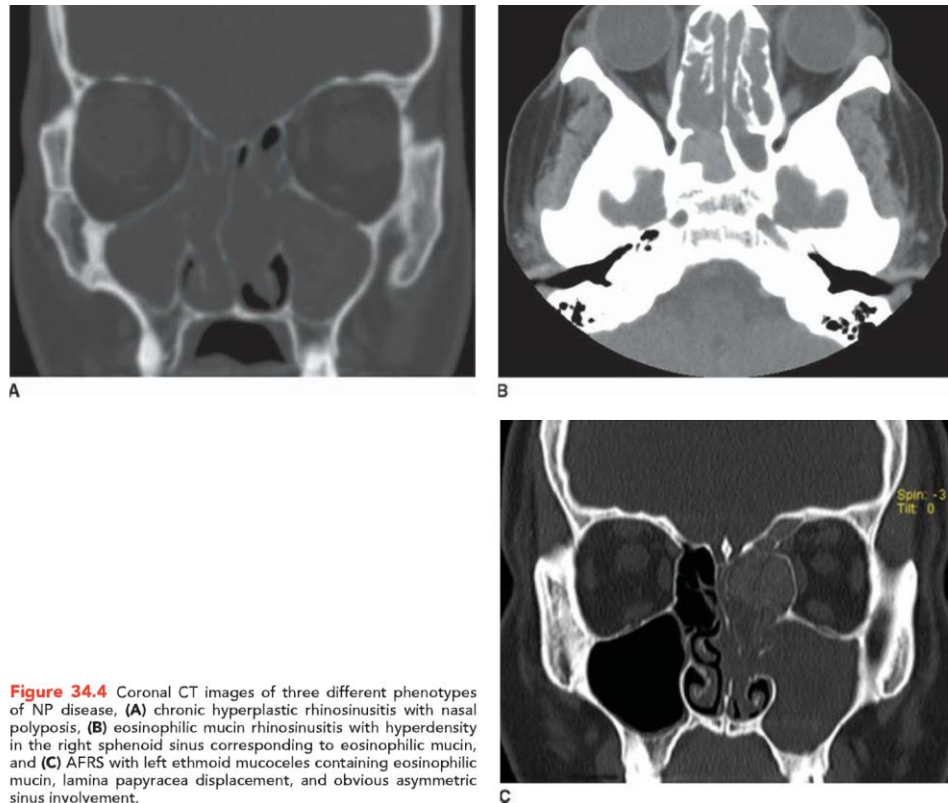


Figure 34.4 Coronal CT images of three different phenotypes of NP disease, (A) chronic hyperplastic rhinosinusitis with nasal polyposis, (B) eosinophilic mucin rhinosinusitis with hyperdensity in the right sphenoid sinus corresponding to eosinophilic mucin, and (C) AFRS with left ethmoid mucocoeles containing eosinophilic mucin, lamina papyracea displacement, and obvious asymmetric sinus involvement.

○ **MRI Sinuses:**

- Improved soft tissue detail.
- Can delineate polyp from retained secretions.
- Poor bone resolution.
- **Indications:**
 - Complicated CRS:
 - Intracranial and Intraorbital extension.
 - Fungal Rhinosinusitis.
- **Findings:**
 - **T1 MRI:**
 - Mucosal thickening is HYPER-intense.
 - Fluid is HYPO-intense.
 - Mucocele is HYPER-intense (Higher protein content).
 - **T2 MRI:**
 - Mucosal thickening is HYPER-intense.
 - Fluid is HYPER-intense.
 - Tumor tissue appears HYPO-intense compared to mucosal thickening.
 - Mucocele is HYPER-intense (Higher protein content).
 - AFRS is HYPO-intense/signal void due to presence of Metallic proteinaceous material, magnesium, iron, and calcium.



- **Ancillary Studies in CRS:**
 - **Middle Meatus Cultures/Biopsy:**
 - Endoscopically-directed middle meatus cultures are well correlated with cultures obtained by direct maxillary sinus aspiration.
 - Cultures can be obtained with a sterile swab or utilizing suction into a sterile trap device.
 - Indications of Culture in CRS:
 - Failure of medical management
 - Complicated CRS.
 - Immunocompromised patient.
 - Indications of Biopsy in CRS:
 - Unilateral disease
 - Suspicious of neoplasm
 - Immunocompromised patient to rule out fungal infection.
 - **Allergy Skin Testing:**
 - Confirm presence of an allergy to either a food or inhaled substance (allergen).
 - **Serum IgE Level:**
 - Total IgE measures all IgE in the blood.
 - Specific IgE (RAST) measures the amount of IgE to a specific allergen.

Assessment of CRS Severity:

- It is important to classify and stage CRS in order to decide the magnitude of the disease and to decide on the optimal management modality.
- Symptomatology, nasal endoscopy and CT evaluation are complementary in the evaluation of the patient with CRS.
- Staging methods of CRS:
 - Questionnaire based assessment.
 - Endoscopic scoring system.
 - CT based grading system.
- **Questionnaire based assessment of CRS:**
 - Disease based questionnaires are more sensitive than very general quality of life questionnaires.
 - Used mainly to assess the patient's response to a treatment modality over a period of time.
 - Should be integrated with objective information to make appropriate adjustments in treatment.
 - Most common used questionnaires:
 - 1. Rhinosinusitis Outcome Measure (RSOM-31):**
 - Validated questionnaire contains 31 questions about rhinosinusitis-specific and general items.
 - The patient is given this questionnaire and is expected to score each item according to its severity and importance to the patient.
 - Estimated completion time is 15 minutes.
 - 2. Sinonasal Outcome Test (SNOT-20)**
 - Validated Condensed modification of Rhinosinusitis outcome measure (RSOM-31).
 - Contains 20 questions about rhinosinusitis-specific and general items.
 - Does not include questions on nasal blockage or smell.
 - Easier to complete with higher patient compliance.
 - Estimated completion time is 10 minutes.
 - Commonly used for QOL measurements in CRS patients.
 - Therapy of CRS with nasal steroids resulted in a reduction of SNOT-20 scores.
 - **SNOT-16:**
 - Validated version.
 - Used to investigate the impact of smoking on post-operative QOL in CRS patients.
 - **SNOT-22:**
 - Non-validated version.
 - Covers all major symptoms of CRS by adding "nasal obstruction" and "loss of smell".

RHINOSINUSITIS OUTCOME MEASURE (RSOM-31)

*To let us know more about your nose and sinus disease, please answer the following questions about the consequences of your nose and sinus disease. There are no "right" or "wrong" answers, and only **you** can provide this information. For each of the statements listed please indicate **how much of a problem** it is to you. If a statement does not apply to you, please circle 0 and go to the next statement.*

Thank you for your time and participation.

	Not present / No problem	Very mild problem	Mild or slight problem	Moderate problem	Severe problem	Problem is as "bad as can be"
1. Stuffy / blocked nose	0	1	2	3	4	5
2. Runny nose	0	1	2	3	4	5
3. Sneezing	0	1	2	3	4	5
4. Decreased sense of taste or smell	0	1	2	3	4	5
5. Postnasal discharge	0	1	2	3	4	5
6. Thick nasal discharge / debris	0	1	2	3	4	5
7. Itchy or watery eyes	0	1	2	3	4	5
8. Sore or swollen eyes	0	1	2	3	4	5
9. Difficulty getting to sleep	0	1	2	3	4	5
10. Wake up during the night	0	1	2	3	4	5
11. Lack of a good night's sleep	0	1	2	3	4	5
12. Wake up tired	0	1	2	3	4	5
13. Fullness in ears	0	1	2	3	4	5
14. Ringing in ears	0	1	2	3	4	5
15. Dizziness	0	1	2	3	4	5
16. Pain in ears	0	1	2	3	4	5
17. Decreased hearing	0	1	2	3	4	5
18. Fatigue / being worn out	0	1	2	3	4	5
19. Reduced productivity	0	1	2	3	4	5
20. Poor concentration	0	1	2	3	4	5
21. Headache	0	1	2	3	4	5
22. Facial pain / pressure	0	1	2	3	4	5
23. Cough	0	1	2	3	4	5
24. Short of breath	0	1	2	3	4	5
25. Inconvenience of having to carry tissues	0	1	2	3	4	5
26. Need to rub nose / eyes	0	1	2	3	4	5
27. Need to blow nose repeatedly	0	1	2	3	4	5
28. Bad breath	0	1	2	3	4	5
29. Frustrated, impatient, or irritable	0	1	2	3	4	5
30. Feeling depressed or sad	0	1	2	3	4	5
31. Embarrassed by your symptoms	0	1	2	3	4	5

I.D.: _____

SINO-NASAL OUTCOME TEST (SNOT-20)

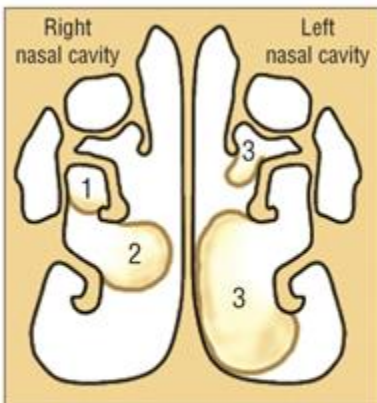
DATE: _____

Below you will find a list of symptoms and social/emotional consequences of your rhinosinusitis. We would like to know more about these problems and would appreciate your answering the following questions to the best of your ability. There are no right or wrong answers, and only you can provide us with this information. Please rate your problems as they have been over the past two weeks. Thank you for your participation. Do not hesitate to ask for assistance if necessary.

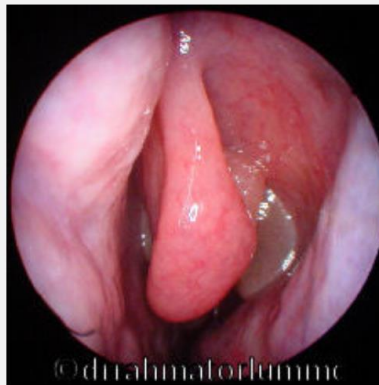
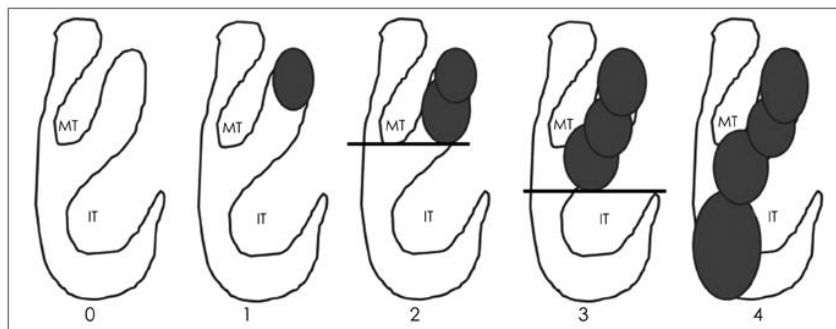
	No problem	Very mild problem	Mild or slight problem	Moderate Problem	Severe Problem	Problem as bad as it can be	5 Most Important Items
1. Considering how severe the problem is when you experience it and how frequently it happens, please rate each item below on how "bad" it is by circling the number that corresponds with how you feel using this scale: →							
1. Need to blow nose	0	1	2	3	4	5	☐
2. Sneezing	0	1	2	3	4	5	☐
3. Runny nose	0	1	2	3	4	5	☐
4. Cough	0	1	2	3	4	5	☐
5. Post-nasal discharge	0	1	2	3	4	5	☐
6. Thick nasal discharge	0	1	2	3	4	5	☐
7. Ear fullness	0	1	2	3	4	5	☐
8. Dizziness	0	1	2	3	4	5	☐
9. Ear pain	0	1	2	3	4	5	☐
10. Facial pain/pressure	0	1	2	3	4	5	☐
11. Difficulty falling asleep	0	1	2	3	4	5	☐
12. Wake up at night	0	1	2	3	4	5	☐
13. Lack of a good night's sleep	0	1	2	3	4	5	☐
14. Wake up tired	0	1	2	3	4	5	☐
15. Fatigue	0	1	2	3	4	5	☐
16. Reduced productivity	0	1	2	3	4	5	☐
17. Reduced concentration	0	1	2	3	4	5	☐
18. Frustrated/restless/irritable	0	1	2	3	4	5	☐
19. Sad	0	1	2	3	4	5	☐
20. Embarrassed	0	1	2	3	4	5	☐

2. Please mark the most important items affecting your health (maximum of 5 items) _____ ↑

- **Endoscopic based scoring system of CRS:**
 - Most useful objective tool for assessing the burden of inflammatory polyps in the nose and paranasal sinuses.
 - Reliable estimation of polyp size is facilitated by the use of a grading system.
 - Most common used scoring system:
 - **Lildholdt's clinical scoring system of nasal polyposis:**
 - **Grade 1:**
 - Polyps within middle meatus.
 - Not reaching inferior border of middle turbinate.
 - **Grade 2:**
 - Polyps extending into nasal cavity.
 - Reaching below inferior border of middle turbinate but not inferior border of inferior turbinate.
 - **Grade 3:**
 - Polyps extending into nasal cavity.
 - Reaching below inferior border of inferior turbinate.

Coronal view		Score	
	Right nasal cavity	Left nasal cavity	
	0	0	No polyps
	≤1	≤1	Polyps in middle meatus, not reaching below inferior border of middle turbinate
	≤2	≤2	Polyps reaching below inferior border of middle turbinate but not inferior border of inferior turbinate
	≤3	≤3	Large polyps reaching to or below lower border of inferior turbinate or polyps medial to middle turbinate
	—	—	Sum of score from both cavities

- **Meltzer's clinical scoring system of nasal polyposis:**
 - **Grade 1:**
 - Polyps within middle meatus.
 - Not reaching inferior border of middle turbinate.
 - **Grade 2:**
 - Polyps within middle meatus.
 - Reaching inferior border of middle turbinate.
 - **Grade 3:**
 - Polyps extending into nasal cavity.
 - Reaching inferior border of inferior turbinate.
 - **Grade 4:**
 - Polyps exceeding inferior border of inferior turbinate and filling up the nasal cavity.



Grade I



Grade II

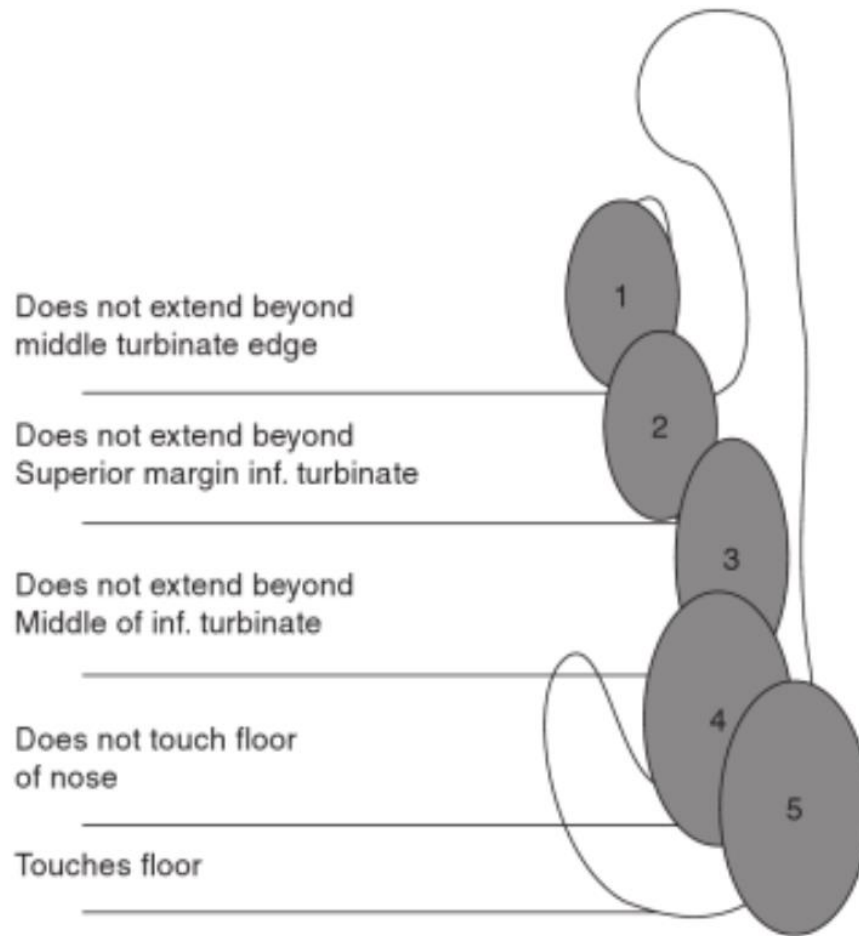


Grade III



Grade IV

- **Hadley's clinical scoring system of nasal polyposis:**
 - Modification of Meltzer's system includes one more stage i.e. Stage 5 which includes polyp touching the floor of the nasal cavity.



- **Lund-Kennedy endoscopic grading system of CRS:**
 - Helps in categorizing nasal polyposis and evaluate post-operative results following endoscopic sinus surgery.
 - Different endoscopic parameters were assessed and given a numeric score:
 - **Polyps:**
 - 0= Absent
 - 1= Polyps in middle meatus only
 - 2= Polyps beyond middle meatus but not obstructing the nasal cavity completely
 - 3= Polyps obstructing the nasal cavity completely
 - **Edema:**
 - 0= Absent
 - 1= Mild
 - 2= Severe
 - **Discharge:**
 - 0= Absent
 - 1= Clear thin discharge
 - 2= Purulent thick discharge
 - **Scarring (Post-op):**
 - 0= Absent
 - 1= Mild
 - 2= Severe
 - **Crusting (Post-op):**
 - 0= Absent
 - 1= Mild
 - 2= Severe

Table 12.4 Lund–Kennedy scoring system

Characteristic	Baseline and follow-up
Polyp, left (0,1,2,3)	
Polyp, right (0,1,2,3)	
Edema, left (0,1,2)	
Edema, right (0,1,2)	
Discharge, left (0,1,2)	
Discharge, right (0,1,2)	
<i>Postoperative scores to be used for outcome assessment only</i>	
Scarring, left (0,1,2)	
Scarring, right (0,1,2)	
Crusting, left (0,1,2)	
Crusting, right (0,1,2)	
Total points	

Polyps: 0 – absence of polyps, 1 polyps in middle meatus only, 2 polyps beyond middle meatus but not blocking the nose completely, 3 polyps completely obstructing the nose. Edema: 0 absent, 1 mild, 2 severe. Discharge: 0 no discharge, 1 clear, thin discharge, 2 thick, purulent discharge. Scarring: 0 absent, 1 mild, 2 severe. Crusting: 0 absent, 1 mild, 2 severe. Reprinted from Lund and Kennedy [6]), with permission from the Annals Publishing Company

- **CT based grading system of CRS:**
 - Most common used scoring system:
 - **Lund-Mackay CT grading system of CRS:**
 - Most widely used grading system.
 - Based entirely on CT findings.
 - Frontal, maxillary, anterior ethmoid, posterior ethmoid, and sphenoid sinuses are graded separately and the OMC is also scored.
 - Different anatomic regions are given a numeric score:
 - 0= No opacification
 - 1= Partial opacification
 - 2= Total opacification
 - Total possible score is 24 (12 each side).

**TABLE
35.3**
**LUND-MACKAY CT STAGING
SYSTEM**

Sinus	Right	Left
Frontal	0–2/2	0–2/2
Maxillary	0–2/2	0–2/2
Anterior ethmoid	0–2/2	0–2/2
Posterior ethmoid	0–2/2	0–2/2
Sphenoid	0–2/2	0–2/2
OMC	0 or 2/2	0 or 2/2
Total	Total score/24	

Each individual sinus is scored: 0 = clear, 1 = partial opacification, 2 = total opacification. Scoring for the OMC: 0 = clear, 2 = occluded. Adapted from Lund VJ, Kennedy DW. Quantification for staging sinusitis. The staging and therapy group. *Ann Otol Rhinol Laryngol Suppl* 1995;167:17–21.

- **Kennedy CT grading system of CRS:**

TABLE 35.4 **KENNEDY STAGING SYSTEM FOR CHRONIC SINUSITIS**

Stage	Findings
I	Anatomic abnormalities All unilateral sinus disease Bilateral disease limited to ethmoid sinuses
II	Bilateral ethmoid disease with involvement of one dependent sinus
III	Bilateral ethmoid disease with involvement of two or more dependent sinuses on each side
IV	Diffuse sinonasal polypsis

Adapted from Kennedy DW. Prognostic factors, outcomes and staging in ethmoid sinus surgery. *Laryngoscope* 1992;102(12 Suppl):1–18.

- **Diagnostic Correlations in CRS:**
 - Correlation between CRS symptoms and nasal endoscopy findings:
 - 70% of patients with CRS symptoms had normal nasal endoscopy.
 - Correlation between CRS symptoms and CT scan findings:
 - 35% of patients with CRS symptoms had negative CT scans.
 - Correlation between nasal endoscopy and CT scan findings:
 - Abnormal endoscopy (Polyps, purulence and mucosal edema) had a positive predictive value of 74% in predicting positive CT scans.
 - 80% of CRS patients with normal CT scan had a normal endoscopic evaluation.
 - Summary:
 - Symptoms did not correlate well with either CT findings or nasal endoscopy score.
 - Although nasal endoscopy and CT scores correlated well, a normal endoscopy could not assure a normal CT scan
 - Symptoms, nasal endoscopy and CT evaluation are complementary in the evaluation of CRS patients with CRS.

- **Treatment of CRS:**

○ **Medical Management:**

1. Saline Irrigations:

- **Strongly recommended as long-term therapy for patients with CRS (CRSSNP mainly).**
- Thins mucus, enhance mucociliary clearance and decrease inflammation.
- Hypertonic saline (Sinomarine) may have superior anti-inflammatory effects compared to isotonic saline.
- High-volume delivery is recommended over low-volume methods.

2. Topical Corticosteroid:

- **Strongly recommended as long-term therapy for patients with CRSSNP or CRSwNP.**
- Significant improvement regarding symptoms and polyp size as determined by endoscopy.
- Off-label use of Mixture of 2 Ampules of Pulmicort (0.5mg/ml) with 500ml Normal saline to be used as nasal irrigation (60ml BID).
- Effective topical therapy relies on:
 - **Surgical state of sinuses:**
 - Distribution of topical solution to non-operated sinuses is limited regardless of delivery device, volume, or head position.
 - <2% of total irrigation volume and <3% Nebulization will reach to non-operated sinuses.
 - Endoscopic sinus surgery is essential to effectively allow topical distribution to the sinuses.
 - Post-surgery distribution is superior with high volume positive pressure irrigation devices.
 - **Delivery Device and Technique:**
 - Simple low volume sprays and drops have very poor distribution and should be considered a nasal cavity treatment only (<50% will reach middle meatus).
 - Large volume squeeze bottles or passive flow devices appear to have the best efficacy post FESS.
- Side Effects:
 - Epistaxis
 - Dry nose
 - Nasal burning and irritation

3. Oral Corticosteroid (Prednisolone):

- The short term use of oral corticosteroids for CRSwNP, including during the perioperative period, is recommended as it improves the short-term outcomes (symptoms and polyp size).
- Oral corticosteroids is considered an option for patients with CRSsNP.
- Lack of long-term improvement limits the utility of oral steroids for CRSwNP because of known side-effect profiles.
- Prednisone for two weeks (30mg for 4 days followed by a 2-day reduction of 5mg).
- Side Effects:
 - Peptic ulcers
 - Sleep disturbance
 - Weight gain
 - Glaucoma
 - DM
 - Hypertension
 - Avascular necrosis of the hip
 - Adrenal suppression

4. Oral Antibacterial Antibiotics:

- Macrolides:
 - Antibacterial and anti-inflammatory effects.
 - Used for as low dose for long term period (>3 months).
 - **CRSsNP:**
 - Significant improvement in symptoms, disease-specific QOL, endoscopy findings in patients with CRSsNP treated with Roxithromycin 150 mg daily for 3 months.
 - An option to offer long-term Macrolide antibiotics for CRSsNP patients with low IgE levels (<200 µg/L) and ongoing symptoms despite topical steroids and saline irrigations.
 - **CRSwNP:**
 - Lack of quality data that demonstrate benefit of using Macrolides in patients with CRSwNP.

- Non-Macrolides:
 - Penicillins, Cephalosporins, Fluoroquinolones and tetracycline.
 - Antibacterial effect only.
 - Used only for short-term period (<4 weeks).
 - **CRSsNP:**
 - Only observational studies report improved outcomes after oral antibiotic in patients with CRSsNP and majority of these studies include adjunctive measures, such as saline irrigations or topical corticosteroid therapies.
 - Short-term treatment is considered option for patients with CRSsNP mainly during exacerbations with a positive culture.
 - **CRSwNP:**
 - Use of Doxycycline (200mg on first day followed by 100 mg daily for 20 days) in CRSwNP patients showed improvement in polyps size and reduction of postnasal discharge.
 - Short-term treatment is considered option for patients with CRSwNP.

5. Intravenous Antibacterial Antibiotics:

- Recommendations against routine use of IV antibacterial antibiotics in CRS due to lack of quality data coupled with high costs and high complication rates.

6. Topical Antibacterial Antibiotics:

- Recommendations against routine use of topical antibacterial antibiotics in CRS.
- Currently, no topical antibiotics are approved by the U.S. FDA for use in the sinuses.
- No standardization with regard to antibiotic preparation, dosage, dosing interval, or delivery method, and a paucity of data exists with regard to systemic absorption, kinetics, and overall safety, both for short-term and long-term administration.

7. Anti-Fungal Medications:

- Topical amphotericin B and systemic anti-fungal medications use in patients with CRS didn't show any significant improvements in symptoms.
- **Strong recommendation against the use of topical or systemic antifungal in patients with CRS, including CRSsNP, CRSwNP, and AFRS.**

8. Bacterial Lysates (OM-85 BV):

- Oral bacterial lysate Broncho Vaxom (OM-85 BV) is an immunostimulant enhance Th1-skewed immune response and results in symptomatic improvement for patients with CRSsNP.
- **Oral OM-85 BV treatment may be considered as an adjunct to standard medical treatment in adults with CRSsNP.**

9. Non-Sedating Anti-Histamine (Loratadine):

- Dries nasal secretions.
- Improves the outcomes in patients with Type I hypersensitivity to inhalant allergens.
- **Not recommended as a routine treatment for CRSsNP or CRSwNP.**

10. Leukotriene Inhibitors (Montelukast):

- Improves the outcomes in patients with Type I hypersensitivity to inhalant allergens.
- **Not recommended as a routine treatment for CRSsNP or CRSwNP.**

11. Monoclonal Antibodies:

- Anti-IgE (Omalizumab) therapy didn't show to improve the outcomes.
- **Not recommended as a routine treatment for CRSsNP or CRSwNP.**

12. Other Medical Therapies:

- **None of the following are supported by quality clinical data, and thus none are recommended for routine use in CRSsNP or CRSwNP:**
 - Immunotherapy (Atopic patients and AFRS).
 - Furosemide therapy (CRSwNP)
 - Systemic immunosuppressants (CRSwNP)
 - Aspirin desensitization (AERD)
 - Baby shampoo (biofilms)
 - Manuka honey
 - Proton pump inhibitors
 - Homeopathic remedies.

TABLE 44-3. Summary of Recommendations for Medical Therapy in Chronic Rhinosinusitis

Medical Therapy	EPOS12*	EBRR†
Oral antibacterial • Nonmacrolide • <3 to 4 weeks	CRSsNP: B+ CRSwNP: C+	CRS: option
Oral antibacterial • Macrolide • ≥12 weeks	CRSsNP: C+ CRSwNP: C+	CRS: option
Intravenous antibacterial	CRSsNP: did not review CRSwNP: did not review	CRS: recommend against
Topical antibacterial	CRSsNP: A– CRSwNP: no data	CRS: recommend against
Oral antifungal	CRSsNP: A– CRSwNP: A–	CRS: recommend against
Intravenous antifungal	CRSsNP: no data CRSwNP: no data	CRS: recommend against
Topical antifungal	CRSsNP: A– CRSwNP: A–	CRS: recommend against
Topical corticosteroid	CRSsNP: A+ CRSwNP: A+	CRSsNP: recommend CRSwNP: recommend
Systemic (oral) corticosteroid	CRSsNP: C+ CRSwNP: A+	CRSsNP: option CRSwNP: recommend
Saline irrigations	CRSsNP: A+ CRSwNP: D+	CRS: recommend
Antihistamines (in allergic patients)	CRSsNP: no data CRSwNP: D+	—
Leukotriene antagonists	CRSsNP: no data CRSwNP: A–	—
Anti-IgE monoclonal antibodies	CRSwNP: A–	—
Anti-IL-5 monoclonal antibodies	CRSwNP: D+	—

*As reported in the European Position Paper on Rhinosinusitis and Nasal Polyps 2012.⁴

†Published evidenced-based reviews with recommendations.^{5,6,61}

+, recommended; –, recommended against; A, directly based on category I evidence; B, directly based on category II evidence or extrapolated from category I evidence; C, directly based on category III evidence or extrapolated from category I or II evidence; D, directly based on category IV evidence or extrapolated from category I, II, or III evidence.

CRSsNP, chronic rhinosinusitis without nasal polyps; CRSwNP, chronic rhinosinusitis with nasal polyps; IgE, immunoglobulin E; IL-5, interleukin 5.

TABLE 44-1. Randomized Controlled Trials Evaluating Antibacterial Antibiotics for Chronic Rhinosinusitis

Study	Year	Design	LOE	Definition of CRS	N	Study Group(s)	Protocol	Clinical End Point(s)	Conclusion
Oral Antibacterial (Nonmacrolide)									
Van Zele et al. ¹⁷	2010	RCT	1B	Bilateral polyps	33	• Doxycycline • Placebo	Doxycycline 200 mg once, followed by 100 mg 1x/day for 20 days	• Polyp size • NPIF • Olfaction • Congestion • Rhinorrhea • Postnasal drainage	• Reduction in polyp size at week 12 • Reduction in postnasal drainage at week 2
Dellamona et al. ¹⁴	1994	RCT	1B	Symptoms + radiography findings	171	• Cefotiam • Cefixime	• Cefotiam 200 mg 2x/day for 10 days • Cefixime 200 mg 2x/day for 10 days	Clinical "cure" or "improvement"	No difference between groups
Legent et al. ¹⁵	1994	RCT	1B	3 months symptoms + CT findings	251	• Amoxicillin/clavulanate • Ciprofloxacin	• Amoxicillin/clavulanate 500 mg 3x/day for 9 days • Ciprofloxacin 500 mg 2x/day for 9 days	• Clinical "cure" • Nasal drainage	No difference between groups
Huck et al. ¹⁶	1993	RCT	1B	"Nonresolving sinus disease"	15	• Cefaclor • Amoxicillin	• Cefaclor 500 mg 2x/day for 10 days • Amoxicillin 500 mg 3x/day for 10 days	• "Success/failure" • Radiography findings	No difference between groups
Oral Antibacterial (Macrolide)									
Videler et al. ²⁰	2011	RCT, blinded	1B	EPOS	60	• Azithromycin • Placebo	Azithromycin 500 mg QD × 3 days, then 500 mg/wk × 11 weeks	• SNOT-22 • Patient Response Rating Scale • VAS symptoms • Nasal endoscopy • NPIF • Olfaction • Short Form-36 • Sinonasal cultures	No significant benefit was found over placebo
Wallwork et al. ¹⁹	2006	RCT, blinded	1B	CRS Task Force Criteria, CT scores	59	• Roxithromycin • Placebo	Roxithromycin 150 mg QD × 3 months	• Symptoms (SNOT-20) • Patient response scale • Peak nasal inspiratory flow • Saccharine transit time • Nasal endoscopy • Olfactory function • Markers from nasal lavage	• Improved patient response, SNOT-20, and endoscopy but no improvement in other outcomes • Better response in patients without elevated IgE
Intravenous Antibacterial*									
Topical Antibacterial									
Videler et al. ³⁶	2008	RCT	1B	3 months symptoms + objective findings + staphylococcal culture	14	• Bacitracin/colimycin • Placebo	• Bacitracin/colimycin 8 mL (830/640 ug/mL) 2x/day for 8 weeks + oral levofloxacin for 2 weeks • Saline 2x/day for 8 weeks + oral levofloxacin for 2 weeks	Nebulizer	• VAS Symptoms • Short Form-36 • Disease-Specific Symptom Score
Desrosiers and Salas-Prado ³⁵	2001	RCT	1B	3 months symptoms	20	• Tobramycin • Placebo	• Tobramycin 4 mL (20 mg/mL) 3x/day for 4 weeks • Saline + 1 mg/mL quinine	Nebulizer	• RQLQ • Pain • Mucosal edema • Secretions • Postnasal drainage • Congestion
Sykes et al. ³⁴	1986	RCT	2B	None	50	• Dexamethasone, tramazoline, and neomycin • Dexamethasone, tramazoline	• Dexamethasone 20 µg, tramazoline 120 µg, neomycin 100 µg per nostril 4x/day for 2 weeks • Dexamethasone 20 µg, tramazoline 120 µg per nostril 4x/day for 2 weeks	Metered dose spray	• Symptoms • Nasal resistance • Mucociliary clearance

*No studies have been performed.

Data from Soler ZM, Over SL, Kern RC, et al. Antimicrobials and chronic rhinosinusitis with or without polyposis in adults: an evidenced-based review with recommendations. *Int Forum Allergy Rhinol* 2013;3(1):31-47.

CRS, chronic rhinosinusitis; CT, computed tomography; EPOS, European Position Paper on Rhinosinusitis and Nasal Polyps; IgE, immunoglobulin E; LOE, level of evidence; NPIF, nasal peak inspiratory flow; QD, daily; RCT, randomized controlled trial; RQLQ, Rhinoconjunctivitis Quality of Life Questionnaire; SNOT, Sino-Nasal Outcomes Test; VAS, visual analog scale.

TABLE 44-2. Summary of Recommendations for the Use of Steroids in Chronic Rhinosinusitis

CRS Patients	Grade of Evidence	Balance of Benefit to Harm	Recommendation	Steroid Protocol
CRSsNP	C	Perceived balance of benefit to harm	Option	
CRSwNP	A	Preponderance of benefit over harm in small, short-term follow-up	Strongly recommend	Consider oral prednisone for short term in CRSwNP
AFS	B	Benefits over harm in short term	Recommend	Consider oral prednisone for patients in AFS
Perioperative use in AFS	B	Benefit over harm, particularly after surgical debridement	Recommend	Consider oral prednisone perioperatively in AFS
Perioperative use in CRSwNP	B	Benefit over harm	Recommend	Consider oral prednisone perioperatively in CRSwNP
Perioperative use in CRSsNP	NA	NA	No recommendation	

Data from Poetker DM, Jakubowski LA, Lal D, et al: Oral corticosteroids in the management of adult chronic rhinosinusitis with and without nasal polyps: an evidence-based review with recommendations. *Int Forum Allergy Rhinol* 2013;3(2):104-120.

AFS, allergic fungal sinusitis; CRSsNP, chronic rhinosinusitis without nasal polyps; CRSwNP, chronic rhinosinusitis with nasal polyps; NA, not applicable.

○ **Surgical Management (FESS):**

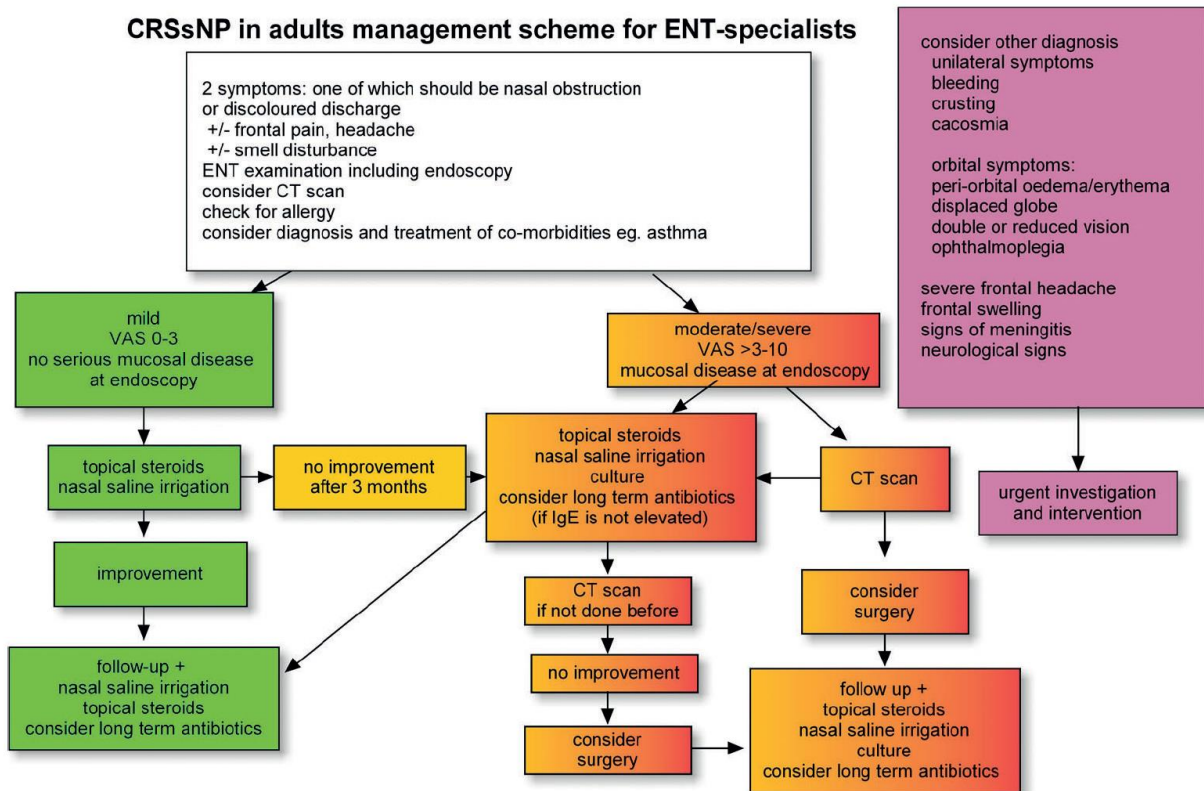
- Indicated in patients with CRSsNP and CRSwNP who failed full medical management.
- Goals of surgery:
 - Reduce the overall burden of inflammatory disease.
 - Provide permanent drainage and ventilation of the sinuses
 - Provide post-operative access to diseased sinuses.
 - Augment postoperative medication regimen (saline irrigations and topical corticosteroids)
- Steps of surgery:
 - Remove nasal polyps
 - Open the obstructed ostia
 - Spare sinonasal mucosa
 - Clear inspissated secretions
- Revision Surgery:
- Indicated in 20% of operated patients who respond unsatisfactorily to initial sinus surgery with concomitant medical therapy.
 - Factors increases risk for requiring revision surgeries:
 - Previous revision surgery
 - Extensive polyps
 - Bronchial asthma
 - ASA intolerance
 - Cystic fibrosis
 - Common reasons to fail primary surgery:
 - Middle turbinate lateralization
 - Adhesions and scar in middle meatus
 - Adhesions and scar in frontal recess
 - Incompletely resected uncinate process
 - Retained ethmoid cells
 - Success rates of revision endoscopic sinus surgery have been reported from 50-70%
 - Complication rates of revision surgery are higher (1-7%) when compared with initial surgery.

- Peri-Operative care:
 - Surgical management is used to decrease amount of inflammation so medical treatment become more effective and recurrence rate reduced.
 - CRSwNP has high tendency for recurrence without aggressive postoperative medical management.
 - **Oral Steroid:**
 - Given for 5 days pre-op and 9 days post-op.
 - Pre-op doses reduce intra-op bleeding and facilitate complete surgery.
 - **Topical Steroid:**
 - Off-label use of Mixture of 2 Ampules of Pulmicort (0.5mg/ml) with 500ml Normal saline to be used as nasal irrigation (60ml BID).
 - **Oral Antibiotics:**
 - Doxycycline (200mg on first day followed by 100 mg daily for 20 days).
- **Difficult-to-treat CRS:**
- CRS patients who do not reach an acceptable level of symptomatic control despite all of the following:
 - Adequate surgery
 - Intranasal corticosteroid treatment.
 - Up to 2 short courses of antibiotics or systemic corticosteroids in the last year.

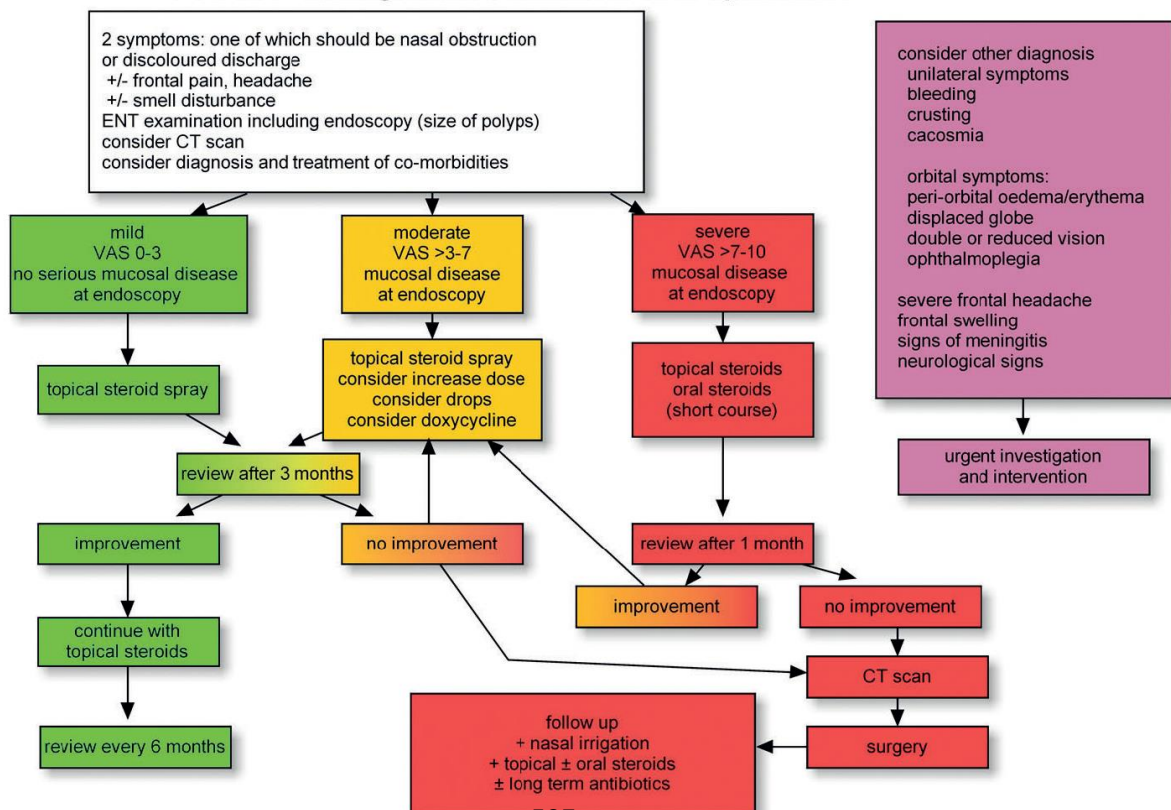
Table 1. Assessment of current clinical control of CRS.

Characteristic	Assessment of current clinical control of CRS (in the last month)		
	Controlled (all of the following)	Partly Controlled (at least one present)	Uncontrolled
Nasal blockage	Not present or not bothersome	Present on most days of the week	Three or more features of partly controlled CRS
Rhinorrhea/ Postnasal drip	Little and mucous	Mucopurulent on most days of the week	
Facial pain/headache	Not present or not bothersome	Present	
Smell	Normal or only slightly impaired	Impaired	
Sleep disturbance or fatigue	Not impaired	Impaired	
Nasal endoscopy (if available)	Healthy or almost healthy mucosa	Diseased mucosa (nasal polyps, mucopurulent secretions, inflamed mucosa)	
Systemic medication needed to control disease	Not needed	Need of up to 1 short course of antibiotics or systemic corticosteroids in the last three months	Need of long term antibiotics or systemic corticosteroids in the last month

CRSsNP in adults management scheme for ENT-specialists



CRSwNP management scheme for ENT-specialists



- **Antrochoanal Polypi:**
- Retention cyst arises from mucosa of Maxillary sinus and protrudes through the Antrum and may enter the Choana and Nasopharynx.
- Usually Unilateral.
- It has three parts:
 1. Antral: Thin stalk.
 2. Choanal: Round and globular.
 3. Nasal: Flat from side to side.
- Clinical Features:
 - Seen in children and young adults.
 - Single and unilateral
 - Unilateral nasal obstruction
 - Bilateral nasal obstruction if polyp grows into Nasopharynx
 - Hyponasality.
 - Nasal discharge, mostly mucoid, may be seen on one or both sides.
 - Signs
- Examination:
 - As Antrochoanal polyp grows posteriorly, it may be missed on Anterior rhinoscopy.
 - Smooth greyish mass
 - Soft and moved with a probe.
 - Posterior rhinoscopy may reveal a globular mass filling Choana or Nasopharynx.
 - Large polyp may hang down behind Soft palate and present in Oropharynx.



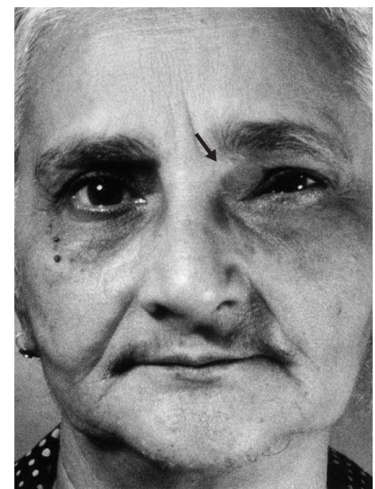
- Histopathology of Antrochanal Polyp:
 - Not Associated with Eosinophilia or typical inflammatory markers of Nasal polyposis.
- Management:
 - Endoscopic removal including the pedicled site within Maxillary Sinus.

	Antrochoanal polypi	Ethmoidal polypi
Age	Common in children	Common in adults
Aetiology	Infection	Allergy or multifactorial
Number	Solitary	Multiple
Laterality	Unilateral	Bilateral
Origin	Max. sinus near the ostium	Ethmoidal sinuses, uncinate process, middle turbinate and middle meatus
Growth	Grows backwards to the choana; may hang down behind the soft palate	Mostly grow anteriorly and may present at the nares
Size & shape	Trilobed with antral, nasal and choanal parts. Choanal part may protrude through the choana & fill the nasopharynx obstructing both sides	Usually small and grape-like masses
Recurrence	Uncommon, if removed completely	Common
Treatment	Polypectomy; endoscopic removal or Caldwell-Luc operation if recurrent	Polypectomy Endoscopic surgery or ethmoidectomy (which may be intranasal, extranasal or transantral)

- **Mucocele:**
- Result of obstruction of sinus ostium with accumulation of mucus and eventual expansion of the sinus and destruction of its bony walls.
- Contents of mucocele are **sterile**.
- **Most common involved sinus:**
 - o **Frontal** > Ethmoid > Maxillary > Sphenoid.
- Types of Mucocele:
 1. Primary:
 - Arises de novo.
 - Mucous Retention Cyst.
 2. Secondary:
 - Arises secondary to Surgery, Trauma, or Polyposis.
- Diagnosis:
 - o **Paranasal Sinuses CT:**
 - Sinus expansion with complete opacification.
 - Bone remodeling (Thinned sinus walls).
 - o **Paranasal Sinuses MRI:**
 - **T1** shows HYPER-intensity due to high protein content
 - **T2** shows HYPER-intensity fluid with central HYPO-intensity mucoid concretion.

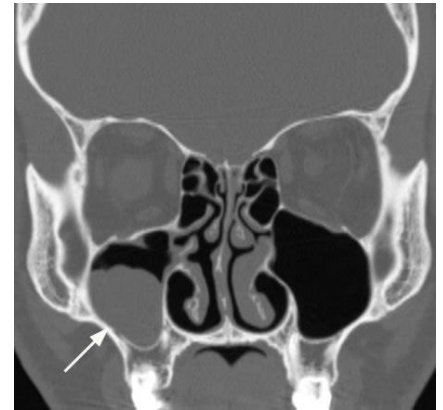


- Frontal Sinus Mucocele:
 - Most common.
 - Presents in Superomedial quadrant of Orbit (90%).
 - Displaces Eyeball forward, downward and laterally.
 - Sometimes, Cystic swelling in Forehead (10%)
 - Swelling is cystic and non-tender, Egg-shell crackling.
 - Patient's complaints are usually mild and may include headache, diplopia and proptosis.
- Ethmoid Sinus Mucocele:
 - Causes expansion of Medial wall of Orbit.
 - Displaces Eyeball forward and laterally.
- Maxillary Sinus Mucocele:
 - Occur as a complication of chronic sinus inflammation when its ostium is blocked.
- Sphenoid sinus Mucocele:
 - Arises from slow expansion and destruction of sphenoid.
 - **Superior Orbital Fissure Syndrome:**
 - Involvement of **CN-III, IV, VI and V1.**
 - **Orbital Apex Syndrome:**
 - Superior Orbital Fissure Syndrome + CN-II, V2.
 - Involvement of **CN-II, III, IV, VI, V1 and V2**
 - Exophthalmos is always present.
 - Pain is localised to Orbit or Forehead.
- Pyocele or Mucopyocele:
 - Similar to mucocele but its contents are purulent.
 - Result from infection of a mucocele of any Sinuses.
- Complications of Mucocele:
 1. Infection (Pyocele)
 2. Rupture (Bacteremia)
 3. Orbital and intracranial involvement
 4. Pituitary abnormalities
 5. Cosmetic deformity
- Management of of Mucoele (Always Surgical):
 1. FESS.
 2. Open:
 - Inaccessible lesions
 - Lateral lesions in Frontal sinus.



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- **Mucous Retention Cyst:**
- Retention cyst due to obstruction of Seromucinous gland duct.
- Does not cause bone erosion.
- Does not cause sinus expansion with complete opacification.
- Infectious or Allergic in origin.
- Typically Asymptomatic.
- Larger cysts may cause dental pain or symptoms from sinus obstruction.
- Diagnosed an incidental finding on CT Paranasal sinuses. (10%)
- No treatment is generally required for Asymptomatic retention cysts.
- Most of them regress spontaneously over a period of time.



- **HIV with Rhinosinitis:**
- 75% of untreated HIV patients develop Rhinosinitis.
- Secondary to:
 - o Impaired Immunity
 - o Mucociliary dysfunction
 - o Atopy
- Pathogens:
 - o CD4 count >200:
 - Similar to Immunocompetent Patients
 - o **CD4 count <200:**
 - Increase risk of unusual and more virulent organisms
 - Fungal, CMV, Pseudomonas, Myobacterium, Pneumocystic Jiroveci (carinii).
- Management:
 - o CD4 count >200:
 - Managed similar to Immunocompetent Patients
 - o **CD4 count <200:**
 - Course of broad spectrum Antibiotics.
 - Typical Sinus regimen (Decongestants, Saline irrigations)
 - Aggressive early work-up (CT/MRI of Paranasal sinuses)
 - Low threshold for sinus aspirate for culture and sensitivity
 - Early surgical management

- **Pediatric Rhinosinusitis:**
- Usually involves Maxillary and Anterior Ethmoid sinuses.
 - o Sphenoid and Frontal sinuses are Less Developed.
- Most Common Pathogens:
 1. Streptococcus Pneumoniae.
 2. Staphylococcus Aureus
 3. Moraxella Catarrhalis
 4. Haemophilus Influenzae
- Clinical Picture:
 - o Similar presentations to URI.
 - o Cough, Halitosis, Persistent Nasal Obstruction, Rhinorrhea, and Fever.
- Persistent Rhinosinusitis in Children:
 1. Adenoid hypertrophy
 2. Cystic Fibrosis
 3. Primary Ciliary Dyskenesia
 4. Immunodeficiencies
- Management:
 1. Antibiotics:
 - o Primary therapeutic agent in pediatric Rhinosinusitis.
 - o Agents used are similar to Adults
 2. Adjunctive Medical Agents:
 - o Saline irrigations.
 - o Nasal corticosteroids
 - o Oral decongestants
 - o Mucolytics
 3. Allergy Management
 4. Management (FESS):
 - o Indications:
 - Severe Rhinosinusitis.
 - Failed extensive medical management.
 - o Relationships of Anterior Ethmoid, Middle Meatus, Uncinate Process, Ethmoid Infundibulum, Hiatus Semilunaris and Bulla Ethmoidalis are Relatively Constant Landmarks throughout Life.
 - o Sphenoid and Frontal sinuses do Not need to be explored since they don't develop until 8-12 years old.
 - o Mini-FESS:
 - Fewer complications
 - Removing Uncinate Process
 - Opening Maxillary Ostium.
 - Taking down Bulla Ethmoidalis or Anterior Ethmoid.
 - Severe Rhinosinusitis.
 - o Balloon Catheterization.

5. Adenoidectomy:

- Controversial relationship with Rhinosinusitis.
- May harbor pathogens and block drainage.
- Considered as an Adjunctive therapeutic option

6. Antral Lavage:

- Controversial efficacy.
- Addresses only Maxillary sinus.
- Typically require multiple lavages

- **Cystic Fibrosis and Rhinosinusitis**

- Autosomal Recessive Multisystem disorder.
- Mutation in CF Trans-membrane conductance Regulator (**CFTR**) gene.
- Located in the Long arm of chromosome 7.
- Leads to impaired Chloride ion transmembranal transport.

- **Clinical Manifestations:**

- Abnormal Exocrine gland function.
- Thick mucus keeps bacteria from being cleared and prevents Antibiotics from being effective.
- Chronic progressive pulmonary disorder
- Pancreatic, Hepatobiliary, and Genitourinary manifestations
- Chronic Rhinosinusitis
- 10% Nasal Polyps

- **Diagnosis:**

- Sweat Chloride Test:
 - >60–90 mmol/L
- Genetic screening

- **Pathogens:**

- Pseudomonal infections
- Burkholderia cepacia
- MRSA
- E.coli
- Aspergillus

- **Management of Rhinosinusitis in CF:**

- **Medical Management:**

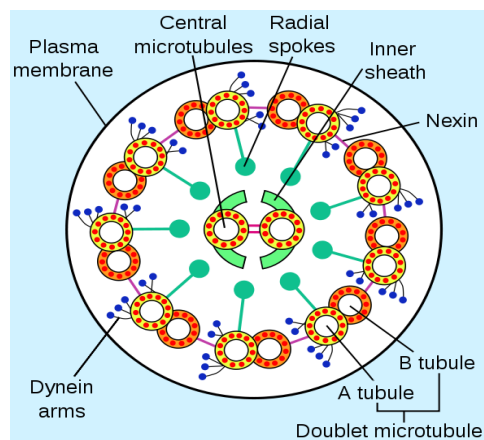
- Aggressive Nasal Hygiene
 - Nasal Saline Irrigation
 - Irrigation with Antipseudomonal Antibiotics.
 - Topical Steroid
- Sinus Aspirate for C/S
- Long term Antibiotics.

- **Surgical Management:**

- Avoid Surgery
 - High Recurrence rate
 - Increases Nasal scarring
 - Patients don't tolerate long GA due to retained pulmonary secretion.
- Indications of Surgery:
 - Uncontrolled Pain.
 - Nasal obstruction
 - Mucocele
 - Unresolved fever
 - Fungal infections
 - Pulmonary Exacerbations that correlate with Sinonasal disease.
- Surgical Considerations:
 - Vit. K supplement
 - Surgery should be less than 1 hour to avoid respiratory problems.
 - Surgery is for relieving obstruction not for cure.
 - Keep landmarks.
 - Wide Maxillary Antrostomies.
 - Unresolved fever
- Post-OP Care:
 - Aggressive Irrigations

- **Primary Ciliary Dyskinesia:**

- Immotile Cilia Syndrome.
- Autosomal recessive disorder.
- Absent **Outer Dynein Arm** causing abnormal ciliary motion and impaired mucociliary clearance.
- **Kartagener's Triad: (50%)**
 1. Chronic Rhinosinusitis.
 2. Bronchiectasis
 3. Situs Inversus



- Clinical Picture:

- Neonatal Pneumonia
- Continuous Rhinorrhoea from 1st day of life
- Chronic Productive Cough.
- Atypical Asthma non-responsive to treatment.
- Bronchiectasis (Most significant morbidity).
- Chronic Rhinosinusitis
- Nasal polyps
- Otitis media with Effusion (OME)
- Prolonged smelly Ear discharge not responding to treatment.
- Male infertility due to immotile sperms

- Diagnosis of PCD:

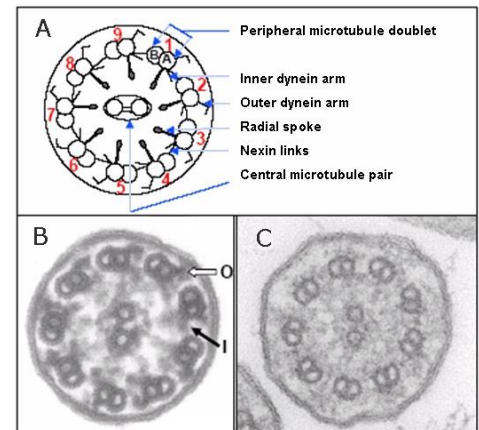
1. Transmission Electron Microscopy:

- Gold standard diagnostic test for PCD
- Requires a biopsy of Respiratory epithelium
- Typically obtained by brushing or scraping inferior surface of Nasal Turbinate or brushing Bronchial surface via Bronchoscopy.

2. High-speed videomicroscopy of Ciliary motility:

- Immotility/dysmotility patterns.

3. Low Nasal Nitric Oxide production.



- Management of Rhinosinusitis in PCD:

- Aggressive Antimicrobial Therapy, Mucolytics, Bronchodilators, and Postural drainage.
- Surgical Management:
 - ESS:
 - "Gravity dependent" surgical inferior antrostomies for refractory sinus disease.
 - Avoid Functional Endoscopic Sinus Surgery (FESS) with standard Antrostomies.
 - FESS doesn't not work since there is No normal Mucociliary clearance.
 - Consider "Gravity dependent" surgical inferior antrostomies for refractory sinus disease.
 - Ventilation tubes for Chronic Otitis Media.

Fungal Rhinosinusitis:

- Main forms of fungi:

1. Yeasts:

- Unicellular organism.
- 3-15 μm in diameter.
- Reproduce asexually by budding.
- If these buds do not detach, a chain of fungal cells results which is known as Pseudohyphae.

2. Molds:

- Multicellular organism
- 2-10 μm in diameter.
- Grow by branching into structures termed Hyphae.

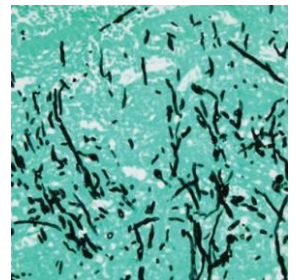
3. Spores:

- Reproductive structures that can be produced in presence of unfavorable conditions.
- Spores can withstand many adverse conditions, and are dispersed widely throughout the environment.
- Once exposed to a favorable environment, they begin to grow.
- Inhalation of spores is thought to be the primary means by which fungal organisms gain access to the sinonasal tract.

- Laboratory diagnosis of Fungal infections:

1. Direct Examination under Microscope (H/P):

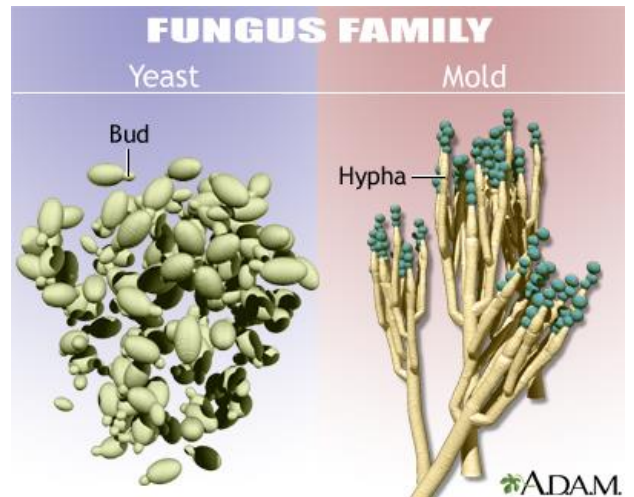
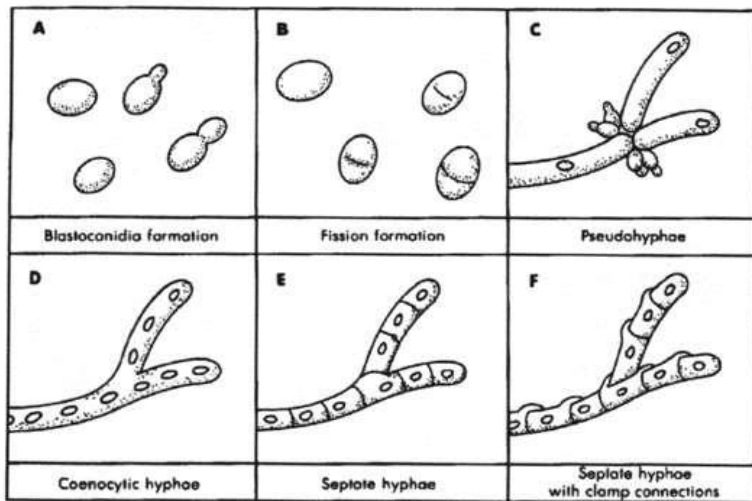
- Most rapid and useful means of diagnosing fungal infection.
- Provide diagnosis in less than an hour.
- Fungal organisms can be identified specifically because they are morphologically distinct.
- Gomori Methenamine Silver (GMS) stain is the best stain used to detect fungi by turning its color into black or dark brown.
 - Fungal elements are recognized for a unique ability to absorb silver.



2. Fungal Culture:

- Fungal cultures should be interpreted with caution.
- They are best used as supportive evidence because of their variable yield (60-100%)
- Allows the identification of the specific etiologic agent.
- Results are not available for days or weeks.
- Examples of fungal media is Sabouraud dextrose agar.

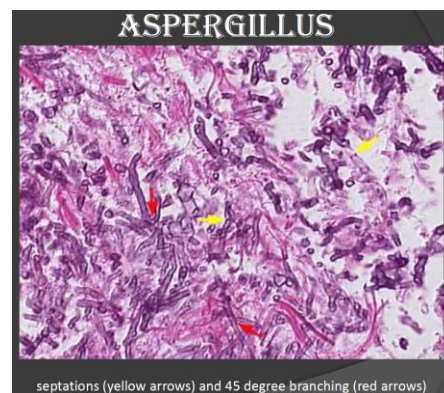




- Microscopic appearance of specific fungi:

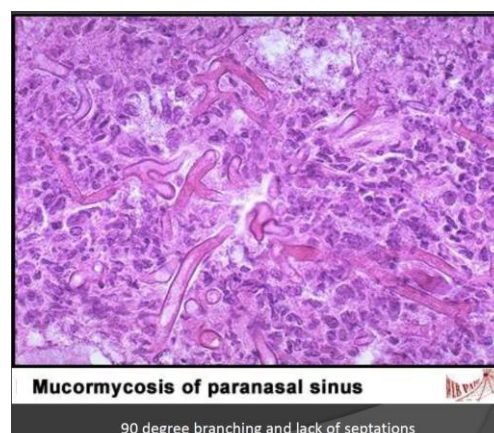
1. Aspergillus:

- Septated hyphae with acute angle branching at 45°



2. Mucromycosis:

- Non-septated hyphae with branching at 90°



- Most common fungi to involve paranasal sinuses:
 1. Aspergillus
 2. Alternaria
 3. Mucor
 4. Rhizopus.
- Classification of Fungal Rhinosinusitis:
 - **Non-Invasive:**
 1. Saprophytic Fungal Infestation.
 2. Sinus Fungal Ball.
 3. Eosinophilic Mucin Rhinosinusitis:
 - Allergic Fungal Rhinosinusitis.
 - Non-Allergic Eosinophilic Fungal Rhinosinusitis.
 - Non-Fungal Eosinophilic Mucin Rhinosinusitis.
 - **Invasive:**
 1. Acute Fulminant Invasive Fungal Rhinosinusitis.
 2. Chronic Invasive Fungal Rhinosinusitis.
 3. Granulomatous Invasive Fungal Rhinosinusitis.

TABLE 47-1. Illustration of How the Manifestation of Fungal Rhinosinusitis Depends on Host Immunologic Status

Host Defense	Immunodeficient		Immunocompetent		Allergic or Altered Cell-Mediated Hyperresponsiveness
Manifestation of fungal rhinosinusitis	Invasive, fulminant	Invasive, chronic	Invasive, granulomatous	Fungus ball	Allergic fungal rhinosinusitis Nonallergic eosinophilic fungal rhinosinusitis

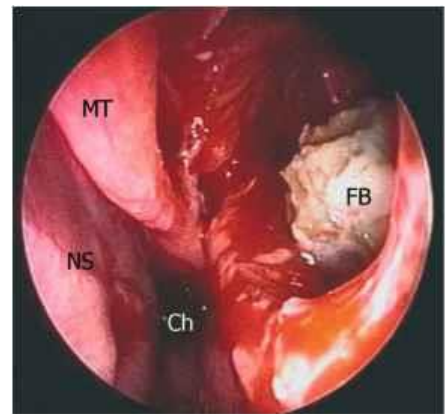
- **Saprophytic Fungal Infestation:**
- Non-invasive localized visible fungal colonization of sinonasal mucosa crusts in otherwise immunocompetent patients.
- Occurs when fungus grows on mucus crusts without the involvement or invasion to the surrounding mucosa.
- Clinical Picture:
 - Patients typically have minimal to no sinonasal symptoms.
 - Commonly found during postoperative endoscopic examinations or as a result of a foul odor from the nose.
- Diagnosis:
 - Made by direct examination with nasal endoscopy.
 - No need for imaging studies.
- Treatment:
 - Weekly endoscopic removal of the crusting with daily sinonasal rinses until the disease process resolves.
 - Antifungal medications are not needed.



- **Sinus Fungal Ball:**
- Non-invasive fungal infection into an otherwise healthy sinus in immunocompetent patients.
- Occurs due to sequestration of fungal elements within a sinus without invasion or granulomatous changes.
 - o Begins with inhalation of spores that become sequestered in a specific sinus only.
- Mycetoma is incorrect term in referring to paranasal fungus balls, because mycetoma refers to a fistulous fungal skin infection.
- Most common sinus involved is **Maxillary sinus** (80%) followed by sphenoid sinus (20%).
 - o Due to presence of favorable conditions for fungal growth such as warmth and humidity.
- Most common fungus responsible is **Aspergillus Species**.
- Bacterial infection may augment fungal growth via purulent secretions that provide valuable nutrient supplies to the fungi.
- Fungal ball is not associated with atopy.

- **Risk factors:**

1. Prior endodontic treatment of maxillary teeth.
 - Strong association.
2. Female predominance.
3. Absence of the disease process in children.
4. Bacterial infection.
 - Augment fungal growth via purulent secretions that provide valuable nutrient supplies to the fungi.



- **Clinical Picture:**

- o Results of mass effect by the fungal ball and sinus obstruction.
 - Unilateral proptosis, facial hypesthesia
- o Mimic chronic rhinosinusitis:
 - Maxillary disease causes facial pain, nasal airway obstruction, purulent rhinorrhea and cacosmia.
 - Sphenoid disease causes vertex headache and postnasal drip
 - Mild to no mucosal inflammation.
 - Polyps in 10% of patients.
- o Prolonged duration of CRS symptoms, unilateral symptoms, and poor response to medical therapy point to Fungal Ball.

- Diagnosis:

1. Non-contrasted CT Sinuses:

- Gold standard imaging study.
- Findings:
 1. Involves single sinus (90%) with maxillary sinus as the most common location (80%).
 2. Heterogeneous complete or subtotal opacification with microcalcifications.
 - Due to increased heavy metal content.
 3. Bony sclerosis and thickening can be seen.
 4. Bony erosion is not usually seen.
 5. Fungal ball is not enhanced in contrasted study.
 - Helpful in differentiating fungal ball from malignant processes associated with sinus opacification and bone erosion.

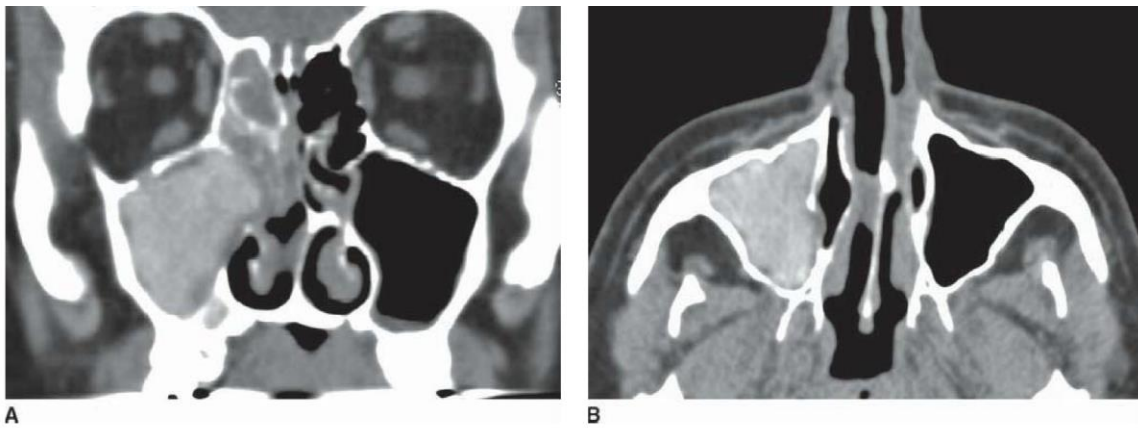


Figure 37.8 A,B: Fungus ball of the paranasal sinuses axial and coronal noncontrast CT scans show complete opacification of the right maxillary sinus, higher attenuation densities within the sinus, and thickening of the paranasal sinus mucosa consistent with chronic inflammation. Image and caption: Jeffrey Bennett, MD. Department of Radiology, University of Florida College of Medicine, Gainesville, FL.



Images show thickening of bony walls (short arrows) and heterodense material within the sinus with calcifications (long arrows)

2. MRI Sinuses:

- HYPO-intense in T1 and T2.
- Chronic inflammatory change of sinus mucosa may enhanced on post contrast T1.



3. Biopsy with Fungal Culture:

- Most common isolated fungus is **Aspergillus Fumigatus**
- Microscopic Appearance of Aspergillus:
 - Septated hyphae with acute angle branching at 45°
 - Absence of tissue invasion, granulomas, or allergic fungal mucin.
- Non-growth on fungal culture occur in 30% of specimens, even though hyphae are present on histopathology.
- Bacterial co-colonization with a wide variety of organisms occurs in 75% of fungus balls.

- Treatment:

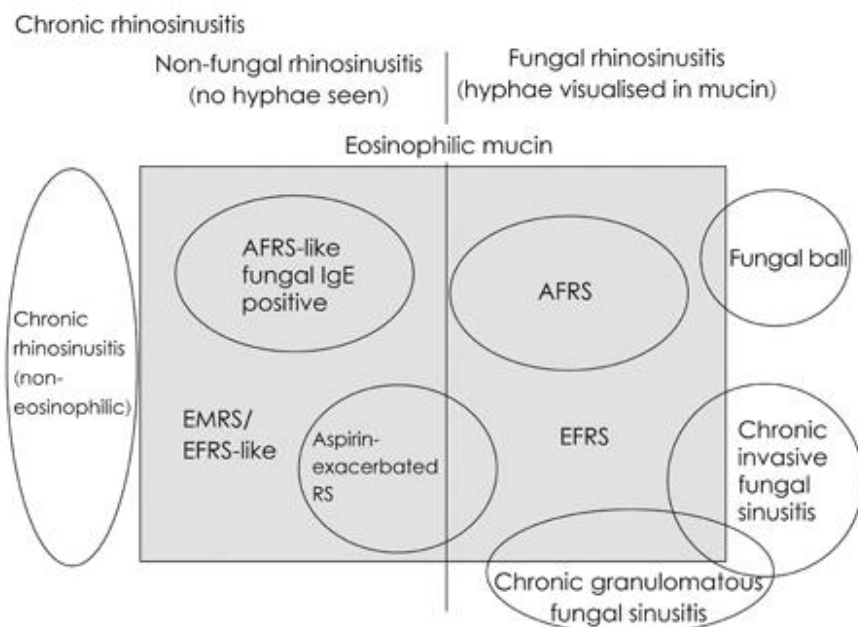
- Complete endoscopic surgical removal of fungal ball with thorough irrigation of the involved sinus.
- External approaches should be considered in only the most challenging cases.
- Antifungal medications:
 - Not needed in immunocompetent patients.
 - Indications start Antifungal medications:
 1. High risk patient for invasive disease (Severely immunocompromised).
 2. Continued recurrence of disease despite proper surgical and medical management.
 - Topical antifungals should be instituted first followed by the least toxic medications if they are required.

- **Eosinophilic Mucin Rhinosinusitis (EMRS):**
- **Includes:**
 1. Allergic Fungal Rhinosinusitis (AFRS).
 2. Eosinophilic Fungal Rhinosinusitis (EFRS).
 3. Eosinophilic Mucin Rhinosinusitis (EMRS).



- All have similar clinical picture and treatment strategies.
- All have associated Eosinophilic mucin present.
- **Key features to distinct between these types:**
 - o Presence/Absence of IgE to fungal antigen
 - o Presence/Absence of fungal hyphae based on microscopic examination (Not on fungal culture).

	Eosinophilic Mucin	Fungal Hyphae	IgE to Fungal Antigen
Allergic Fungal Rhinosinusitis (AFRS)	Present	Present	Present
Eosinophilic Fungal Rhinosinusitis (EFRS)	Present	Present	<u>Absent</u>
Eosinophilic Mucin Rhinosinusitis (EMRS)	Present	<u>Absent</u>	<u>Absent</u>



- **Allergic Fungal Rhinosinusitis (AFRS):**
- Fungal colonization of sinonasal cavity resulting in non-invasive allergic inflammation in otherwise immunocompetent patients.
- Endemic in areas of high humidity.
- **Pathophysiology:**
 - IgE mediated hypersensitivity response to fungal protein (Aspergillus).
 - Development of AFRS requires that an individual genetically predisposed to fungal allergy be exposed by inhalation of the mold spore.
 - Production of sticky allergic mucin resists clearance by normal mucociliary action and promotes growth of nasal polyps.
 - If the mold spore randomly becomes lodged in just one side of the nose, unilateral AFRS will develop.
- **Clinical Picture:**
 - Young immunocompetent patient.
 - Positive history of asthma or atopy (40%).
 - Unilateral or asymmetric symptoms of chronic rhinosinusitis with allergic component:
 - Nasal obstruction (gradual)
 - Nasal polyposis.
 - Rhinorrhea
 - Facial pressure/pain
 - Sneezing, watery/itchy eyes
 - Periorbital edema
 - Recurrent symptoms with history of multiple surgeries.
- **Diagnosis:**
 - **Bent and Kuhn Diagnostic Criteria of AFRS:**
 - Standard for diagnosis.
 - Patients must meet ALL Major criteria for diagnosis.
 - Minor criteria serve to support the diagnosis only.

Table 1. Bent and Kuhn Diagnostic Criteria	
Major	Minor
Type I hypersensitivity	Asthma
Nasal polyposis	Unilateral disease
Characteristic CT findings	Bone erosion
Eosinophilic mucin without invasion	Fungal cultures
Positive fungal stain	Charcot-Leyden crystals
	Serum eosinophilia
CT, computed tomography	

○ **Major Criteria (Diagnostic):**

1. Type I hypersensitivity:

- History.
- Positive skin test.
- Positive RAST and ELISA test.
- Elevated total serum IgE level (> 1000 IU/mL).

2. Nasal polyposis.

3. Characteristic CT findings:

- Unilateral or asymmetric involvement (80%)
- Involvement of ethmoid sinus.
- Heterogeneous opacification due to deposition of heavy metals (iron and manganese) and calcium salts within eosinophilic mucin.
- Remodeling and thinning of sinuses bony walls.
- Bony erosion in advance disease due to pressure, but not due to fungal invasion.

4. Presence of eosinophilic mucin containing non-invasive fungal hyphae:

- Confirm the diagnosis.
 - Most reliable indicator of AFRS (Pathognemonic).
 - Thick, tenacious and highly viscous
 - Tan to brown or dark green in appearance
 - Microscopic examination
 - Branching fungal hyphae
 - Sheets of eosinophils
 - Charcot-Leyden crystals (Products of eosinophilic breakdown of cells).
- 5. Positive fungal stain of sinus contents:**
- Gomori Methenamine Silver (GMS) stain.
 - No evidence of necrosis, giant cells, granulomas, or invasion into surrounding structures.

○ **Minor Criteria (Supportive):**

- 1.** History of Asthma
- 2.** Unilateral Predominance
- 3.** Radiographic Evidence of Bone erosion
- 4.** Fungal cultures
- 5.** Presence of Charcot-Leyden crystals in surgical specimens
- 6.** Serum Eosinophilia

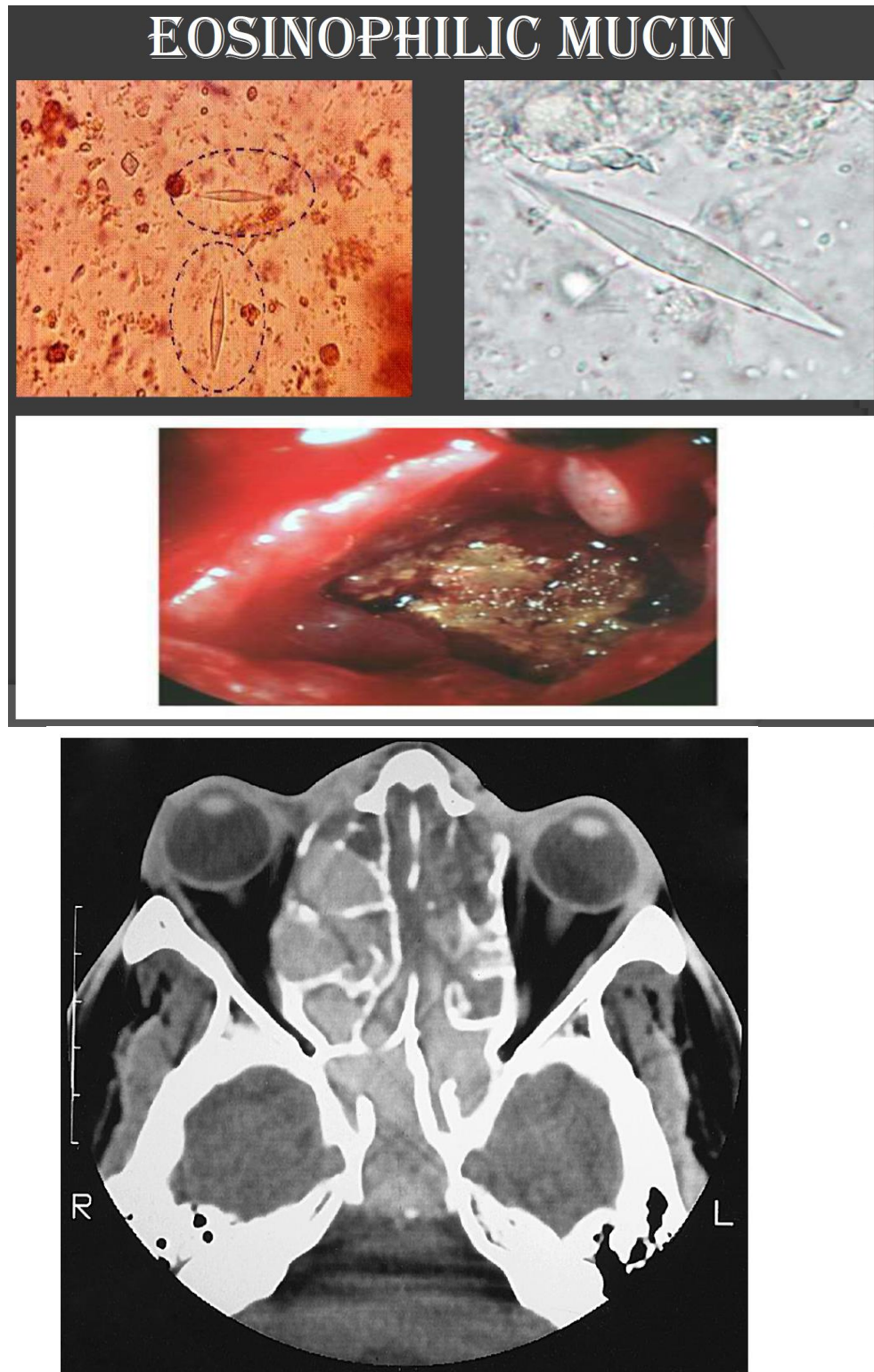


FIGURE 47-7. Axial computed tomography scan of a patient with allergic fungal rhinosinusitis. Note the heterogeneity of densities in the ethmoid and bony erosion of right lamina papyracea and the posterior wall of the sphenoid sinus.



**TABLE
37.3**

**RADIOGRAPHIC CHARACTERISTICS
OF AFRS**

CT Characteristics

1. Disease tends toward unilateral or asymmetric distribution
2. Involved sinuses are expanded
3. Bone bordering involved sinuses may demonstrate attenuation or erosion (best demonstrated on bone-weighted algorithms)
4. Adjacent anatomic spaces may be displaced
5. Signal heterogeneity within the involved sinuses (best demonstrated with soft tissue algorithm)

MRI Characteristics

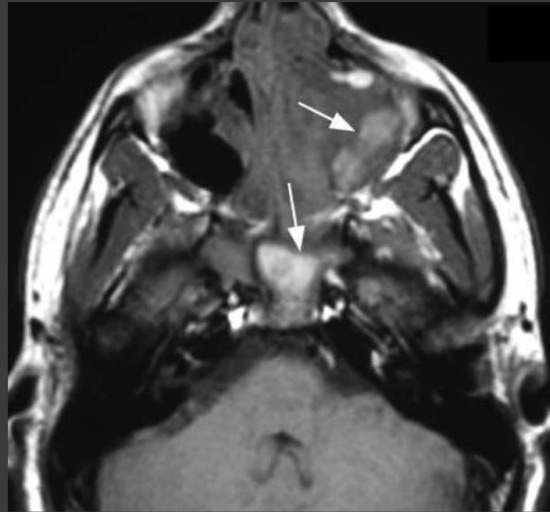
T1

1. Involved paranasal sinuses demonstrate variable signal intensity.
2. Enhancement of periphery of the involved paranasal sinuses (mucosal edema)

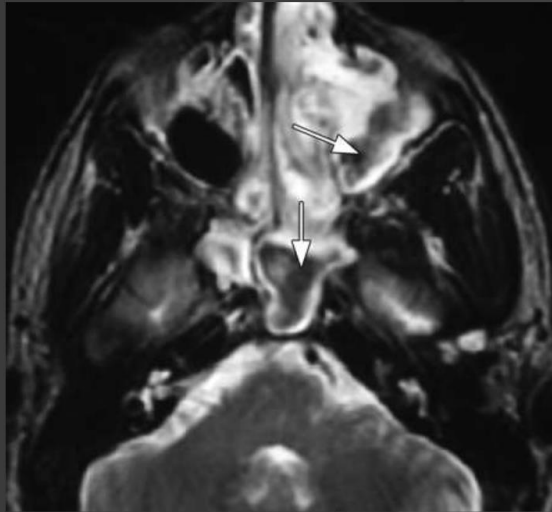
T2

1. Hypointensity of signal within involved paranasal sinuses (indication of dehydrated state of mucin)
2. Enhancement of periphery of the involved paranasal sinuses (mucosal edema)

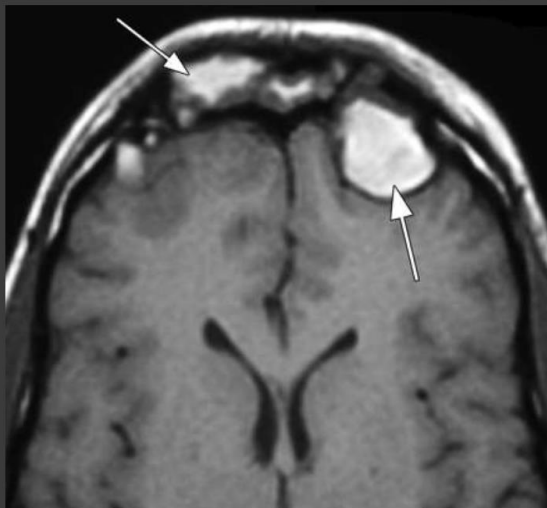
- Characteristic MRI findings in AFRS:
 - **T1:**
 - Central variable intensity.
 - Peripheral high intensity due to mucosal edema.
 - **T2:**
 - Central signal void due to high protein and low water concentration of eosinophilic mucin in conjunction with heavy-metal precipitate.
 - Peripheral high intensity due to mucosal edema.



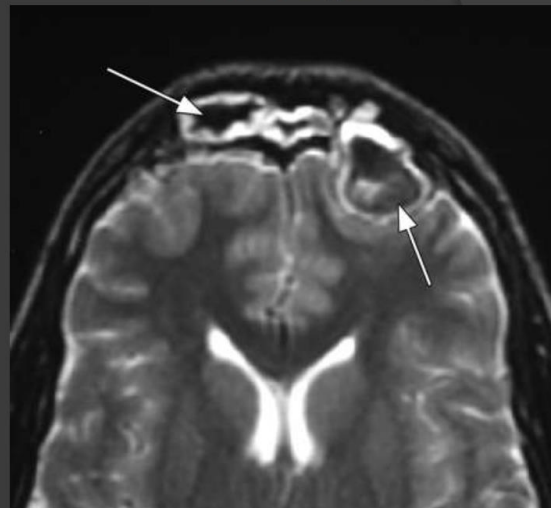
T1 MRI – high signal intensity of debris



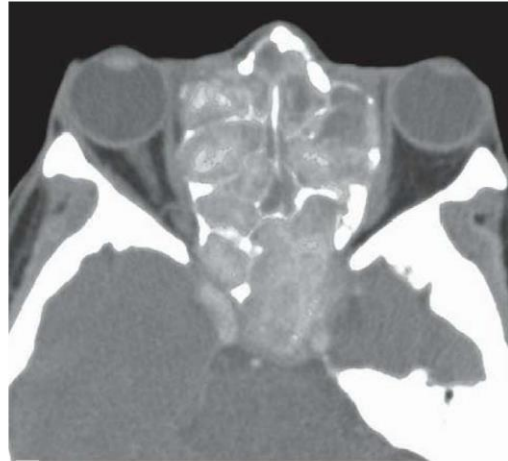
T2 MRI – central area of low intensity surrounded by high intense signal



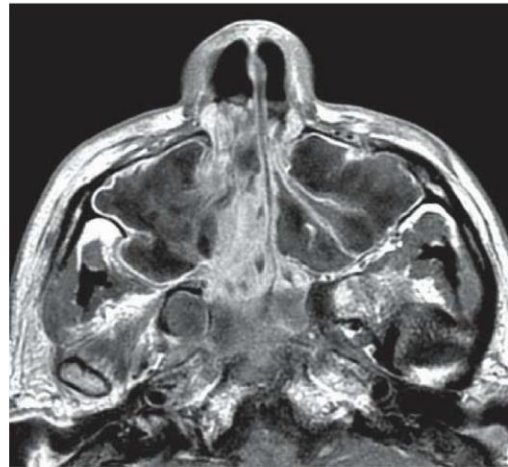
T1 MRI – high signal intensity of debris



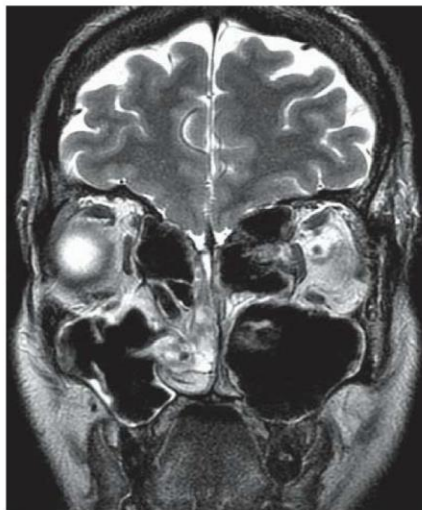
T2 MRI – central area of low intensity surrounded by high intense signal



A



B



C

Figure 37.10 A–C: AFRS Axial noncontrast CT scan shows pan-sinus opacification and expansion, with areas of increased attenuation corresponding to fungal colonization and chelated metals. Axial T1-weighted gadolinium-enhanced MRI shows peripheral mucosal enhancement and internal low signal. Coronal T2-weighted MRI demonstrates areas of signal void within the involved ethmoid sinuses. Image and caption: Jeffrey Bennett, MD. Department of Radiology, University of Florida College of Medicine, Gainesville, FL.

- Treatment of AFRS:

○ **Surgery:**

- Cornerstone of treatment.
- Complete mucosal-sparing endoscopic surgical debridement of involved sinuses
- Goals of surgery:
 1. Removal of all allergic mucin.
 2. Provide permanent drainage and ventilation of effected sinuses.
 3. Provide post-operative access to diseased areas.

○ **Medical:**

1. Topical treatment (steroid and saline irrigation):

- Mainstay of medical management.
- Saline irrigation helps to facilitate mucociliary clearance, loosen and dislodge secretions, and reduce mucosal exposure to allergens and other pro-inflammatory irritants.
- Daily use of topical intranasal steroids is safer than long-term oral corticosteroids.
- Topical intranasal steroids are often used with high doses delivered via saline irrigation.
 - 2 Ampules of Pulmicort (0.5mg/1ml) mixed with 500 ml NaCl to be used as 60ml BID within 2 days.

2. Systemic steroids:

- Pre-op steroids reduce bleeding and facilitate complete surgery.
- Post-op steroids decrease rate of recurrence (Prednisolone 35mg daily for one week then taper over one more week)
- Longer courses had better results, but more side effects.
- Brief courses of systemic steroids are used to treat exacerbations or polyp regrowth

3. Immunotherapy:

- Decrease recurrence.
- Alleviate need for steroids.

4. Other Medications:

- Insufficient data for Antifungal therapy, Leukotriene inhibitors or macrolide antibiotics in the medical management of AFRS.
- Each of these classes of agents has a theoretic rationale and may eventually prove worthwhile in diminishing the overall inflammatory state within the nose and sinuses.

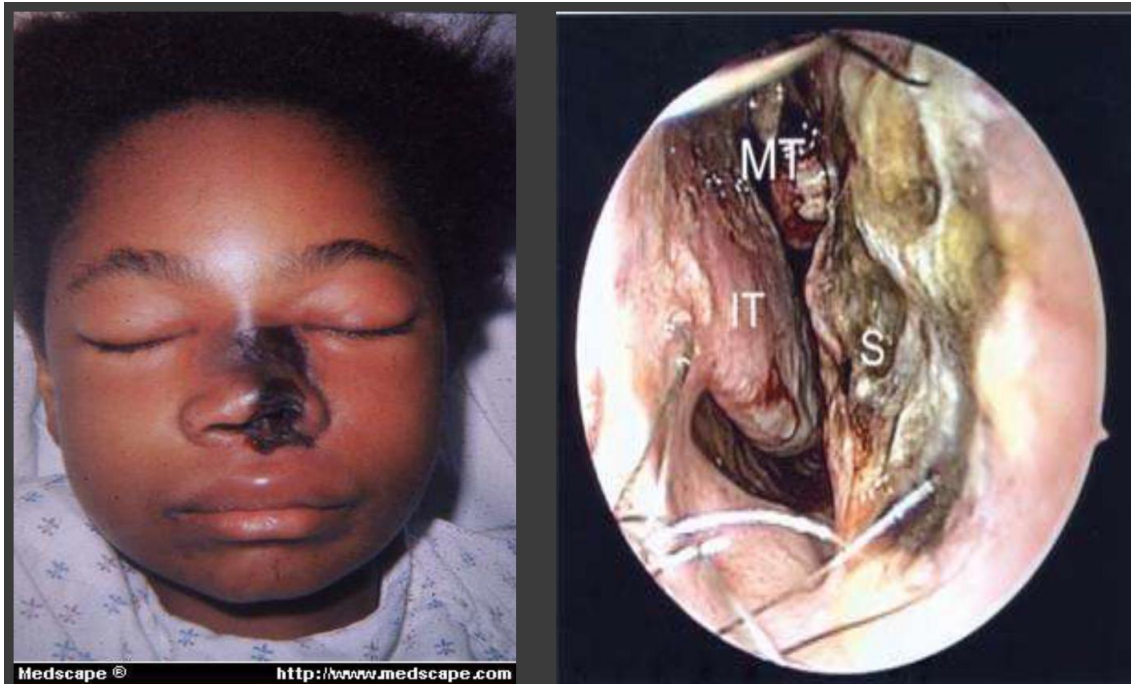
TABLE 47-6. Therapy of Allergic Fungal Rhinosinusitis

Definitely Helpful	Probably or Possibly Helpful	Not Helpful or Unclear
Steroids, systemic or topical Surgery Nasal lavage	Immunotherapy Oral antifungals* Anti-immunoglobulin E*	Topical amphotericin B Leukotriene modulators Calcineurin inhibitors Antibiotics

*Demonstrated efficacy in allergic bronchopulmonary aspergillosis.

- **Invasive Fungal Rhinosinusitis:**
- Defined by fungal elements invading host sinonasal tissue.
- Diagnosed by histopathologic evidence of fungi invading nasal tissue with hyphal forms within sinus mucosa, submucosa, blood vessel or bone.
- Specific histopathologic features, immunocompetence of the host, and disease progression allow further classification of invasive fungal rhinosinusitis disease into three forms:
 1. Acute fulminant invasive fungal rhinosinusitis.
 2. Chronic invasive fungal rhinosinusitis.
 3. Granulomatous invasive fungal rhinosinusitis.
- Accurate classification of these different disease entities is an important with regard to their clinical presentation, pathogenesis, diagnosis, and treatment.
- A time course of 4 weeks separates acute from chronic disease.
- Acute invasive and chronic invasive fungal rhinosinusitis typically occur in patients with some degree of immunocompromised.
- Granulomatous invasive fungal rhinosinusitis is limited to apparently immunocompetent patients.
- **Any immunocompromised patient with fever and one other sinonasal symptom should undergo evaluation for fungal sinusitis.**

- **Acute Fulminant Invasive Fungal Rhinosinusitis:**
- Incidence rates from 1.7% to 2.6%.
- Almost always seen in immunocompromised patients:
 - Insulin-dependent Diabetes and Diabetic ketoacidosis
 - AIDS
 - Organ transplantation
 - Systemic steroids
 - Chemotherapy
 - Hematologic malignancies.
 - Aplastic anemia
- Most common fungi:
 - Aspergillus (**Most Common**)
 - Mucormycosis (**Most Fulminant**)
 - 1.** Mucor
 - 2.** Rhizopus
 - 3.** Absidia
- Pathophysiology:
 - Fungus gain access to the sinonasal cavity typically by way of spores that are inhaled.
 - Grows and begins to invade neural and vascular structures.
 - Leads to thrombosis of vessels with resultant mucosal necrosis and loss of sensation.
 - Acidotic environment of tissue ischemia and necrosis provide an ideal medium for fungal growth and further propagation of the fungal infestation.
 - Extends beyond the affected sinus via bony destruction, perineural and perivascular spread.
 - 50% mortality with CNS or cavernous sinus involvement.
- Clinical Picture:
 - Symptoms are similar to acute bacterial rhinosinusitis.
 - Rhinorrhea. Headache, nasal congestion or facial pain.
 - Fever is the most frequent finding (90%).
 - Early findings are often subtle.
 - High index of suspicion for invasive disease should be maintained in immunocompromised patient with fever and symptoms of rhinosinusitis.
 - Extra-sinus extension:
 - Destruction and necrosis of nasal and palate mucosa.
 - Facial anesthesia
 - Ophthalmoplegia, Proptosis and Visual loss
 - Cranial nerve deficits
 - Mental status changes
 - Sepsis
 - Death



- Diagnosis:

1. High index of suspicion:

- Presence of fever with one additional symptom of sinonasal inflammation in immunocompromised patient should prompt both nasal endoscopy and imaging studies.

2. Nasal Endoscopy:

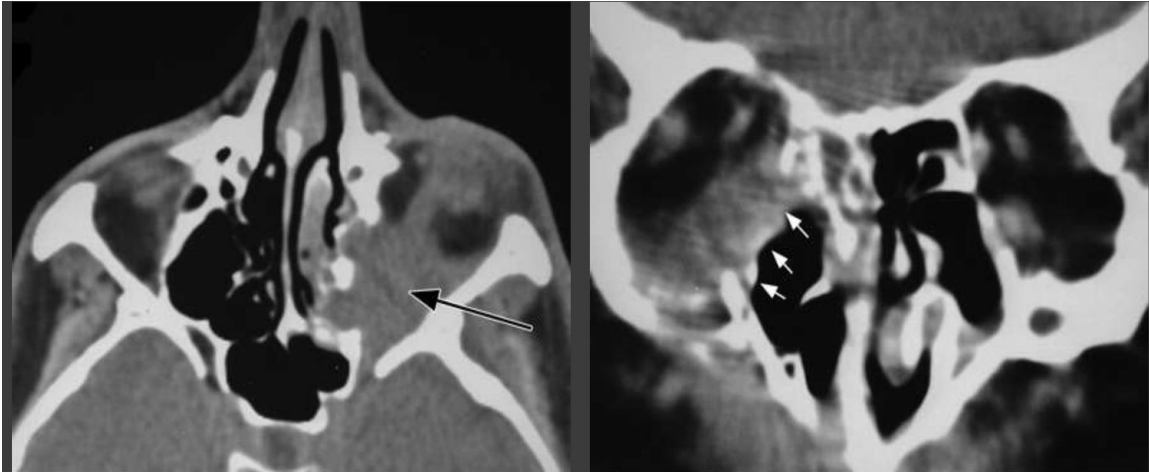
- Changes in appearance and anesthesia of sinonasal mucosa are the most consistent findings.
 - Pale mucosa in early stage.
 - Black necrotic mucosa in late stage.
- Serial examinations are required.

3. Biopsy and Culture:

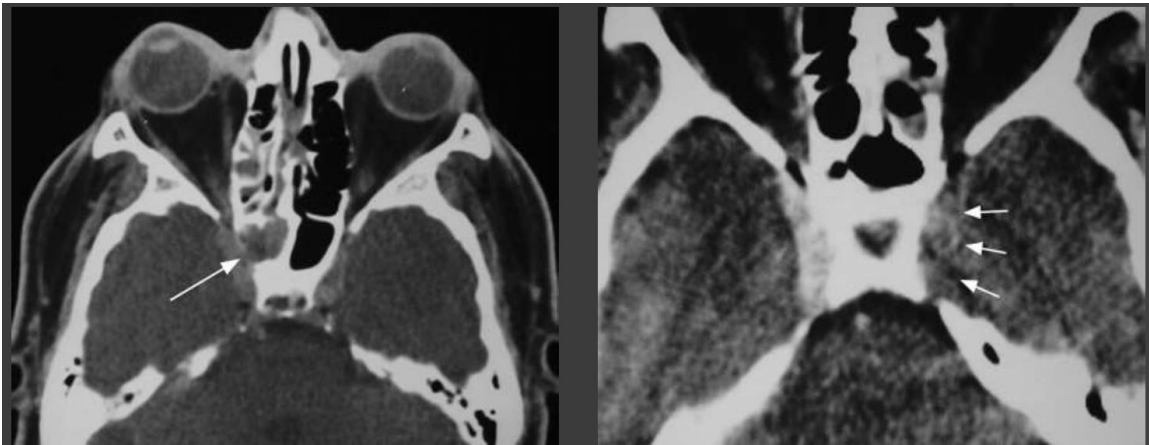
- Biopsy should be obtained whenever one suspects fungal disease or notes changes in mucosal appearance or sensation.
 - Should be taken from:
 1. Diseased mucosa (pale, insensate, ulcerative, black).
 2. Middle turbinate and nasal septum in normal appearance mucosa (Most common sites of involvement).
- Fungal culture is the gold standard for identifying the responsible fungi and guiding the choice of antifungal therapy.
 - Difficult to get positive culture result, especially with Mucormycosis.

4. CT Sinuses:

- Imaging studies are supportive, but not diagnostic of acute invasive fungal rhinosinusitis.
- Findings:
 - Bone erosion with extra-sinus extension (Mainly)
 - Severe, unilateral mucosal thickening.
 - Thickening of peri-antral fat planes.



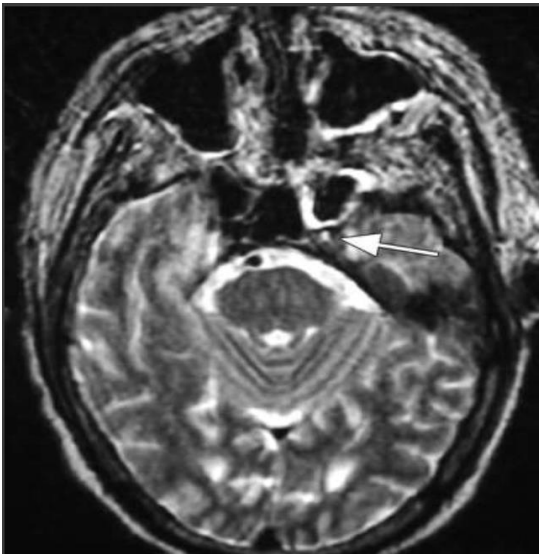
CT scans; **Left image:** Destruction of medial wall of orbit with extension of disease into the orbit. **Right image:** Destruction of medial and inferior walls of the orbit with extension of disease into the orbit



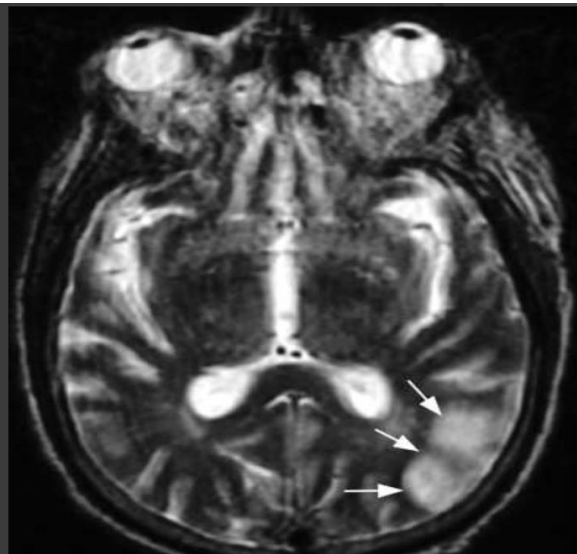
Axial CT scans. **Left image:** invasion through lateral wall of the sphenoid sinus and into the cavernous sinus. **Right image:** lack of enhancement of the cavernous sinus due to fungal thrombosis

5. MRI Sinuses:

- Imaging studies are supportive, but not diagnostic of acute invasive fungal rhinosinusitis.
- More sensitive than CT for detecting characteristic invasion into paranasal sinus tissue.
- Findings:
 - Obliteration of the periantral fat
 - Leptomeningeal enhancement (intracranial extension)
 - Granuloma formation
 - Hypointense on T1 and T2
 - Extrasinus extension
 - Cavernous sinus involvement
 - Absent flow void of carotid
 - Soft tissue thickening of the involved sinus



Axial MRI, T2 – left sphenoid sinus with central hypointense region with surrounding hyperintensity. Flow void in left cavernous sinus absent (arrow)



Axial MRI, T2 – Acute infarction of the left temporal lobe in same patient

- Treatment:

○ **Prevention:**

- High index of suspicion for disease and preventive measures constitute the first step in management in the at-risk population.
- Treatment is most successful when aggressive multimodality therapy can be instituted in the earliest stages of disease.

- Methods:

1. Decreasing environmental exposure to fungi:

- Hospital rooms equipped with high-efficiency particulate air filtration have been shown to reduce fungal burdens.

2. Hospital-based screening protocols for Invasive Fungal Sinusitis:

- CT scans sinuses in high-risk patients with sinonasal symptom or unexplained fever, followed by endoscopic examination and intranasal biopsy of all patients with radiographic abnormalities.

3. Prophylactic Antifungal drugs:

▪ Indications:

1. High-risk patients undergoing additional immunosuppressive therapy.
2. Patients with a known history of invasive fungal disease who will require further immunosuppressive therapy

▪ Medications:

1. Amphotericin B:

- Demonstrates a reduction in the rate of recurrent infection.

2. Posaconazole:

- Second-generation extended spectrum triazole.
- Superior to standard azole therapy in preventing invasive fungal infections in immunocompromised hosts.
- High-risk patients undergoing additional immunosuppressive therapy.

○ **Medical:**

1. Correction of underlying compromised state:

- Most important step in treatment.
- Control DM and treat DKA and underlying dehydration:
 - Associated with 80% improvement of survival.
- Restoration of Neutropenia:
 - Absolute Neutrophil Count (ANC) ≤ 1000 is associated with poor prognosis.
 - WBC transfusion and administration of Granulocyte colony stimulating factor is done to increase ANC > 1000 .

2. Systemic Anti-Fungal therapy:

- Instituted immediately while correcting of the underlying compromised state.
- Medications:

1. Amphotericin-B Infusion:

- Drug of choice for systemic treatment of invasive and disseminated fungal infections.
- 1mg/kg/day.
- Broad spectrum antifungal.
- Used mainly in Mucormycosis.
- Serious side effects limited its use in critically ill patients
 - **Myelosuppression**
 - **Ototoxicity**
 - **Nephrotoxicity (80%)**

2. Lipid-based Amphotericin B:

- More expensive.
- Less toxic.
- Can achieve higher concentrations of drug.

3. Extended Spectrum TriAzoles:

- Voriconazole, Itraconazole and Posaconazole.
- Used as treatment and prophylaxis of invasive fungal sinusitis in patients with immunocompromised state.
- Less toxic than Amphotericin B.
- Used for Aspergillus pathogens but resistant with Mucormycosis.

4. Echinocandins:

- Caspofungin, Micafungin, and Anidulafungin.
- Has fungicidal activity against *Candida* and fungistatic activity against.
- Potential role in the combination therapy of invasive fungal disease.

3. Topical Amphotericin B Nasal Rinses:

- Has minimal systemic risk and possibly of great benefit.
- Used as adjunctive measures in management of Acute invasive fungal sinusitis.

4. Hyperbaric Oxygen:

- Reduces ischemia and acidosis which are needed for fungus growth.
- Adjuvant therapeutic option.

○ **Surgical:**

- Less important step than intact immunity and appropriate medical therapy.
- Goals of surgery:
 1. Confirm the diagnosis through tissue biopsy.
 2. Debridement of devitalized tissue.
 3. Decrease pathogen load.
 4. Provide drainage and ventilation of effected sinuses.
 5. Provide post-operative access to diseased areas for monitoring.
- Principles of surgery:
 - Endoscopic debridement of devitalized tissue until clear healthy bleeding tissue is obtained which will improve the host defenses to clear the infection by:
 - Well-perfused tissue can deliver systemic antifungal medications.
 - Restoration of normal tissue pH.
 - Augment local neutrophil numbers.
 - Open surgical approach should be limited to disease cannot be removed endoscopically:
 - Advance orbital, facial, palatal, and intracranial fungal disease.
 - Prognosis of the patient should be considered when extensive resection is going to be required.
 - Once disease has gone intracranial, prognosis is very poor (70% mortality).

- **Prognosis:**
 - Overall mortality rate for is 20-80%
 - **Factors affecting mortality rate:**
 - 1. Time onset of treatment:**
 - Early treatment patients do much better than those who present with advanced disease.
 - 2. Absolute Neutrophil Count (ANC):**
 - ANC < 1000/mm³ is associated with a worse prognosis.
 - Recovery from neutropenia is the most predictive indicator for survival.
 - 3. Intra-cranial involvement:**
 - Single most predictive indicator for mortality.
 - 70% mortality rate.
 - 4. Type of Fungus:**
 - Mucor infection tends to worse than Aspergillus as Mucor tends to be more aggressive with earlier orbital and intracranial involvement.
 - 5. Type of immunocompromised state:**
 - Diabetics do worse than those of other immunocompromised states because mucormycosis is more often seen in diabetics.

TABLE 47-2. Various Fungal Agents and Antifungal Drugs of Choice

Antifungal	Dosage	Activity	Comments
IV amphotericin B Lipid and nonlipid	0.25 up to 1.0 mg/kg qd IV in 5% dextrose Dose varies with lipid formulation	Therapy is active against most pathogenic fungi but is variable against some <i>Aspergillus</i> species and Zygomycetes <i>Pseudallescheria boydii</i> and <i>Fusarium</i> species are resistant	Nephrotoxicity is reduced with more expensive lipid formulations
Topical amphotericin B	100 µg/mL, 20 mL bid	As above	Not absorbed via oral route; efficacy is unproven in noninvasive fungal rhinosinusitis
Ketoconazole	400 mg PO qd	<i>Candida</i> species and <i>Pseudallescheria boydii</i>	Hepatotoxic; monitor liver function
Itraconazole	100 mg 1 to 2 PO bid with meals	Dermatiaceous fungi and variable activity against <i>Aspergillus</i> species Not active against Zygomycetes	Best absorbed in acidic stomach; coadminister with cola or cranberry juice; if used long term, monitor liver function
Posaconazole	Oral suspension of 40 mg/mL; dosage is 400 mg bid	Indicated for the treatment of a broad range of invasive fungal infections including Zygomycetes and <i>Fusarium</i> and <i>Aspergillus</i> spp. in patients refractory or intolerant of other antifungal therapies	Safety under age 13 not assessed; use with caution with other drugs metabolized through the CYP3A4 system
Terbinafine	250 mg PO qd	Variable in vivo antifungal activity; utilized chiefly for dermatophytosis	Not effective in a randomized controlled trial of noninvasive CRS
Voriconazole	200 mg PO bid 1 hr before or following meals; IV formulation given as loading dose followed by 4 mg/kg bid	Indicated for treatment of a broad range of fungal pathogens	Monitor liver functions; an unusual adverse effect is visual disturbance

bid, twice daily; CRS, chronic rhinosinusitis; IV, intravenous; PO, by mouth; qd, daily.

- **Chronic Invasive Fungal Rhinosinusitis:**
- Rare condition in which invasive fungal sinusitis progresses gradually over time (months to years).
- Has similar clinical appearance of acute fulminant invasive fungal sinusitis.
- Occurs mainly in immunocompetent and mild immunocompromised patients (steroid treatment, diabetes mellitus, HIV).
- Most common fungi:
 - o Aspergillus Fumigatus (Most common >80%)
 - o Bipolaris
 - o Candida
 - o Mucormycosis
- Clinical Picture:
 - o Nonspecific chronic rhinosinusitis (CRS) symptoms:
 - Nasal congestion, rhinorrhea, facial pressure, headaches, polyposis.
 - Long duration, slow progression, refractory to standard antibiotic therapy and worsen with steroids.
 - o Unilateral blood streaks within nasal drainage is the most common symptom.
 - o Ocular symptoms are indication of extent and aggressiveness of the disease.
- Diagnosis
 - 1. High index of suspicion:**
 - If patient presented with CRS that is unresponsive to antibiotics or presented with epistaxis, facial anesthesia, visual changes, altered mental status and seizures should all increase suspicion for fungal infection.
 - 2. Nasal Endoscopy:**
 - Nasal endoscopic findings are very subtle and include:
 - Mucosal edema, crusting, and the presence of polyps.
 - Rarely will ulceration and eschar formation.
 - 3. Biopsy and Culture:**
 - Required for the diagnosis.
 - Identification of submucosal invasion of fungal elements.
 - Few if any inflammatory cells:
 - o Major difference between acute and chronic invasive disease
 - No Granuloma formation:
 - o Main difference between chronic invasive and granulomatous invasive fungal disease.

4. CT Sinuses:

- No pathognomonic imaging findings.
- Findings:
 - Unilateral paranasal sinus involvement.
 - Homogenous opacification
 - Bone erosion limited to sites of extrasinus extension

5. MRI Sinuses:

- MRI findings are very similar to that found with Acute fulminant invasive sinusitis except there will be no granuloma formation.

- Treatment:

- Similar to Acute fulminant invasive sinusitis with a combination of surgical and medical treatments.
- Cure rates of nearly 90%.

○ **Anti-Fungal drugs:**

- Systemic and topical Amphotericin B should be started until cultures prove that the offending agent is not a Mucor species.
- If not a Mucor species, Voriconazole, Itraconazole and Posaconazole is used to limit the side effects.
- Most recommend duration of therapy is 3-6 months.

○ **Surgical debridement:**

- Should include all involved tissues until clear, bleeding margins are obtained.

○ **Close Follow-up visits:**

- Patients should be followed closely with examination and debridement.
- Biopsies should be taken from any suspicious areas as asymptomatic recurrence is common.

- **Granulomatous Invasive Fungal Rhinosinusitis:**

- Similar clinical picture, work-up and treatment to chronic invasive fungal sinusitis.

- Main differences:

- Caused by Aspergillus Flavus.
- Almost exclusively found in North Africa and Southeast Asia.
- Presence of multinucleated giant cell granulomas on microscopic examination.



Figure 37.4 A,B: GFR Axial and Coronal CT scans show expansion of the involved right frontal sinus with long-standing invasive fungal infection. Bone of the skull base has become dehiscent and there is direct extension of GFR into the brain. Image and caption: Jeffrey Bennett, MD. Department of Radiology, University of Florida College of Medicine, Gainesville, FL.

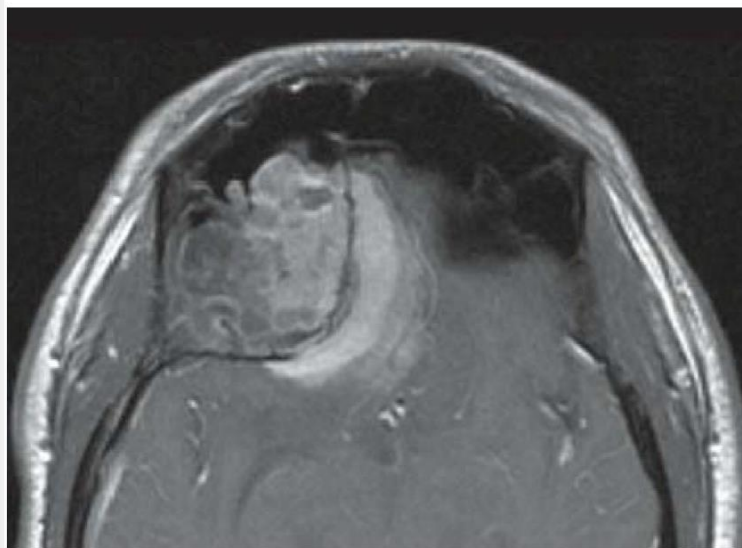


Figure 37.5 GFR Axial postcontrast T1-weighted MRI of the same patient shown in Figure 37.4 demonstrates enhancement within the granulomatous mass and the adjacent involved brain. Image and caption: Jeffrey Bennett, MD. Department of Radiology, University of Florida College of Medicine, Gainesville, FL.

**TABLE
37.2****FEATURES OF INVASIVE FUNGAL RHINOSINUSITIS**

Syndrome	Histopathology	Immunocompetence
Granulomatous	– Invasive fungal hyphae are sparse and found within noncaseating granulomas	– Immunocompetent
Invasive	– Dense fibrosis and mild inflammatory cell infiltrate in surrounding tissue	– Diabetic cases reported rarely
Fungal Rhinosinusitis	– <i>A. flavus</i> is most common	
Chronic Invasive	– Invasive fungal hyphae and necrotic tissue with mixed acute and chronic inflammatory cellular response.	– Diabetes Mellitus
Fungal Rhinosinusitis	– Matted hyphae can resemble sinus FB	
	– <i>A. fumigatus</i> , many others	
AIFR	– Invasive fungal hyphae with prominent angioinvasion, thrombosed vessels, and necrotic tissue.	– Immunocompromised as a result of hematologic malignancies, therapeutic immunosuppression, diabetes mellitus
	– Moderate inflammatory cell infiltrate (predominantly neutrophils)	
	– <i>Aspergillus</i> and Family Mucoraceae are common	– Immunocompetent cases reported rarely

Modified from Chakrabarti A, Das A, Panda NK. Controversies surrounding the categorization of fungal sinusitis. *Med Mycol* 2009;47:S299–S308; Das A, Bal A. Chakrabarti A, et al. Spectrum of fungal rhinosinusitis: histopathologist's perspective. *Histopathology* 2009;54:854–859; DeShazo RD. Syndromes of invasive fungal sinusitis. *Med Mycol* 2009;47:S309–S314.

Complications of Rhinosinusitis:

- Rhinosinusitis complications arise when infection spreads into or beyond bony wall of sinuses.
- Results in devastating consequences if they are not promptly recognized and treated.
- Rare as most episodes of ABRS respond to appropriate medical treatment.
- **Classification of rhinosinusitis complications:**
 1. Orbital Complications **(60-75%)**
 2. Intracranial Complications **(15-20%)**
 3. Bony Complications **(5-10%)**
- **Location and timing of rhinosinusitis complications are directly relate to:**
 1. Development of the paranasal sinuses:
 - Development of ethmoid and maxillary sinuses starts in utero.
 - Maxillary sinuses have a rapid phase of growth until age 4 years and continue to expand through puberty.
 - Ethmoid sinuses are the most developed sinuses at birth and gradually increase in volume through puberty.
 - Contributes to the higher rate of orbital complications in younger children.
 - Frontal sinuses first become visible radiographically around age 5 years and increase in size through puberty.
 - Contributes to the higher rate of intracranial complications in adolescents.
 - Sphenoid sinuses are the last to develop at around age 6 years, but are rarely involved in any complications of acute rhinosinusitis.
 2. Proximity of paranasal sinuses to the orbit and brain:
 - Orbital complication occurs mainly after sinusitis involving the ethmoid sinuses.
 - Intracranial complication occurs mainly after sinusitis involving the frontal sinuses.

- **Risk Factors of developing rhinosinusitis complications:**

1. Winter months:

- From January through March.
- Higher rates of upper respiratory tract infections.

2. Age:

- Pediatric populations below 7 years:
 - Most common populations involved in orbital complications due to:
 - Higher rates of upper respiratory tract infections.
 - Ethmoid sinuses are the most developed sinuses at the time of birth which is in close proximity to the orbit.
- Adolescence and young adulthood:
 - Most common populations involved in intracranial complications due to:
 - Frontal sinuses first become visible radiographically around age 5 years and increase in size through puberty which is in close proximity to the brain.

3. Gender:

- Male gender in intracranial and post-septal orbital complications.
- Gender has not been identified as a risk factor in pre-septal orbital complications.

- **Diagnostic tools of rhinosinusitis complications :**

- **Blood tests:**
 - For base line analysis:
 - CBC, ESR, CRP.
 - Leukocytosis was seen in 50% only of patients with orbital complications.
- **Blood culture:**
 - Indications:
 - Severe orbital complications
 - Intracranial complications
 - Overall positive blood cultures overall 2.7–6%.
- **Nasal Endoscopy and Middle meatus culture:**
 - Should be done for all patient presenting with rhinosinusitis complications.
 - Endoscopically-directed middle meatus cultures should be obtained with a sterile swab or utilizing suction into a sterile trap device.
 - Endoscopically-directed middle meatus cultures are well correlated with cultures obtained by direct maxillary sinus aspiration.
 - Helps to direct the antibiotic coverage to specific microorganism.

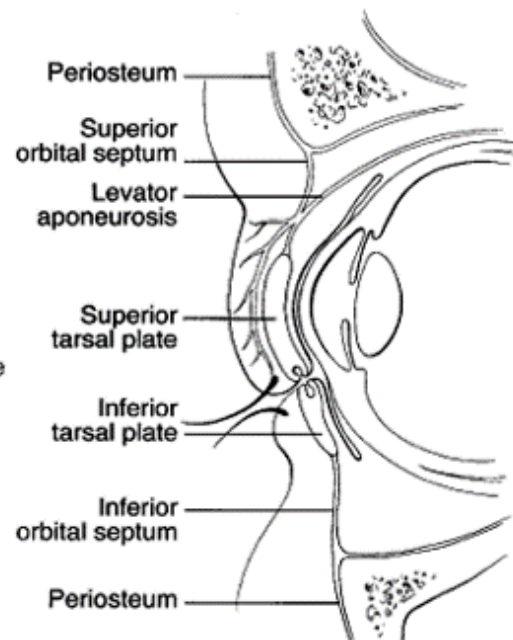
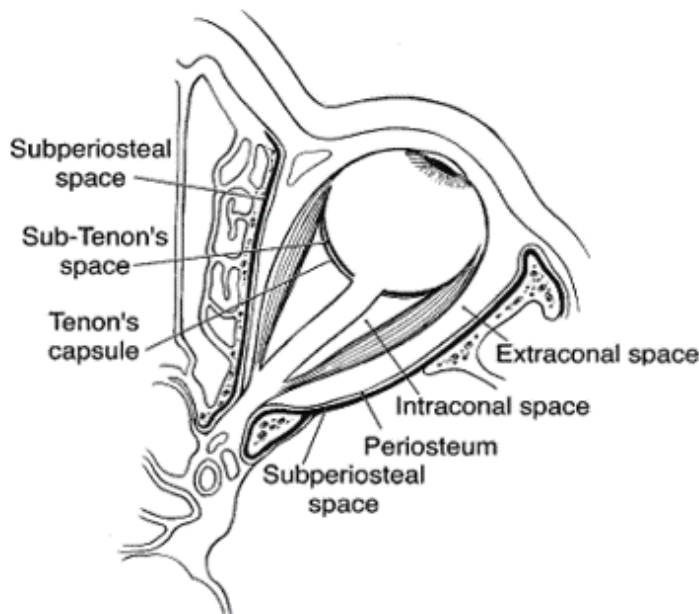
- **CT scan with contrast:**
 - Indicated for all patients with post-septal orbital complications and intracranial complications.
- **MRI with gad:**
 - Indicated mainly for all patients with intracranial complications.
- Microbiology of rhinosinusitis complications:
 - Orbital complications are polymicrobial (**75%**)
 - Most common pathogens are:
 - *Strep. viridians* (including *Strep. Milleri*)
 - *Staph. aureus*
 - Immunocompromised patients are susceptible to atypical pathogens and fungus with development of invasive infections

**TABLE
38.2**
MICROBIOLOGY OF SINUSITIS AND RELATED COMPLICATIONS

Acute sinusitis (children)	More common	<i>Streptococcus pneumoniae</i> , <i>Haemophilus influenzae</i> , <i>Moraxella catarrhalis</i> , and other <i>Streptococcus</i> species
	Less common	Anaerobic organisms and <i>Staphylococcus aureus</i>
Acute sinusitis (adults)	More common	<i>S. pneumoniae</i> , <i>H. influenzae</i> , other <i>Streptococcus</i> sp., and anaerobic organisms
	Less common	<i>M. catarrhalis</i> and <i>S. aureus</i>
Orbital complications of sinusitis	More common	<i>Streptococcus viridans</i> (including <i>Streptococcus milleri</i> group), <i>S. aureus</i> , and <i>S. pneumoniae</i> ; these infections are commonly polymicrobial
	Less common	Anaerobic organisms (especially <i>Bacteroides</i> sp., <i>Peptostreptococcus</i> , and <i>Eikenella</i>), other <i>Streptococcus</i> sp., <i>H. influenzae</i> , and other gram negative bacilli
Intracranial abscesses (intracerebral, epidural, subdural abscesses)	More common	<i>S. viridans</i> (including <i>S. milleri</i> group), <i>S. aureus</i> , other <i>Streptococcus</i> sp., other anaerobic organisms (<i>Peptostreptococcus</i> , <i>Bacteroides</i> sp., others), and <i>S. pneumoniae</i> ; commonly polymicrobial infections
	Less common	Coagulase-negative <i>Staphylococcus</i> and gram negative bacilli (<i>H. influenzae</i> , others)
Meningitis	More common	<i>S. pneumoniae</i> , <i>S. aureus</i> , and other <i>Streptococcus</i> sp.
	Less common	Anaerobic organisms (<i>Fusobacterium</i> sp., others), gram negative bacilli, and <i>H. influenzae</i>
Pott's puffy tumor (acute osteomyelitis)	More common	<i>S. viridans</i> (including <i>S. milleri</i> group) and <i>S. aureus</i> ; commonly polymicrobial infections
	Less common	Anaerobic organisms (<i>Bacteroides</i> sp., etc.) and, much less commonly, gram negative bacilli (<i>Proteus</i> sp., etc.)

Note: This compilation represents the observed commonalities and average incidence of groups of organisms identified in series published over the last 10 y; the exact predominance of each organism in the listed disease processes varies among different published series.

- **Orbital Complications: (60-75%):**
 - Most common rhinosinusitis complications in pediatrics and adults.
 - Most common in children below 7 years.
 - Orbital complications are still considered a serious threat to essential functions of the eye, including loss of vision, and at worst, life threatening symptoms.
 - Most common origin of orbital complication is ethmoiditis.
 - Followed by frontal and maxillary sinusitis.
- Main anatomical barriers of spreading the infections into the orbit:
 - 1. Lamina Papyracea:**
 - Paper-thin bony plate separate ethmoid cells from orbit.
 - Contains several perforations through which valveless blood vessels and nerves travel.
 - 2. Periorbita:**
 - Periosteum of the internal orbit and covers the bony orbital walls from the anterior aperture of the orbital cavity back to the cone enveloping the optic canal.
 - Only soft tissue barrier between the sinuses and the orbital contents.
 - 3. Orbital septum:**
 - Reflection of the periorbita at the margins of the orbit and attaches into the upper and lower eyelids at the levator aponeurosis superiorly and the tarsal plate inferiorly.
 - Lacks lymphatic channels and forms a barrier limiting infections from passing directly through eyelids into orbit.
 - Forms a boundary between pre and post-septal infections.



- Important anatomical spaces related to the orbit:

1. Sub-periosteal space:

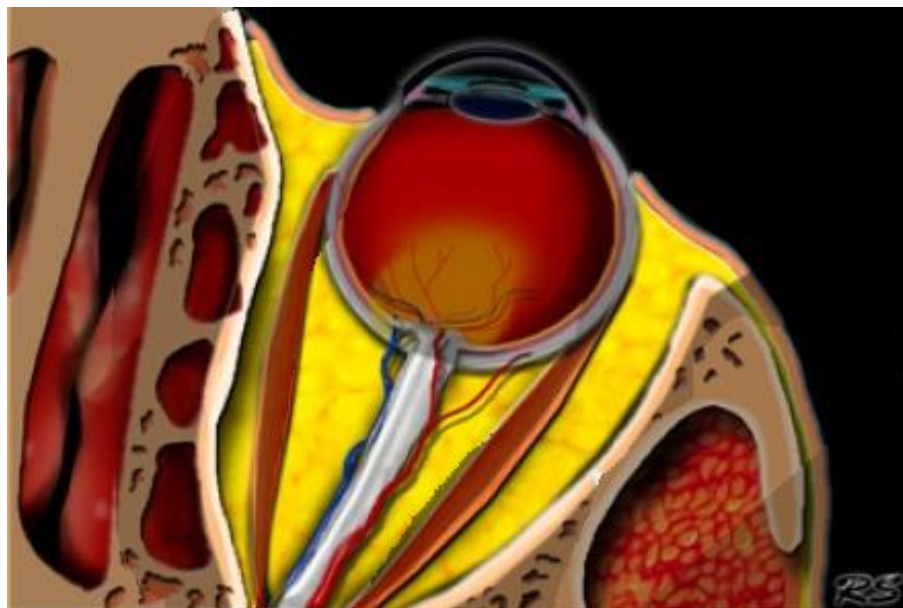
- Potential space lying between the orbital bones and the periorbita.
- Can be fill with blood after orbital fracture or with abscesses when associated with a paranasal sinusitis.

2. Extra-conal space:

- Potential space lying within the periorbital but outside the intermuscular septum.
- Contains:
 - Extra-conal fat.
 - Superior oblique muscle and trochlea
 - Trochlear nerve (CN IV)
 - Lacrimal gland
 - Lacrimal and frontal branches of Ophthalmic nerve (V1)

3. Intra-conal (Retro-bulbar) space:

- Potential space lying within the periorbital but inside the intermuscular septum.
- Contains:
 - Intra-conal fat
 - Optic nerve (CN II)
 - Oculomotor nerve (CN III)
 - Abducens nerve (CN VI)
 - Nasociliary branch of Ophthalmic nerve (V1)
 - Ophthalmic artery



- Pathways of orbital involvements:

1. Thrombophlebitis:

- Main pathway of orbital complications.
- Superior and inferior ophthalmic veins are valveless, allowing direct communication between the nose, ethmoid sinuses, face, orbit, and cavernous sinus.
- Thrombophlebitis interferes with the venous drainage of the orbital contents.

2. Direct extension:

- Ethmoid sinusitis can extend through the thin lamina papyracea or through congenital, surgical, or traumatic dehiscence of lamina papyracea.

- Classification of orbital complications:

- Classified primarily by Hubert in 1937.
- Chandler categorized them in 1970, followed by later modifications by Moloney.

Table 1. The most commonly accepted orbital classifications of sinuses

Group	Chandler	Moloney
First	Inflammatory oedema	Preseptal cellulitis
Second	Orbital cellulitis	Subperiosteal abscess
Third	Subperiosteal abscess	Orbital cellulitis
Fourth	Orbital abscess	Orbital abscess
Fifth	Cavernous sinus thrombosis	Cavernous sinus thrombosis

[Int Forum Allergy Rhinol.](#) 2012 Jul-Aug;2(4):331-5. doi: 10.1002/alr.21029. Epub 2012 Mar 12.

Ophthalmic manifestations of paranasal sinus disease: a clinical grading system.

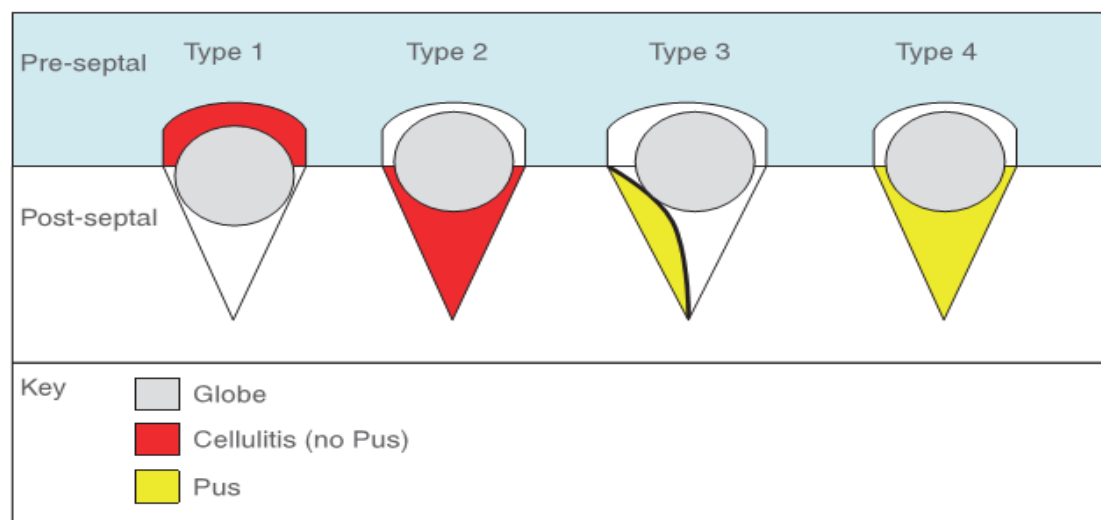
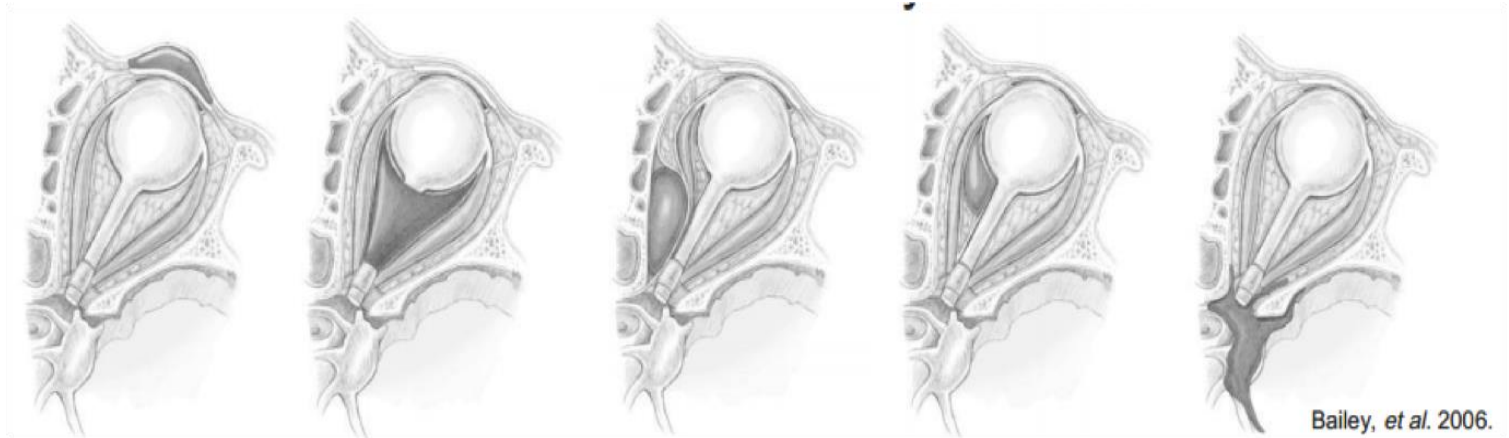
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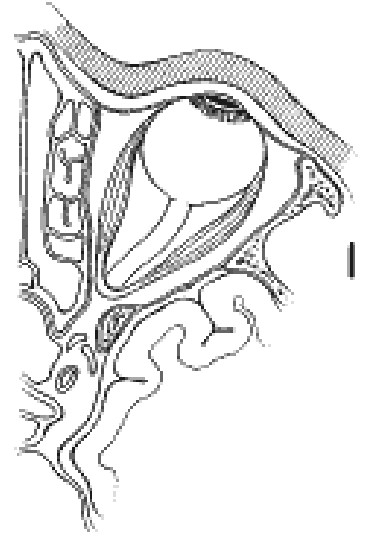
- Chandler classification of orbital complications:

- Most widely utilized classification in clinical practice.
- Each of these complications should be considered separately and treatment individualized.
- Multiple complications can occur in the same patient.
- Five groups of rhinosinusitis-related orbital complications:
 1. Pre-septal cellulitis
 2. Orbital cellulitis,
 3. Sub-periosteal abscess
 4. Orbital abscess
 5. Cavernous sinus thrombosis

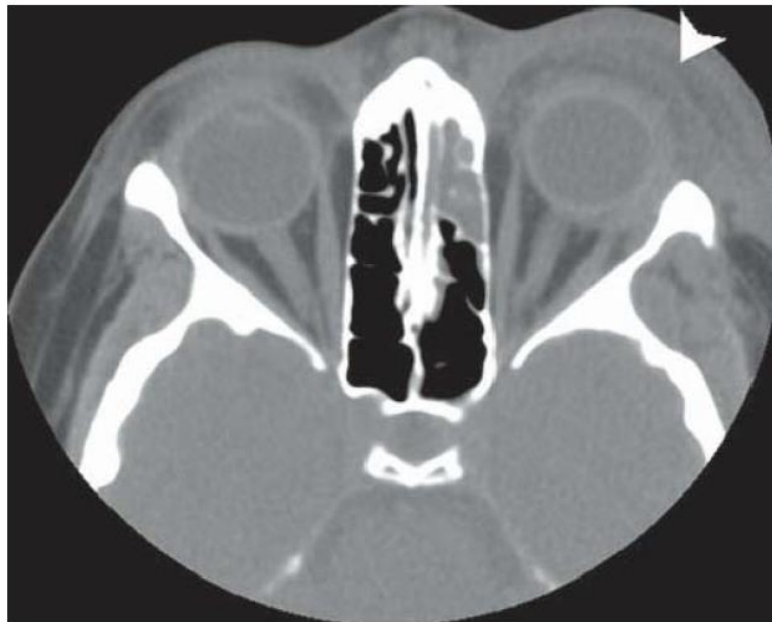


I. Peri-orbital (Pre-septal) Cellulitis:

- Infection limited to skin and subcutaneous tissues of the eyelid anterior to the orbital septum.
- Inflammatory edema cause the eyelids to swell.
- Most common form of all orbital complications of rhinosinusitis **(70%)**.
- Least severe complication.
- Caused by impaired venous drainage of the ethmoidal vessels that are obstructed by inflammation and pressure.
- Clinical Presentation:
 - Unilateral eyelid edema, erythema and tenderness.
 - Small eyelid abscess may encountered.
 - Fever.
 - No involvement of post-septal structures:
 - No impairment of visual acuity
 - No chemosis
 - No proptosis
 - No restriction of extraocular muscle movement.



- Diagnosis:
 - **CT scan with contrast:**
 - Usually unnecessary in patients with pre-septal infections.
 - Indications:
 1. Failure of improvement or worsening of symptoms within 24-48 hours despite medical therapy.
 2. Presence of signs and symptoms of post-septal infection.
 3. Suspicious of intracranial complications.
 - Findings:
 - Diffuse thickening of the eyelids.
 - Abscess formation may be seen.



A

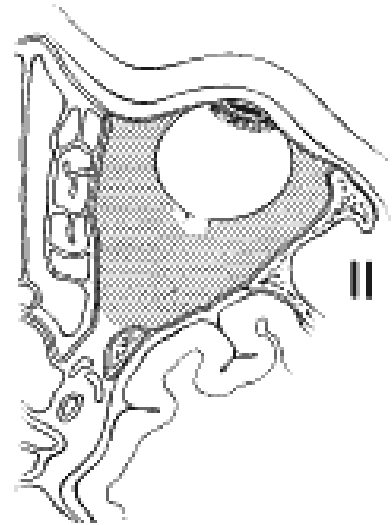


B

Figure 38.2 Preseptal abscess secondary to preseptal cellulitis. **A:** Axial CT scan taken on admission showing the patient with left ethmoid sinusitis and left preseptal cellulitis (*arrow*); patient also had frontal sinusitis (*not pictured*). **B:** Axial CT scan after 4 days of IV antibiotic treatment showing progression of infection to left preseptal abscess (*arrow*). The patient also developed an orbital subperiosteal abscess, scalp abscess, and small epidural abscess.

II. **Orbital Cellulitis:**

- Infection occurs within the orbit proper posterior to the orbital septum.
- Orbital contents show diffuse edema with inflammatory cells and fluid, without distinct abscess formation.
- Swelling of orbital contents occurs when an increase in sinus venous pressure is transmitted to the orbital vasculature, resulting in transudation and leakage through the vessel walls.
- Orbital cellulitis is more concerning than pre-septal cellulitis because it can evolve into an orbital abscess.
- More severe infections such as invasive fungal sinusitis should be considered in diabetic patients with ketoacidosis and immunocompromised patients.
- Clinical Presentation:
 - Unilateral eyelid edema, erythema and tenderness.
 - Fever.
 - Signs of involvement of post-septal structures:
 - Chemosis
 - Proptosis
 - Pain
 - Ophthalmoplegia might present due to impaired extraocular muscle movement.
 - **NO Impairment of visual acuity**



- Diagnosis:
 - **CT scan with contrast:**
 - Indicated in all patients with post-septal infection.
 - Findings:
 - Diffuse thickening of the eyelids.
 - Enhancement of edematous extra-conal fat maximally adjacent to the most severely affected sinus.
 - Enlargement and enhancement of the adjacent rectus muscle.
 - Intra-conal fat may be radiographically normal.
 - Discrete abscess should not be encountered.

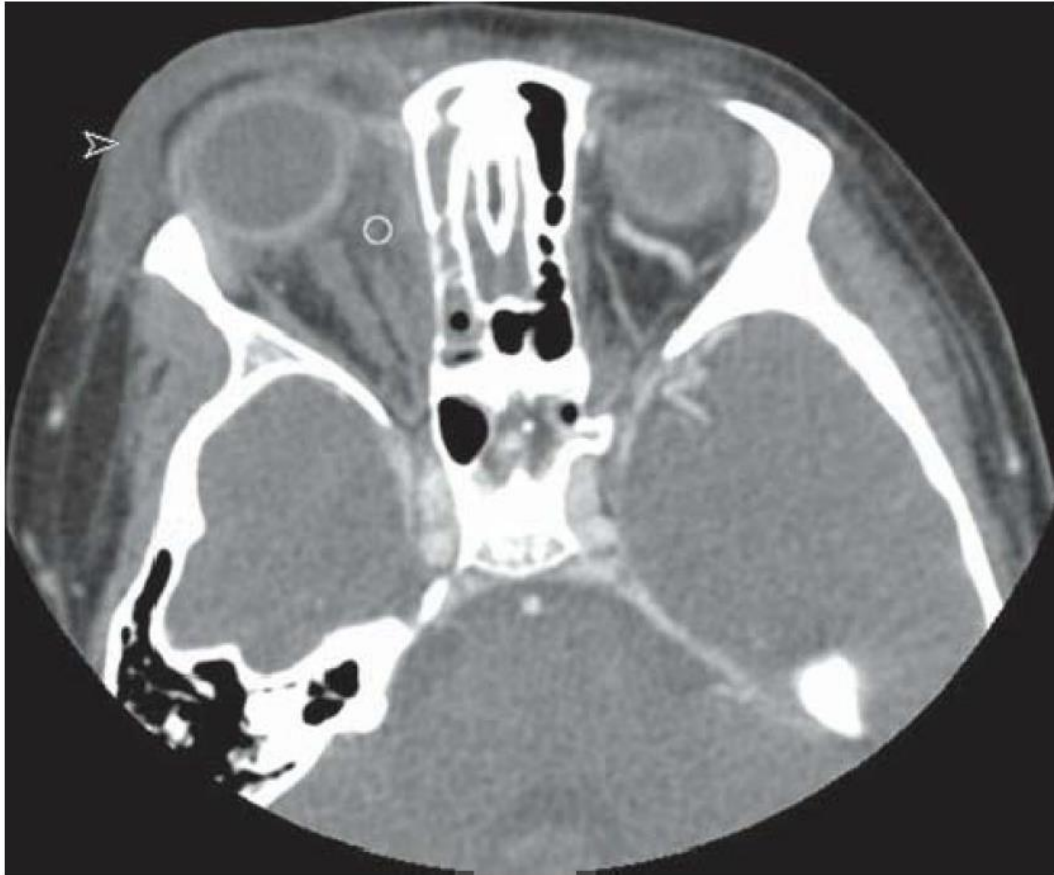


Figure 38.4 Axial CT scan showing right orbital cellulitis with diffuse orbital inflammatory changes that are intra- and extraconal (*open circle*); there is concurrent preseptal edema and inflammatory changes (*arrow*).

- [Treatment of pre-septal and orbital cellulitis:](#)
 - o **Medical management:**
 - 1. Ophthalmologist evaluation:**
 - Indicated immediately for all patients with post-septal involvement for:
 - Daily assessments of:
 - o Visual acuity
 - o Color vision
 - o Pupillary reaction
 - o Extraocular motility.
 - Surgical planning if needed.
 - 2. Systemic Broad-spectrum Antibiotics:**
 - Oral antibiotics are appropriate for:
 - Adults with mild pre-septal cellulitis and when daily follow-up is ensured to rule out developing signs and symptoms of orbital cellulitis.
 - IV antibiotics are appropriate for:
 - All pediatric patients.
 - Adults with severe pre-septal cellulitis.
 - Adults with mild pre-septal cellulitis but daily follow-up can't be ensured.
 - All adults with orbital cellulitis.
 - Choices of Antibiotics:
 - No standard rules on type of antibiotics due to great decline in culture-positive isolates.
 - Empirical antibiotic treatment should target the most common pathogens including Staphylococci, Streptococci and Anaerobes and should cross blood brain barrier.
 - **IV Antibiotics:**
 - o Clindamycin + Ceftriaxone.
 - o Levofloxacin or Ciprofloxacin can be used instead of Ceftriaxone for patients with allergy to penicillin.
 - **Oral Antibiotics:**
 - o Can be started after improvement on IV antibiotics (afebrile and improvements of orbital findings).
 - o If definitive culture data are available:
 - Oral antibiotics should be directed against the infecting organisms.
 - o If no results, appropriate antibiotic is:
 - Clindamycin
 - o Duration of at least 2-3 weeks.

3. Aggressive local care to improve sinus drainage:

- Normal saline irrigation
- Topical steroids
- Topical decongestants

4. Warm compresses

5. Elevating the head of the bed.

6. Daily clinical re-assessment:

- Daily reassessment of orbital signs and symptoms to ensure the clinical improvement on medical management and rule out further progression.

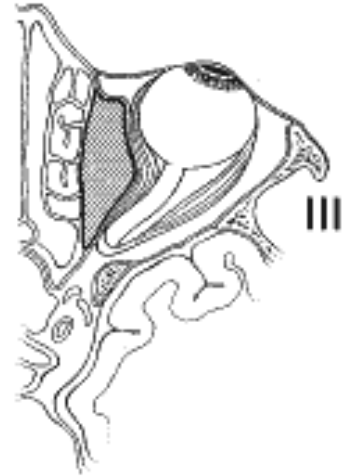
▪ No evidence supporting the routine use of systemic steroids in patients with orbital complications.

○ **Surgical management:**

- Medical therapy is typically sufficient to adequately treat cases of pre-septal and orbital cellulitis.
- Pre-septal cellulitis usually doesn't require any surgical intervention except incision and drainage of eyelid abscess if present.
- Indications of surgical drainage in orbital cellulitis:
 1. Progression of symptoms despite medical therapy.
 2. Lack of improvement within 48 hours despite medical therapy.
 3. Visual acuity of 20/60 or worse on initial evaluation.
 4. Severe orbital complications (Blindness or an afferent pupillary defect) on initial evaluation.
- Surgical treatment include:
 - Adequate endoscopic drainage of the infected sinuses.
 - Incision and drainage of eyelid abscess if present.

III. Sub-periosteal Abscess:

- Abscess collection between periorbita and lamina papyracea.
- Second most common orbital complication of rhinosinusitis.
- Most commonly located at:
 1. Medial wall of the orbit with superomedial or inferomedial extensions from ethmoid sinusitis.
 2. Inferior wall of the orbit from maxillary sinusitis.
 3. Superior wall of the orbit from frontal sinusitis.
- Sub-periosteal abscess develops when infection breaks through the lamina papyracea or travels through the anterior or posterior ethmoidal foramina.
- It can expand rapidly leading to blindness by compromising optic nerve function by:
 1. Direct optic nerve compression
 2. Elevation of intra-orbital pressure
 3. Proptosis causing a stretch optic neuropathy
- 15-30% patients with sub-periosteal abscess will develop various visual sequelae even with aggressive medical and surgical intervention.
- Sub-periosteal abscess can be suspected when a patient who has orbital cellulitis develops:
 - Worsening proptosis.
 - Gaze restriction.
 - Loss of red/green perception due to increasing intraorbital pressure.
 - Deterioration of visual acuity.
- Clinical Presentation:
 - Unilateral eyelid edema, erythema and tenderness.
 - Fever.
 - Abscess may rupture through septum and present in eyelids
 - Signs of involvement of post-septal structures:
 - Chemosis
 - Proptosis
 - Pain
 - Ophthalmoplegia due to impaired extraocular muscle movement.
 - Displacement of the orbit downward and laterally.
 - Impairment of visual acuity.



- Diagnosis:
 - **CT scan with contrast:**
 - Indicated in all patients with post-septal infection.
 - Findings:
 - Rim-enhancing hypodensity in extraconal space adjacent to lamina papyracea with mass-effect and lateral displacement and enlargement of medial rectus muscle.

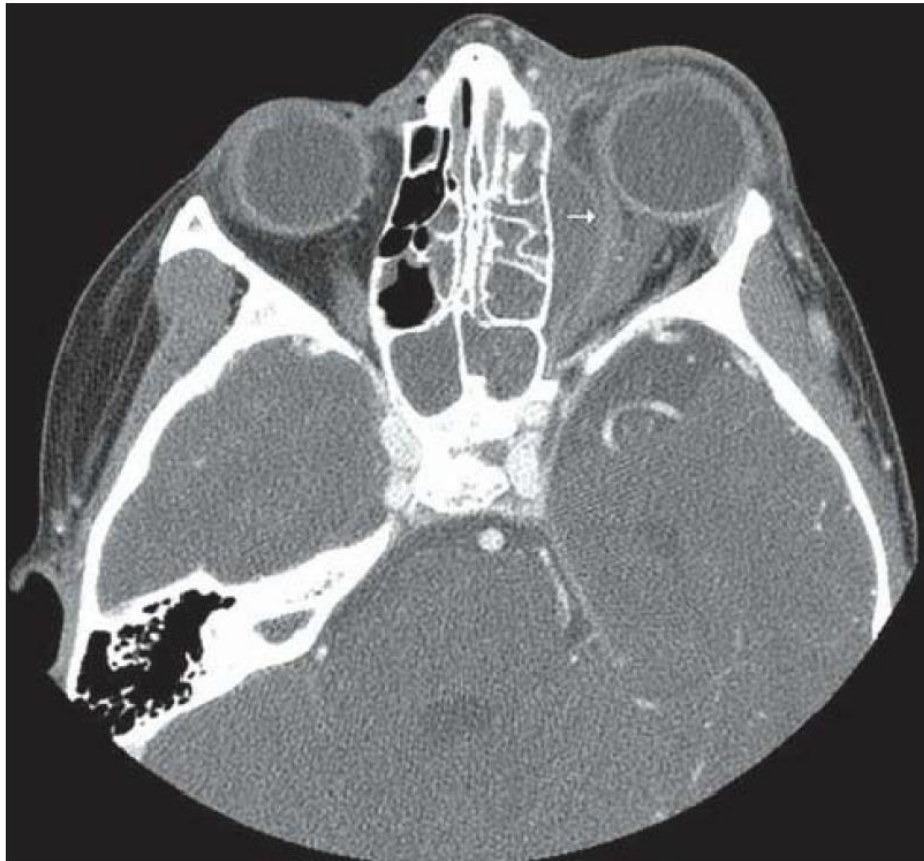


Figure 38.3 Axial CT scan demonstrating a left subperiosteal orbital abscess. The abscess is immediately adjacent to the lamina papyracea (the *arrow* is inside the abscess and identifies the lateral edge of the abscess); note the proptosis and the displacement of the orbital periosteum and medial rectus muscle.

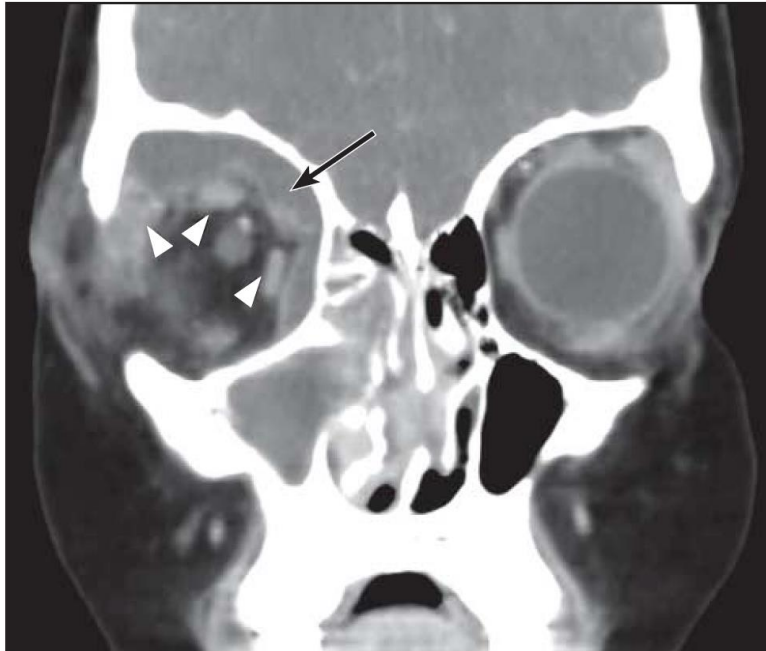
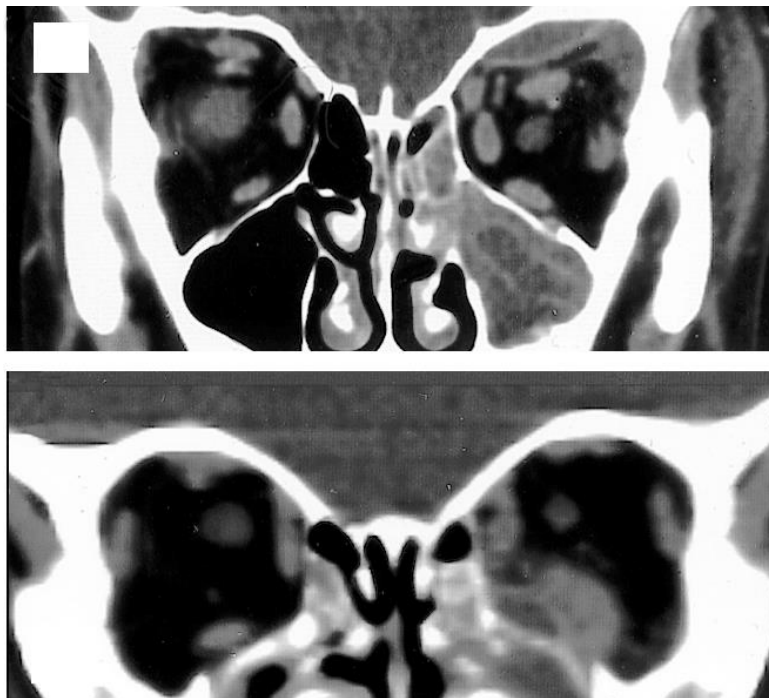
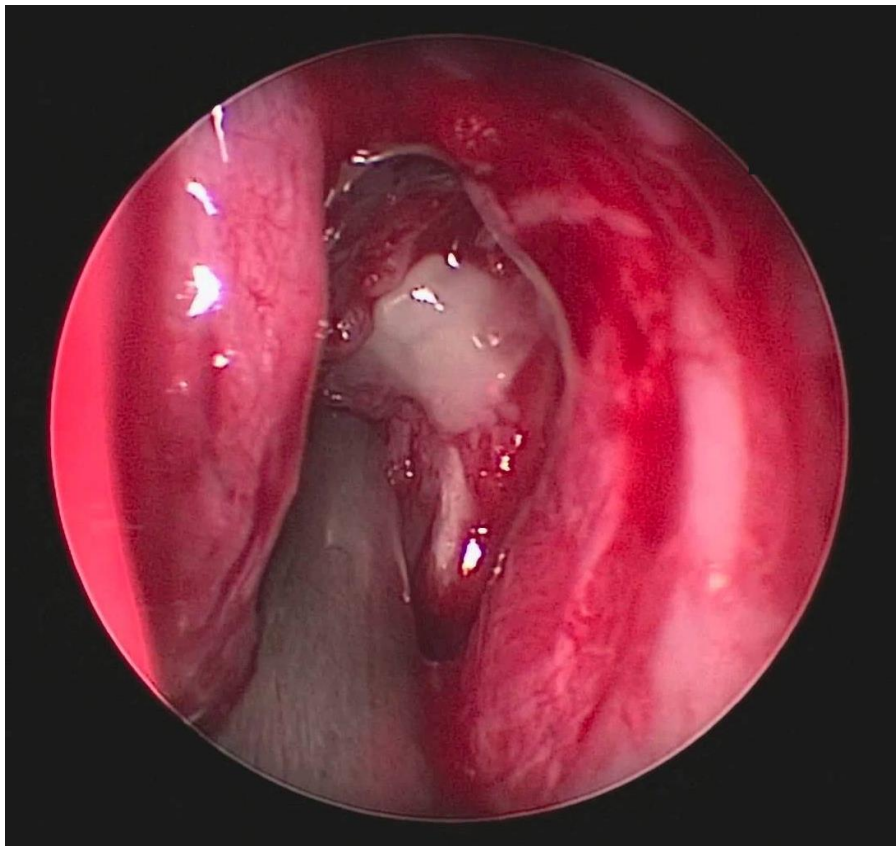


Figure 2. Right pediatric subperiosteal orbital abscess with superolateral extension (black arrow) and involvement of 3 extraocular muscles (arrowheads) drained externally.



- Treatment of sub-periosteal abscess:
 - **Medical management:**
 - Medical management only with close ophthalmologic and otolaryngologic assessment can adequately treat sub-periosteal in 50-60% of patients.
 - Oxford and McClay proposed the following criteria to consider medical management only:
 1. Medial Sub-periosteal abscess.
 2. Abscess width $\leq 4\text{mm}$.
 3. Normal visual acuity, pupil, and retina.
 4. No ophthalmoplegia.
 5. Intraocular pressure $< 20\text{mm Hg}$.
 6. Proptosis $\leq 5\text{mm}$.
 - Todman et al. show that volume of subperiosteal abscess seemed to be the most important criterion in determining medical versus surgical management.
 - Abscess volume of $<1,250\text{ mm}^3$ did not require surgical management.
 - **Surgical management:**
 - Combined surgical and medical treatment achieve complete resolution in 95-100% of cases.
 - Immediate surgical intervention for patients started of medical therapy only is warranted if:
 1. Progression of symptoms despite medical therapy.
 2. Lack of improvement within 48 hours despite medical therapy.
 - Goals of surgical management:
 - Removing lamina papyracea to drain the abscess.
 - Opening ethmoid cells to facilitate sinus drainage.
 - Surgical approach:
 1. **Endoscopic Trans-Nasal:**
 - Gold standard approach for medial sub-periosteal abscess.
 - Intra-operative measurement of orbital pressures is extremely helpful and dictates the extent of orbital decompression.

- Steps:
 - Uncinectomy.
 - Anterior ethmoidectomy.
 - Skeletonizing of lamina papyracea
 - Cracking the lamina with a Cottle or Freer elevator.
 - Drainage of subperiosteal collection.
 - Periorbita should not be violated.
- Advantages:
 - Avoiding facial scar
 - Removal of polypoidal tissue, polyps, and necrosed debris.
- Disadvantages:
 - Increase risk of bleeding from the acutely inflamed mucosa.



2. External Frontoethmoidal Orbitotomy/Lynch :

- Indicated for superior-medial or superior sup-periosteal abscess if endoscopic approach is not feasible.
- Steps:
 - Frontoethmoidal "Lynch" incision is marked halfway between the medial canthus and bridge of the nose.
 - Extends superiorly and inferiorly in a curved fashion approximately 2-3cm.
 - Incision is extended deeply to periosteum.
 - Angular vessels are controlled with bipolar cautery.
 - Supratrochlear bundle is preserved superiorly.
 - Sub-periosteal dissection is performed and the periosteum is preserved to ensure the integrity of medial canthal ligament, trochlea of superior oblique, and lacrimal sac as well as to prevent herniation of orbital fat.
 - As the medial wall of the orbit is widely exposed, the anterior ethmoidal artery is encountered in the frontoethmoidal suture line 24mm posterior to the anterior lacrimal crest.
 - This vessel is dipped or coagulated with a bipolar electrocautery.
 - Periorbita is protected with a malleable retractor to prevent herniation of orbital fat into the field.
 - Periorbital elevation is not routinely performed posterior to the anterior ethmoidal artery.
 - Additional elevation demonstrates the posterior ethmoidal artery 12mm posterior to anterior artery and 6 mm anterior to the optic foramen.
 - These proportions are not constant, and great care must be exercised in dissecting the area of the posterior ethmoidal artery to avoid retrobulbar hemorrhage and injury to the optic nerve.

- After a wide exposure of the medial orbital wall, ethmoidectomy is done and through the lamina papyracea below the level of skull base which is defined by frontoethmoidal suture line.
- Careful dissection is necessary to prevent injury to lateral lamella of the cribriform and a CSF leak.
- External drain is inserted if no ethmoidectomy being performed.
- Advantages:
 - Ensure adequate drainage of the abscess when combined with ESS.
- Disadvantages:
 - Facial scar.

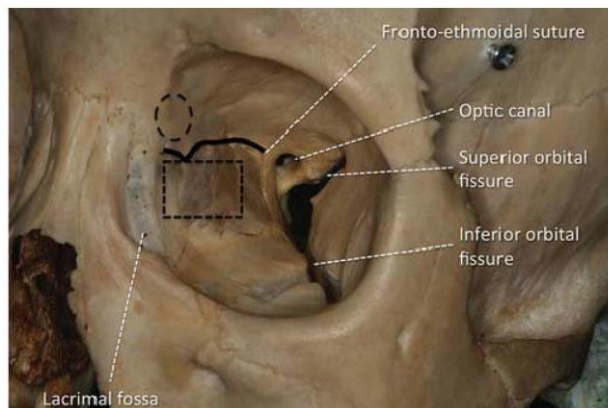
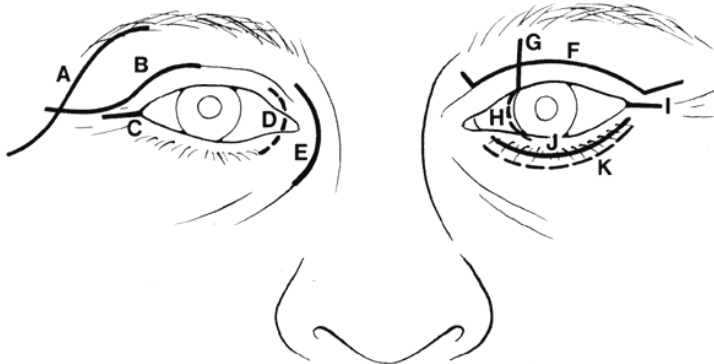
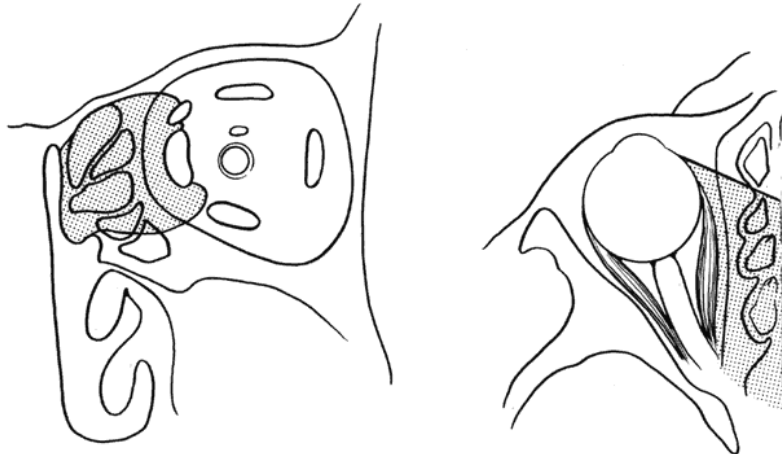
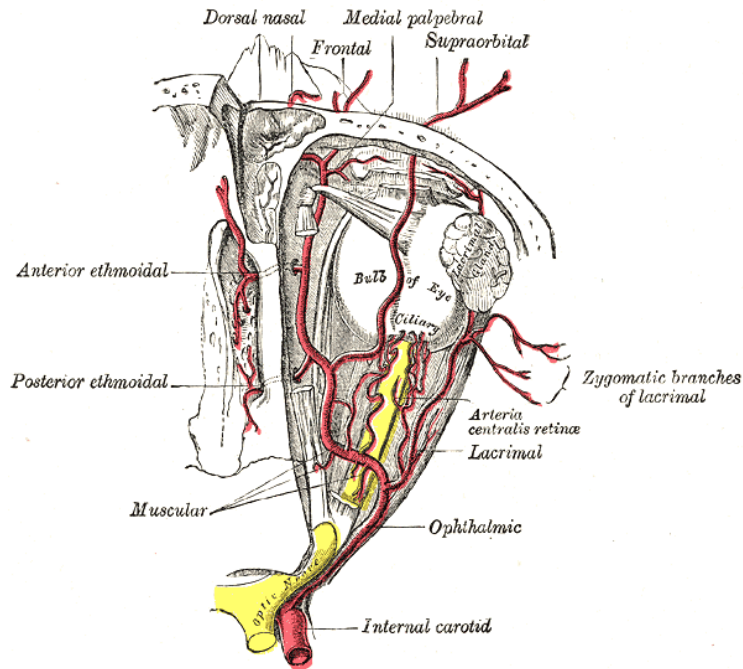


Figure 41.3 Dry skull to show the regions approached through a Lynch incision. The lateral wall of the frontal recess is exposed and the frontal sinus can be accessed (*circle*). External ethmoidectomy is performed after a wide exposure of the medial orbital wall and the ethmoid is entered through the lamina papyracea. The level of the skull base is defined by the frontoethmoidal suture line and bone is removed inferior to this line (*rectangle*).



Incisions used for orbital surgery:

Deep Approaches:

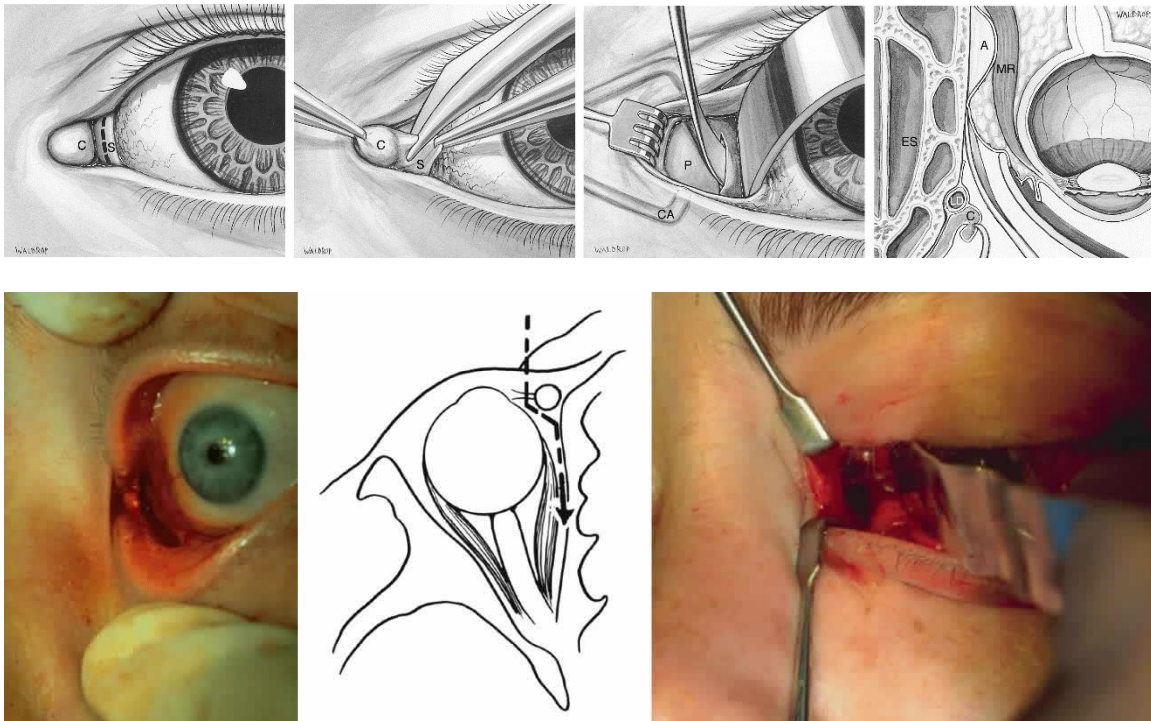
- A: Stallard-Wright lateral orbitotomy incision
- B: Lid crease with lateral extension
- C: Modified Berke lateral canthotomy incision
- D: Transcaruncular incision
- E: Frontoethmoidal "Lynch" incision

Anterior Approaches:

- F: Upper lid crease incision
- G: Vertical lid split incision
- H: Transconjunctival medial orbitotomy
- I: Lateral canthotomy incision
- J: Lower lid percutaneous incision
- K: Transconjunctival lower lid incision.

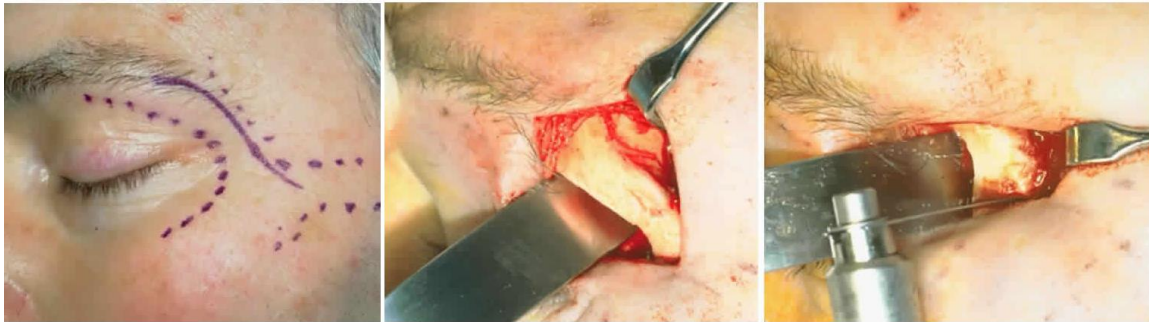
3. Transcaruncular Medial Orbitotomy:

- Indicated for medial sup-periosteal abscess if endoscopic approach is not feasible.
- Steps:
 - Transconjunctival incision is made with tenotomy scissors just lateral to the caruncle.
 - Globe is protected with a retractor and dissection remain lateral to the lacrimal sac but medial to the globe and medial rectus muscle.
 - Posterior to the sac, dissection is carried to the medial bony wall, where periosteum can then be incised and elevated posteriorly until the abscess cavity is encountered.
- Advantages:
 - No cutaneous scar.
 - Allows easy access and visualization of the abscess cavity from the orbital side of the lamina papyracea.
 - Allows confirmation of complete drainage from nasal and orbital sides when used in conjunction with ESS.



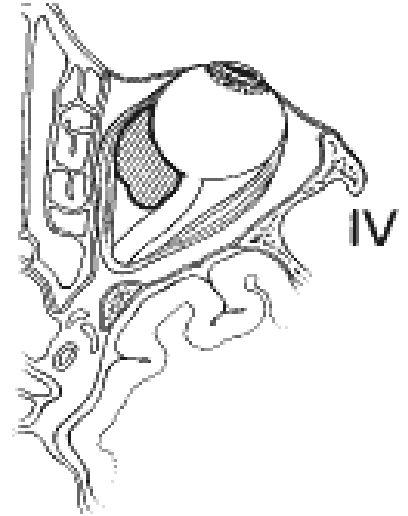
4. Lateral Eyebrow Orbitotomy:

- Indicated for sup-periosteal with substantial superior and lateral extension.
- Steps:
 - Skin incision is made just below the inferior border of the eyebrow overlying the lateral orbital rim.
 - Periosteum is incised and elevated along the lateral rim.
 - Large malleable retractor is placed between the orbital contents and the medial surface of the lateral orbital rim to protect the orbital contents.
 - Periorbital along the medial surface of the lateral wall is elevated posteriorly within the orbit until the abscess cavity is encountered.

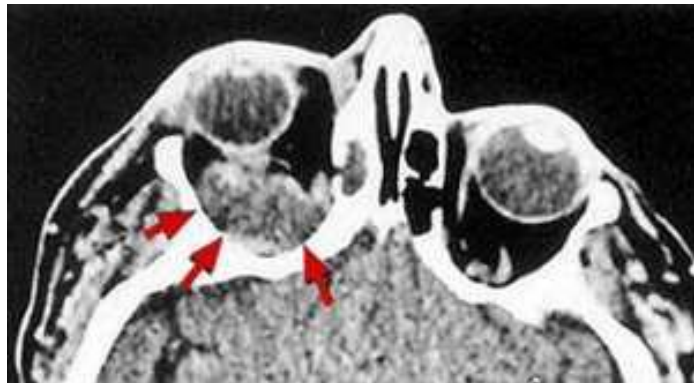
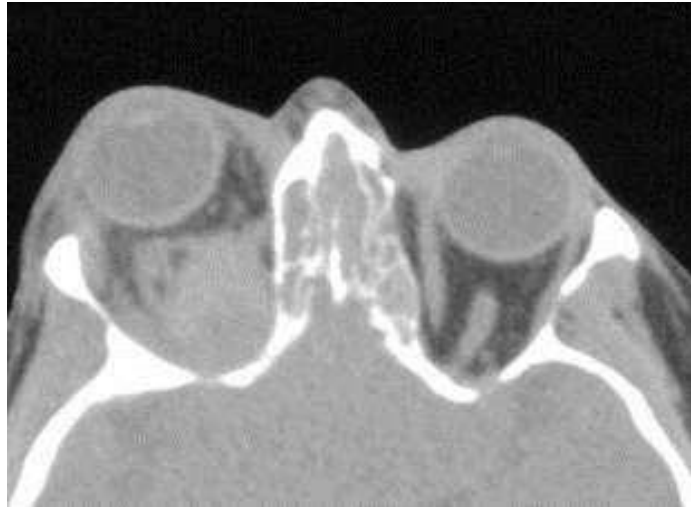


IV. Orbital Abscess:

- Abscess collection within the orbital soft tissue.
- Occurs when orbital cellulitis coalesces into a discrete collection of pus within the orbital tissues.
- Progression to this stage is linked to either:
 - Delay in diagnosis and therapy.
 - Immunocompromised state.
- Presents similar to sub-periosteal abscess but with more severe manifestations.
- Clinical Presentation:
 - Unilateral eyelid edema, erythema and tenderness.
 - Fever.
 - Abscess may rupture through septum and present in eyelids
 - Signs of involvement of post-septal structures:
 - Severe chemosis
 - Severe proptosis
 - Severe pain
 - Complete ophthalmoplegia due to impaired extraocular muscle movement.
 - Impairment of visual acuity.
 - Risk of irreversible blindness.



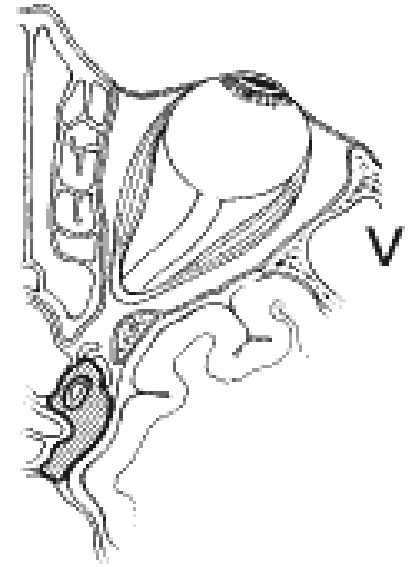
- Diagnosis:
 - **CT scan with contrast:**
 - Indicated in all patients with post-septal infection.
 - Findings:
 - Diffuse infiltration of the intraconal and extraconal orbit fat with areas of cavitations.
 - Massive proptosis, extraocular enlargement and gas formation.
 - **MRI with Gad:**
 - True necrotic abscess appears as HYPOintensity on T1 and HYPERintensity on T2.



- [Treatment of orbital abscess:](#)
 - **Combined Medical and Surgical management:**
 - Goals of surgical management:
 - Removing lamina papyracea to drain the abscess.
 - Opening ethmoid cells to facilitate sinus drainage.
 - Surgical approach:
 - 1. Endoscopic Trans-Nasal:**
 - Preferred alone for drainage of medial extra-conal orbital abscess.
 - Should be combined with external approach if intra-conal abscess is present with the assistance of ophthalmologist to avoid the significant risk of irreversible blindness.
 - Steps:
 - Similar to technique as for sub-periosteal abscess except:
 - Incision of periorbital with sickle knife from posterior to anterior is necessary to drain extra-conal orbital abscesses.
 - Posterior ethmoidectomy is indicated if there is significant posterior ethmoid disease and extension of the abscess toward the orbital apex.
 - A wide exposure and decompression of the medial orbital wall is need if orbital pressures continue to elevate substantially.
 - 2. External approach:**
 - Should be combined with Endoscopic approach if intra-conal abscess is present with the assistance of ophthalmologist to avoid the significant risk of irreversible blindness.
 - Different approach can be utilized depends on ophthalmologist perefences.

V. Cavernous Sinus Thrombosis:

- Serious orbital and intracranial complication of rhinosinusitis.
- Results from retrograde spread of infection from orbit and sinonasal cavities (Sphenoid > Ethmoid > Frontal) through the valveless superior and inferior ophthalmic veins connecting the cavernous sinus.
- Extension of phlebitis posteriorly into the ipsilateral cavernous sinus and through intercavernous sinuses to the contralateral cavernous sinus occurs within 24-48 hours of the initial presentation and results in a progression of symptoms in the opposite eye, which is the hallmark feature of cavernous sinus thrombosis.
- Carotid thrombosis may follow cavernous sinus thrombosis with concomitant strokes, subdural empyema, brain abscess or meningitis.
- Staph. aureus is the most common pathogen.
- High mortality rate reaching up to 30% even with rapid recognition and treatment.



Clinical Presentation:

- Picket fence spiking fevers.
- Toxemia.
- Severe bilateral orbital involvement:
 - Severe chemosis
 - Severe proptosis
 - Severe pain
 - Complete ophthalmoplegia and paralysis of extraocular muscles due to involvement of CN-III, IV, VI.
 - Papilledema due to obstruction of venous drainage from the retina.
 - Fixed dilated pupils.
 - Impairment of visual acuity with irreversible blindness.



Agayev A, Yilmaz S. Cavernous sinus thrombosis.
N Engl J Med 2008; 359:2266.

- Diagnosis:
 - **CT scan with contrast:**
 - Findings:
 - Identify source of infection
 - thickening of superior ophthalmic vein
 - Irregular filling defects in the cavernous sinus.
 - Aneurysm of the internal carotid artery as a complication of cavernous sinus thrombosis.
 - **MRI with Gad/MRV:**
 - More sensitive method for diagnosing cavernous sinus thrombosis.
 - Findings:
 - Engorgement of the cavernous sinus, ophthalmic veins, and extraocular muscles.
 - Heterogeneous intensity in the abnormal cavernous sinus with HYPER-intense signal of thrombosed vascular sinuses on all sequences.
 - Cavernous portion of the internal carotid artery is usually deformed.
 - Aneurysm of the internal carotid artery as a complication of cavernous sinus thrombosis.

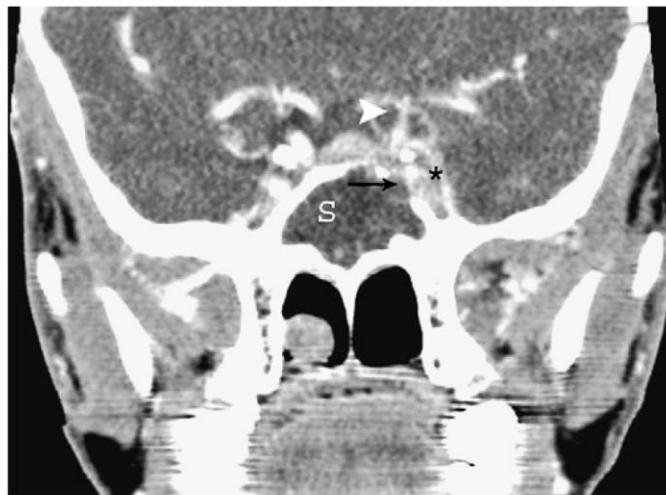
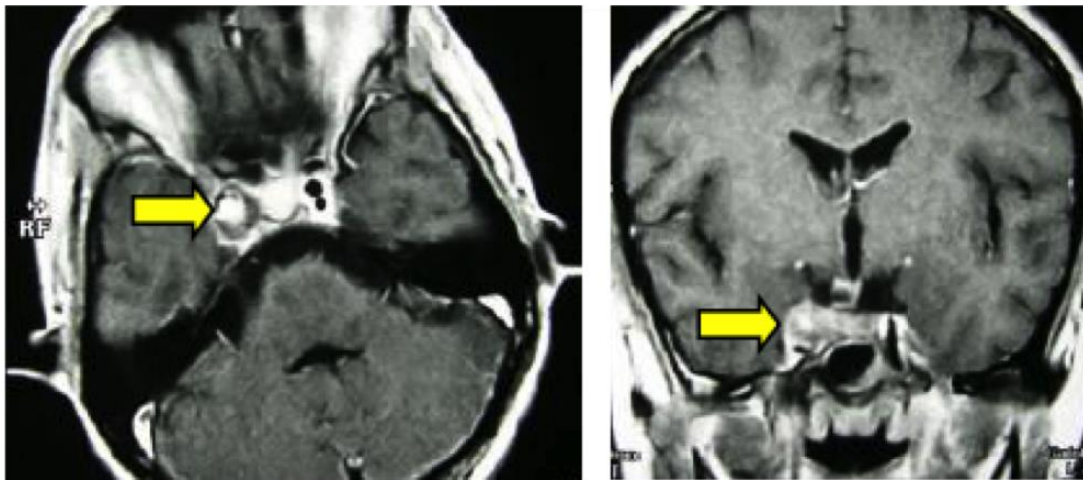


Fig. 6. Coronal CT sinuses with contrast demonstrating left sphenoid sinusitis (S) with bone erosion (*black arrow*). Note the filling defect and expansion of the left cavernous sinus consistent with CST (*asterisk*), and an abscess formed around the supraclinoid internal carotid artery (*arrowhead*). (*Courtesy of Ilana Seligman, MD, Chicago, IL.*)



- Treatment of cavernous sinus thrombosis:
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease.
 - Empiric treatment may include Nafcillin, Ceftriaxone, Clindamycin, or Vancomycin if methicillin-resistant *S aureus* is a concern.
 - Intravenous antibiotics should continue for 3-4 weeks, or 6-8 weeks if intracranial complication is evident.
 - Anti-coagulants:
 - Controversy.
 - May decrease the progression of thrombosis.
 - Risk of fatal hemorrhagic cerebral infarction and subarachnoid hemorrhage.
 - Retrospective reviews of published reports showed that hemorrhage caused by anticoagulation is rare, and early adjunctive anticoagulation is beneficial in these patients if commenced after excluding the hemorrhagic sequelae of CST radiologically.
 - One study suggested that heparin use in conjunction with antibiotics early in the course of CST may reduce morbidity, not evidence found that it reduces mortality.
 - **Surgical management:**
 - Endoscopic drainage of the affected sinuses is advisable in most circumstances once the patient is stable.

Table 38-2. Source and route of infection in cavernous sinus thrombosis

Source	Disease	Route
Nose and danger area of face	Furuncle and septal abscess	Pharyngeal plexus
Ethmoid sinuses	Orbital cellulites or abscess	Ophthalmic veins
Sphenoid sinus	Sinusitis	Direct
Frontal sinus	Sinusitis and osteomyelitis of frontal bone	Supraorbital and ophthalmic veins
Orbit	Cellulitis and abscess	Ophthalmic veins
Upper lid	Abscess	Angular vein and ophthalmic veins
Pharynx	Acute tonsillitis or peritonsillar abscess	Pharyngeal plexus
Ear	Petrositis	Petrosal venous sinuses

- **Other Orbital Complications:**

1. Superior orbital fissure syndrome:

- Infection of sphenoid sinus can rarely affect structures of superior orbital fissure.
- Deep orbital pain
- Frontal headache
- Progressive paralysis of **CN-VI, III and IV.**

2. Orbital apex syndrome:

- Superior orbital fissure syndrome with additional involvement of Optic nerve and Ophthalmic division of Trigeminal (V₂)
- **CN-VI, III and IV + II and V1**

TABLE
38.1

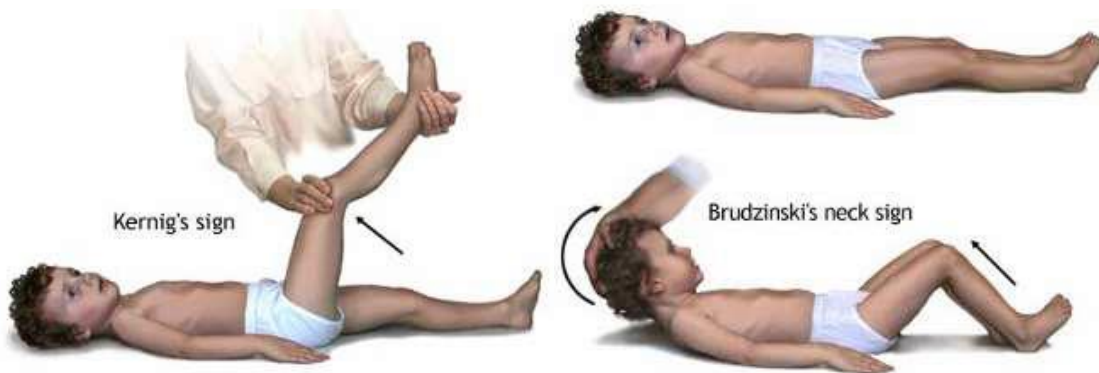
ORBITAL COMPLICATIONS OF SINUSITIS

	Typical Findings	Treatment
Preseptal cellulitis	Edematous, erythematous eyelids Extraocular muscles (EOM) intact Normal vision	Medical therapy (Rarely, drainage of secondary abscess)
Orbital cellulitis	More diffuse orbital edema ± Impaired EOM Usually normal vision until later in disease course	Medical therapy ± Sinus drainage ^a
Subperiosteal abscess	Edematous, erythematous eyelids; Proptosis Impaired EOM Usually normal vision, esp in the case of small abscesses Visual changes more likely with larger abscesses	Medical therapy ± Sinus drainage ± Abscess drainage
Orbital abscess	Severe exophthalmos, chemosis, Ophthalmoplegia, common Visual impairment, common	Medical therapy Sinus drainage, often Abscess drainage, usually
Cavernous sinus thrombophlebitis	Bilateral orbital pain, chemosis, proptosis Ophthalmoplegia CN III, IV, V1, V2, V3, VI can be affected	Medical therapy Sinus drainage, often ± Anticoagulation (controversial)

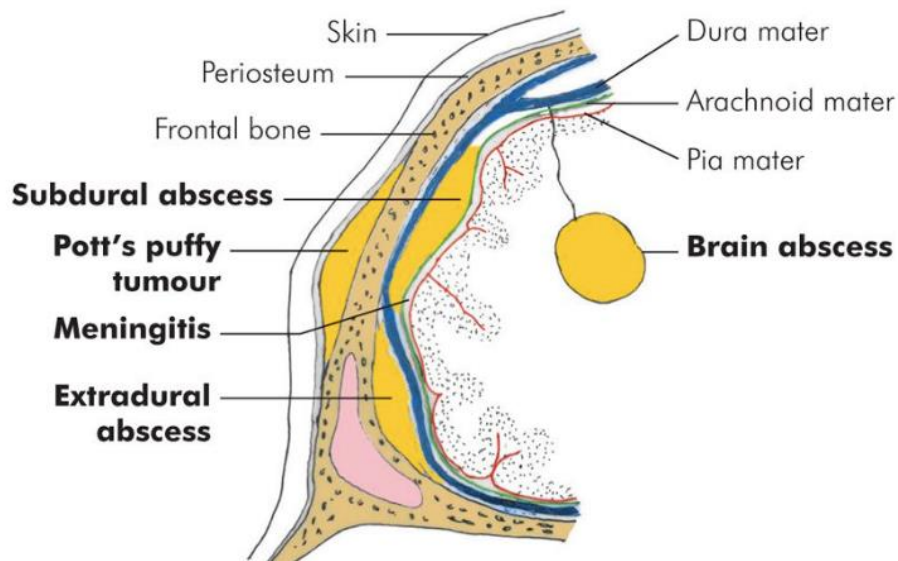
^aSurgical sinus drainage may be limited to maxillary sinus aspiration or may include endoscopic or open sinus surgery; its necessity depends on severity of symptoms, physical examination, duration of medical treatment and need for cultures to direct antibiotic therapy.

- **Intracranial Complications: (15-20%)**
- Second most common complications of rhinosinusitis.
- More common in male and adolescents.
 - Adolescents have developed their frontal and sphenoid sinuses compared to children and more to URTI compared to adults.
- Incidence, morbidity, and mortality of suppurative intracranial complications secondary to sinusitis have dramatically decreased due to widespread use of antibiotics and improved intensive care medicine.
- 4% overall mortality rate of intracranial complication.
- 13–35% overall morbidity rate, including long-term neurologic deficits, cognitive impairment, cranial nerve palsy, aphasia, epilepsy, hydrocephalus, and visual and hearing loss.
- Intracranial complications occur due to:
 - Close relationship between paranasal sinuses and anterior and middle skull base.
 - Complex venous network that traverses this area.
- Pathways of intracranial involvements:
 - 1. Thrombophlebitis:**
 - Main pathway of intracranial complications.
 - Retrograde septic thrombophlebitis through the valveless diploic veins of skull and ethmoid bone or communicating veins.
 - The intracranial venous system is also valveless, allowing further spread of thrombophlebitis and septic emboli.
 - 2. Direct extension:**
 - Occurs via preformed routes such as congenital or traumatic dehiscences, through sinus wall erosion or traumatic fracture lines, and by existing neurovascular foramina such as those for the optic and olfactory nerves.
- Risk factors for intracranial extension in patients with orbital complications of rhinosinusitis:
 - 1.** Age $\geq$ 7 years
 - 2.** Failure to improve after appropriate therapy
 - 3.** Changes in neurologic status
 - 4.** Frontal sinus opacification on CT
 - 5.** Superior or lateral position of orbital abscess
 - 6.** Need for surgical intervention to drain orbital abscess
 - 7.** Male sex
 - 8.** African-American race.

- Clinical picture of intracranial complications:
 - **High grade fever.**
 - **Increased intracranial pressure:**
 - Headache
 - Nausea and vomiting
 - Papilledema
 - Change in mental status
 - **Meningism:**
 - Neck stiffness
 - Photophobia
 - Kernig's sign:
 - Pain is elicited with hip flexion and leg extension.
 - Brudzinski's sign:
 - Flexion at neck causes a reflexive flexion of the legs.
 - **Neurologic deficits:**
 - Cranial nerve palsy
 - Epilepsy
 - Visual and hearing loss.



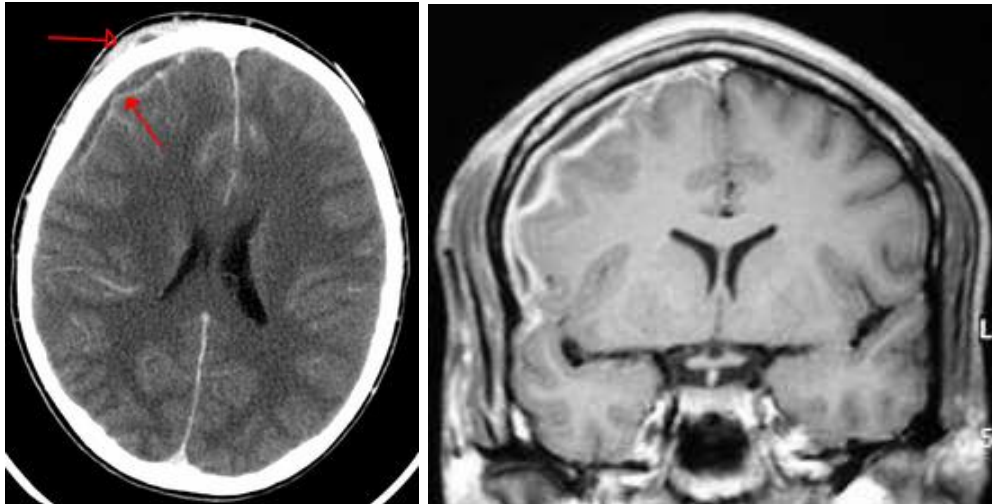
- Classification of intracranial complications:
 - All associated with significant morbidity and mortality and warrant immediate attention.
 - Multiple complications can occur in the same patient either concurrently or sequentially.
 - Intracranial complications include:
 1. Subdural abscess
 2. Intracerebral abscess
 3. Meningitis
 4. Epidural abscess
 5. Venous sinus thrombosis



I. Subdural Abscess:

- Most common intracranial complication (60%).
- Subdural abscesses form between the dura and arachnoid mater of the meninges.
- Most commonly results from frontal sinusitis.
- Due to lack of anatomic barriers, subdural abscess spreads diffusely within the subdural space (over the cerebral convexities, interhemispheric fissure, tentorium, posterior fossa, spinal canal)
- Life-threatening emergency because patients can deteriorate quickly.
- Have higher mortality (25%) and morbidity (30%) rates compared to other intracranial complications.
- Patients present with rapidly clinical deterioration with symptoms of increased intracranial pressure, meningeal irritation, cerebritis, neurological deficits and septic shock.

- Diagnosis:
 - **CT scan with contrast of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Concave (crescentic) hypodensity with strong peripheral contrast enhancement.
 - Extra-axial, Not bounded by sutures, Not crossing the midline.
 - **MRI with Gad of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - **T1:** Concave HYPO-intensity.
 - **T1 + Gd:** Strong peripheral contrast enhancement.
 - **T2:** Concave HYPER-intensity.
 - **DWI:** Concave HYPER-intensity.
 - **Lumbar puncture is contraindicated due to high risk of cerebellar tonsillar herniation due to increased intracranial pressure.**

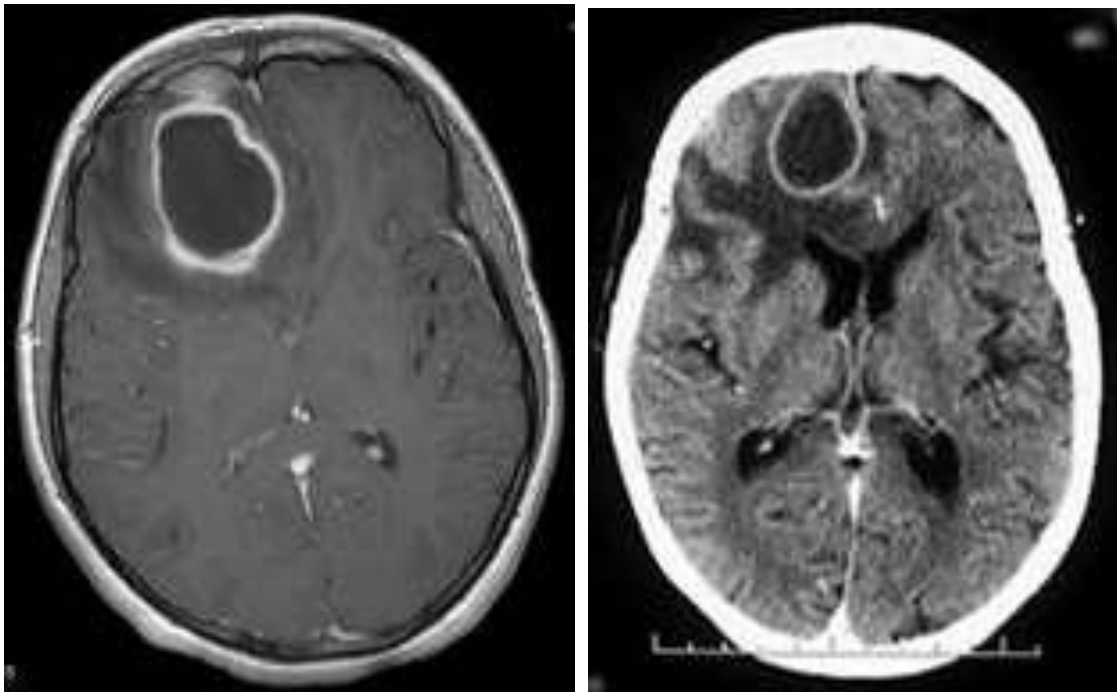


- Treatment:
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 2-6 weeks depending on presence of osteomyelitis.
 - Systemic steroid and Anti-convulsants:
 - Adjunctively administered to treat any secondary cerebral edema and to reduce the risk of seizures.
 - Some argue against steroid use because it impairs the abscess encapsulation process, increase necrosis, reduce antibiotic penetration into the abscess, and alter the appearance on CT scans.
 - **Surgical management:**
 - Required immediate drainage of subdural abscess.
 - Concurrent drainage of sinuses can be done if the patient is stable.
 - Sinuses can be addressed through ESS, frontal sinus trephination, or combined approaches.
 - Drainage of the subdural abscess can be accomplished through craniotomy or burr holes; controversy exists in the neurosurgical literature regarding the preferred intervention.

II. Intracerebral Abscess:

- Second most common intracranial complication (20%)
- Most common locations for intracerebral abscess are frontal and frontoparietal lobes.
- Most commonly results from frontal sinusitis.
- Thrombophlebitis and septic implantation in areas of sluggish venous flow seem to be the predominant routes of parenchymal abscess formation, with the junction of white and gray matter at highest risk.
- Abscesses localized to the frontal lobe are associated with more subtle symptoms, such as mood and behavioral changes.
- Abscess rupture is suggested by worsening headache with meningismus and is often fatal.
- Associated with high mortality (20-30%) with 60% of children who survive developing neurologic and developmental sequelae.

- [Diagnosis:](#)
 - **CT scan with contrast of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Intracerebral hypodensity with strong peripheral contrast enhancement.
 - **MRI with Gad of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - **T1:** HYPO-intense intracerebral collection.
 - **T1 + Gd:** Strong peripheral contrast enhancement.
 - **T2:** HYPER-intense intracerebral collection.
 - **DWI:** HYPER-intense intracerebral collection.
 - **Lumbar puncture is contraindicated due to high risk of cerebellar tonsillar herniation due to increased intracranial pressure.**



- Treatment:
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 2-6 weeks depending on presence of osteomyelitis.
 - Systemic steroid and Anti-convulsants:
 - Adjunctively administered to treat any secondary cerebral edema and to reduce the risk of seizures.
 - Some argue against steroid use because it impairs the abscess encapsulation process, increase necrosis, reduce antibiotic penetration into the abscess, and alter the appearance on CT scans.
 - **Surgical management:**
 - Required immediate drainage of intracerebral abscess.
 - Concurrent drainage of sinuses can be done if the patient is stable.
 - Sinuses can be addressed through ESS, frontal sinus trephination, or combined approaches.
 - Drainage of the intracerebral abscess can be accomplished through stereotactic aspiration and drainage or open surgical excision.

III. Meningitis:

- Commonly associated with other complications of Rhinosinusitis.
- Isolated meningitis is a relatively uncommon complication of Rhinosinusitis. (10%)
- Rhinosinusitis itself is an unusual cause of meningitis.
- The involved sinuses are typically the ethmoid and sphenoid sinuses.

- [Diagnosis:](#)
 - **CT scan with contrast of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Normal CT brain.
 - **MRI with Gad of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Dural enhancement, most evident along the falx cerebri, tentorium, and convexity dura.
 - **Lumbar Puncture:**
 - Indicated in all patients with intracranial complications with no evidence of increased intracranial pressure to avoid risk of brain herniation.
 - CSF samples should be sent for cytological and laboratory analysis.

Table 4. Normal CSF characteristics and main pathological alterations

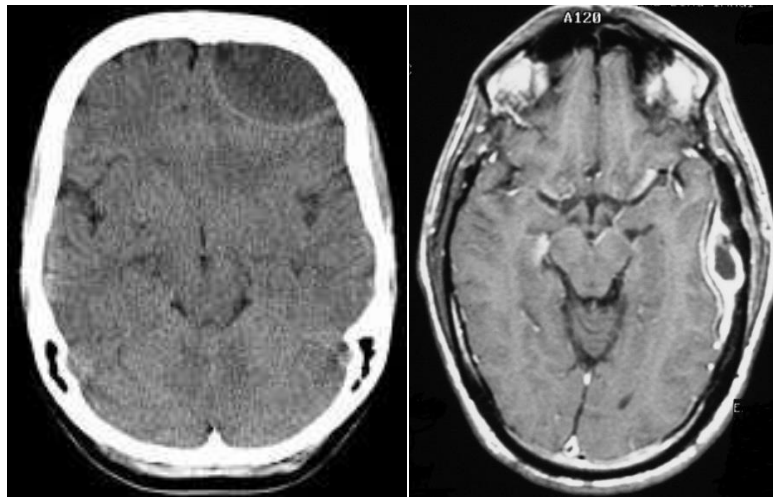
Characteristics	Normal	Meningitis		
		Acute bacterial	Viral	Chronic
Pressure (mm/H ₂ O)	100-200	N or ↑	N or ↑	N or ↑
Aspect	Clear	Turbid/purulent	clear/cloudy	Clear/cloudy
Color	Clear	white	Clear	Clear/white
Cytology (mm ³)	Until 4	>1,000	500-1,000	<500
Cell type	Lymphocytes	Neutrophils	Lymphocytes	Lymphocytes
Protein (mg/dL)	V 5-10 SO 10-25 L 15-45	↑↑	Normal or ↑	Normal or ↑
Glucose (mg/dL)	2/3 from serum	↓	Normal	Normal or ↓
Lactic acid (mmol/L)	<3.5	>3.5	<3.5	<3.5

- Treatment:
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Meningitis is amenable with medical treatment.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 2-6 weeks depending on presence of osteomyelitis.
 - Systemic steroid and Anti-convulsants:
 - Adjunctively administered to treat any secondary cerebral edema and to reduce the risk of seizures.
 - **Surgical management:**
 - Most institutions recommend Endoscopic surgical drainage of the sinuses if clinical improvement is not evident 24-48 hours after the start of intravenous antibiotics, provided the patient has been stabilized.

IV. Epidural Abscess:

- Epidural abscesses form between the skull and dura.
- Expand slowly because of the tight adherence of dura to bone.
- Seen almost exclusively in patients who have frontal sinusitis.
 - High degree of venous communication and loosely adherent dura in frontal area.
- Patients may have a prolonged period, up to several weeks, of nonspecific symptoms until either the infection penetrates the dura or the abscess becomes large enough to cause elevated intracranial pressure.
- Have a favorable prognosis than other intracranial complications regardless of origin of infection.

- Diagnosis:
 - **CT scan with contrast of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Biconvex (lentiform) hypodensity with strong peripheral contrast enhancement adjacent to skull inner table.
 - Extra-axial, bounded by sutures, can cross the midline.
 - **MRI with Gad of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - **T1:** Biconvex HYPO-intensity.
 - **T1 + Gd:** Strong peripheral contrast enhancement.
 - **T2:** Biconvex HYPER-intensity.
 - **DWI:** Biconvex HYPER-intensity.
 - **Lumbar puncture is contraindicated due to high risk of cerebellar tonsillar herniation due to increased intracranial pressure.**

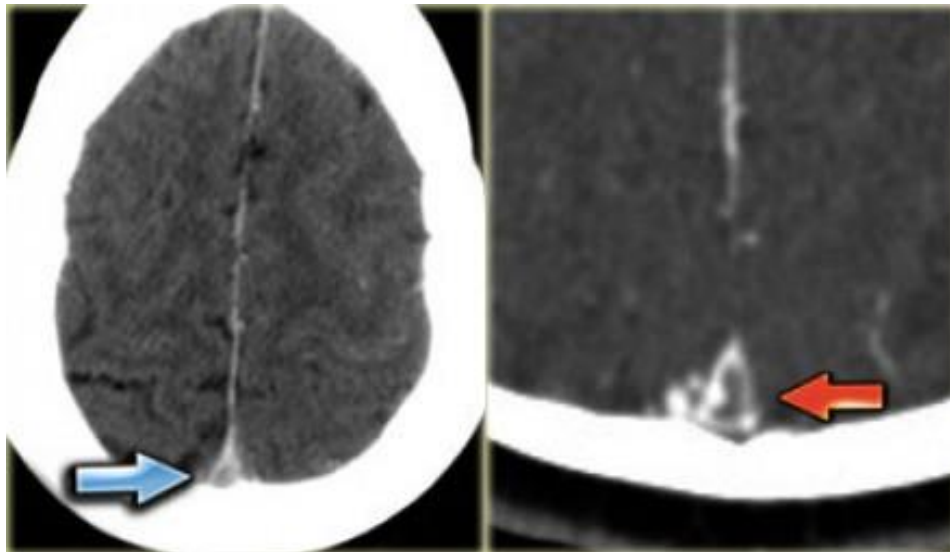


- Treatment:
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 2-6 weeks depending on presence of osteomyelitis.
 - Systemic steroid and Anti-convulsants:
 - Adjunctively administered to treat any secondary cerebral edema and to reduce the risk of seizures.
 - Some argue against steroid use because it impairs the abscess encapsulation process, increase necrosis, reduce antibiotic penetration into the abscess, and alter the appearance on CT scans.
 - **Surgical management:**
 - Required immediate concurrent drainage of the sinuses and epidural abscess once the patient is stable.
 - Sinuses can be addressed through ESS, frontal sinus trephination, or combined approaches.
 - Epidural abscess is most commonly addressed through craniotomy.
 - In rare cases, cranialization of frontal sinus may be necessary if inspection of the posterior table reveals marked osteitis with perforation or necrosis.

V. Venous Sinus Thrombosis:

- Similar to the cavernous sinus thrombosis encountered with orbital-pertaining complications, any of the dural venous sinuses may be affected by rhinosinusitis.
- Superior sagittal sinus is the most commonly involved due to retrograde thrombophlebitis from frontal rhinosinusitis.
- Acute occlusion of the dural sinus is not well tolerated and lead to massive cerebral edema, venous congestion, and infarction.
- Patients are extremely ill, presenting with high spiking fevers, meningeal signs, or other serious neurologic complications.
- Almost always have an associated intracranial abscess formation.

- [Diagnosis:](#)
 - **CT scan with contrast of PNS and Brain:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - **Empty delta sign** (specific to SSST):
 - Triangular area of enhancement with a relatively low-attenuating center, which is the thrombosed sinus (filling defect).
 - Occurs due to recanalisation around an organising clot
 - Gyral enhancement
 - Prominent intramedullary veins
 - **MRI with Gad of PNS and Brain/MRA/MRV:**
 - Indicated in all patients with intracranial complications.
 - Findings:
 - Decreased cavernous carotid artery flow void
 - **T1:** Clot is ISO to HYPER-intense.
 - **T2:** Clot is HYPO-intense.

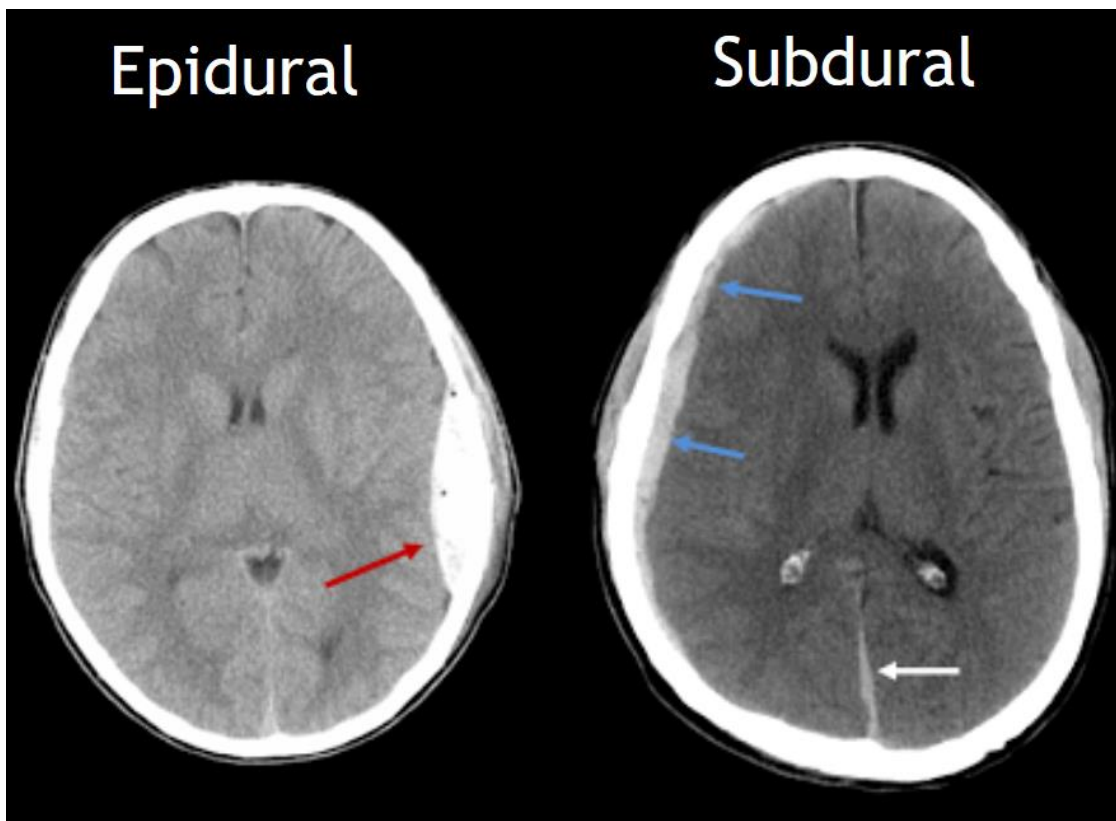
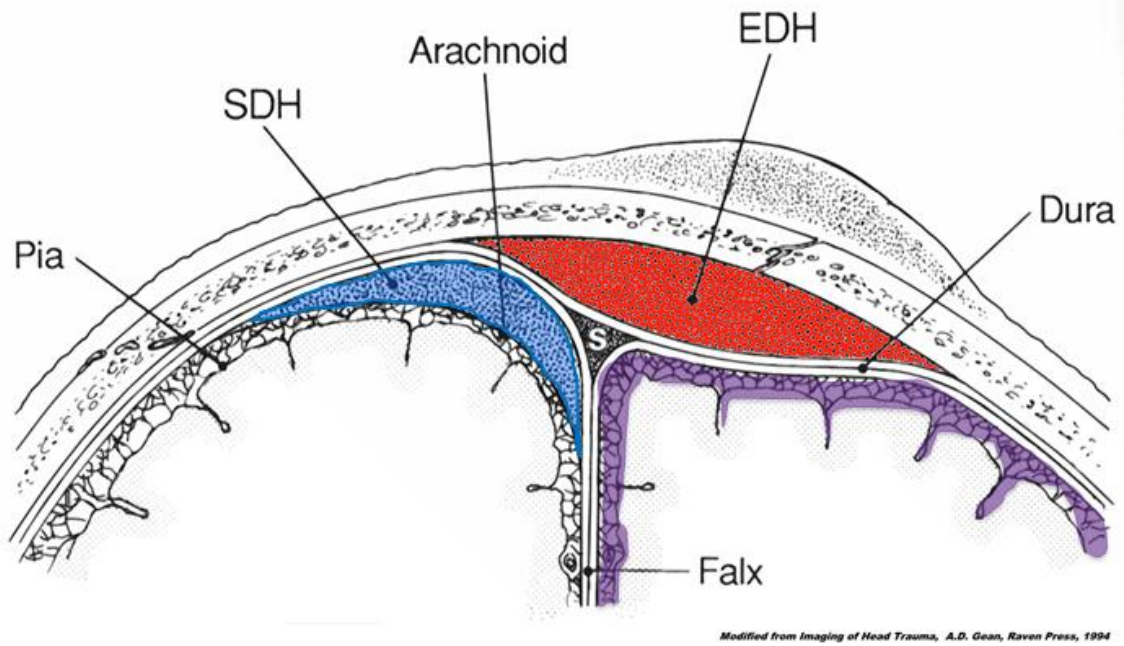


- **Treatment:**
 - **Medical management:**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 2-6 weeks depending on presence of osteomyelitis.
 - Anti-coagulants:
 - Controversy.
 - May decrease the progression of thrombosis.
 - Risk of fatal hemorrhagic cerebral infarction and subarachnoid hemorrhage.
 - When used, the treatment is continued until there is radiologic evidence of resolution of the thrombus.
 - **Surgical management:**
 - Endoscopic drainage of the affected sinuses is advisable in most circumstances once the patient is stable.

**TABLE
38.3****INTRACRANIAL COMPLICATIONS OF SINUSITIS, TREATMENT HIGHLIGHTS**

	Sinus Source^a	Highlights of Disease Process and Treatment
Meningitis	Ethmoid sinus; Sphenoid sinus	High incidence neurologic sequelae (e.g., hearing loss) Medical treatment only, usually
Epidural abscess	Frontal sinus	Aggressive medical treatment; surgical drainage of the abscess, usually Sinus drainage, usually, when medically stable
Subdural abscess	Frontal sinus	Highest rate of neurologic morbidity and mortality Aggressive medical treatment (often includes steroids and anticonvulsants) Surgical drainage of the abscess, usually; sinus drainage, usually, when medically stable
Intracerebral abscess	Frontal sinus (less commonly: ethmoid sinus, sphenoid sinus)	High neurologic morbidity and mortality; usually frontal lobe so symptoms may be subtle Aggressive medical treatment (often includes steroids and anticonvulsants) Surgical drainage of the abscess, usually; sinus drainage, usually, when medically stable
Venous thrombophlebitis	Frontal sinus	Aggressive medical treatment (often includes steroids and anticonvulsants) Anticoagulation is controversial Surgical drainage of the sinuses, sometimes

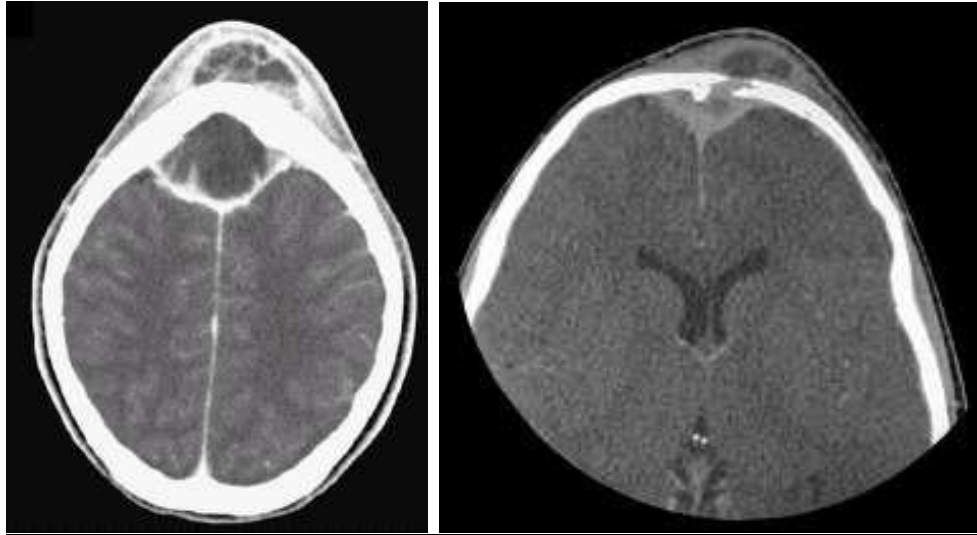
^aMost patients with intracranial complications have unilateral or bilateral pansinusitis; the sinus most commonly implicated as the source of intracranial spread of infection is reported.



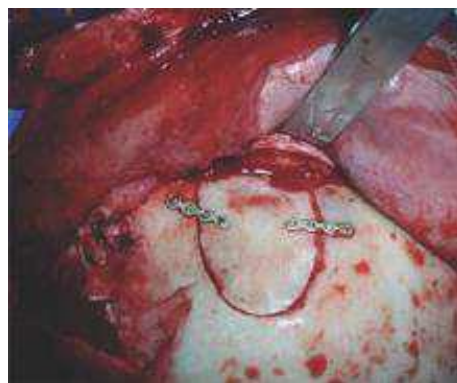
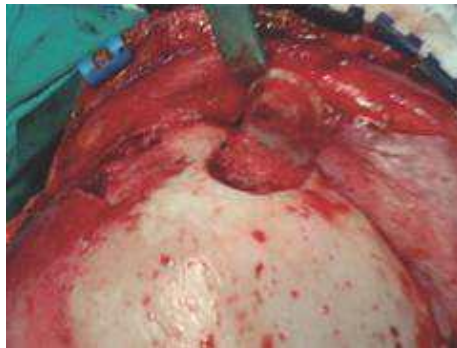
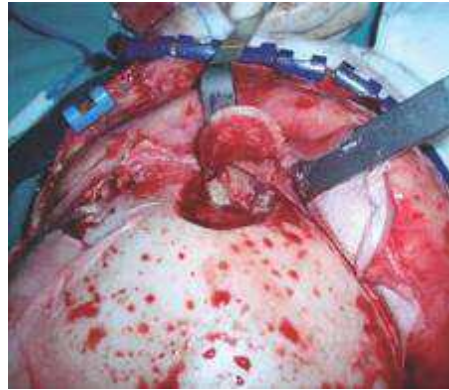
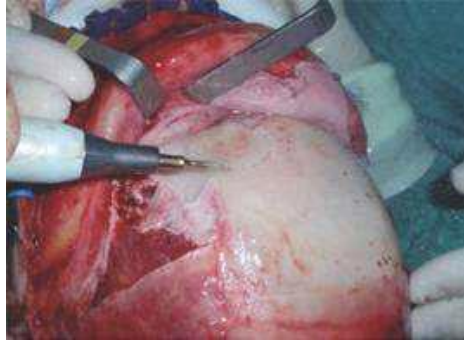
- **Bony Complications: (5-10%)**
- Least common rhinosinusitis complications in pediatrics and adults.
- Osteomyelitis following sinusitis, involves either Maxilla or Frontal bone.
- Osteomyelitis of maxillary bone is more common in children.
- Osteomyelitis of frontal bone is more common in adults.
- **Pott Puffy Tumor:**
 - Acute osteomyelitis of frontal bone followed by subperiosteal abscess formation in the forehead as a complication of frontal sinusitis.
 - Results from acute infection of frontal sinus either directly or through the venous spread.
 - It can also follow trauma or surgery of frontal sinus in the presence of acute infection.
 - Pus form externally under the periosteum as soft doughy swelling (Pott's puffy tumor), or internally as epidural abscess.
 - Associated with other intracranial complications in 60% of cases, especially epidural abscess.
 - In addition to headache, fever, nasal complaints, and neurological findings, Pott's Puffy Tumor is well recognized for the prominent frontal swelling on physical examination.
 - Rare and only 25 cases reported in post-antibiotic era.
 - Clinical picture of intracranial complications:
 - Severe headache
 - Fever
 - Nasal complaints
 - Neurological findings
 - Prominent frontal swelling
 - Frontocutaneous fistula



- Diagnosis:
 - **CT scan with contrast of PNS and Brain:**
 - Confirm the diagnosis and rule out intracranial involvement.
 - **MRI with Gad of PNS and Brain:**
 - Confirm the diagnosis and rule out intracranial involvement.



- Diagnosis:
 - **Medical management::**
 - Required multidisciplinary team including ENT, Neurology, Neurosurgery, Infectious disease, ICU.
 - Systemic High-dose Broad-spectrum Antibiotics:
 - Should cross the blood–brain barrier.
 - Directed at the most common pathogens associated with the disease (Staph aureus and strep. Pneumoniae)
 - Empiric treatment include Ceftriaxone plus Clindamycin, or Vancomycin if methicillin-resistant S aureus is a concern.
 - Duration of IV antibiotics ranging from 6 weeks.
- **Surgical management:**
 - Required concurrent drainage of sinuses and the abscess and resection of the infected bone by raising bicoronal flap.
 - Frontal sinus may be obliterated in situations of recurrence.



Functional Endoscopic Sinus Surgery:

- Endoscopic surgery has made a great contribution towards management of sinus disease.
- Replace other types of sinus surgeries.
- Minimally invasive surgery and does not require skin incisions or removal of intervening bone to access the disease.
- At all times care should be taken to minimize trauma to nasal mucosa.
- **Primary objectives of FESS:**
 - Restore paranasal sinus function by re-establishing the physiologic pattern of ventilation and mucociliary clearance.
 - Reduce the overall burden of inflammatory disease.
 - Provide post-operative access to diseased sinuses.
 - Augment postoperative medication regimen.
- **Main steps of FESS:**
 1. Removal of irreversibly diseased mucosa and bone.
 2. Removal of bony septations.
 3. Judiciously widen the true natural ostia of the sinuses.
 4. Preservation of normal mucosa.
- **Extent of surgery:**
 - Studies showed that surgical results and post-op symptoms were similar in patients underwent limited surgery compared with patients underwent extended surgery (Full-house FESS), suggesting that a conservative approach may be sufficient.
- **Radiological assessment:**
 - Current protocol is to obtain non-contrasted CT scan with thin axial slice thicknesses <3 mm (ideally 1 mm is required for accurate image-guidance compatibility).
 - High-resolution multiplanar reconstruction in coronal and sagittal planes.
 - Contrast CT is needed in the following cases:
 - Sinonasal neoplasm
 - Complicated CRS
 - Intracranial and orbital extension
 - Review of the imaging should be done at least three times:
 - During preoperative planning
 - Night before surgery
 - Immediately prior to surgery.

TABLE 49-1. Key Factors to Be Reviewed on Preoperative Computed Tomography Scan Before Endoscopic Sinus Surgery

Factor	Details
Disease	Extent and pattern Clinical correlation
Bony integrity (erosion, expansion, dehiscence)	Skull base Lamina papyracea Optic canal Carotid canal
Skull base	Height Symmetry Slope of cribriform plate and fovea ethmoidalis Maxillary/posterior ethmoid height ratio
Maxillary sinus	Location and attachment of uncinate process to medial orbital wall Pneumatization and height (hypoplastic maxillary sinus has low orbital floor) Presence of infraorbital cells
Ethmoid sinus	Location of anterior and posterior ethmoid arteries Height of posterior ethmoid cells (determines slope of the skull base) Large sphenoethmoid (Onodi) cells and their relationship to the optic nerve
Sphenoid sinus	Location of sphenoid ostium Sphenoid septations and relation to carotid canal
Frontal sinus	Extent of pneumatization (deeper fovea ethmoidalis noted on side with a frontal cell and/or hyperpneumatized frontal sinus) Natural drainage pathway Presence of agger nasi/frontal/supraethmoid cells Anteroposterior diameter of frontal recess in sagittal section
Miscellaneous	Septal deviations and their clinical correlation Concha bullosa Abnormalities within the orbit

TABLE 28.3**INFORMATION OBTAINED FROM THE DIFFERENT PLANES OF SINUS CT**

Sinus CT Plane	Helpful Information
Coronal	Septal deviation OMC Uncinate attachment type (A,B,C) Middle turbinate variations Lamina papyracea anatomy Ethmoid anatomy Skull base anatomy/lateral lamella length Anterior ethmoid artery anatomy Sphenoid anatomy/sphenoethmoid cell (Onodi) Identify optic nerve and carotid artery dehiscence.
Sagittal	Frontal recess anatomy Slope of the skull base Sphenoid pneumatization pattern
Axial	Sphenoid anatomy/attachment of superior turbinate

- **Indications for Endoscopic Sinonasal Surgery:**

- Rhinosinusitis:
 - CRS (with or without nasal polyps) that failed to improve after maximum medical management.
 - Recurrent ARS (>4 episode per year).
 - Complications of ARS.
- Septoplasty and turbinoplasty.
- Mucoceles.
- Silent sinus syndrome.
- Intractable Epistaxis.
- Skull base and CSF leak repair
- Removal of foreign bodies.
- Sinonasal tumor Excision.
- Trans-sphenoidal approach to pituitary
- Orbital decompression.
- Dacryocystorhinotomy (DCR)
- Grave's ophthalmopathy
- Choanal Atresia repair

Box 49-2. INDICATIONS FOR PRIMARY SINONASAL SURGERY

Inflammatory Sinonasal Disease

Chronic rhinosinusitis (with or without nasal polyposis)
Recurrent acute rhinosinusitis
Complications of rhinosinusitis
Sinonasal polyps
Noninvasive fungal ball and eosinophilic fungal rhinosinusitis
Invasive fungal rhinosinusitis
Mucoceles

Other

Intractable epistaxis
Cerebrospinal fluid rhinorrhea and anterior meningoencephaloceles
Foreign body removal
Choanal atresia repair
Headaches and facial pain
Silent sinus syndrome
Sinonasal tumors
Expanded transnasal approaches to the skull base and orbit

- **Contraindications:**

- No absolute contraindication to endoscopic approach.
- The decision to use an external or an endoscopic approach depends on the exposure needed and the surgeon's training and experience.
- Lateral frontal sinus disease may not be accessible endoscopically.

- **Pre-operative medical care:**
 - Works by reducing the inflammation from purulent CRS or nasal polyposis and reactive lower airway disease.
 - Advantages:
 - Improve surgical field visualization
 - Decrease surgical bleeding
 - Decrease surgical time
 - Better postoperative results,
 - Decrease disease recurrence rate
 - Decrease need for revision.
 - Medications:
 - **Oral Steroid:**
 - Prednisone 30mg PO OD.
 - Given for 5-7 days pre-op.
 - The efficacy of perioperative steroid therapy is supported only in patients with CRS with nasal polyps (CRSwNP).
 - **Oral Antibiotics:**
 - Doxycycline (200mg on first day followed by 100 mg daily for 20 days) can be used as anti-inflammatory in patient with CRS with nasal polyps (CRSwNP).
- **Anesthesia & Position:**
 - General Anaesthesia is preferred by most of the surgeons.
 - Use of Total Intravenous Anesthesia (TIVA) significantly improve the surgical field during ESS compared with inhalational anesthesia.
 - Laryngeal mask airways (LMA) have also been substituted for endotracheal intubation to decrease the hemodynamic response to endotracheal intubation, minimize positive end expiratory pressure, and improve venous return.
 - Hypotensive medications have been used to reduce the mean arterial pressure during ESS to decrease blood flow.
 - β -blockers have been shown to be advantageous because they decrease blood pressure by lowering cardiac output rather than by lowering systemic vascular resistance.
 - A throat pack is optional if general anesthesia is used to prevent aspiration.
 - Eyes should be lubricated and taped in a vertical fashion with the medial part of the eye kept clear for observation.
 - Pre-operative dose of an antibiotic that covers common sinus pathogens should be considered, particularly in the presence of an active infection.
 - The patient is put in reverse Trendelenburg position and is rotated toward the surgeon to help to reduce blood loss and to provide a more comfortable position for the surgeon.

- **Techniques of ESS:**
 - **Anterior to Posterior (Messerklinger's technique):**
 - Begins with removal of uncinate process to expose the infundibulum and maxillary ostium, removal of the ethmoid bulla, exposure of the frontal sinus ostium, and identification of the roof of the ethmoid, removal of the remaining anterior ethmoid cells, removal of the posterior ethmoid cells, opening of the sphenoid sinus.
 - **Posterior to anterior (Wigand's technique):**
 - Begins with partial resection of the middle turbinate, opening of the posterior ethmoid cells, and removal of the anterior wall of the sphenoid sinus.
 - Once the skull base is identified within the sphenoid sinus, dissection is continued anteriorly through the posterior and anterior ethmoids.
 - The major advantage of this technique is early exposure of the skull base.
 - **Combination of these techniques is typically used in practice.**
- **Steps of a FESS for CRS:**
 1. Examination of both nasal cavities (3 Passes):

TABLE 28.1 RIGID SINONASAL ENDOSCOPY 3-PASS TECHNIQUE	
Endoscopic Pass	Structures Visualized
1st Pass Medial to the inferior turbinate (along floor of nose)	Inferior Turbinate Inferior aspect of Middle turbinate Nasopharynx Eustachian tube torus
2nd Pass Medial to the middle turbinate	Septal relationship with middle turbinate (TS1-4) Superior turbinate Sphenoethmoidal recess / sphenoid os
3rd Pass Medial to the middle turbinate (within the middle meatus)	Lateral portion of middle turbinate Accessory fontanelles

2. Application of decongestants and local anesthesia:

- Lidocaine with epinephrine 1:100,000 may be injected into lateral wall just below the axilla of middle turbinate, the inferior aspect of uncinate, and tail of middle turbinate.

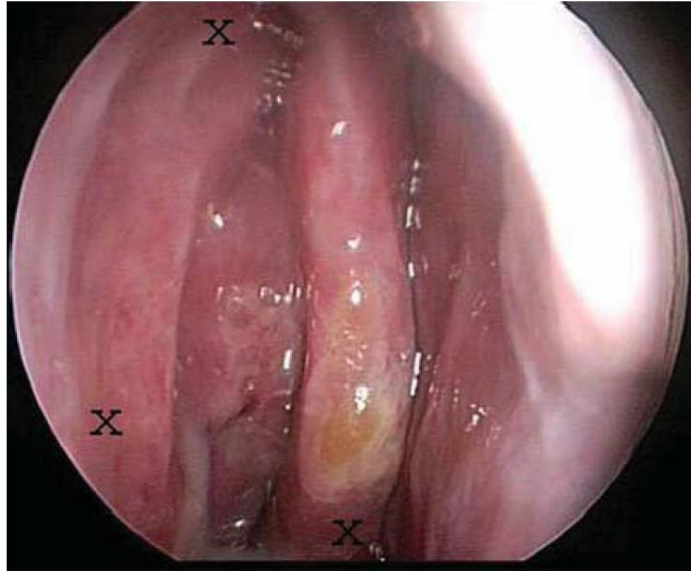


Figure 40.1 Standard three right nasal cavity injection sites.

3. Medializing the middle turbinate:

- Done gently with a Freer elevator to create better visualization of the middle meatus.
- Aggressive manipulation of the middle turbinate should be avoided, because it can lead to destabilization of the turbinate and fracture at the skull base.
- Should be done gently due to transmission of force to the lateral lamella of the cribriform plate.
- If concha bullosa is present, its lateral wall is resected with sickle knife and scissor posteriorly to the basal lamella without destabilizing the middle turbinate.
- Damage to superior medial aspect of middle turbinate can lead to olfactory impairment.

4. Retrograde Uncinectomy:

- Posterior free margin of UP is medialized with a ball probe followed by multiple bites moving forward with pediatric back-biting forceps beginning between the inferior one third and superior two thirds of UP to avoid inadvertent orbital penetration and is also likely to correspond to the natural maxillary sinus ostium location.
- Incision is continued anteriorly until the harder lacrimal bone is encountered, endoscopically seen as the anterior maxillary line.
- Penetration of this bone can damage the lacrimal duct.
- Lacrimal duct can be as close as 4 mm anterior to the natural ostium of the maxillary sinus.
- UP is removed superiorly (with shaver or up-biting ethmoid forceps) up to the axilla of middle turbinate.
- The inferior aspect is rotated medially with a ball probe and is removed with the combination of a downbiting instrument and a microdebrider.
- The inferior portion of UP is composed of medial nasal mucosa (which can be removed by shaver) and residual uncinate bone (which can be dissected and elevated from the sinus side mucosa with a double ball probe) and the lateral mucosa adjacent to the maxillary sinus (which is left in place).
- Alternately, an anterograde uncinectomy can also be performed with a sickle knife, or an elevator can be used to resect the uncinate process via a vertical incision that starts on its anterior aspect and is continued inferiorly and posteriorly along the crescent-shaped anterior margin.

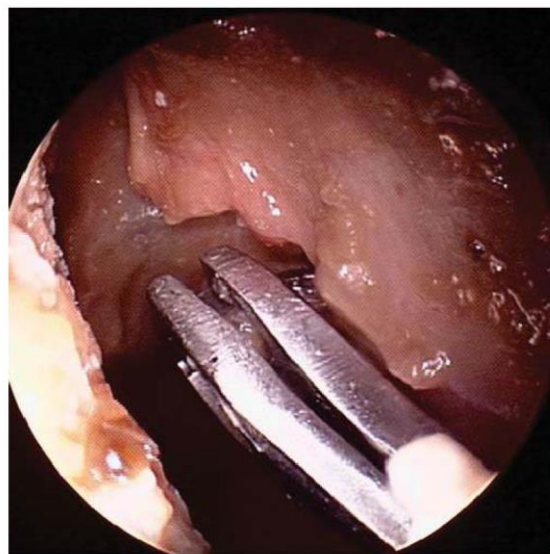


Figure 40.2 Left nasal cavity. Pediatric backbiter removing uncinate process creating the uncinete window.

5. Maxillary Antrostomy:

- Removal of UP exposes infundibulum lateral to it.
- Drainage of the maxillary sinus occurs through the natural ostium which is elliptical and opens at 45-degree angle in the floor of the infundibulum lateral to the lower third of UP.
- Maxillary sinus ostium is best visualized and manipulated with the help of an angled 30- or 45-degree endoscope.
- A ball probe passed gently into the infundibulum can identify the natural ostium and can be used to dilate it posteriorly.
- To avoid penetration of the orbit, the probe is never forced through the bony wall.
- A straight punch can then be used to open it further both posteriorly and inferiorly if necessary.
- Accessory ostia are circular and found within the posterior fontanelle.
- Opening an accessory ostium of the sinus and missing the natural ostium leads to mucus re-circulation and persistent infection.
- Larger antrostomies are preferred for patients with polyps, and the antrostomy can be widened inferiorly toward the inferior turbinate and posteriorly toward the posterior maxillary wall.
- It is important not to strip mucosa off, so as to prevent scarring and mucociliary dysfunction.
- Once the maxillary antrostomy is complete, the sinus is carefully examined with angled 30-, 45-, or 70-degree endoscopes.
- Disease in the sinus can be removed with curved endoscopic instruments, a microdebrider, or irrigation. Any pus or debris should be cultured.

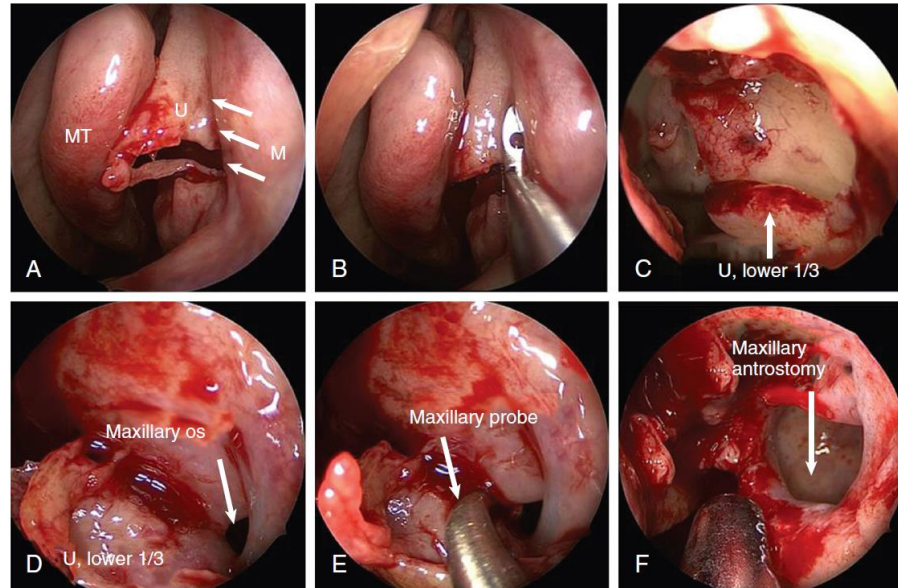


FIGURE 49-16. Steps in left-sided retrograde uncinectomy and maxillary antrostomy: Steps **A**, **B**, and **C** are done with a zero-degree endoscope view; for steps **D**, **E**, and **F**, a 30-degree angled endoscope has been used. **A**, Uncinectomy is initiated with a pediatric backbiter punch at the junction of the upper two thirds and lower one third, extending anteriorly until the anterior maxillary line (M, arrows). **B**, The upper part of the uncinate (U) is removed. **C**, View of lower one third; the natural ostium of the maxillary sinus is lateral to it. **D**, Using a 30-degree endoscope angled inferolaterally brings the elliptical natural maxillary ostium into optimal view. **E**, The natural ostium is gently dilated posteriorly with a maxillary seeker. **F**, The lower third of the uncinate is removed, and a pear-shaped antrostomy is thus created. MT, middle turbinate.

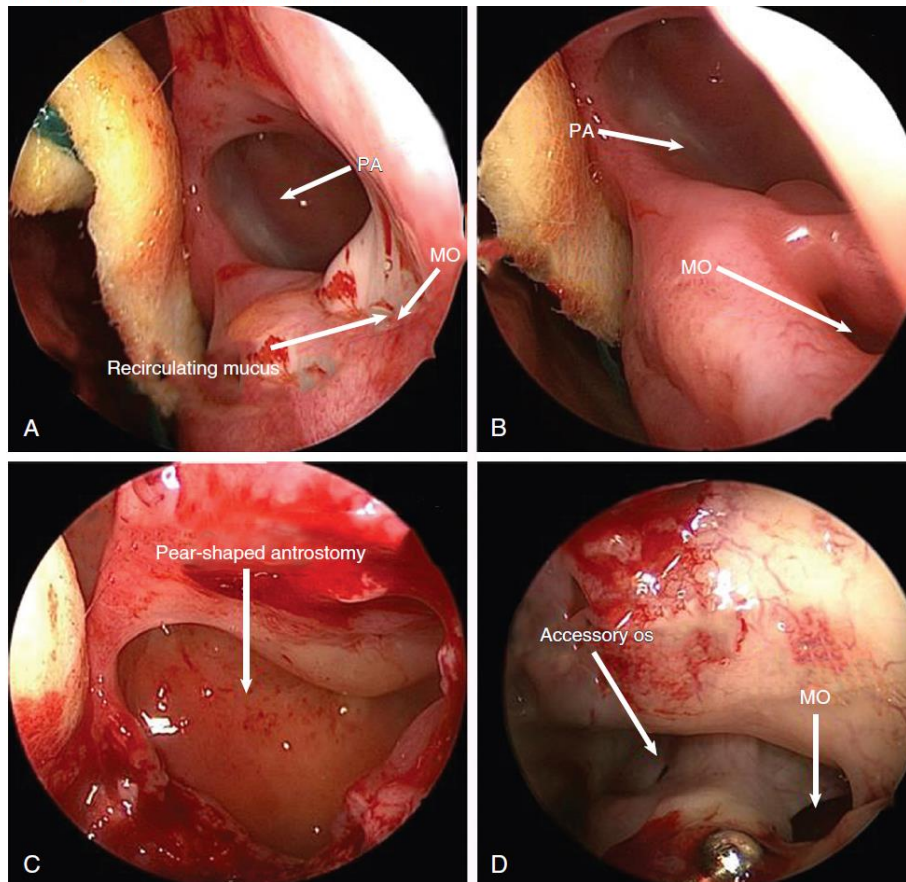


FIGURE 49-17. An antrostomy in the posterior fontanelle (PA) is not helpful, because the maxillary sinus mucociliary clearance continues to the natural ostium (MO). **A**, Mucus recirculates back into the left maxillary sinus via the posteriorly placed surgical antrostomy. **B**, Scar and tissue exists between the two openings. **C**, Once this tissue is removed, the desired pear shape of the maxillary antrostomy is verified with a 30-degree endoscope. **D**, An accessory ostium in the posterior fontanelle must similarly be incorporated into the maxillary antrostomy.

6. Anterior Ethmoidectomy:

- Ethmoid air cells are divided into anterior and posterior at the basal lamella of the middle turbinate.
- Ethmoid bulla is the largest cell of the anterior ethmoid complex and the first, most prominent cell, seen as a bulge within the middle meatus.
- If there is a retrobullar space, ethmoid bulla can be removed in retrograde fashion.
- If no retrobullar space exists, a curette, microdebrider, or a punch forceps can be used to enter the ethmoid bulla along its inferior and medial aspect.
- Next, the anterior and medial walls are removed to expose the posterior wall.
- Lamina papyracea forms the lateral wall of the bulla and is easily identified.
- Mucosa should be preserved on the lamina papyracea.
- Opening the agger nasi and suprabullar cells completes the anterior ethmoidectomy, but the opening is performed only after the skull base is identified.
- Dissection should never be carried out medial to superior vertical attachment of the middle turbinate, because such extension carries the risk of penetrating the cribriform plate and fovea ethmoidalis.
- Dissection should continue posteriorly, until the basal lamella is identified, which is the posterior limit of the anterior ethmoid.
- Microdebrider should always face superiorly, inferiorly, or medially and never laterally when working along the lamina papyracea to prevent inadvertent orbital injury.

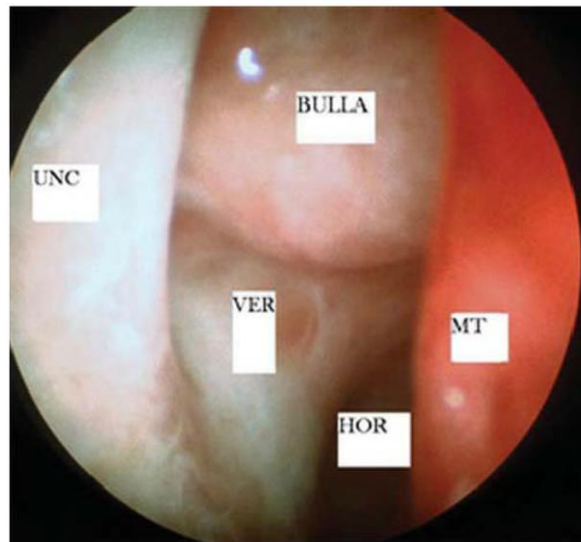


Figure 40.7 Left-sided endoscopic overview. UNC, uncinate; MT, middle turbinate; VER, vertical basal lamella; HOR, horizontal basal lamella; BULLA, ethmoid bulla.

7. Posterior Ethmoidectomy:

- To open posterior ethmoids, inferior and medial aspects of the vertical basal lamella must be removed with the microdebrider or curette.
- Boundaries of the posterior ethmoidectomy are between the lamina papyracea laterally and the superior turbinate medially.
- Care is taken not to open the horizontal basal lamella, because this will destabilize the middle turbinate and expose the vessels coming from sphenopalatine artery, causing immediate or delayed hemorrhage.
- As one proceeds in an anterior to posterior direction, the skull base slopes inferiorly several millimeters, which can lead to entry into the skull base within the posterior ethmoid air cells.
- Posterior ethmoid cells open into the superior meatus (present between the superior turbinate and the middle turbinate oblique lamellae).
- Once the superior turbinate and the superior meatus have been identified, posterior ethmoid cells can be removed from the medial aspect toward the lamina papyracea, staying low until the skull base is identified.
- Anterior face of sphenoid can be identified with help of computerized navigation system or calibrated probe.
- In an average adult, this structure is approximately 7 cm from the nasal sill at a 30-degree angle.
- Skull base can then be identified anterior and superior to the sphenoid face.
- Last posterior ethmoid cell lies superolateral to the sphenoid sinus.
- Sometimes, a large Onodi cell may be mistaken for the sphenoid sinus.
- If the floor of this cell can be visualized by a zero-degree endoscope, this is usually a posterior ethmoid cell and not the sphenoid sinus.
- Also, the natural sphenoid ostium is almost always medial to the superior turbinate attachment.
- During the initial dissection through the posterior ethmoids, septations and smaller cells found superiorly should not be explored until skull base is clearly identified.
- These are removed later, as skull base dissection is continued superiorly in a posterior-to-anterior direction.

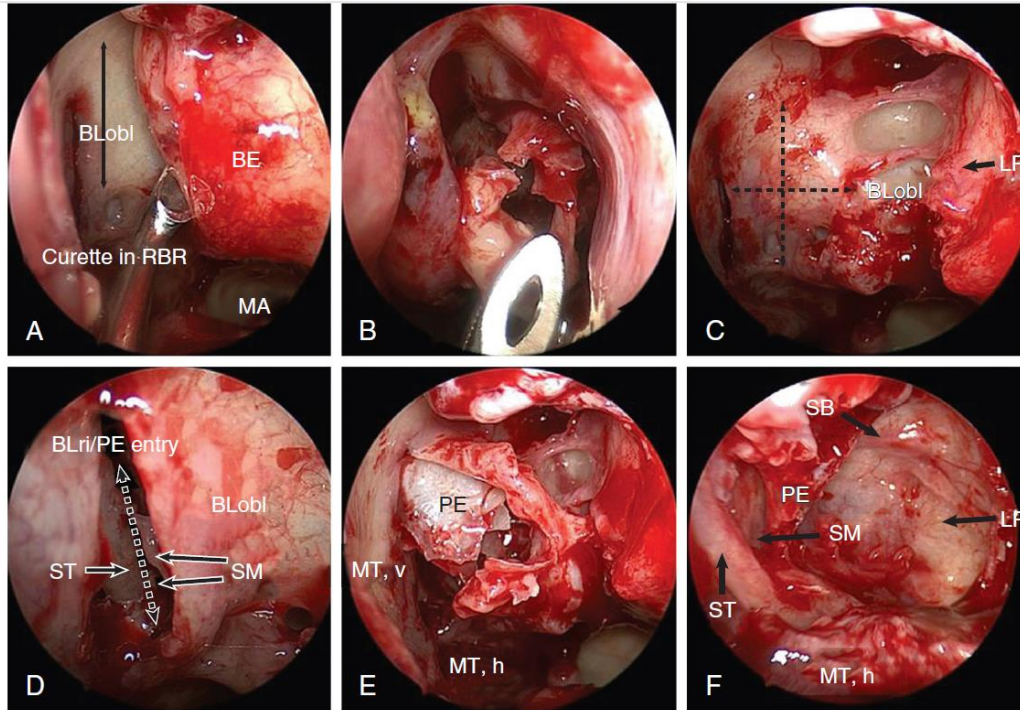


FIGURE 49-18. Left ethmoidectomy. **A** through **C**, Anterior ethmoidectomy. **D** through **F**, Posterior ethmoidectomy. **A**, The ethmoid bulla may be removed by entering the retrobulbar recess (RBR) and fracturing it forward (shown) or entering the inferomedial part of the bulla (BE) with a microdebrider or curette. **B**, The bulla is removed with a microdebrider or other instruments. **C**, This exposes the lamina papyracea (LP) in its lateral wall and the oblique part of the middle turbinate basal lamella (BLobl) posteriorly. The orbit is identified through the maxillary antrostomy. **D**, The inferomedial aspect of the vertical basal lamella is removed with a microdebrider or Freer elevator to open the posterior ethmoid. Alternatively, if a basal lamella relaxing incision (BLRi) had been performed (this corresponds to the inferomedial vertical basal lamella), this may also be used to initiate the posterior ethmoidectomy (PE). **E**, Posterior ethmoid (PE) cells are then removed via the superior meatus (SM) with a curette or microdebrider, staying low and lateral, dissecting between the lamina papyracea laterally and the superior turbinate (ST) medially. The SM lies behind the BLobl of the middle turbinate and lateral to the ST. MT,v: middle turbinate vertical aspect; MT,h: middle turbinate horizontal aspect. **F**, The skull base (SB) can be identified in the large, nonpolypoid posterior ethmoid cells. The anterior vertical (MT,v) and inferior horizontal (MT,h) attachments of the middle turbinate are carefully preserved. MA, maxillary antrostomy.

8. Sphenoid Sinusotomy:

- Ostium of the sphenoid sinus can be approached by:
 - Trans-ethmoidally:
 - Lateral to middle turbinate.
 - Done by either entering the sphenoid in the inferomedial part of the most posterior ethmoid or through the sphenoethmoidal recess after exposing the superior turbinate and superior meatus through the middle turbinate basal lamella removal.
 - Trans-nasally:
 - Medial to the middle turbinate.
 - Natural ostia is identified 1.5 cm above arch of choana between septum and superior turbinate.
- In either approach, inferior third to half of the superior turbinate may have to be removed for full visualization of the sphenoethmoidal recess and the sphenoid ostium just medially or directly posteriorly.
- Over-resection is avoided to preserve olfaction; removal of inferior third does not impact olfaction.
- Natural ostium can be located by gentle sliding a probe along sphenoid face until it easily passes and never be forced to avoid injury to skull base, optic nerve, and ICA.
- Once sphenoid ostium is identified, it should be enlarged by inserting a small curette or straight forceps, with opening directed first medially and inferiorly.
- A sphenoid punch or microdebrider can then be used to safely widen the ostium as needed.
- A circumferential opening should be avoided to prevent postoperative stenosis.
- As ostium is enlarged, small branches from posterior septal branch of sphenopalatine artery may be present medially or laterally that could cause bleeding which can be controlled by suction cautery.
- Pedicle of a potential pedicled nasoseptal flap is based on a posterior septal branch of the sphenopalatine artery.
- If a large inferior extension of ostium is needed (fungal sinusitis, polyposis, mucocoele), mucosa of inferior sphenoid face should be lifted off the bone before removing the bone to preserve the flap for future use.
- Instrumentation to remove disease posteriorly, posterolaterally, or superiorly in sphenoid sinus should be avoided unless guided by computer navigation system to avoid injury to the ICA, optic nerve, and skull base.
- Septations should not be unnecessarily removed, because they may attach posteriorly to a dehiscence or weak carotid wall.

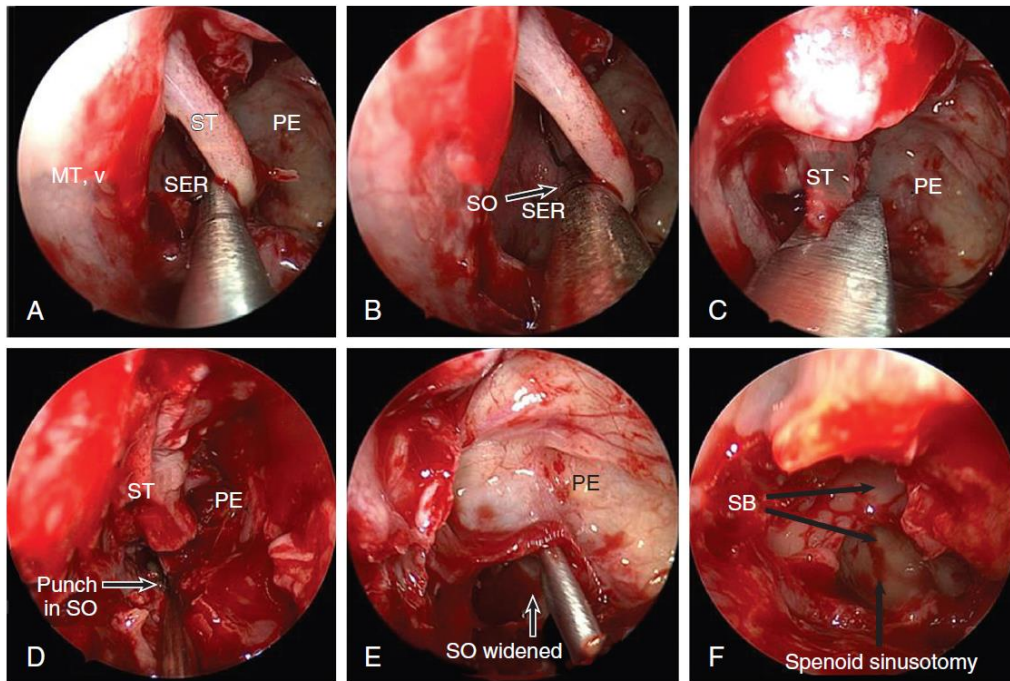


FIGURE 49-19. Left sphenoid sinusotomy. **A**, A calibrated probe or suction is used to identify the anterior wall of the sphenoid sinus, which is approximately 7 cm from the nasal sill at a 30-degree angle. The sphenoid ostium (SO) is located by gently sliding a probe along the anterior wall until it passes easily. **B**, The SO is located approximately halfway up the anterior sphenoid wall, in the sphenoid ethmoidal recess (SER), which lies between the septum medially and the superior turbinate (ST) laterally. **C**, Once the level of the SO has been determined, the inferior third to half of the ST is resected to identify the ostium. **D**, The SO is widened, first inferomedially with a curette or small punch. **E**, As needed, the SO can be widened toward the skull base (SB), lamina papyracea, septum, and sphenoid floor. **F**, The SB is identified both within the sphenoid sinus and anterior to its front face in the posterior ethmoid (PE) space. MT,v: middle turbinate, vertical aspect.

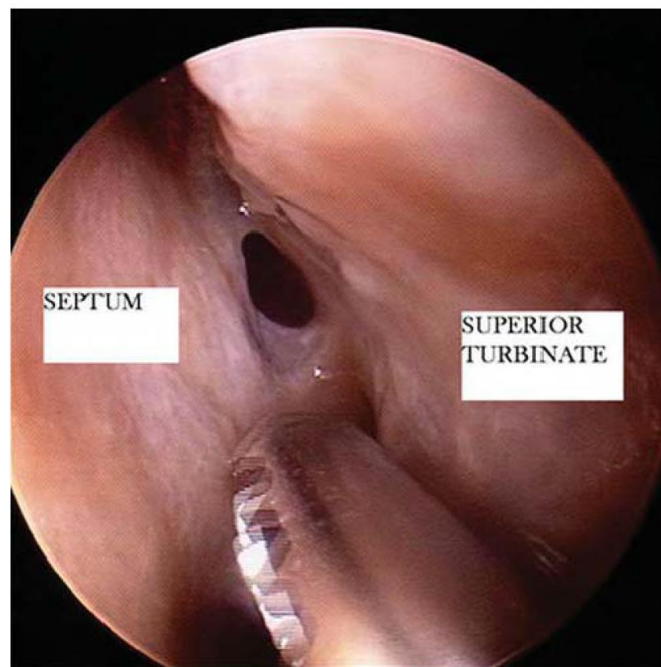


Figure 40.8 Natural os of the sphenoid sinus medial to the superior turbinate.

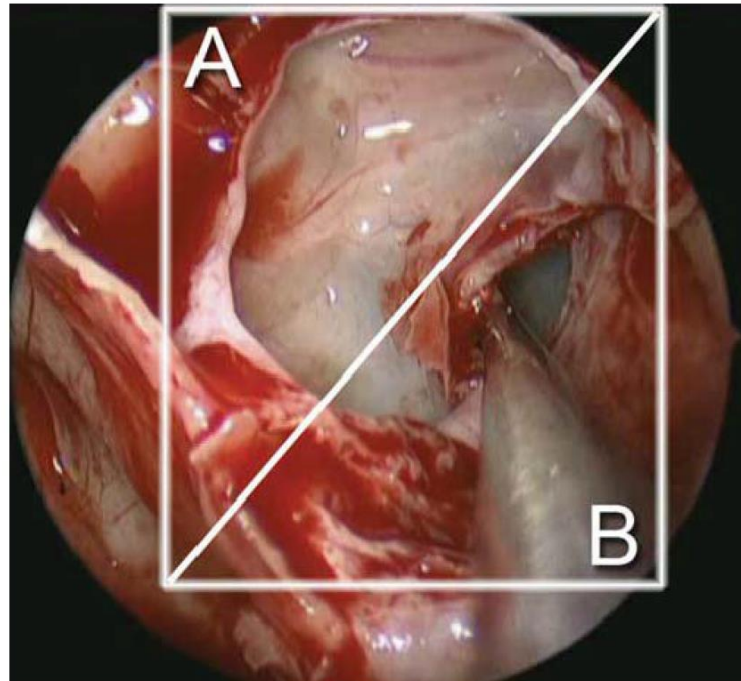


Figure 44.13 An endoscopic view of a patient undergoing primary endoscopic sinus surgery of the sphenoid sinus. Safe entry into the sphenoid sinus may be performed through the inferior and medial aspect of "Bolger's Box," which is an imagined box with the following boundaries: lateral being the *lamina papyracea*, superior being the skull base, inferior being the ground lamella, and medial being the septum and superior turbinate.

Figure 44.6 Beaded probe measurements to various areas of the nose from the nasal opening (nasal spine). (From Stankiewicz J. Complication of endoscopic sinus surgery. *Otolaryngol Clin North Am* 1989;22:749, with permission.)

Bulla ethmoidalis-5 cm

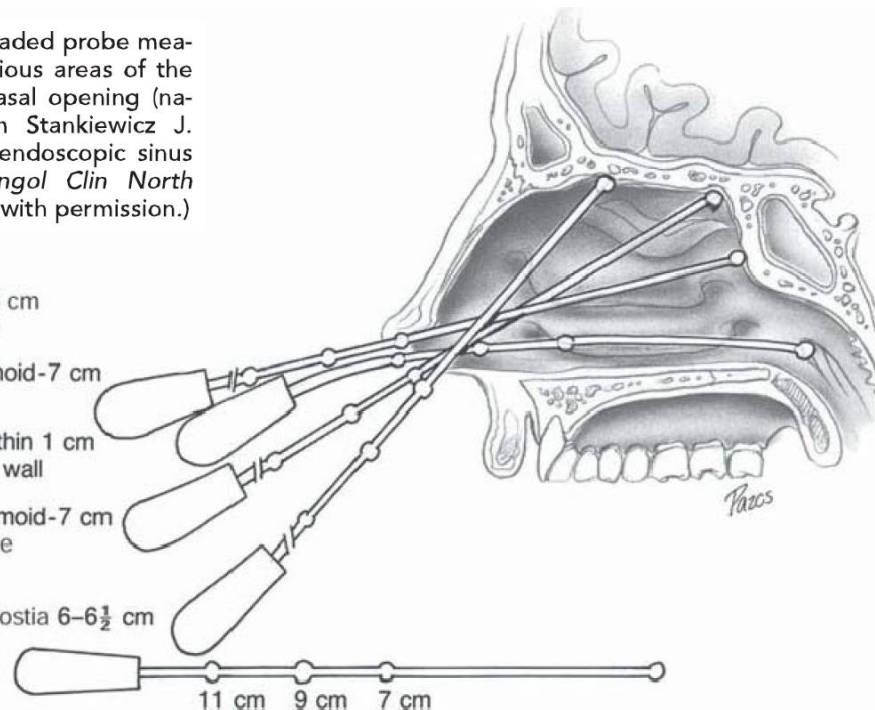
Basal lamella-6cm

Anterior wall sphenoid-7 cm

Nasopharynx $\cong$ within 1 cm
posterior sphenoid wall

Posterior ethmoid-7 cm
and skull base

Nasofrontal ostia 6-6½ cm



9. Completion of Ethmoidectomy and skull base dissection:

- Skull base can be easily visualized within and superior to sphenoid ostium or within the posterior ethmoid.
- Once orbit and skull base identified, posterior-to-anterior dissection of the superior ethmoids can be done with true cut instrument.
- This maneuver is safe, because as the skull base rises superiorly, the instrument will go up and behind the superior ethmoid projections, which are then pulled forward.
- The tip of the instrument should always be behind a partition before its removal; otherwise, the surgeon might be biting on the skull base.
- This dissection can be done with 30 or 45-degree endoscope with cutting forceps or blunt instruments such as a curette or curved suction tube.
- Dissector is never thrust upward if it appears flush with a structure; only tangential dissections are performed.
- Ethmoid is wider posteriorly as the orbit narrows, the surgeon should carefully avoid penetrating the lamina papyracea.
- Application of gentle pressure on the orbit externally during visualization of the lamina with the scope allows identification of any dehiscent areas.
- Herniation of orbital fat during this maneuver should raise concern for orbital trauma.
- As ethmoidectomy approaches the most anterior extent, suprabullar cells are opened.
- Frontal recess and agger nasi area are opened last in a complete or anterior ethmoidectomy, because bleeding from above can reduce endoscopic visualization and may hamper dissection of lower cells.
- This area is also most at risk for scarring and iatrogenic injury, and so it requires meticulous dissection.
- Superior aspect of agger nasi may lie close to the skull base, and the lateral or anterior aspect may be contiguous with the lacrimal sac and orbit.
- It is safely opened from the inferior, medial, or posterior to anterior aspect after the skull base has been identified.

10. Frontal Sinusotomy:

- In general, if the frontal sinus is not diseased, it should be left alone.
- If frontal disease is present, opening the frontal recess is warranted, with care taken to minimize mucosal injury and prevent middle turbinate lateralization.
- The risk of causing frontal sinus symptoms in a patient who has been without them should be considered.
- Computerized image guidance can be very helpful in conducting frontal sinus dissection.
- 30-, 45-, and 70-degree endoscopes may be necessary for dissection higher up in the recess.
- Frontal instruments must be available.
- If there are remnants of UP superiorly, it should be carefully removed.
- Superior attachment of UP should be determined through pre-op CT to help identify the frontal recess.
- If UP attaches to middle turbinate or to fovea ethmoidalis, frontal recess is found draining into infundibulum.
- Removal of most superior aspect of UP provides access to the frontal recess.
- When UP attaches to lamina papyracea (commonly), frontal recess is found between middle turbinate and superior extent of UP.
- Once frontal recess anatomy has been defined, a curved frontal probe may be passed into the sinus.
- Frontal "giraffe" instruments and curved curettes are utilized to perform meticulous, mucosal preserving dissection to remove cellular partitions around the recess to widen it.
- Presence of agger nasi, frontal, supraorbital, and suprabullar cells are noted, as is the drainage path of the frontal sinus.
- Dissection is performed by first opening up agger nasi cell and then sequentially removing subsequent frontal sinus cells.
- Large agger nasi, frontal, or supraorbital cell can easily be confused with the frontal recess.
- Once the frontal recess is identified, any remaining polyps and disease should be removed to provide an adequate drainage area.
- Position and stability of the middle turbinate should be examined to ensure that it will not scar into a lateral position that may obstruct the frontal recess.
- If necessary, measures should be taken to prevent middle turbinate lateralization.

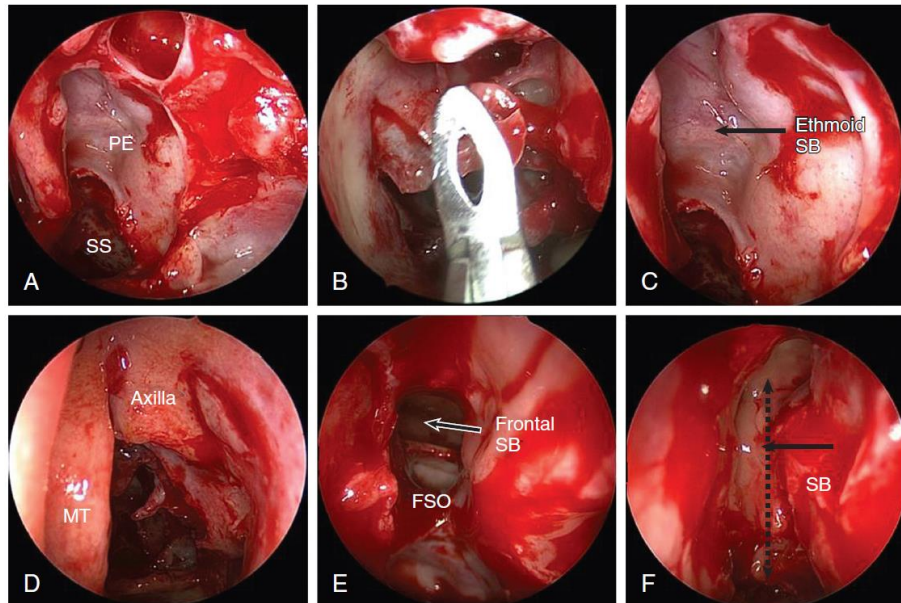


FIGURE 49-20. Completing left-sided ethmoidectomy with skull base (SB) dissection. **A**, The SB is identified in the sphenoid sinus (SS) and posterior ethmoid (PE). **B**, Remaining septations and cells of the posterior and anterior ethmoids are removed in a posterior-to-anterior direction, with the tip of the instrument always kept behind a partition before removing it. **C**, The SB is opened in the suprabullar area. **D** and **E**, Frontal sinusotomy (FSO) is performed next to identify the SB in the posterior frontal table. **F**, The ethmoid partitions are then removed in the posterior frontal recess area (solid arrow) to create a smooth transition from the posterior frontal table to the anterior ethmoid SB (dashed arrow). MT, middle turbinate.

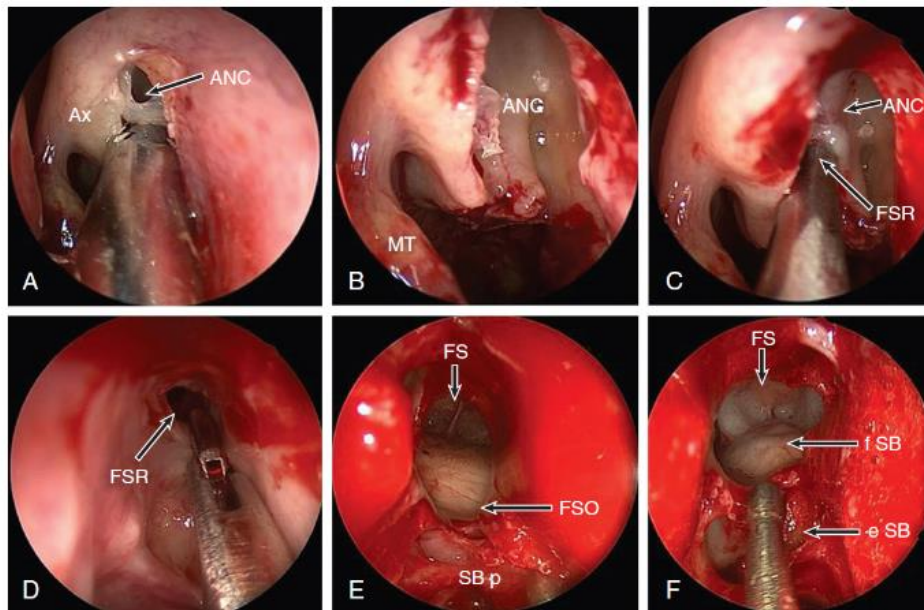


FIGURE 49-22. Basic left frontal sinusotomy carried out with a 45-degree endoscope. A thorough review of the frontal recess anatomy is necessary before undertaking any frontal sinusotomy. **A**, The axilla (Ax) of the middle turbinate is opened to identify the agger nasi cell (ANC). **B**, The ANC anterior wall is removed to identify its medial wall, posterior wall, and roof. **C**, The frontal sinus recess (FSR) is cannulated with a probe behind the posterior wall of the ANC. **D**, The posterior wall and cap of the ANC are removed to start opening into the frontal sinus. **E**, After removal of the ANC, the frontal sinus ostium (FSO) is identified as the narrowest part of the frontal sinus (FS) recess using a 70-degree endoscope. **F**, In this 70-degree endoscope view, ethmoid partitions around the posterior frontal recess are removed to create a smooth transition from the frontal (f) skull base (SB) to the ethmoid (e) SB. MT, middle turbinate.

11. Treatment of Middle turbinate:

- Vertical attachment and posterior horizontal attachments of middle turbinate should be carefully preserved to prevent its destabilization and lateralization.
- Lateralized middle turbinates cause post-op obstruction of the middle and superior meatus and block sinus drainage, resulting in persistent inflammation and infection and a need for revision surgery.
- Middle turbinate should be preserved and its partial or complete removal is considered only when disease has caused severe destabilization.
- Controlled adhesion of the middle turbinate to the septum can be promoted by debriding a small area of mucosa on medial aspect of middle turbinate and a corresponding area on the nasal septum.
- A small pack may be placed into the middle meatus to keep the middle turbinate medialized.
- Various materials, both dissolvable and non-dissolvable, are available as middle meatal spacers as should be kept for 4-7 days post-op.
- Alternatively, suture medialization can be performed, and no packing may be necessary.

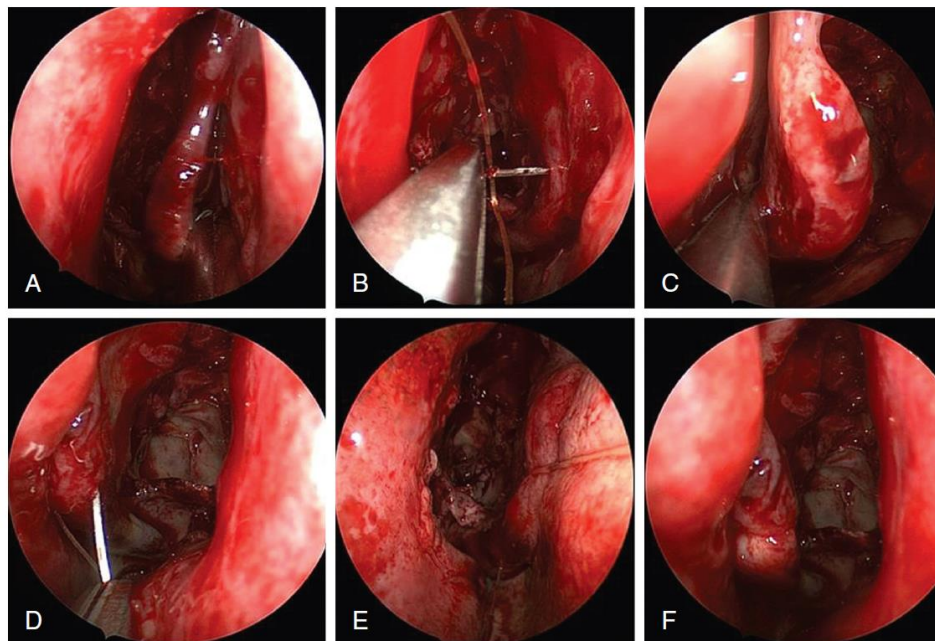


FIGURE 49-23. Suture medialization of the middle turbinate. **A**, The anterior-inferior end of the middle turbinate and the corresponding area on the septum are abraded on each side (right is shown here). **B**, Suture on a Keith needle is passed through the right middle turbinate and septum. **C**, The needle is grabbed medial to the left middle turbinate. **D**, The needle is then passed through the left middle turbinate back into the septum to create a figure-of-eight suture. **E**, The suture is then tied down to the septum. **F**, Note the medialized position of the right (**E**) and left middle turbinates at the conclusion of the procedure. No packing is necessary.

12. **Concluding the procedure:**

- Before procedure is terminated, all small bone fragments are removed, and hemostasis is achieved.
- Pack soaked in an epinephrine can be placed over bleeding areas.
- Areas of sphenopalatine, posterior septal, anterior ethmoidal, and posterior ethmoidal arteries are carefully examined.
- If these vessels have been injured, they should be cauterized.
- Anticoagulant gels and packing products are used for persistent oozing, but some can cause granulations and scarring.
- Nasal packing is not necessary for hemostasis after ESS.
- If a septoplasty has been performed, quilting sutures can substitute for the use of splints or nasal packing material.
- Inferior packing in the nasal cavity can usually be removed before the patient is discharged or in 24-72 hours, greatly improving patient comfort.

Box 49-3. BASIC STEPS IN ENDOSCOPIC SINUS SURGERY

1. Patient positioning
2. Diagnostic nasal endoscopy (Video 49-2)
3. Topical anesthetic injections (Video 49-3)
4. Medialization of the middle turbinate to expose the ostiomeatal complex (basal lamella relaxing incision is optional)
5. Uncinectomy with a zero-degree endoscope
6. Maxillary antrostomy (Video 49-4): use a 30- or 45-degree endoscope to identify the maxillary sinus ostium, identify the floor of the orbit, and then follow it up to the medial orbital wall (lamina papyracea)
7. Removal of the ethmoid bulla; identification of the lamina papyracea in its medial wall
8. Identification of the basal lamella of the middle turbinate in the horizontal and oblique segments
9. Removal of the inferomedial part of the vertical middle turbinate basal lamella to penetrate into the posterior ethmoid sinus
10. Ethmoidectomy (Video 49-5); stay low, between the superior turbinate medially and the lamina papyracea laterally
11. Identification of the sphenoid face
12. Identification of the posterior skull base, either in the posterior ethmoid (possible in the absence of polyps or in a single tier of cells) or following the sphenoid face up to the skull base
13. Clearance of skull base in a posterior to anterior direction, removing ethmoid partitions (Video 49-7)
14. Sphenoid sinusotomy (if needed) through the medial inferior triangle of the posterior ethmoid box or by identifying it in the sphenoethmoidal recess (Video 49-6)
15. Optional: Identification of the frontal recesses, frontal sinusotomy
16. Optional: Medialization of the medial middle turbinate by creating adhesion between medial surface and septum (suture or pack)
17. Optional: Placement of a middle meatal spacer

**TABLE
44.1****SUMMARY OF IMPORTANT
ANATOMIC RELATIONS**

- The *lamina papyracea* is superior to and just lateral to the natural ostium of the maxillary sinus.
- Ethmoid dissection is performed lateral to the middle turbinate, never medial or superior.
- The maxillary antrostomy is performed just superior to the inferior turbinate with the surgical instrument lying on top of the inferior turbinate.
- The maxillary antrostomy should not be more anterior than the anterior end of the middle turbinate. A backbiter is used to incise the uncinate process and create an "uncinate window." The natural ostium of the maxillary sinus is in the lower half of the infundibulum behind the uncinate process.
- The maxillary antrostomy is at the level of the inferior orbital rim.
- The frontal recess lies at 6–6.5 cm from the *limen nasi*.
- In adults, the anterior ethmoidal artery and base of the skull (posterosuperior *fovea ethmoidalis*) are 6 cm from the nasal opening or 5.5 cm from the *limen nasi*.
- In adults, the sphenoid sinus ostium is 7 cm from the nasal opening.
- In adults, the basal lamella of the middle turbinate is 6 cm from the nasal opening (posterior ethmoid bones lie behind this).
- The nasopharyngeal wall approximates the posterior sphenoid wall to within 1 cm.
- The surgeon should identify and cannulate the sphenoid ostium, if possible. The superior turbinate is the gateway to the sphenoid sinus since the ostium is just medial to the vertical midline of the superior turbinate. The ostium lies one-third of the way up the anterior wall from the choana just next to the septum.
- If the middle turbinate must be removed, the surgeon should remove only the inferior or anterior part of the turbinate and preserve the superior part as an anatomic landmark.

- **Endoscopic Septoplasty:**

- Significant septal deviations can cause lateral displacement of the middle turbinate and obstructs OMC and makes ESS technically difficult.
- If septoplasty makes the endoscopic procedure simpler, or makes postoperative endoscopy and debridement easier, it should be performed during ESS.
- It is easier to perform ESS on the side with adequate exposure and then perform a septoplasty and continue the procedure on the other side.
- Schaitkin classification to describes the endoscopic turbine-septal (TS) relationship following nasal decongesting:

1. TS-1:

- Medial and lateral aspect of middle turbinate is visualized.

2. TS-2:

- Anterior attachment of middle turbinate is partially obscured by septum.
- Require a septoplasty for access during endoscopic sinus surgery (ESS).

3. TS-3:

- Anterior attachment of middle turbinate completely obscured by septum.
- Require a septoplasty for access during endoscopic sinus surgery (ESS).

4. TS-4:

- Septum contacting the lateral nasal wall.
- Require a septoplasty for access during endoscopic sinus surgery (ESS).



Figure 28.1 Endoscopic appearance of various septal deviations. A: TS1 view, (B) TS2 view, (C) TS3 view.

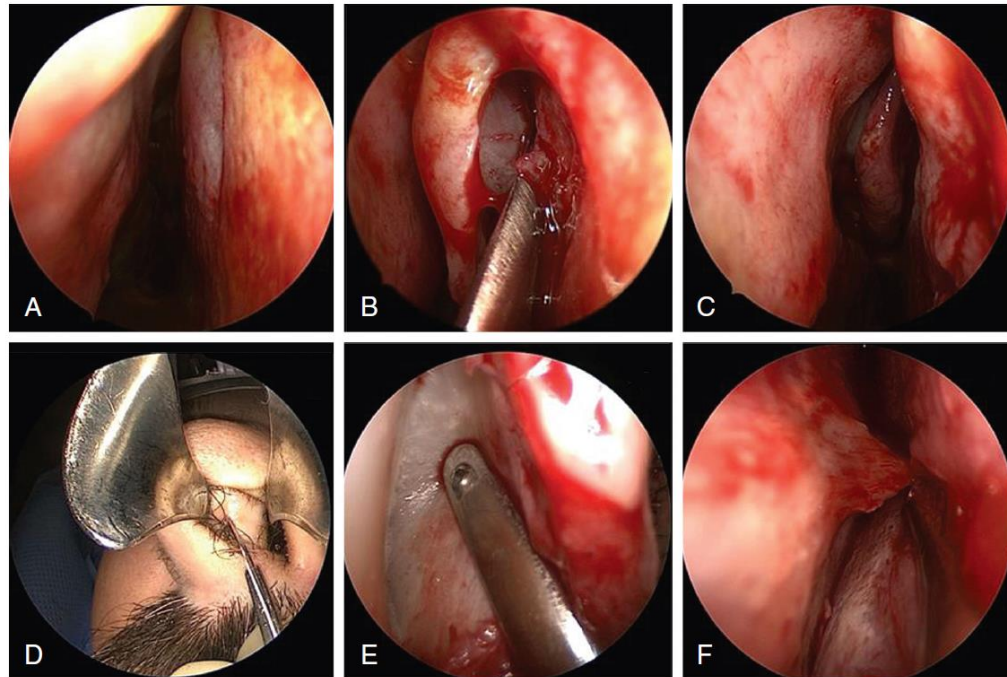


FIGURE 49-24. Endoscopic “direct” septoplasty. **A**, A mucosal incision is made just anterior to the focal, rightward septal deviation. **B**, The offending spur is removed after elevating bilateral submucoperichondrial and submucoperiosteal flaps. **C**, The mucosal flap is laid down; the middle meatus is now satisfactorily visualized for endoscopic sinus surgery to proceed to endoscopic “assisted” septoplasty. **D**, A conventional hemitransfixion is made under headlamp guidance. **E**, The endoscope and a suction Freer elevator can be used, once a 1-cm long submucoperichondrial flap has been elevated. **F**, Dissection then proceeds as in conventional septoplasty, but under endoscopic guidance.

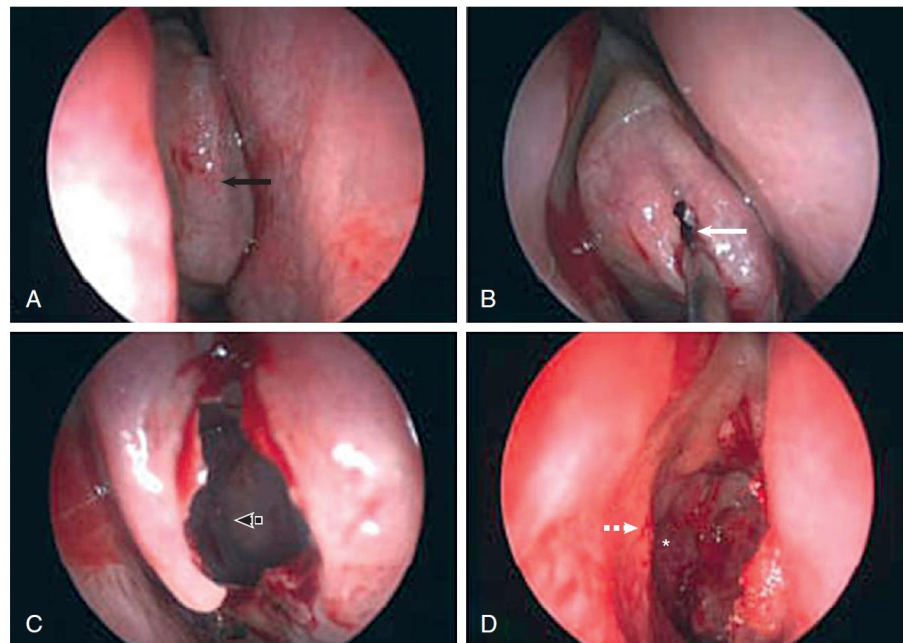


FIGURE 49-25. **A**, Right concha bullosa (arrow). **B**, A vertical incision is made with a sickle knife (arrow). **C**, Inner mucosal lining of the concha bullosa (arrow). **D**, The lateral part of the concha bullosa has been removed, exposing the uncinate process (arrow) and the bulla (asterisk).

- **Post-op Care:**

- In the recovery room, a quick assessment of vision and mental status is performed.
- Most healthy patients can be discharged home at the same day of ESS.
- Medications:
 - Saline nasal irrigation
 - Short course of nasal decongestant
 - Oral antibiotics (If nasal packing is inserted)
 - Patients with CRS may be given short course of oral steroid and nasal steroid care to reduce post-op mucosal edema and the rate of recurrence.
- Instructions:
 - Patients are advised to avoid strenuous activity, nose blowing, and any medications that may increase the risk for bleeding.
- Follow ups:
 - Post-op period is almost as critical to the success of surgery as the surgery itself to ensure that crusting and scarring are not obstructing the sinus ostia and that the middle turbinate is not lateralized.
 - First post-operative visit should be done 3-7 days after surgery:
 - Any packing is removed
 - Sinuses are examined endoscopically
 - Loose crusts and clots are removed to promote mucosal healing
 - Fixed clots or crusts should not be removed due to such removal can damage the mucosa, cause bleeding and scar, and slow healing.
 - Frequency and thoroughness of postoperative debridement is not standardized.
 - Follow-up visits, endoscopic examinations, and additional debridements are individually tailored for each patient to help achieve favorable results.
 - Patients who have undergone minimal surgery with minor bleeding may need only daily irrigations to clean debris from the sinuses.
 - More frequent debridement may be necessary for patients who had extensive surgery, polyps, AFRS, or who underwent frontal sinus surgery.
 - Other problems, such as a lateralized middle turbinate, can often be dealt with in this period, thus avoiding the need for revision surgery.

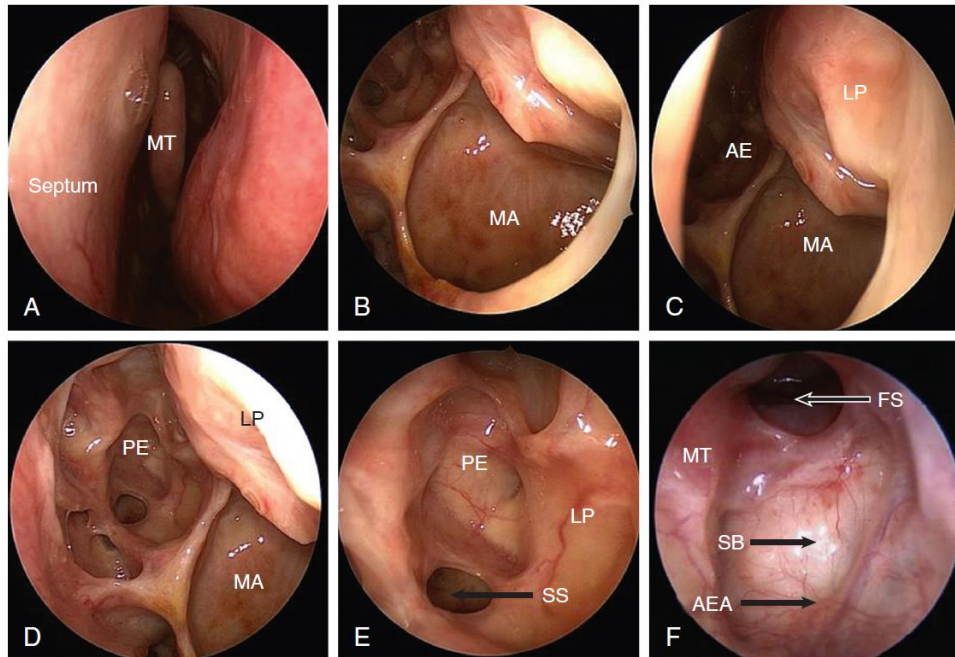


FIGURE 49-27. A well-healed left sinonasal cavity 3 months after endoscopic sinus surgery, examined with a 30-degree endoscope. **A**, Favorable medial position of the right middle turbinate (MT). **B**, Well-healed pear-shaped maxillary antrostomy (MA). **C**, Widely patent and healed anterior ethmoid (AE) cavity. **E**, Patent and healed sphenoid sinusotomy (SS). **F**, Widely patent and healed frontal sinusotomy (FS). **D**, Widely patent and well-healed left posterior ethmoid (PE) cavity. AEA, anterior ethmoid artery; LP, lamina papyracea; MA, maxillary antrostomy; MT, middle turbinate; SB, skull base.

- **Image Guidance ESS:**

- No level-1 evidence for the use of image guidance in endoscopic sinus procedures.
- In 2002, AAO-HNS released statement on indications for use of image guidance in ESS:
 1. Revision sinus surgery.
 2. Extensive sinonasal polyposis.
 3. Pathology involving frontal, posterior ethmoid, or sphenoid sinuses.
 4. Disease abutting the cranial base, orbit, optic nerve, or carotid artery.
 5. Distorted sinus anatomy of development (postoperative or traumatic) which includes surgery in absence of normal landmarks or in presence of anatomical pitfalls.
 6. CSF rhinorrhea and skull base defect.
 7. Benign and malignant sinonasal neoplasms.

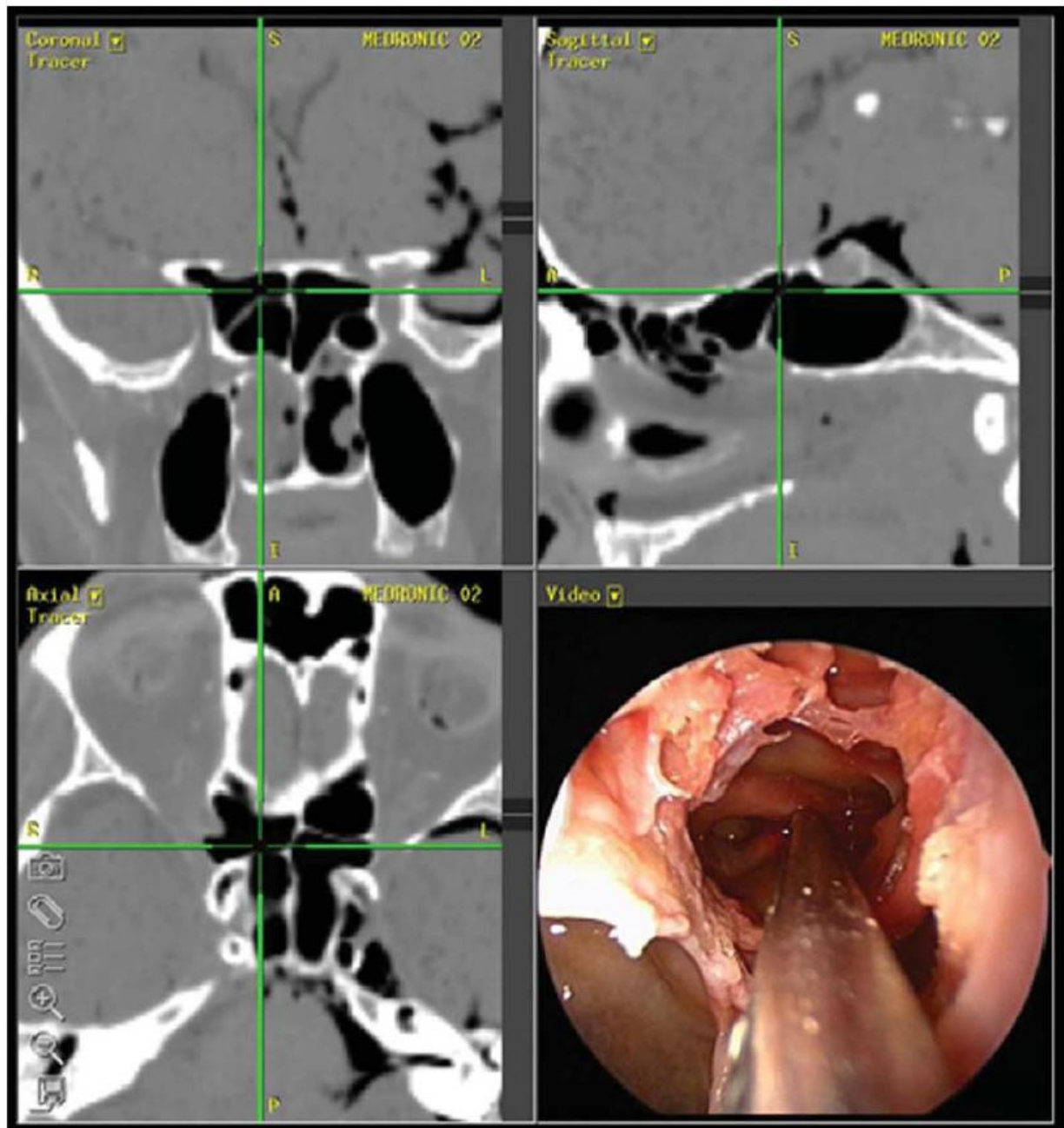


Figure 44.2 Triplanar images of an endoscopic dissection performed on a cadaver. The probe is pointing to the location of the opticocarotid recess where the carotid artery and the optic nerve are prominent within an Onodi (sphenothmoid) cell.

- **Balloon Sinuplasty:**

- Approved by FDA in 2005 for the use in the paranasal sinuses.
- Principle of this technique is to provide a minimally invasive option to open the ostia of paranasal sinuses with avoidance of significant mucosal trauma.
- Can be used on maxillary, frontal, and sphenoid sinuses.
- If combined with ethmoidectomy performed by traditional means the procedure is termed "Balloon assisted/hybrid".
- The procedure involves insertion of a balloon dilation catheter over a guidewire that has been positioned within the involved paranasal sinus under endoscopic visualization with or without fluoroscopy or a light-assisted guidewire.
- Inflation of the balloon results in dilation of the targeted ostium.
- This procedure may facilitate the insertion of drug-eluting stents in selected patients.
- Indication for sinus balloon sinusotomy:
 - Mild to moderate disease in maxillary, ethmoid, or sphenoid sinuses.
 - Severe CRS disease (CRSwNP or AFRS) is controversial.
- Contraindications:
 - Dehiscence in skull base and orbit is considered a relative contraindications because of the likelihood of dural, intracranial or orbital injury.
- Results:
 - Efficacy and utility of balloon sinuplasty is still unclear and prospective randomized trials are needed.
 - Complication rates from balloon procedures are
 - noted by authors to be similar to ESS, but long-term data are not yet available.

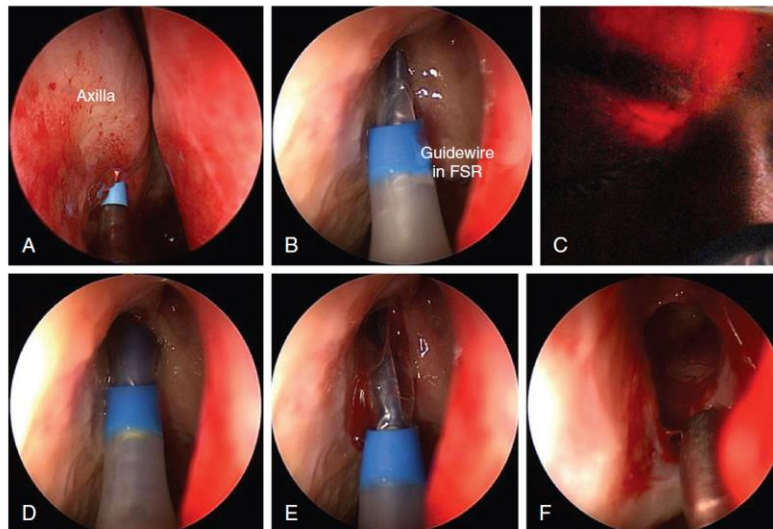


FIGURE 49-26. Balloon-assisted right frontal sinusotomy. **A**, Ethmoidectomy has been performed, and the agger nasi cell has been opened. **B**, The guidewire is used as a probe within the frontal sinus recess (FSR). **C**, After the guidewire slides into the frontal sinus, transillumination is confirmed by viewing the forehead. A sharp, bright spot of light from the illuminated catheter, rather than a diffuse dull light, should be noted. This confirms that the probe is in the frontal sinus and not in the agger nasi or an adjacent frontal recess cell. Alternatively, fluoroscopy may be used. **D**, Once the position of the probe inside the frontal sinus is confirmed, the balloon is inflated to a predetermined pressure for 10 to 12 seconds. **E**, The balloon is then deflated, and the device is removed. **F**, Endoscopic visualization is then used to verify the dilation of the frontal sinus tract. A longer balloon or sequential dilation with shorter balloons may be necessary. Residual bone fragments are carefully removed at this time, as with conventional surgery.



Step 1. A balloon catheter is inserted into the inflamed sinus.



Step 2. The balloon is inflated to expand the sinus opening.



Step 3. Saline is sprayed into the inflamed sinus to flush out the pus and mucus.



Step 4. The system is removed, leaving the sinuses open.

- **Outcome of FESS in CRS:**

- FESS carries a high success rate when combined with appropriate medical therapy and postoperative care.
- Results of FESS are difficult to compare because of variations in disease severity, procedures and techniques used, and length of patient follow-up.
- Overall success in relieving sinus symptoms is 98% at early follow-up and 91% at 4-year follow-up.
- Most series report success rate greater than 80%.
- The basis for successful FESS revolves around proper patient selection, comprehensive medical management, and good surgical technique.

- **Revision Surgery in CRS:**

- The first surgery represents the patient's greatest chance for long-term success.
- Review of the causes of failure is critical in reducing the number of patients who remain symptomatic postoperatively.
- Revision sinus surgery requires forethought and extensive experience.
- Causes of failure of primary sinus surgery:
 1. Poor patient selection
 2. Medical factors:
 - Persistent microbial disease.
 - Persistent inflammatory disease.
 - Systemic comorbidities.
 3. Environmental factors:
 - Smoking.
 - Lower socioeconomic status.
 4. Surgical factors:
 - Incomplete surgery.

- **Poor patient selection:**

- Non-sinogenic etiologies must be considered in patients with main symptoms of headache and facial pain in the absence of clinical or radiological findings of mucosal inflammation.
 - Multiple studies have shown that majority of patients diagnosed with CRS but with normal CT actually meet the criteria for migraine headache.
 - These patients will respond to medical therapy and surgery should be avoided because instrumentation of the mucosa in underlying trigeminal hypersensitivity may lead to worsening of primary headache symptoms.
- Odontogenic and TMJ etiologies must be considered in patients with main symptoms of maxillary or frontal pressure.
- Patients with postnasal drip and minimal sinus disease on exam, reflux should be kept in mind.
- Even when true CRS is present, acid reflux has been independently shown to correlate with a worse prognosis following FESS, it should be concomitantly managed.

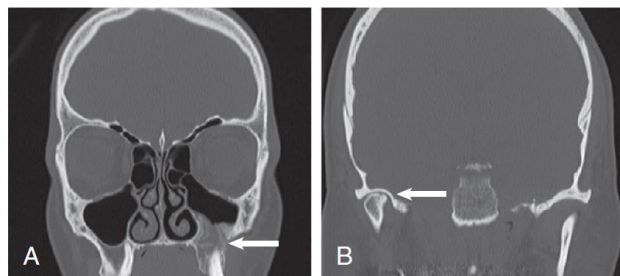


FIGURE 50-1. Incidental computed tomography findings of patients seen initially with nonsinogenic facial pain. **A**, Left odontogenic cyst (arrow). **B**, Degenerative changes of the right temporomandibular joint (arrow).

- **Persistent microbial disease:**
 - In patients who have persistent mucopurulence despite FESS, endoscopic-guided culture should be done to exclude the presence of antimicrobial-resistant organisms.
 - Superantigens:
 - Gram-positive *Staphylococcus aureus* are capable of the release of enterotoxins, which can function as superantigens that lead to type 2 T-helper cell polarized inflammation and multiclonal IgE production.
 - Current evidence suggests that superantigens may contribute to persistent inflammation in CRSwNP despite primary surgery.
 - Biofilms:
 - Organized communities of adherent bacteria which aggregate within an extracellular polymeric matrix.
 - Presence of biofilm-forming bacterial species has been associated with poor outcomes following primary FESS.
 - Biofilms is resistant to traditional antimicrobial therapies.
 - Multiple methods of chemical and mechanical disruption of biofilm matrix are being explored, including:
 - Regular antibiotic irrigation
 - Prolong oral antibiotics
 - Photodynamic therapy
 - Osteitis:
 - Inflammatory spread through bony haversian canals results in decrease of bone viability as a result of haversian canal obliteration.
 - Post-op success rates were diminished in patients with evidence of osteitis on CT scan.
 - Intraosseus bacterial entrapment or intraosseus inflammation lead to persistent mucosal irritation.
 - Comprehensive removal of the osteitic bone and prolonged antibiotics may be of benefit.

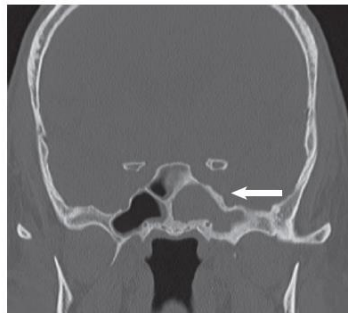


FIGURE 50-2. Severe osteitis of the left sphenoid sinus (*arrow*) with associated recalcitrant sinusitis.

- **Persistent inflammatory disease:**

- Patients with high levels of mucosal inflammation have been shown to have a higher risk for disease recurrence and a greater need for multiple surgeries.
- All CRS patients have some persistent asymptomatic inflammation post-op which needs to be managed medically to avoid revision surgery.
- CRSwNP:
 - CRSwNP clearly have higher revision rates than CRSsNP.
 - CRSwNP patients have greater evidence of residual disease on both objective and subjective measures even with more extensive primary surgery and postoperative medical therapy.
- Asthma:
 - CRSwNP patients with asthma have worse outcomes following surgery compared to CRSwNP without asthma.
 - These patients typically require aggressive concomitant medical therapy.
- Aspirin Sensitivity:
 - CRSwNP patients with aspirin sensitivity and asthma (Samter's triad) have an independent predictor of recalcitrant disease and may reduce the likelihood of primary surgical success.
 - Pathophysiology involves a reduction in prostaglandin synthesis following aspirin-mediated inhibition of the cyclooxygenase enzyme.
 - These patients have higher Lund-Mackay scores than non-sensitive patients.
 - Recurrence rates between 36-96%.
 - Post-op very aggressive medical therapy is required, including a prolonged course of low-dose oral steroids in addition to aggressive local care.
 - Aspirin desensitization is an additional consideration in the management of these patients long term.

- **Systemic Comorbidities:**

- Systemic diseases may associate with sinonasal findings.
- Some of these comorbidities represent relative contraindications to FESS and may significantly impact surgical outcomes if unrecognized or undertreated.
- Wegener granulomatosis:
 - Multiorgan disease that demonstrates granulomatous inflammation of upper and lower airways.
 - Seen between 30-50 years of age.
 - Head and neck is the presenting site in 95% of patients.
 - Nasal manifestations include purulent infections, involvement of septal and lateral nasal wall and destruction of nasal septum.
 - Positive cytoplasmic antineutrophil cytoplasmic antibody (ANCA) test and elevated erythrocyte sedimentation rate ESR are highly suggestive.
 - Not all patients will have a positive ANCA test.
 - Diagnosis is made by histologic examination.
 - Treatment is primarily directed at management of the systemic disease using cytostatic agents and steroids.
 - Instrumentation of the nose exacerbate the nasal disease.
 - Patients do poorly with both primary and revision surgery.
- Sarcoidosis:
 - Granulomatous disease that affects the lower airway.
 - 9% of patients presents with head and neck symptoms.
 - Rates of sinonasal involvement 0.7-6%.
 - Patients seen initially with crusting and mucosal edema, which progress to septal perforation and saddle nose deformity.
 - Diagnosis is be made via elevated angiotensin-converting enzyme (ACE), chest radiography with hilar lymphadenopathy.
 - Biopsy of affected mucosa showing non-caseating granulomas is very helpful in establishing the diagnosis.
 - Sinus surgery leads to worsening edema, obstruction, and synechiae formation.
 - Revision surgery should be avoided unless becomes necessary in profound nasal obstruction
 - Patients should be followed by a rheumatologist for maximal medical management prior to surgery.
 - Sarcoidosis patients with sinonasal manifestations tend to have less remission and require more long-term systemic therapies.

- Churg-Strauss syndrome:
 - Necrotizing granulomatous vasculitis.
 - Affects small- to medium-bore blood vessels.
 - Involve sinonasal mucosa in 70-75%.
 - Presence of asthma and eosinophilia are among the diagnostic criteria.
 - Prodromal phase is complicated by presence of nasal polyps and recurrent rhinosinusitis.
 - Distinguished from CRSwNP based on additional findings of extravascular eosinophilic infiltrates and mononeuropathies or polyneuropathies.
 - Surgery in Churg-Strauss syndrome does not seem to worsen disease outcome, patients will often require revision procedures.
 - Decision to undergo revision surgery should always be preceded by a consultation with a clinical immunologist for potential systemic treatment.
- Cystic fibrosis:
 - Ciliary dysmotility due to chlorine channel dysfunction with resultant mucostasis and recurrent infection.
 - Autosomal recessive
 - Should be suspected in children with CRSwNP.
 - Endoscopic medial maxillectomy has been advocated because these patients tend to fail traditional FESS approaches as a result of a lack of functional mucociliary clearance pathways.
- Primary ciliary dyskinesia:
 - Ciliary dysfunction is a hallmark of primary ciliary dyskinesia.
 - Autosomal recessive
 - Should be suspected in children with CRSwNP.
 - Diagnosed using a brush biopsy with examination of the cilia by electron microscopy.
 - Endoscopic medial maxillectomy has been advocated because these patients tend to fail traditional FESS approaches as a result of a lack of functional mucociliary clearance pathways.
- Immunodeficiency:
 - Suspected in presence of recurrent or multifocal bacterial infections despite optimal medical and surgical therapy.
 - Patients will fail to respond to primary or revision surgery, unless their underlying immunodeficiency is adequately addressed.

- **Smoking:**
 - Patients who continue to smoke are at increased risk for poor outcomes.
 - Smoking was one of the critical factors in determining whether a patient would require revision surgery over time.
 - Smoke exposure impairs mucociliary clearance, promotes epithelial inflammation, increases nasal resistance and impair wound healing.
- **Low socioeconomic status:**
 - Non-compliance to post-op medical management secondary to economic or motivational factors contributes to poor surgical results.
 - The issue of patient compliance is worsened by the fact that early persistent sinus disease following surgery is typically asymptomatic, making medication compliance an issue across all socioeconomic strata.
- **Incomplete surgery:**
 - A common cause of primary surgical failure may be due to submaximal dissection of the instrumented sinuses.
 - Although a complete “full-house” FESS is not always indicated, once the decision has been made to address a particular area, the sinus in question should be completely dissected while preserving the mucoperiosteal covering of the adjacent bone.
 - Failure to fully clear a particular region to its natural anatomic boundaries results in unresolved or iatrogenic post-op
 - Most common technical causes of surgical failure:
 1. Lateralization of middle turbinates
 2. Failure to incorporate natural maxillary ostium into the middle meatal antrostomy, resulting in recirculation.
 3. Maxillary ostium stenosis
 4. Residual air cells and adhesions in the ethmoid area.
 5. Frontal recess scarring.



FIGURE 50-4. **A,** Coronal computed tomography image of a patient following primary functional endoscopic sinus surgery (FESS) with recalcitrant rhinosinusitis in the setting of residual uncinate tissue and ethmoid partitions. **B,** Endoscopic appearance of the same patient demonstrates mucopurulence emanating from the maxillary sinus (arrow). **C,** Endoscopic appearance following revision FESS with wide maxillary antrostomy (arrow) and removal of residual ethmoid partitions.

- **Clinical approach for revision surgery:**
 - Rate of revision surgery has been reported at 10% with a 78% success rate.
 - Image guidance can be a useful adjunct but the use of anatomic landmarks and spatial relationships is still the most important component of successful revision sinus surgery.
 - Detailed analysis of endoscopic exam and preoperative imaging is essential to direct the revision procedure and to minimize the risk of a second failure.
 - During nasal endoscopy:
 - Entire sinonasal cavity should be examined using both zero-degree and angled rigid endoscopes.
 - Identify any exacerbating features such as retained UP, mucus recirculation and inferior antral windows.
 - Presence and location of inflammation, synechiae, biofilms, or mucopurulence should be noted.
 - Appropriate endoscopically directed cultures should be obtained.
 - Pre-op CT scan must be reviewed:
 - Comparison with the patient's initial imaging prior to their first surgery is helpful in delineating primary from iatrogenic disease.
 - Retained bony partitions, regions of neo-osteogenesis and drainage pathway of the involved sinuses should be assessed to determine the need for traditional versus non-anatomic techniques.
 - Continuity of orbital wall and skull base, location and continuity of bony covering of carotids and optic nerves within sphenoid sinus and presence of any dehiscence should be always checked prior to sinus surgery.
 - Anatomical landmarks that can be found at the time of revision sinus surgery and serve as a guide to progress in a safe manner:
 - 1. Anterior arch of the middle turbinate:**
 - Formed between the posterior superior aspect of lacrimal bone and the anterior superior attachment of the middle turbinate.
 - Defines the anterior border and entrance into the ethmoid air cells and is present even with removal of the middle turbinate.
 - 2. Maxillary sinus antrostomy:**
 - It is key at the time of revision surgery to ensure that the natural ostium of the maxillary sinus is patent.
 - All retained uncinate must be removed and the natural os identified.

3. Lamina papyracea:

- The easiest way to identify the lamina is to follow floor of orbit medially.
- Lamina is located just superior to the maxillary antrostomy.
- Once within ethmoid air cells, keeping laterally along the lamina will decrease the risk of penetrating the ethmoid roof, as the frontal bone cap of the ethmoid is thickest laterally.

4. The Ridge:

- Once maxillary sinus antrostomy has been opened widely, one can identify both the lamina papyracea and the ridge.
- The ridge is bony extension of orbital floor between maxillary antrostomy inferiorly and lamina superiorly.
- Benefit of this landmark is the posterior ethmoid air cells will be superior to this ridge and the sphenoid sinus inferior.
- This will play a key role in polyp patients who have undergone multiple prior procedures with dramatic changes to their native anatomy.

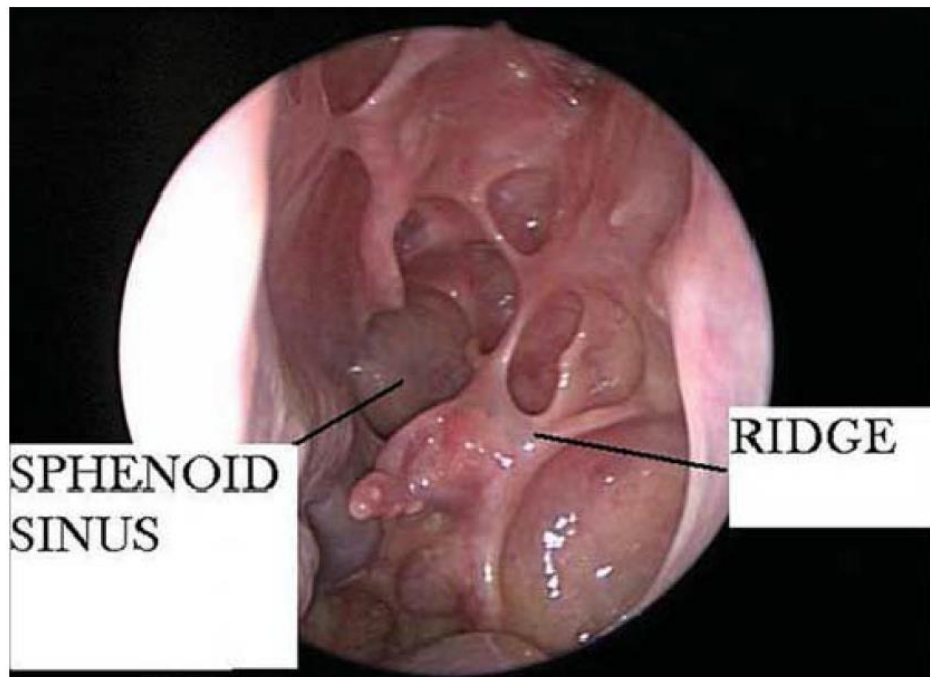


Figure 40.9 Left-sided endoscopic view after maximal endoscopic surgery showing ridge and relation of opened sphenoid sinus extending below the ridge.

5. Posterior choanal arch:

- Allows for identification of the sphenoid sinus and safe entry in scarred or difficult-to-identify sphenoid ostium.
- Mucosa over face of sphenoid can be dissected inferiorly to displace the posterior nasal branch of the sphenopalatine artery.
- 1.5 cm above the arch and near the septum, the surgeon can enter the sphenoid safely through the newly exposed bone.
- Safest point of entry into the sphenoid is medial and inferiorly.
- This opening is then enlarged laterally and superiorly to incorporate the natural ostium.
- In case of stenosis of sphenoid os, once the sinus is reopened a septal flap can be used to line the floor of the sinus at the conclusion of the revision surgery to prevent stenosis.

6. Sphenoid sinus roof:

- Ethmoid and sphenoid sinuses share a common roof.
- Once sphenoid has been entered and opened to the roof, one can proceed in a posterior to anterior direction clearing residual ethmoid air cells to the level of the skull base because the most inferior limit of the skull base has now been defined.
- This is an effective method for avoiding entrance at the skull base posteriorly.

Complications of ESS:

- **Risk factors:**

1. Advanced sinus disease with more extensive surgical approach.
2. Revision surgery.
3. Patients with anatomical abnormalities (missing anatomical landmarks).
4. Increased intraoperative bleeding.
5. Lack of conception/manual experience of the surgeon.
6. Surgical approaches from the right side (by a right handed surgeon as they tends to drift medially).

- **Classifications (According to European Rhinologic Society):**

1. Minor Complications
2. Major Complications

Localization/overall type of injury	“minor complication”	“major complication”
Orbital complication	- Orbital emphysema - Ecchymosis of the eyelid	- Orbital hematoma - Reduced visual acuity / blindness - Enophthalmos - Injury of the nasolacrimal duct
Intracranial complication	Uncomplicated CSF fistula	- CSF leak (Tension-) pneumocephalus - Encephalocele - Brain abscess - Meningitis - Intracranial (subarachnoid) hemorrhage - Direct injury of brain tissue
Bleeding	minor bleeding (stopped with nasal packing, no need for blood transfusion)	- Injury of the ant. ethmoidal artery - Injury of the sphenopalatine artery - Injury of internal carotid artery - Bleeding in need of transfusion
other	- Synechiae - Slight exacerbation of pre-existing bronchial asthma - Hyposmia - Local infection (osteitis) - Postoperative MRSA-infection - Atrophic rhinitis - Paraffinoma - Myospherulosis - Temporal irritation of the infraorbital nerve - Hypoesthesia of the lip or teeth	“Toxic shock syndrome” - Anosmia - Severe exacerbation of a pre-existing bronchial asthma or bronchospasm - Death

- **Synechia and Scarring:**

- Minor complications.
- Most common complication of endoscopic sinus surgery.
- Release of adhesions can be done in the clinic in some cases.
- Locations:

1. Between middle turbinate and lateral nasal wall:

- Occurs after ethmoidectomy.
- Scar-induced lateralization of a detached vertical lamella of middle turbinate occurs in 10-40%.
- Lateral synechia arise in the medial orbital wall in 7%.
- Affects the drainage of maxillary and frontal sinuses and considered one of the causes for revision ESS.
- Floppy turbinate can be prevented by:
 - Removal of agger nasi and lateral wall of concha bullosa to widening the meatus.
 - Intra-op controlled synechia to the nasal septum.
 - Inserting middle meatus spacer for 7-14 days.
 - Fixation of the lamella via suture to nasal septum but can lead to a further destabilization of the turbinate in some cases.
 - Partial resection of anterior 1/3 of middle turbinate if the vertical lamella fractured or destabilized during surgery.
 - Good postoperative care (nasal irrigation, topical steroids, endoscopic debridement).

2. Frontal sinus ostium:

- Frontal sinus ostium is prone to unwanted scarring, regardless of a high success rate of 75–93% in frontal sinus surgery.
- Scar-induced narrowing of the neo-ostium by at least 30% is considered normal after Draf-III frontal surgery.
- Prevented by
 - Mucosal sparing surgery.
 - Placing mucosal grafts onto exposed bone to avoid reactive ostitis with secondary thickening of bone.
 - Good postoperative care (nasal irrigation, topical steroids, endoscopic debridement).
- Treatment of frontal sinus obstruction:
 - Frontal rescue procedure (Draf 2b)

3. Maxillary sinus ostium:

- Rate of stenosis after middle meatus is 10%.
- The size of an enlarged primary maxillary ostium is not decisive for the condition of the maxillary sinus mucosa.
- Normal function occurs if ostium diameter of > 2mm.
- Prevented by
 - Non absorbable nasal packing.
 - Good postoperative care (nasal irrigation, topical steroids, endoscopic debridement).

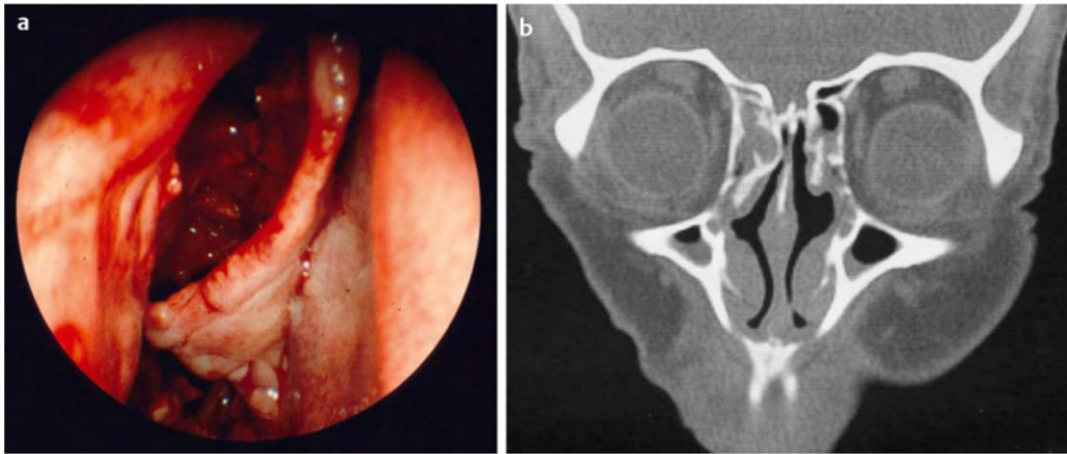


Figure 3: a. Intraoperative picture of a "floppy turbinate" (right side).
b. CT-scan of a patient having been subjected to anterior ethmoidectomy. Lateralization of the right sided vertical lamella of the middle turbinate causing inflammatory retentions in the ethmoidal cavity.

- **Paresthesia and hypesthesia:**
- Minor complications.
- 3% of all cases.
- Injury to infra-orbital nerve at the roof of maxillary sinus results in facial sensibility.
- Injury to alveolar nerves result in upper teeth sensibility.
- In most cases, these are temporary complications and resolve 3-6 months after surgery.
- **Risk factors:**
 - o Electrocautery inside the maxillary sinus.
 - o Bony dehiscence in the channel of the infraorbital nerve.

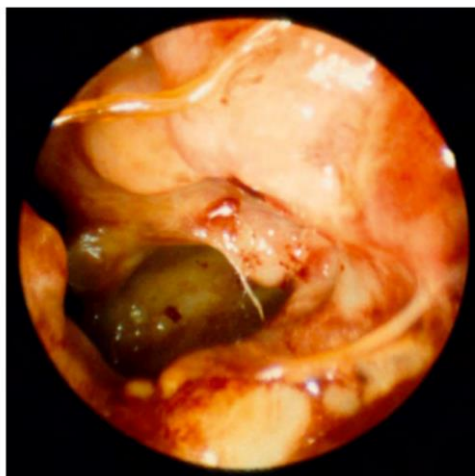


Figure 5: View of the left maxillary sinus after middle meatal anastomostomy (70° angled optical device). The inferior orbital n. reveals signs of partial destruction (small nerve ending is hanging down from the orbital roof).

- **Hyposmia:**
- Minor complication.
- 3% of all cases.
- Olfactory region is formed by:
 - o Anterior $\frac{3}{4}$ bony part of the common vertical lamella of middle and superior turbinates (conchal plate).
 - o Dorsal part of the roof of the nasal cavity and the adjoining parts of the nasal septum.
- Post-op hyposmia may occur from:
 - o Direct mechanical trauma to olfactory mucosa.
 - o Removal of olfactory mucosa accompanied by scar formation.
 - o Modification of the nasal air passage.
- Most common cause of hyposmia is disease recurrence.
- In case of recurrent nasal polyps and repeated surgeries, smell will diminish over time.
- Partial resection of lower third of anterior middle nasal turbinate does not affect the ability to smell.
- **Anosmia:**
- Major complication.
- 1% of all cases.
- One of the most frequent reasons for a legal dispute in the US.
- May result from generous resection of middle and superior turbinate.
- **Secondary Atrophic Rhinitis (Empty Nose Syndrome):**
- 0.4% of routine sinus surgery cases.
- 10% of patients underwent trans-sphenoidal approaches.
- Occurs after extensive nasal surgery:
 - o Resection of inferior, middle or superior nasal turbinate.
 - o Removal of larger areas of mucosa
- Represent 10% of the patients with secondary atrophic rhinitis.
- Develops with a latency period of several years post-op.
- Pathophysiology:
 - o Results from loss of physiological nasal functions (humidification, warming and cleansing of inhaled air) due to reduced mucosal area inducing proportional loss of the sensory, tactile and thermal receptors needed for inhaled air treatment.
- Clinical picture:
 - o Paradoxical nasal obstruction in presence of an objectively wide nasal cavity.
 - o Dry feeling in nose and pharynx
 - o Hyposmia
 - o Depression.
 - o Pain if sphenopalatine ganglion is intensively exposed towards nasal airflow after extensive tissue resection.

- **Treatment:**
 - Mainly conservative with intensive moistening, local care with the administration of ointments or oils.
 - Endonasal microplasty can be considered for severe cases to reduce nasal cavity volume by positioning an implant on the septum, floor or lateral wall.

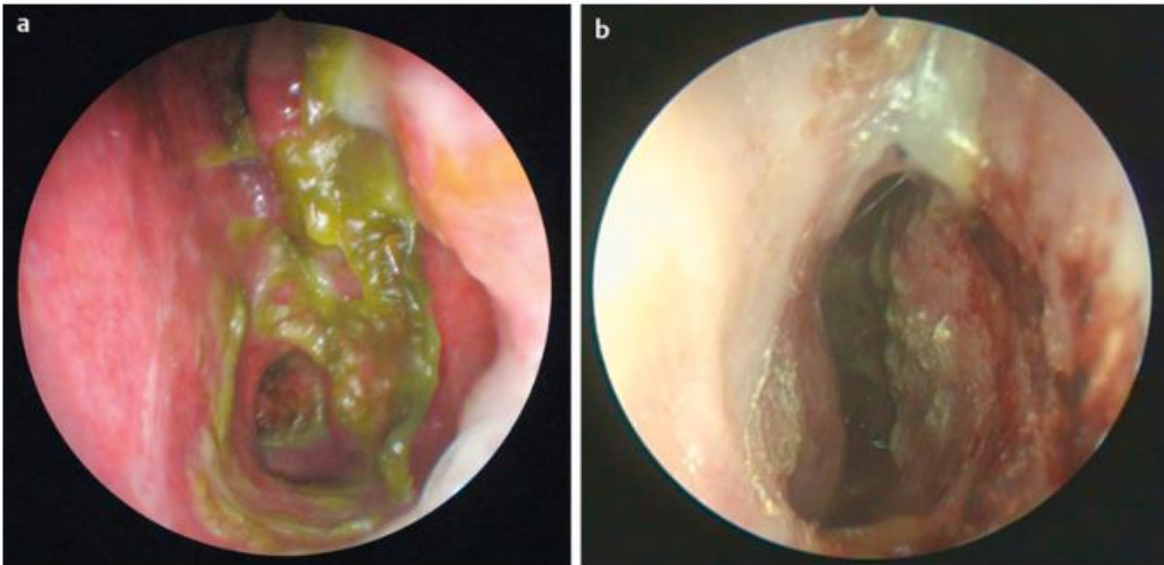


Figure 4: a. Left nasal cavity of a patient having been subjected to extensive ("radical") sinus surgery for chronic rhinosinusitis leading to "empty nose syndrome".
b. Right nasal cavity of a patient having been subjected to a rhino-neurosurgical intervention for craniopharyngeoma with application of a naso-septal flap. Irrespective to use of a "reverse flap" for amelioration of postoperative crusting, significant atrophic rhinitis is seen.

- **Injury to lamina papyracea:**
- Minor complication.
- Occurs while performing uncinectomy or maxillary antrostomy.
- Incidence is around 2%.
- If there is functional nor an aesthetic consequence for the patient the injury does not count as statistic complication.
- **Risk Factors:**
 - Less experienced surgeons
 - Right side (for Right sided surgeons) because ethmoids appear more lateral.
 - Maxillary sinus hypoplasia (4%)
 - Ethmoid hypoplasia (10%)
 - Congenital or acquired defects of medial orbital wall (0.5%)

- **Sequels:**

- **Injury to periorbita with orbital fat prolapse:**
 - Pressure to eyeball produces corresponding movements of the bulging fat.
 - To confirm it, place it into water (fat swims in water, tissue does not).
- **Pre-septal Bleeding:**
 - Small venous bleeding of the orbit beneath the skin of the eyelid (Ecchymosis).
 - No other ocular symptoms (proptosis or chemosis)
- **Orbital Emphysema:**
 - Post-op emphysema of eyelid (Mostly upper eyelid).
 - No other ocular symptoms.
 - Following:
 - Nose blowing, sneezing
 - After ventilation with mask ventilation.

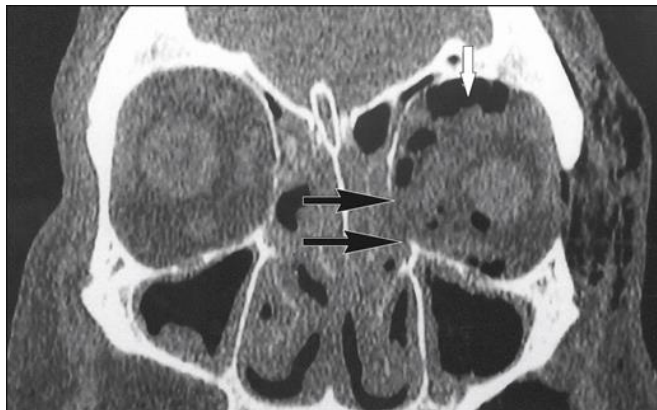


A



B

Figure 44.15 Photographic depictions of patients who sustained injuries to the *lamina papyracea*. The patient featured on the top (**A**) developed orbital emphysema after blowing his nose. The patient on the bottom (**B**) developed superior lid ecchymosis due to a bony and soft tissue injury of the orbit.



▪ **Treatment:**

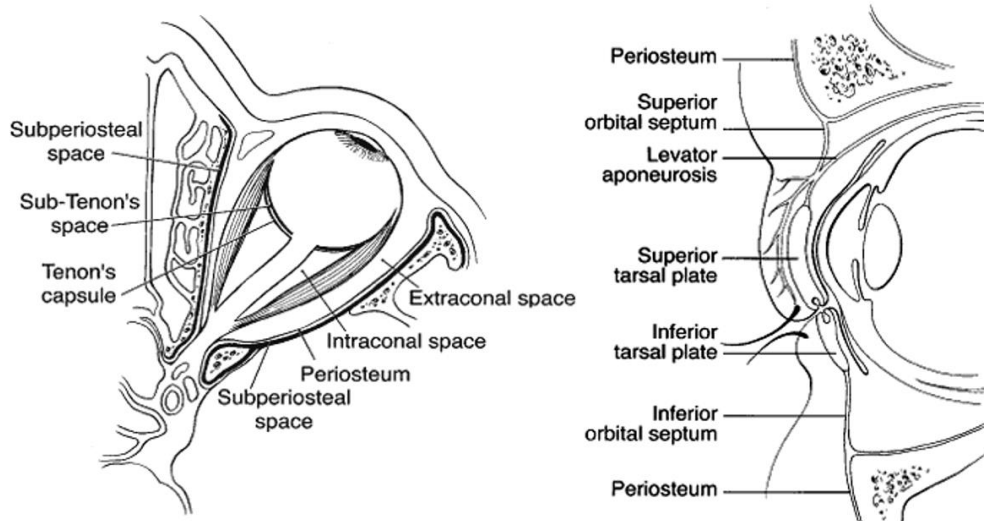
- Early recognition of the injury prevents secondary damages.
- Surgeon can complete the planned surgery.
- Avoid further intra-op trauma (suction or shaver).
- Avoid tight nasal packing.
- Antibiotics may be given in order to prevent an orbital or periorbital infection.
- Observe for vision changes, proptosis, or restricted ocular gaze.
- Ophthalmic exam is recommended, but is not mandatory in every case.
- Patient should avoid to blow his nose or undergo physical activities.
- Ecchymosis and emphysema spontaneously resorbs and improve within 7-10 days.
 - Reported case of blindness due progressive emphysema after 48 hours of the surgery with compressing to the optic nerve.

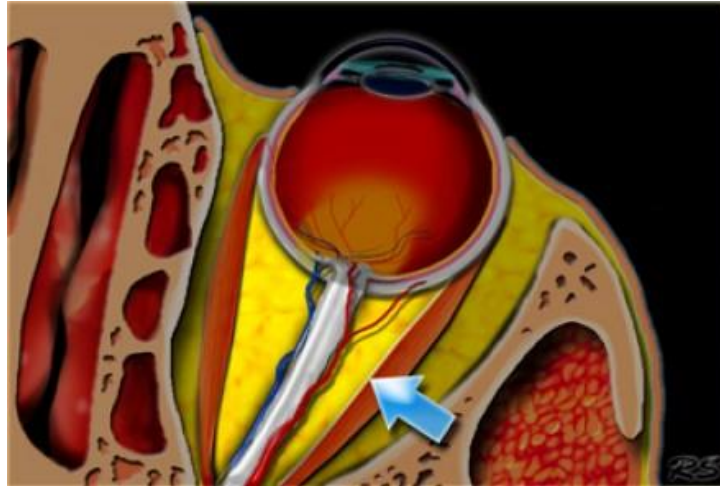
- **Orbital Hematoma:**

- Major complication.
- Post-septal injury of retrobulbar (intraconal) space.
- Incidence of orbital hematomas is **0.1%**.
- Accumulation of 5 ml of blood can lead to a dangerous intraorbital increase in pressure, causing a loss of vision.
- Orbital hematoma is a clinical diagnosis and one should not wait for a radiological confirmation.
- For the purpose of early diagnosis the patient's eyes should remain accessible during surgery for intra-op control.

- **Types:**

1. Fast (Arterial) Hematoma
2. Slow (Venous) Hematoma





- **Arterial orbital Hematoma:**
- Rapid accumulation of blood in the retrobulbar space with rapid development of symptoms.
- Most commonly from retraction injury of Anterior Ethmoid Artery (AEA) which causes increased orbital pressure that compresses vascular supply to Optic nerve.
- Appears intra-op or even with delay, e.g. in the recovery room.
- Clinical picture:
 - Rapidly progressive proptosis.
 - Chemosis.
 - Ecchymosis.
 - Disturbed ocular motility.
 - Reduced or absent pupil reaction.
 - High resistance of orbital tissue during palpation
 - High intraocular pressure reaching 30-40mm Hg (Normal range is 10-20 mm. Hg)

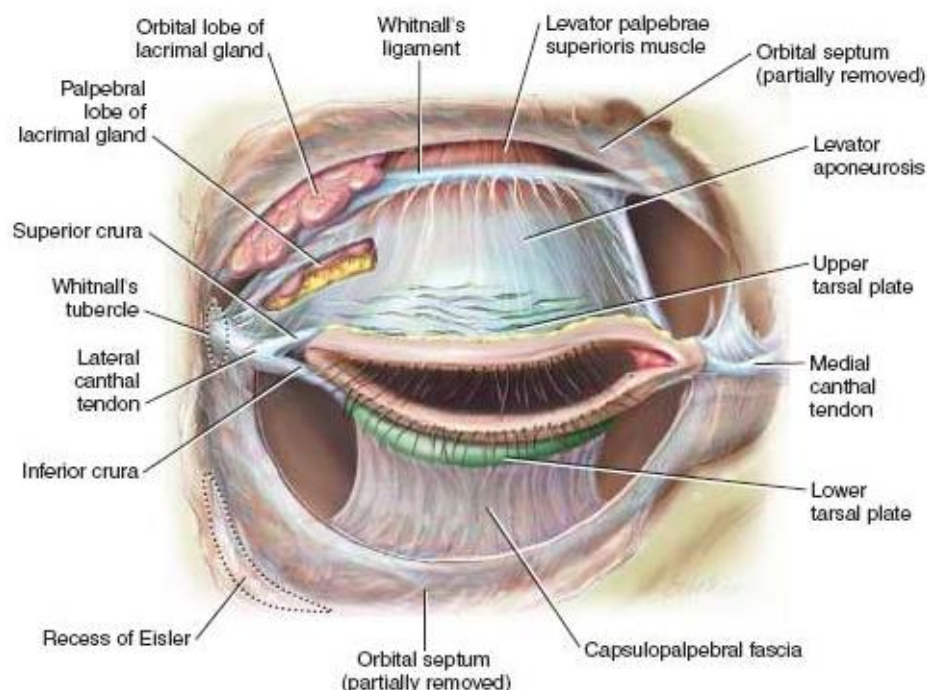


Figure 1. Patient with post-traumatic retrobulbar hemorrhage of the right eye and orbital compartment syndrome. Note the eyelids forced open, the hemorrhagic chemosis and the inability of the eye to move inferiorly while the patient is in down gaze.

- Irreversible changes in vision may occur in 30 minutes or less if there is an arterial bleed.
- High risk of blindness due to:
 - A pressure-related occlusion or a spasm of the ophthalmic or the central retinal artery.
 - Direct compression of the optic nerve.
 - Discontinuation in the immediate vascularization of the optic nerve.
- **Venous orbital hematoma:**
- Slow developing orbital hematoma from venous injury near lamina papyracea.
- Mostly observed with a delay i.e. post-op progressive exophthalmos within 24-48 hours.
- Color vision, eye mobility and visual acuity must be evaluated repeatedly.
- In cases of retrobulbar venous hematoma the retina can tolerate extreme intra-ocular pressure for 60-90 minutes.
- **Emergency management of Retrobulbar Hematoma in OR:**
 - Initial surgery is shortened or terminated.
 - Urgent ophthalmology consultation for full assessment.
 - Conservative measures:
 - Head of the bed is raised to 45 degree.
 - Cooling compresses (ice) are administered.
 - Digital ocular massage of affected eye or manual pressure is recommended which will lead to redistribution of the hematoma (contraindicated in perviously operated eye).
 - Any nasal packing should be removed to prevent pressure on the orbit.
 - Medications:
 - Mannitol 20% (1-2 g/kg over 30 minutes).
 - Dexamethasone (8-10 mg i.v q8h to 4 doses).
 - Acetazolamide (500 mg i.v).
 - Timolol eye drops (0.5 %, 1-2 drops 2x daily).
 - These conservative measures are adjunctive and do not replace an active investigation of the site of bleeding and immediate relief of intraocular pressure.

▪ Lateral Canthotomy and Cantholysis:

- Simple and every sinus surgeon should handle it.
- Emergency indication is assumed for an IOP above 40 mmHg.
- However, in daily routine the indication mainly takes place clinically, the pressure conditions can be estimated via comparative bilateral palpation.
- Surgeon should not hesitate to perform a lateral canthotomy and cantholysis as it can save a patient's vision.
- Lateral canthotomy results in a reduction of intra-ocular pressure by **15 mmHg**.
- If additional pressure needs to be relieved, a cantholysis should be performed.
- Cantholysis leads to a decompression of **20 mmHg**.
- Many authors suggest to perform canthotomy followed by inferior cantholysis only.
- Others recommend an additional superior cantholysis if canthotomy with inferior cantholysis is not effective.
- Wound of lateral canthotomy/cantholysis should be left and sutured with a delay of 2 to 5 days, e.g. with Vicryl 7 x 0.
- If further relief of pressure is still necessary, an endoscopic orbital decompression can be performed.



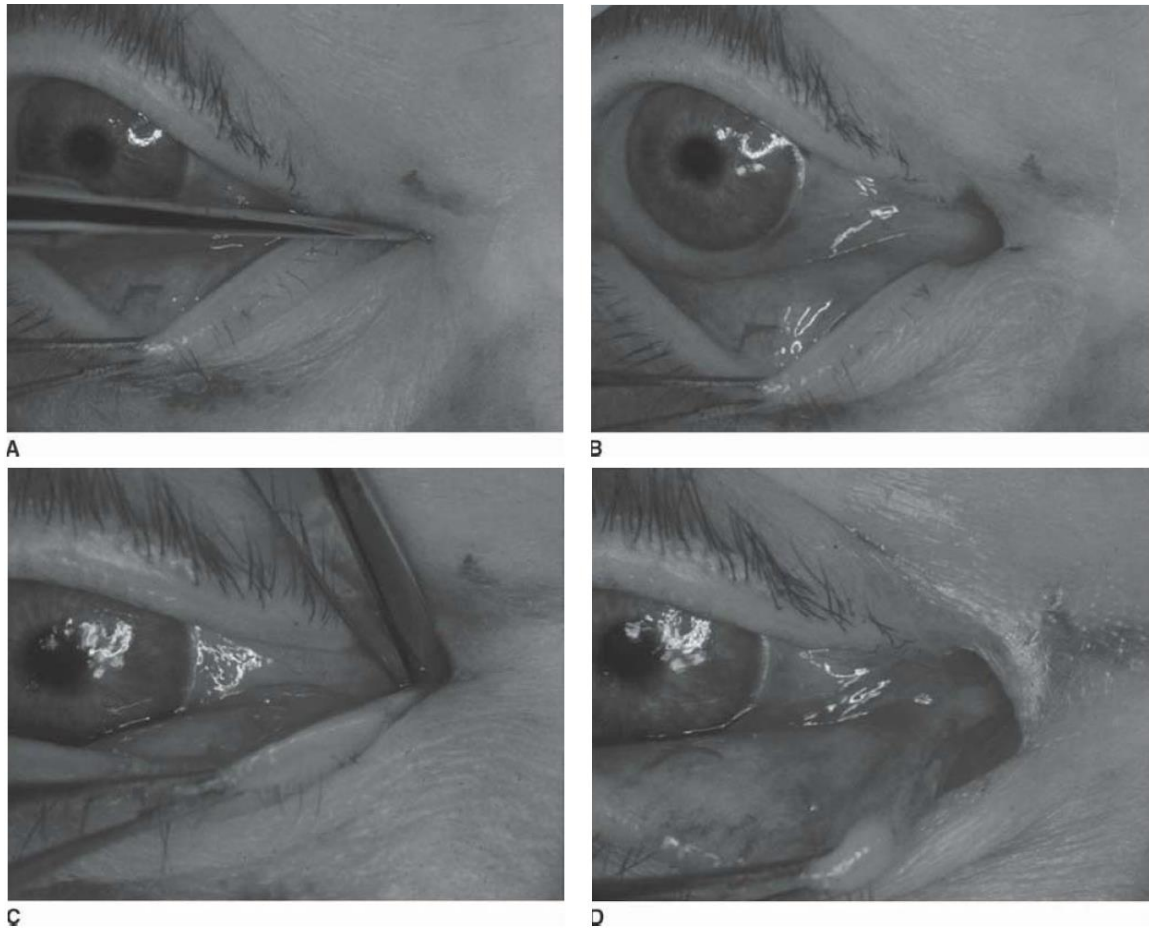


Figure 44.17 Photographic steps of a lateral canthotomy and cantholysis. Scissors or a blade is used to incise from the lateral commissure through the lateral canthus (**A**). With the lid retracted (**B**), this maneuver alone with help achieve up to 15mm Hg release. If further relief of pressure is required, the superior and inferior ligaments are divided (**C**). The result is a lateral canthotomy and cantholysis (**D**), which can relieve up to 20 mm Hg of additional pressure. This maneuver can be performed in less than 1 minute. All surgeons performing endoscopic sinus surgery should know how to perform this relatively simple procedure, as it can save a patient's vision.

<https://www.youtube.com/watch?v=MhGQ1ikN93M>

- Endoscopic orbital decompression:
 - Medial orbital wall decompression can be done by removing lamina papyracea and incising transversely the periorbital to allow prolapse of extraconal fat into the sinonasal cavity.
 - Medial orbital floor could be partially removed as well.
 - In cases of extended decompressions, postoperatively enophthalmos must be expected to occur.
 - Orbital decompression may cause intra-ocular pressure reduction of **10 mmHg**.

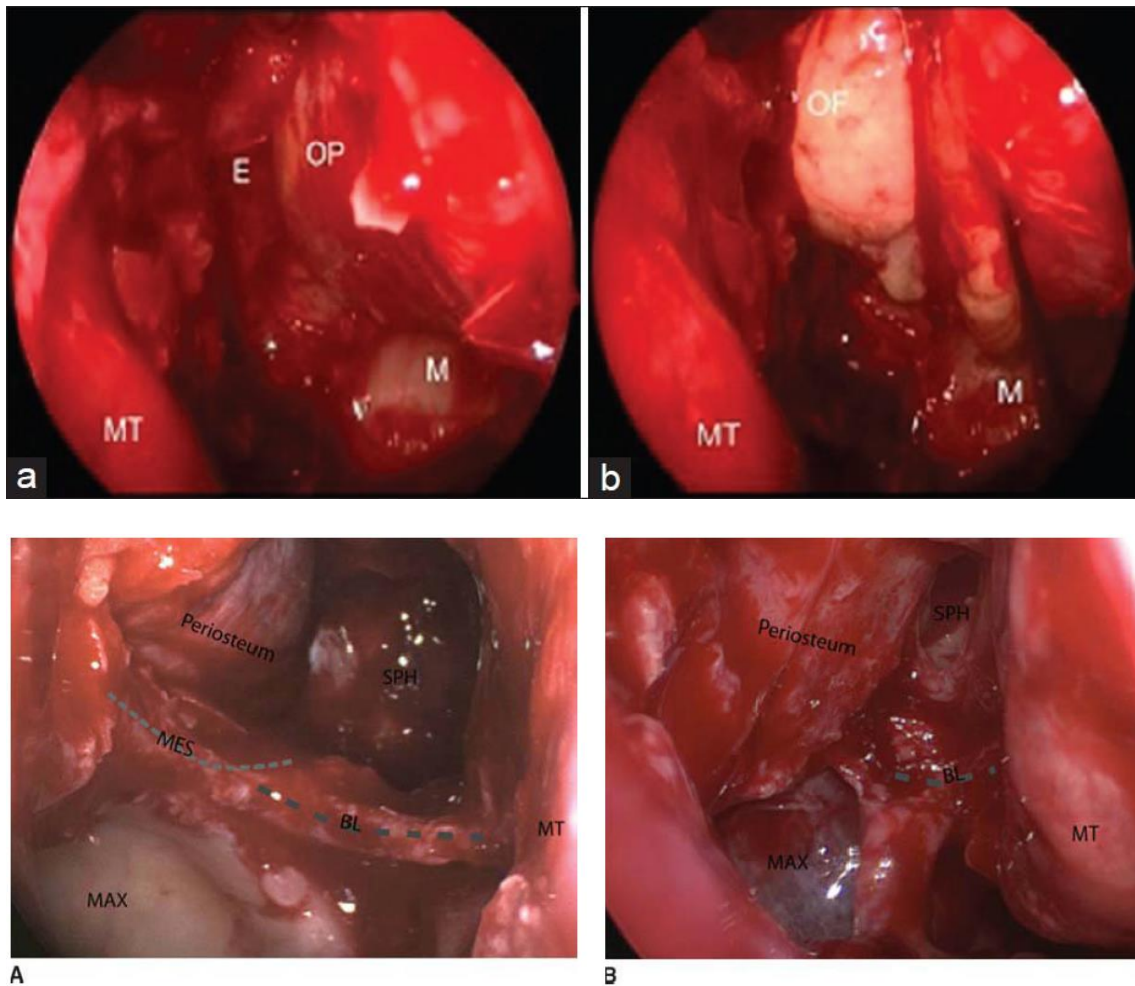


Figure 43.12 Right endoscopic view after (A) isolated medial wall and (B) medial/floor decompression. Note the increased exposure of the orbital periosteum in (B) due to removal of the maxilloethmoid suture (MES) and orbital floor lateral to V2. View includes maxillary (MAX) and sphenoid (SPH) sinuses as well as the middle turbinate (MT) and the base of the basal lamella (BL).

- Once intra-ocular pressure is relieved, the source of bleeding should be identified:
 - Monopolar cautery should be avoided due to risk of injury to neurovascular structures or extraocular muscles.
 - Endoscopic ligation with bipolar cautery can be performed for anterior or posterior ethmoidal arteries.
 - If these vessels are embedded within skull (90%), medial external (Lynch) procedure should be done.

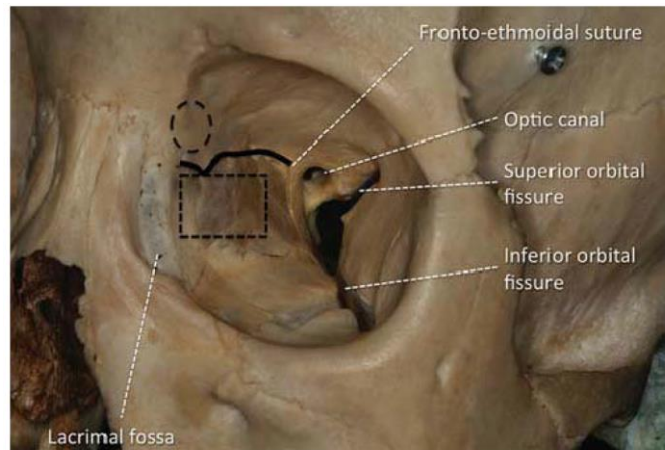
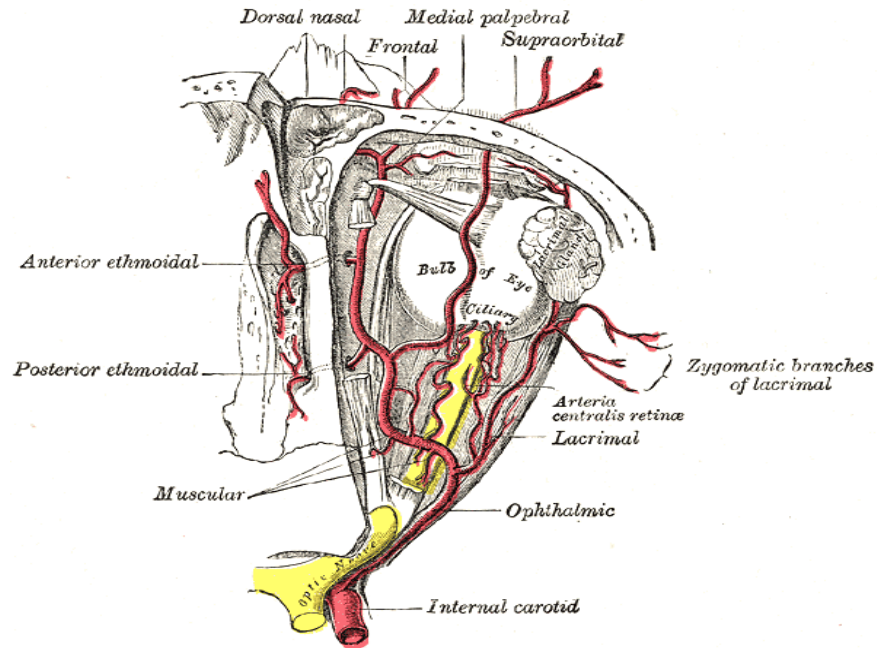


Figure 41.3 Dry skull to show the regions approached through a Lynch incision. The lateral wall of the frontal recess is exposed and the frontal sinus can be accessed (*circle*). External ethmoidectomy is performed after a wide exposure of the medial orbital wall and the ethmoid is entered through the lamina papyracea. The level of the skull base is defined by the frontoethmoidal suture line and bone is removed inferior to this line (*rectangle*).


**TABLE
44.3**
MANAGEMENT OF SUDDEN LOSS OF VISION
Immediate (fast arterial hematoma: >30 min decompression time)

1. Orbital injury	Proptosis ^a Pupillary change ^a Vision loss ^a Yes ↓	No	→	Observe
2. Medical treatment	Eye consultation Mannitol ^b Orbital massage High-dose steroid ^d No success	Success	→	Observe; bed rest; sedation; acetazolamide ^c ; ophthalmology follow-up
3. Surgical treatment	Lateral canthotomy Medial external (Lynch) Decompression Endoscopic decompression Control bleeding artery	Success	→	Observe as in medical treatment

Delayed (venous slow hematoma: 60–90 min decompression time)

Proptosis Vision loss Yes ↓	No	→	Observe
Medical treatment (same as above) No success ↓	Success	→	Observe
Surgical treatment (same as above)			

^aLocal or general anesthesia.

^bIntravenous dose of mannitol, 1–2 g/kg.

^cAcetazolamide used at discretion of ophthalmologist.

^dHigh-dose steroid works within a few minutes.

Adapted from Stankiewicz JA. Blindness and intranasal endoscopic ethmoidectomy: prevention and management. *Otolaryngol Head Neck Surg* 1999;120:841, with permission.

- **Prognosis:**
- Risk of vision loss in patients with retrobulbar hematoma is reaching 50%.
- Vision recovery takes place within a time frame of approx. 30 hours.
- Prognosis for younger patients is better.
- **Blindness:**
- Major complication.
- Catastrophic complication of sinus surgery.
- Caused by:
 - **Orbital hematoma.**
 - **Vasoconstrictive agents:**
 - Permanente.
 - Results from injection or application of adrenaline-soaked (e.g. 1:5000) packs into the nasal cavity which will cause spasm of the vessel network around the optic nerve and ischemic neuropathy.
 - **Direct injury to Optic Nerve:**
 - In the sphenoid sinus or onodi cell.
 - Posteriorly, orbit converges and permits passage of neurovascular structures through optic canal.
 - Optic canal narrows and can form indentation in sphenoid sinus at opticocarotid recess.
 - Dehiscence of optic nerve occurs in 4-8% of cases.

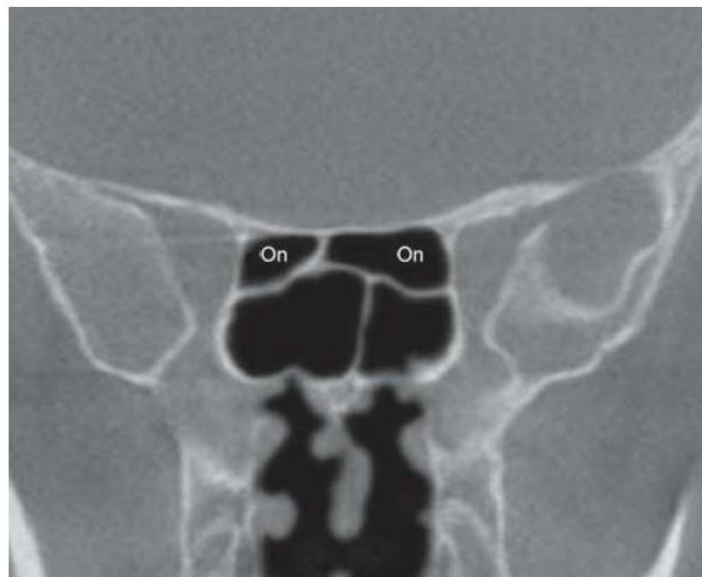


Figure 44.12 A coronal CT of the sphenoid sinus. Transverse partitions within the sphenoid sinus may indicate the presence of Onodi (sphenothmoid) cells (*asterisk*). These ethmoid cells pneumatize the sphenoid bone lateral to and superior to the sphenoid sinus. They are typically related to the optic nerve and may feature the optic nerve prominently (see Fig. 44.2).

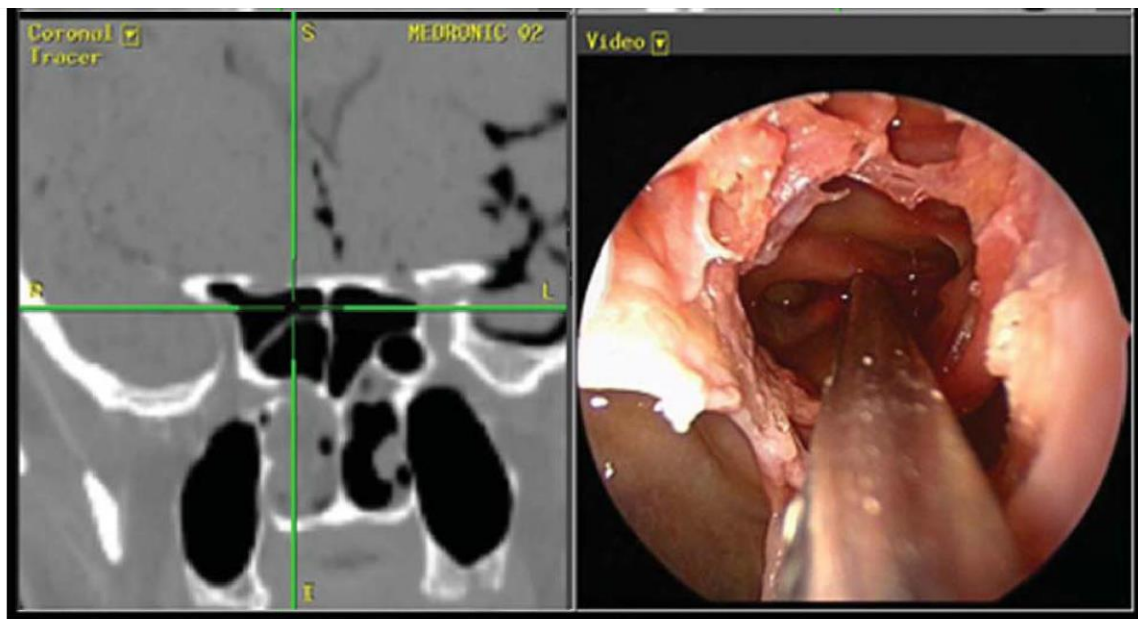
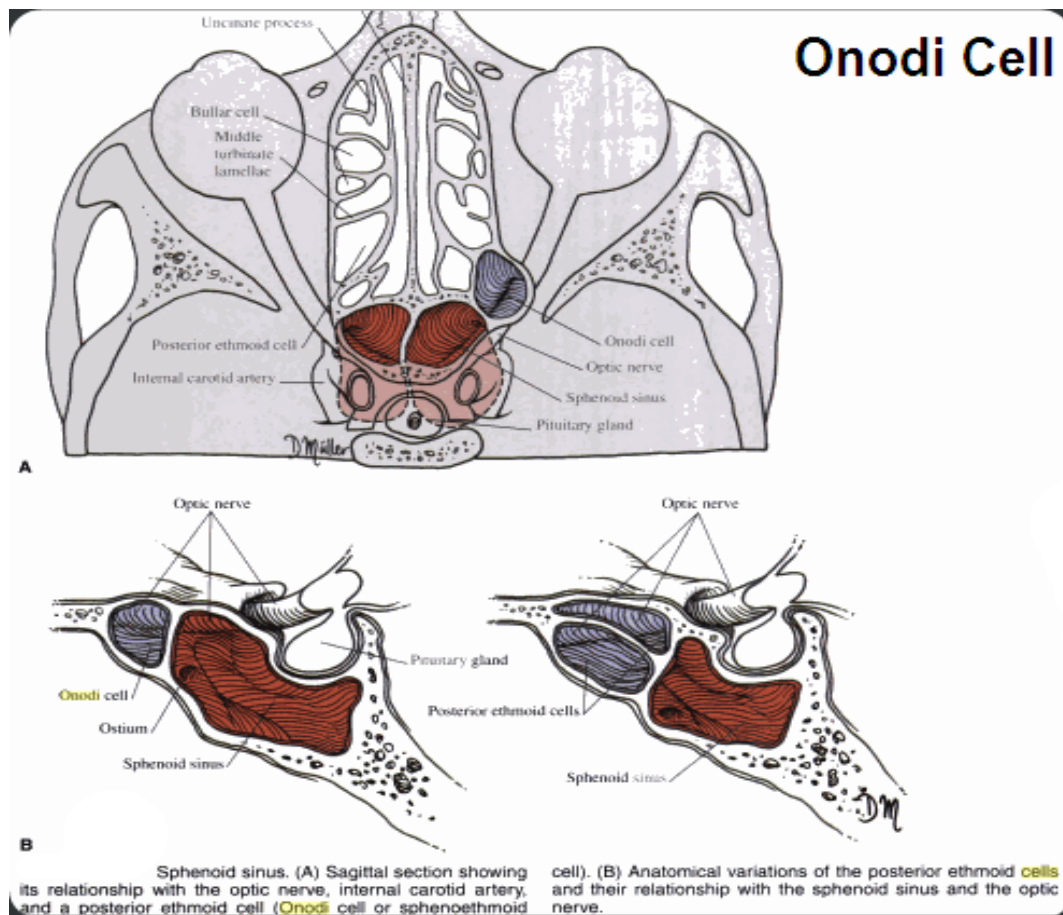


Figure 44.2 Triplanar images of an endoscopic dissection performed on a cadaver. The probe is pointing to the location of the opticocarotid recess where the carotid artery and the optic nerve are prominent within an Onodi (sphenothmoid) cell.



- **Treatment:**
 - After every post-op noticed or supposed visual reduction, an ophthalmological emergency consultation should occur.
 - Imaging (MRI) is strongly recommended.
 - If neurapraxia or a hematoma is suspected, a high dose corticosteroid treatment is administered (IV dexamethasone 0.5-1 mg/Kg).
 - After mechanical injury of the nerve, collateral damage has to be searched for in the orbital apex or at the skull base.
 - If the optic nerve is visibly cut through, there is no specific treatment.
- **Diplopia:**
- Major complication.
- Double vision caused by injury to extraocular muscles, mainly medial rectus muscle.
- Medial rectus is immediately lateral to the periorbital along the transverse midpoint of the lamina papyracea.
- Superior oblique muscle is higher in the orbit just lateral to ethmoidal roof and it may be lacerated in extended endonasal frontal sinus surgery.
- Incidence of approximately 1/1000.

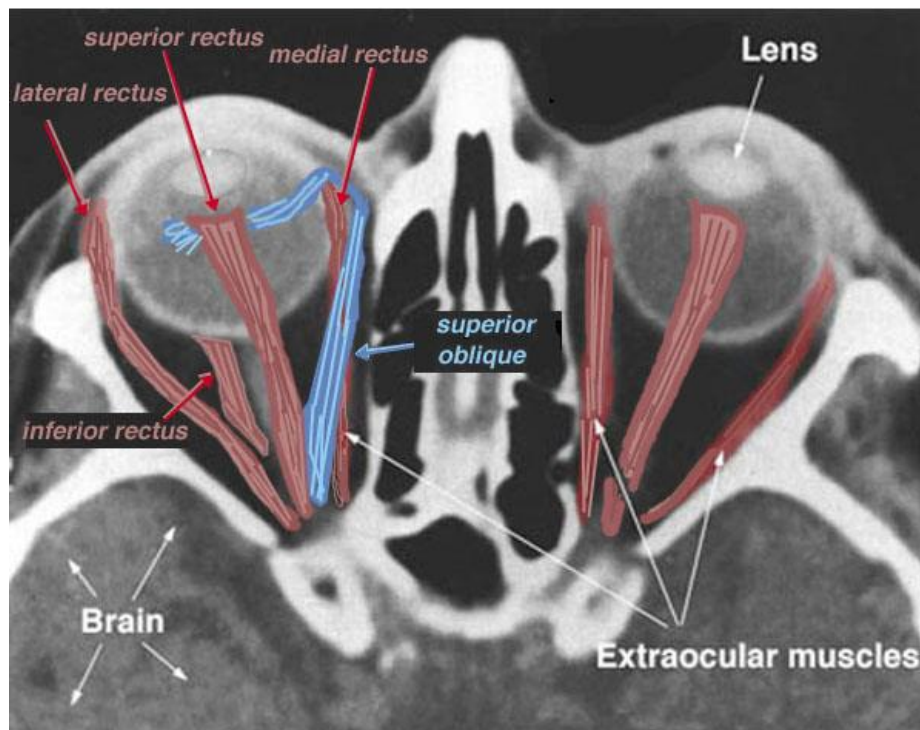
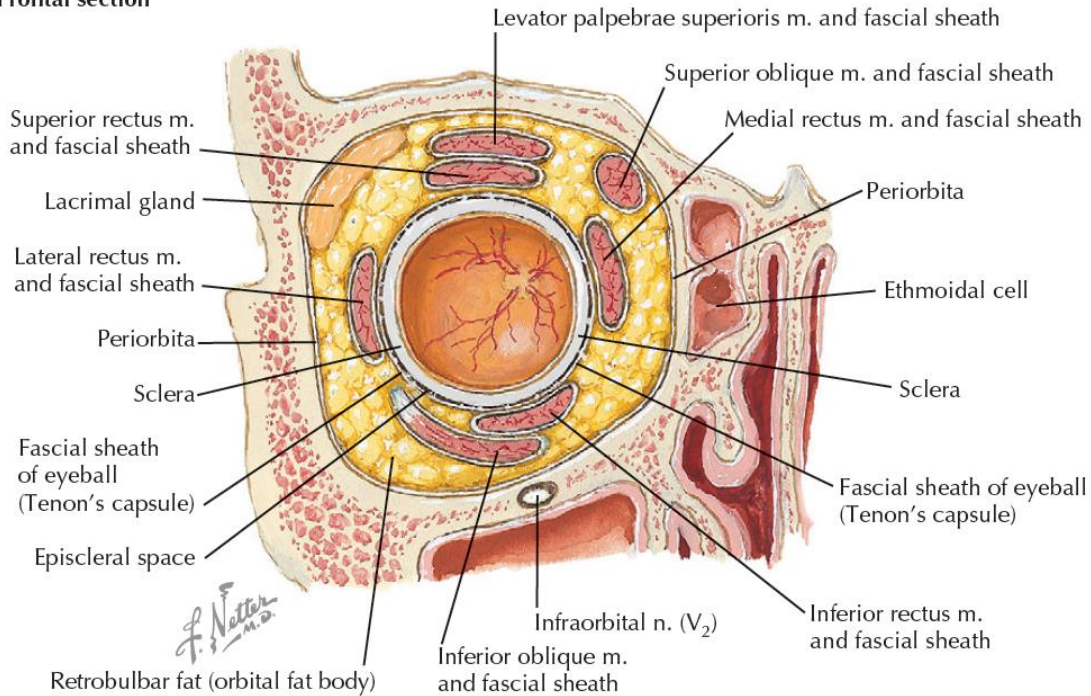


Fig. 3. CT Horizontal transverse scan at the plane of the brain, orbits and nose of the human head.

Frontal section



▪ **Causes for a post-op diplopia:**

1. Partial or complete transection of the muscle:
 - Direct injury to the muscles after penetration of the lamina papyracea with powered instrumentation.
 - Middle or posterior ethmoid is most at risk – as hardly any fat is situated between the muscle and bony orbital wall.
2. Contusion with hematoma of muscle tissue.
3. Impairment of CN-III (At the point of between middle and dorsal third of medial rectus muscle).
 - Heat conduction from a cautery or laser through a dehiscence in the lamina papyracea.
 - Bipolar cautery is advised when topical thrombotic agents are ineffective.
 - Unintentional injection of local anesthesia into the orbit can cause temporary diplopia.
4. Prolapse and incarceration of muscle parts into a defect of orbital wall.
5. Destruction of intra-orbital fascia with irregular scarring.

- **Diagnosis of post-op diplopia:**
 1. **Eye muscle damage is NOT noticed intra-op (50%).**
 2. **Symptoms appears immediately post-op.**
 - Horizontal diplopia
 - Restriction of adduction.
 3. **CT Orbit:**
 - Evaluate medial orbital wall.
 - Evaluate gross appearance of MR.
 - Rule out orbital hemorrhage.
 4. **MRI Orbit:**
 - Better for evaluation of the status of MR.



- **Management of post-op diplopia:**
 - Immediate ophthalmology consultation.
 - Early surgical intervention should be performed within 1-2 weeks, if a muscle was completely intersected or if an incarceration of tissue or a skewering of bone fragments into the muscle is suspected clinically or via imaging.
 - If the muscle is partially damaged or only affected by bruising, neural or vascular damages, it may be justified to wait for 3–12 months.
 - Overall prognosis for persistent diplopia is poor.
- **Lacrimal duct injury / Epiphora:**
 - Major complication.
 - Incidence of about 0.1-1.7 %.
 - Mostly occurs during uncinectomy (backward cutting punch) and surgery on anterior frontal recess.
 - Agger nasi cells are adjacent to lacrimal sac, whereas the ethmoid sinus and natural maxillary sinus ostia are related to the lacrimal duct.
 - True maxillary sinus ostium is less than 5 mm posterior to nasolacrimal duct.
 - Hasner valve opens lacrimal apparatus into inferior meatus within 1 cm of the anterior end of the inferior turbinate or pyriform.
 - As a rule, Ethmoid sinuses or maxillary ostia should not be opened anterior to anterior end of middle turbinate or into hardened bone separating the maxillary os from the nasolacrimal duct.
 - During the course of a routine sinus operation, frequently parts of lacrimal bone or frontal process of maxilla are removed in an undirected manner, without any direct malfunctions resulting.
 - May result in iatrogenic lacrimal duct stenosis with epiphora appearing directly post-op or with a delay of 2-3 weeks.

▪ **Management:**

- Patients should be observed initially because most cases of epiphora resolve spontaneously.
- Dacryocystorhinostomy (DCR) if no improvement.

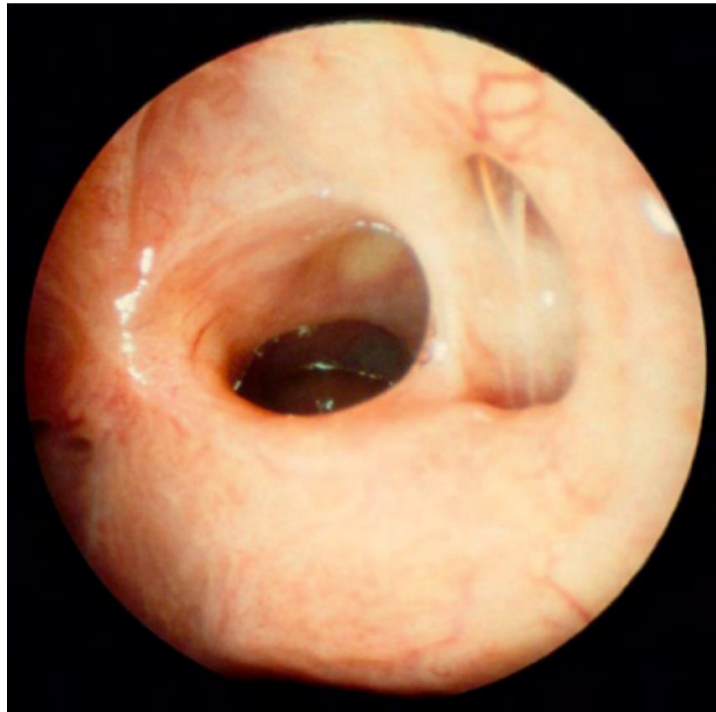
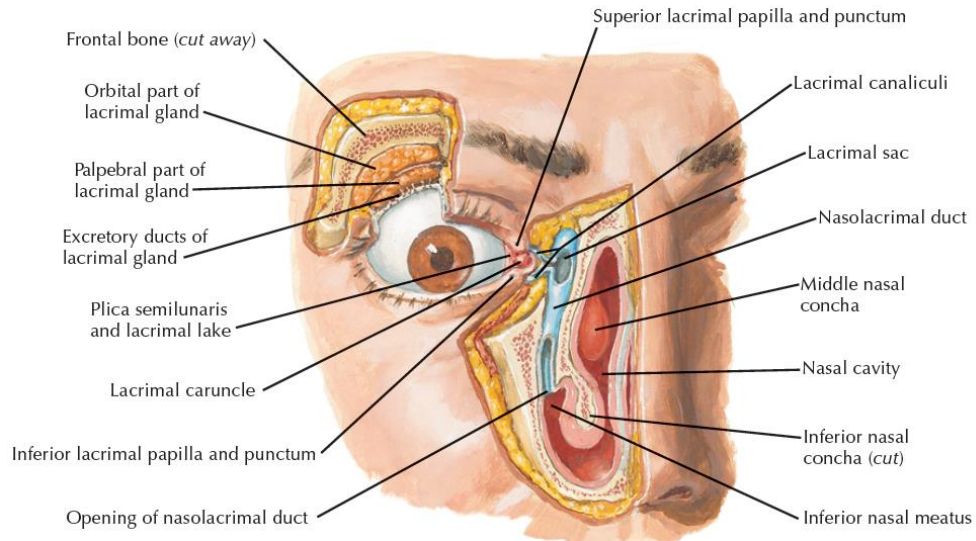
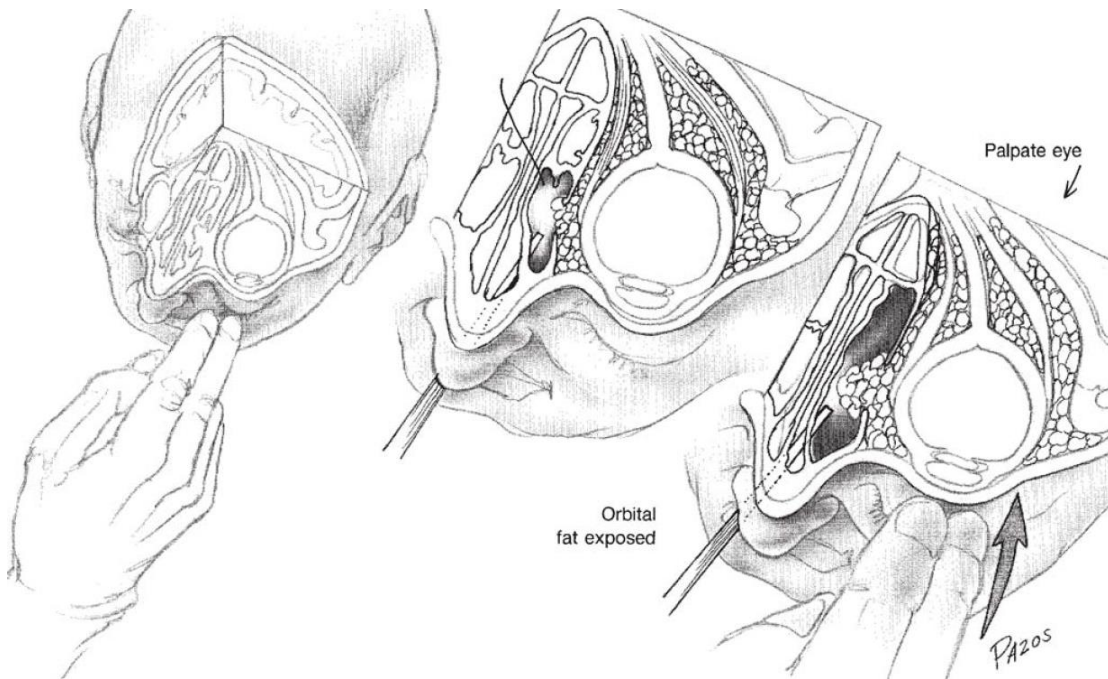
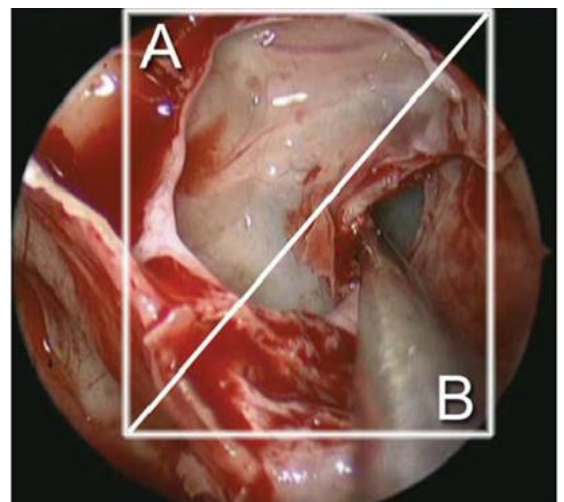


Figure 11: Endoscopic aspect of the left-sided nasal cavity with a middle meatal antrostomy (70° angled optical device: some crusts are shown inside the maxillary sinus). Anterior to the neo-ostium the nasolacrimal duct is shown, having been subjected to “unintended dacryocystorhinostomy”. This event had no functional sequel.

- **Prevention of orbital complications:**
- Eyes should always remain free from covering during the procedure, so complications can be noticed early.
- **Eye Pressure Test:**
 - o Should be done prior to surgery to assess how much tension is present prior to the procedure.
 - o So no confusion will be when suspecting retrobulbar hematoma and the globe is tense.
 - o Surgeon should perform the pressure test to determine if there is any dehiscence of LP and fat is visible.



- Entry into the sphenoid sinus through the natural os or through the inferior and medial half of "Bolger's box" is considered safe and unlikely to injure the carotid artery or optic nerve.



- **Mucosal bleeding:**
- Minor complication.
- Bleeding in the surgical area hinders visibility, cause delays, improper performance of the operation or even surgical complications.
- Mean arterial pressure is essential for arterial bleeding, whilst for venous bleeding it is the pressure in the venous vascular territory.
- For capillaries the blood flow in the respective vascular bed of the capillary is the determining factor.
- Generally, the capillary is the substrate of an uncomplicated mucosal bleeding during paranasal sinus surgery.
- Medical condition and drug history should be obtained before proceeding with ESS.
- Bleeding often only counts if it terminates the surgical procedure or requires a specific nasal packing.
- **In routine ESS:**
 - o 5% are persistently disturbed by a hemorrhage.
 - o 0.4% procedure is cancelled.
- **Risk factors:**
 - o Simultaneous inferior turbinate surgery.
 - o Extensive polypoidal sinusitis
 - o Revision surgeries
- **Measures to improve mucosal bleeding:**
 - 1. Pre-op Steroids:**
 - Systemic (30 mg/day prednisone for 5 days) and topical cortisone treatment can lead to a clear and unobstructed operating field with less bleeding which consequently reduces the duration of surgery.
 - Visibility within the surgical area gets improved via anti-inflammatory and anti-edematous effects of steroids.
 - A preoperative antibiotics can support this effect.
 - 2. Patient positioning:**
 - Patients put in 20 degree reverse Trendelenburg position was found to have the best endoscopic field view with lowest blood loss.
 - 3. Vasoconstriction agents:**
 - A solution of lidocaine 1% with epinephrine 1:100,000 can be injected into the nasal mucosa before starting the surgery to induce vasoconstriction and reduce mucosal bleeding.
 - After injection, cotton pledgets soaked in cocaine, tetracaine-ephedrine, or epinephrine can be placed into the nasal cavity for additional topical anesthesia and vasoconstriction.

- Subjectively, after injection of (epinephrine 1:100,000/1:200,000) visibility is reported to be improved.
- This advantage could not be proven clearly, compared to a sodium chloride injection or to the application of additional topical decongestion.
- Hypertensive crisis and cardiac shock were reported after combined application of epinephrine saturated packs (1:1,000) with injection of 1% lidocaine with epinephrine (1:100,000).
- Care should be taken with an exposed optic nerve as application of pads moistened by adrenaline have been reported to cause optic nerve damages and blindness.
- Topical application of epinephrine 1:1,000 can only be deemed to be safe in adults without previous cardiac damage.
- For children below 12 years of age, 0.05% oxymetazoline is recommended to be used.

4. Controlled hypotension anesthesia:

- Controlled hypotension is recommended to reduce the mean arterial pressure (MAP) during ESS.
- The aim is MAP of 50-60 mmHg for healthy patients and MAP of 80 mmHg for elderly patients.
- At the same time, MAP should not be lowered to less than 66% or 85% of the initial value (risk of post-op cognitive deficiencies).
- The recommended heart rate is 60/minute.
- Severe complications including organ ischemia have been observed in 0.02–0.06% of cases.
- However, there should be no risk for healthy patients (ASA I) in general, if the mentioned rules are respected.

$$\mathbf{BP = CO \times SVR}$$

- β -blockers (metoprolol) shown to decrease BP by lowering cardiac output rather than by lowering systemic vascular resistance (avoiding reflex tachycardia and the increase cardiac output occurs with pure vasodilator like sodium nitroprusside).
 - Leads to a short positive effect regarding bleeding.
 - Visibility in the surgical area tends to drop as the operation time gets extended.

5. Total Intravenous Anesthesia (TIVA):

- Recent systematic review of literature showed that 4 out of 7 studies demonstrated a statistically significant improvement in surgical field grade during ESS when patients received total intravenous anesthesia (TIVA) compared with inhalational anesthesia.
- Propofol reduces cardiac output and might contribute to a better objective local bleeding via alpha-adrenergic mediated vasoconstriction).
- If the operation lasts longer than 45 minutes, adverse effects on the platelet function become apparent.

6. Tranexamic Acid:

- Synthetic derivative of the amino acid lysine that has anti-fibrinolytic effect.
- Administration methods:
 - Peri-op administration of TA (1g TID PO for 5 days, starting 2 hours pre-op) showed significant less operative and postoperative bleeding compared with controls.
 - IV (10 mg/kg) administration at beginning of the sinus surgery, shows significant improvement of the bleeding in the surgical area.

7. Warm Saline Irrigations:

- Warm saline irrigation (40°C) helps maintaining platelet function and showed to be beneficial in improving surgical field of view and reducing blood loss.

- **Bleeding from Sphenopalatine Artery:**

- Major complication.

- Anatomy:

- In 80% of cases, sphenopalatine foramen is located in superior nasal meatus or in the transition area between the middle nasal meatus and the superior nasal meatus, directly behind or below the cresta ethmoidalis of the palatine bone.
- In 15% of cases, several ostia are found.
- In 97% of cases, sphenopalatine artery is divided into two or more branches.
- In 64% of cases, 3-10 branches enter the lateral nasal wall, above as well as below the ethmoidal crest.
- Terminal branches of sphenopalatine artery:
 - Pharyngeal artery (via palatovaginal canal to nasopharynx).
 - Posterior septal artery (via lower third of face of sphenoid to nasal septum).
 - Descending palatine artery.

- Sites of injury:
 - **Maxillary antrostomy:**
 - If opening of the maxillary sinus is enlarged in dorsal direction to the level of posterior wall of the maxillary sinus, in individual cases, it will cut through a branch of the sphenopalatine artery.
 - **Surgery at the posterior process of middle turbinate:**
 - Injury to the root of the sphenopalatine artery.
 - **Endoscopic neurectomy of the vidian nerve:**
 - Injury to the root of the sphenopalatine artery.
 - **Extended surgical procedures in the infratemporal fossa:**
 - Injury to the root of the sphenopalatine artery and maxillary artery.
 - **Sphenoid sinostomy:**
 - Injury to the posterior septal artery in the lower third of face of sphenoid.
 - **Floor of sphenoid:**
 - Injury to the pharyngeal branch of sphenopalatine artery in the palatovaginal canal at the floor of sphenoid during skull base surgery.
- Management:
 - Monopolar or bipolar endoscopic cauterization provides good control of sphenopalatine artery bleeding.

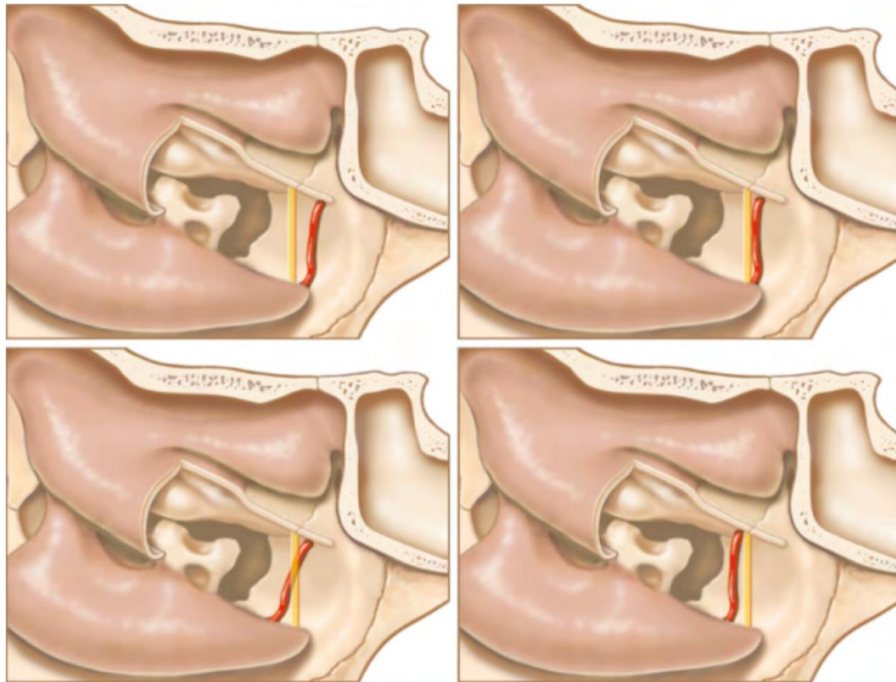


Figure 8: Individual anatomical relation of the posterior wall of the maxillary sinus in the frontal plane (depicted in yellow) and branches of the sphenopalatine a. supplying the interior turbinate (depicted in red) (refer to: [150, 654]). The diagram illustrates that generous fenestration of the maxillary sinus in the middle nasal meatus inevitably will cause relevant bleeding in some cases.

- **Bleeding from Anterior Ethmoid Artery:**
- Major complication.
- Anatomy:
 - Originates from Ophthalmic Artery in Orbit.
 - Cross medial rectus and penetrates lamina papyracea from posterior-laterally to anterior-medially between the second and third ground lamella.
 - AEA coursed in direct contact with the skull base in 85% of cases.
 - AEA coursed freely through the anterior ethmoidal cells in only 11% of cases.
 - Mainly in presence of supraorbital cells (pneumatization of roof of orbit postero-laterally to the frontal recess).
 - AEA bony canal dehiscence found in 25% of cases.

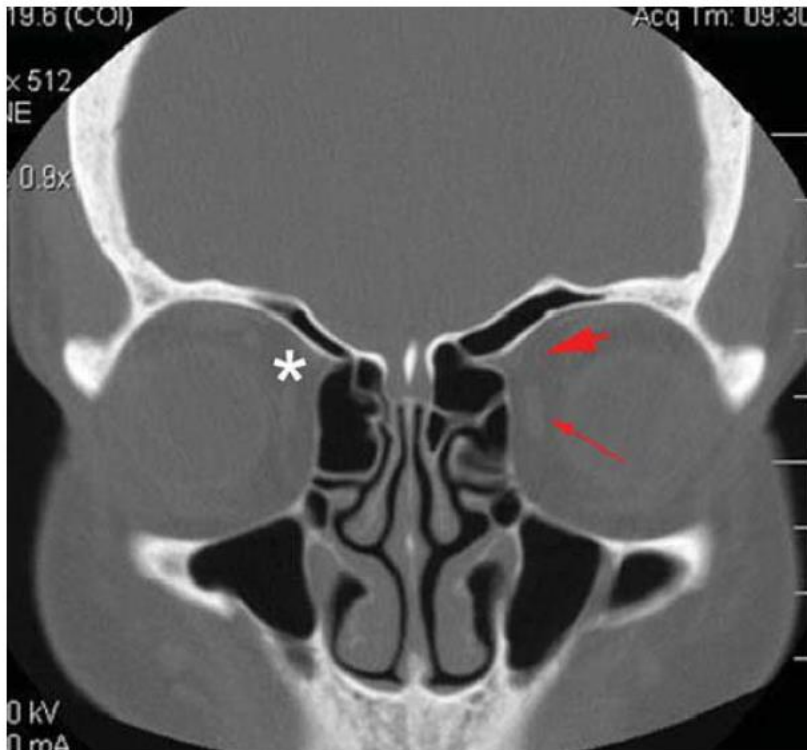


Figure 44.1 A coronal CT of the paranasal sinuses featuring the location of the anterior ethmoidal artery (*asterisk*) near the convergence of the medial rectus (*arrow*) and superior oblique (*arrow-head*) muscles. Note that in this patient, the anterior ethmoidal artery (*asterisk*) is located within a bony mesentery, suspended from the skull base.

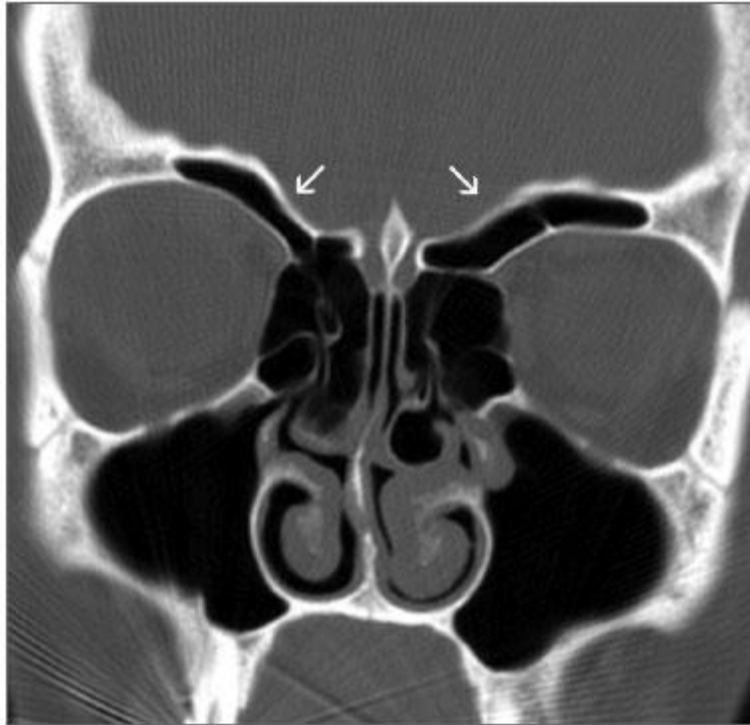
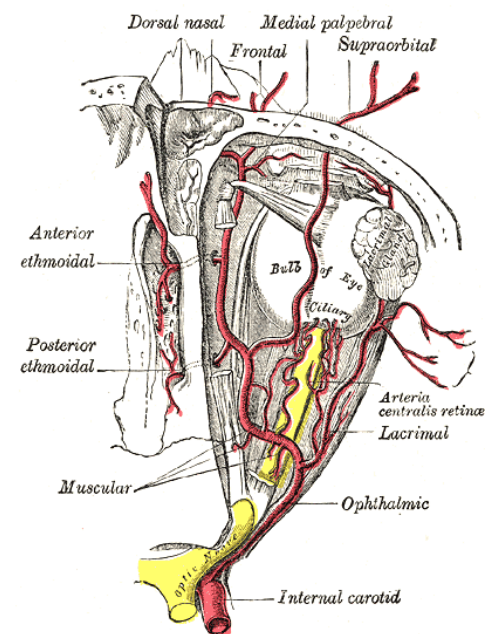


Figure 3. Supraorbital pneumatization - ample bilateral supraorbital pneumatization (arrows)

- Sites of injury:
 - **Anterior ethmoidectomy:**
 - Risk factors:
 - Larger distance to the skull base.
 - Bony dehiscence.
 - Arteries running within a ground lamella.
 - Lateral injuries can result in a threatening orbital hematoma, after retraction of the vessel stump.
- Management:
 - Management of orbital hematoma.
 - Bipolar coagulation is used to stop the bleeding.
 - Avoid monopolar coagulation at the skull base due to possible secondary damage to the meninges.



- **Bleeding from Posterior Ethmoid Artery:**
- Major complication.
- **Anatomy:**
 - Originates from Ophthalmic Artery in Orbit.
 - Smaller than the anterior ethmoidal artery.
 - Runs almost regularly (in 90% of cases) at the skull base few millimeters anterior to sphenoid face and is therefore more difficult to be identified endoscopically.
- **Sites of injury:**
 - **Posterior ethmoidectomy**
 - **Sphenoid sinostomy**
- **Management:**
 - Management of orbital hematoma.
 - Bipolar coagulation is used to stop the bleeding.
 - Avoid monopolar coagulation at the skull base due to possible secondary damage to the meninges.

- **Bleeding from Internal Carotid Artery:**
- The most feared and disastrous complication.
- Result in overwhelming blood loss and place the patient's life at imminent risk.
- Neurophysiologic monitoring of cortical and brainstem function during surgery can be helpful in the event of major bleeding to assess cerebral blood flow.
- **Rate of injury:**
 - 0.3% in surgery of diffuse CRS.
 - 1% in pituitary surgery.
 - 5% in extensive rhinoneurosurgical approaches (craniopharyngiomas, clival chordomas, and chondrosarcomas).
- **Most common segments injured are:**
 - Cavernous (interdural/intracavernous) segment (in left side).
 - Clinoid (interdural/paracavernous) segment.
- **Identifying at-risk patients is the mainstay of carotid artery injury prevention:**
 - **Anatomical Factors:**
 - ICA bulges out in anterior lateral wall of sphenoid sinus in 80% of cases.
 - The bony wall overlying ICA is < 0.5 mm thick and is not sufficient to protect the artery.
 - 20% rate of lateral sphenoid wall dehiscence with only dura and sphenoid sinus mucosa overlying the ICA.
 - ICA that approaches the midline also predispose to injury.
 - In the majority of patients, the bony sphenoid septum or sphenoid septation inserts onto ICA canal wall, and surgery on this septation may place the artery at risk.
 - Unrecognized preoperative cavernous ICA aneurysms may results in ICA rupture (confirmed by pre-op MRA).

- **Tumor Factors:**
 - Tumors closely adherent to the ICA require careful attention as tumor dissection can predispose the vessel to injury and vasospasm, which can result in altered mental status and/or hemiparesis.
 - Recommended to use blunt instruments, such as suction Freer dissectors and pituitary ring curettes, when working in close proximity to the artery.
 - If bone is required to be removed, then a grasping and twisting motion should be avoided.
 - Diamond burrs should be utilized in preference to cutting burrs.
- **Patient Factors:**
 - Risk factors include:
 - Previous radiotherapy
 - Revision surgery
 - Bromocriptine therapy.
 - Acromegalic patients as they tend to have more tortuous and ectatic carotid arteries with tumor in contact or surrounding the carotid.
- These patients with risk factors may warrant preoperative assessment of collateral cerebral circulation by Balloon occlusion test.
 - Help to identify the subset of patients who will not tolerate permanent carotid occlusion.
 - After arterial occlusion has been established, the patient's neurologic status is assessed and monitored continuously throughout the 15-minute period of endovascular occlusion.
 - If there is a change in neurologic status, the balloon is immediately deflated and the procedure terminated.
 - Patients who fail BOT have been shown to be at extremely high risk of stroke associated with internal carotid artery sacrifice or prolonged temporary occlusion.
 - 100% of these have experienced an associated neurologic deficit.
 - Patients who clinically pass BOT have demonstrated a relatively low risk of stroke associated with internal carotid artery sacrifice.
 - 5% of these have developed permanent stroke.

- Intra-op management of ICA bleeding:
 - **Controlling the Surgical Field:**
 - Two surgeons are engaged, allowing one surgeon to control the bloodstream, directing it away from the endoscope, while the other obtains visualization to attempt hemostasis.
 - Two large-bore (10F) suction devices and a lens cleaning system for the endoscope should be used.
 - The second surgeon uses suction downside the nose with predominant bleeding to direct flow away from the other side.
 - The primary surgeon places the endoscope down the contralateral side, using the posterior septal edge as a shield from the blood flow.
 - The primary surgeon clears blood ahead of the endoscope using the second suction device.
 - A pedicled septal flap should also be cleared and pushed into the nasopharynx.
 - The second surgeon is then free to “hover” the suction device directly over the site of injury to help gain visualization for the primary surgeon.

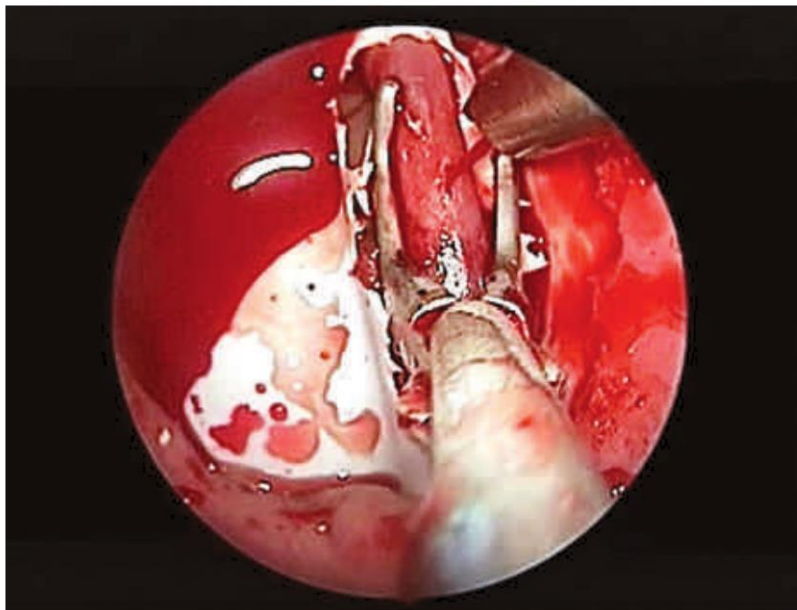


Fig. 1 Controlling the surgical field: suction device directs blood flow away from endoscope to allow attempt at hemostasis with Wormald vascular clamp (Medtronic, Jacksonville, Florida, United States).

○ **Hemostasis:**

1. Compression of ICA:

- Effect of compression of ipsilateral carotid is not reliable in presence of a good collateral circulation.
- Compression of both carotids for no longer than 2 minutes, to allow time for the emergency treatment within the sphenoid sinus or for inserting nasal packing.
- Normotension should be maintained through resuscitative measures to preserve adequate cerebral perfusion.

2. Emergent insertion of a tight nasal packing.

- Several packing agents have been described including Teflon, fibrin glue, Gelfoam, oxidized cellulose packing thrombin-gelatin matrix, oxygel, and glue and muslin gauze.
- Packing is contraindicated if dura is opened due to risk of developing subdural hematoma.
- Over packing may results in ischemia and storke due to:
 - ICA occlusion
 - ICA stenosis

3. Muscle pack:

- Crushed muscle patch is superior to other types of packing in gaining hemostasis.
- Muscle is harvested from the thigh (usually prepared for fascia lata graft in skull base cases) or SCM and then crushed.
- It is placed directly over the injury site with Blakesley forceps with enough force to stay in contact with the vessel injury site but should not compress or occlude the vessel.
- It takes up to 12 minutes to gain hemostasis.
- If the carotid is likely to be exposed to the nasal cavity, the muscle patch should be reinforced with an overlying septal flap.
- If the vessel is intracranial, the patch should be secured with oxidized cellulose and fibrin glue and tightened with packing.
- It has been associated with destabilization and pseudoaneurysm if used as the lone treatment.

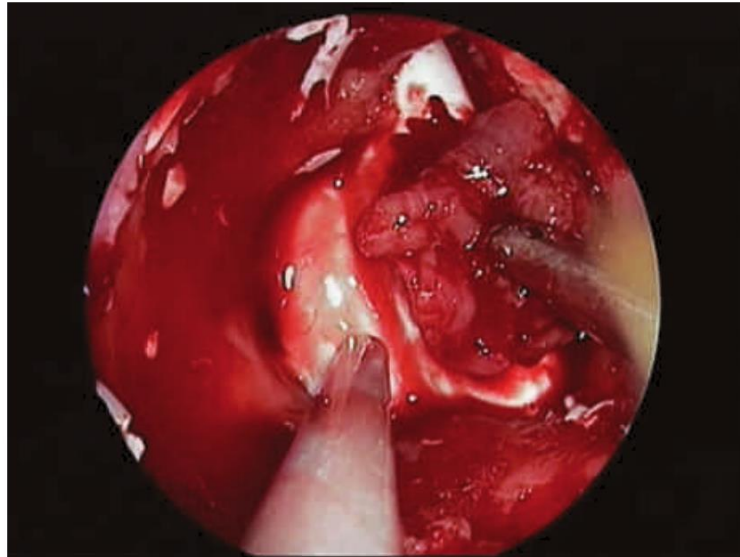


Fig. 2 Muscle patch placed over carotid injury site with Blakesley forceps.

4. Direct Vessel Closure:

- If there is adequate exposure of vasculature during surgery and the vessel injury site is not enclosed by bone or difficult to access, direct closure of the injury is possible.
- Methods:
 - **U-clip** anastomotic device was used to repair the injury site after clamping with a Wormald endoscopic vascular clamp and found it to be very effective in gaining hemostasis in an animal model of carotid catastrophe.
 - **T2 Aneurysm Clip** was used and it was able to gain hemostasis in all injury types as well as prevent pseudoaneurysm occurrence in an animal model of carotid bleeding.
 - **Bipolar electrocauterization** was tried on different injury types in an animal model of bleeding and it was effective in gaining hemostasis, however it is not recommended due to high risk of delayed secondary hemorrhage, total carotid occlusion and worsen the injury by enlarging the defect.

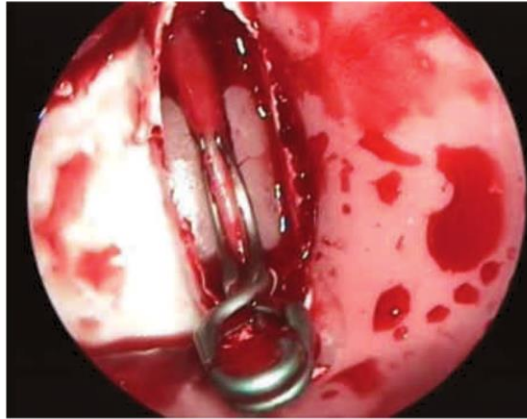


Fig. 3 T2 aneurysm clip (Mizuho, Tokyo, Japan) placement on carotid injury site.

5. Endovascular intervention:

- If hemostasis was not achieved, urgent transfer for endovascular intervention must be done.
- The aim is either to occlude the vessel or maintain vascular flow.
- Tight nasal packing prevent identification of injured vessel during angiography and it should be loosen.
- Methods:
 - **Endovascular occlusion of the artery:**
 - Performed using a balloon or coil at the wall defect to prevent extravasation of blood from both anterograde and retrograde vessel filling.
 - Risk of distal migration due to the high-pressure, high-flow environment of the artery which places the ophthalmic artery at imminent risk due to its location distal to the cavernous ICA.
 - Assessment of collateral circulation should done before the occlusion if time permits.
 - If occlusive intervention is planned and time permits, prior assessment of collateral circulation by balloon occlusion test (BOT) of the ICA should done.
 - However, 5% of patients passed the balloon occlusion test developed permanent stroke after the occlusion.

- **Endovascular stenting of the artery:**
 - An alternative to occlusion intervention.
 - Placement of a stent graft to seal the injury site and maintain vascular flow.
 - Technically challenging to place in the tortuous cavernous carotid siphon.
 - Associated with risk of distant migration and ICA spasm.
 - 5% risk of stroke within the first 30 days of stent placement.
 - Requires placing the patient on concurrent anticoagulation therapy.



Figure 9: Laceration of the left-sided internal carotid a. as a complication of routine paranasal sinus surgery.

a. Angiogram of the internal carotid artery revealing a lesion located at the anterior bending (anterior 'genu').

b. Occlusion of the artery by coils.

c. Revision surgery of the respective sphenoid sinus revealing a partly exposed coil (green arrow: coil; white arrow: suction device).

d. Post-treatment axial CT scan depicting the coils.

6. External ligation of carotid artery:

- Considered obsolete due to high risk of neurological complications.

○ **Treatment of Blood loss:**

- Establishing several extensive intravenous lines and emergency blood transfusions.
- Maintenance of the circulation is strongly recommended by normotension, not to endanger the cerebral circulation disproportionately.

- **Post-op care of ICA bleeding:**

- Involves the prevention of potential complications of carotid artery injury, which include:
 - **Pseudoaneurysm formation:**
 - Tear through all layers of an artery with persistent flow outside the vessel into aspace contained by surrounding tissue.
 - Incidence after carotid injury reaches up to 60%.
 - Developed in average after three weeks of carotid injury.
 - High risk of rupture for up to 3 months.
 - May result in secondary, massive hemorrhage or in thromboembolism.
 - Immediate mortality rate in case of bleeding from pseudoaneurysm is about 30%.
 - **Carotidocavernous fistula:**
 - Fistula between the carotid sinus and the cavernous sinus.
 - Results in chemosis, proptosis, orbital pain, diplopia and bruit (a humming sound within the skull due to high blood flow through the AV fistula).
- Prompt identification and treatment is required for successful long-term management.
- Once hemostasis has been achieved intra-op, the patient should be transferred for urgent angiographic investigation to assess the repair and ascertain if further endovascular intervention is required.
- If the immediate post-op angiogram is normal, then monitoring the patient in the ICU until the packing is removed and another angiogram is performed at 1 week post-op.
- If the follow-up angiogram is normal, then the angiogram is repeated at 6 weeks, 3 months, and 1 year to rule out presence of complications.

- If pseudoaneurysm or carotidocavernous fistula are detected, treatment options:
 - **Stent-graft placement:**
 - The safest option.
 - **Endovascular occlusion of aneurysm lumen:**
 - Associated with increased risks of complications during pseudoaneurysm surgery as the pseudoaneurysm lacks a wall on which the coil/balloon can sit, and can result in rupture or dissection of the ICA wall
 - Detachable balloons may be used in carotidocavernous fistula to occlude the fistula while maintaining parent vessel patency.
 - **Surgery (bypass or aneurysmal clipping):**
 - Has a relatively high complication rate.

**TABLE
44.4**



**TREATMENT
CAROTID DRILL**

Tertiary care center

1. For every sinus operation, have available, on momentary notice, two long polyvinyl acetate (Merocel) sponges, petroleum-impregnated gauze, two 18F Foley catheters and umbilical clamps for nasal packing.
2. Pack nose immediately at the first sign of severe hemorrhage.
3. Compress the carotid artery in the neck on the affected side.
4. Begin anesthesia to induce controlled hypotension.
5. Ready blood for transfusion.
6. Call neurosurgeon immediately.
7. If the patient's condition is unstable, the neuroradiologist should perform intraoperative arteriography. If the patient's condition is stable, transfer the patient to the neuroradiology suite.
8. Perform balloon occlusion under EEG surveillance.
 - a. If there is no evidence of a change in perfusion or lateralization, ligate the carotid artery.
 - b. If changes are present on the EEG (dangerous recording), deflate the balloon, maintain packing, and observe.
9. Insert Swan-Ganz catheter. Put the patient in a hyperdynamic state by using high molecular weight starch (hetastarch) to increase cerebral perfusion.
10. After cerebral perfusion has been increased, reinflate the balloon and check the EEG.
 - a. If lateralization occurs, try carotid bypass or barbiturate coma.
 - b. If bypass is possible, reinflate the balloon and ligate the carotid artery.

Nontertiary hospital

1. Call neurosurgeon.
2. Expose the carotid artery in the neck.
3. Temporarily occlude the carotid artery with a clamp or tape.
4. Ligate the carotid artery.
5. Perform a trapping procedure: Clip the carotid artery below the anterior communicating artery to isolate this segment from blood flow.

EEG, electroencephalogram.

- **Uncomplicated CSF fistula:**
- Small and isolated CSF fistulas, which treated at once successfully, count statistically as "minor complication".
- Rate of unexpected dura exposure is 0.2%.
- Rate of manifested clinically relevant CSF fistulas is 0.2–0.8%.
- Weakest part of the anterior skull base is at the lateral lamella of olfactory fossa in which the bone thickness is only 0.05 mm.
- Avoid cribriform injury by staying Lateral to middle turbinate.
- Certain anatomic variants of skull base favor injuries:
 - Position of ethmoid roof beneath roof of the orbit.
 - Asymmetry of the ethmoid roof.
 - Asymmetry regarding the height of the ethmoid roof.
 - Deep position of the cribriform plate, i.e. high lateral lamella of the olfactory fossa.
 - Larger angle between skull base and horizontal line through the sagittal plane.
- Diagnosis:
 - Intra-op diagnosis:
 - Clear fluid flows in the operating field.
 - Topical intranasal 5% fluorescein (CSF will change the fluorescein from yellow to green).
 - Post-op diagnosis:
 - CSF rhinorrhea, recurrent meningitis.
 - Beta 2 transferrin
 - Beta-trace protein
 - High resolution CT
 - MR cisternography
- Management:
 - Timing:
 - If detected intra-op, it should be repaired immediately.
 - If detected post-op, small CSF leaks can close with conservative management in 1/3 of the patients, but surgery should be considered as soon as possible for any persistent leak lasting longer than 2 weeks.
 - Technique:
 - For small defects, free autogenous mucosal grafts are preferred as onlay/overlay technique.
 - Larger defects above 5 mm in diameter are closed in several layers, partly with cartilage or bone and fibrin glue.
 - Sinus surgery may be continued after an isolated CSF fistula.

- Post-op care:
 - Anesthesia:
 - No positive pressure ventilation
 - Deep extubation technique
 - Avoiding coughing and straining
 - Instructions:
 - Bed rest for 1-5 days
 - Elevation of head of bed 30 degree
 - No coughing or sneezing (Sneezing with open mouth)
 - No heavy weight lifting.
 - Medications:
 - Antibiotics (IV ceftriaxone to cross blood brain barrier)
 - Antihistamine
 - Laxatives
 - Lumbar drain:
 - Indicated in high flow CSF leak with large defects and high ICP.
- **Meningitis:**
 - Postoperative meningitis is rare.
 - Spreads through dural lesions, perivascular or vascular paths or via perineural spaces of olfactory fibers.
 - Frequently develop on the basis of a preexisting inflammation of mucosa in the paranasal sinuses.
 - Meningitis may occur with a delay of one week after routine surgery.
 - Symptoms:
 - Fever, laboratory diagnostics indicating major inflammation, headache and neck pain, as well as an impaired consciousness.
 - When suspecting meningitis, CT scan has to be ordered immediately followed by lumbar puncture.
 - Active CSF fistula needs to be detected.
- **Encephalocele:**
 - Skull base defect can cause a secondary herniation of brain tissue.
 - Iatrogenic encephalocele can develop slowly within months and might only become apparent though meningitis.
 - Diagnosis:
 - CT and MRI will confirm the presence of encephalocele.
 - CT angio or MRA should be done to rule out presence of vascular loop inside the encephalocele.
 - Management:
 - Endoscopic resection followed by skull base reconstruction with neurosurgical team.
 - If encephalocele doesn't contain vascular loop, cauterized it with bipolar diathermy with use of copious saline irrigation.
 - If encephalocele contains vascular loop, it should pushed intracranially without cauterization to avoid risk of intracranial bleeding.

- **Tension Pneumocephalus:**
- Pneumocephalus is the presence of gas (air) in the cranial cavity.
- In most cases, it is based on a communication between extracranial and intracranial space.
- Air can be present in epidural, subdural, subarachnoid, intraventricular or intracerebral spaces.
- **Pathophysiology:**
 - A minor defect of the dura may act like a “valve” leading to accumulation and trapping of air intracranially during intermittently increased pressure in the upper respiratory tract (e.g. during sneezing or during mask respiration).
 - Air may be sucked in, after CSF has been discharged.
 - ICP increases gradually and tension pneumocephalus develops.
- **Symptoms:**
 - Altered state of consciousness, restlessness, headache, nausea, vomiting, eye motility disorders, ataxia, and spasms.
 - If the underlying process is not interrupted, a pressure effect in the interhemispheric fissure (close to the motor cortex) might induce a diplegia.
 - Additionally rupture of bridging veins may cause sub dural hematomas and finally cardiac arrest.
 - Neurological symptoms may have latent period of several days.
- **Diagnosis:**
 - CT scan:
 - “Mount Fuji” sign is seen in axial plane due to the compression with tip-like protrusion of frontal poles.
 - The mass effect of air does not always have to be spectacular and is not always bilateral.
 - Intracerebral tension pneumocephalus occur subcortically in the frontal brain with radiological evidence of expansive intraparenchymal frontal air bubble was identified.
 - Lumbar drainage is contraindicated in case of a prominent pneumocephalus due to high ICP.
- **Management:**
 - Immediate neurosurgical decompression and simultaneous closure of the skull base defect.



Figure 13: Axial CT-scan revealing tension pneumocephalus caused by a skull base perforation during routine paranasal sinus surgery (note: “Mount Fuji sign” of the anterior cerebral poles).

- **Direct mechanical cerebral trauma:**
- Fatal, partially lethal complications with mechanical destruction of cerebral tissue are limited to extremely rare cases in routine paranasal sinus surgery.
- Corresponding reports are mostly from earlier decades.
- Intraoperatively, the surgeon is mostly not aware of life-threatening brain damage, often only a "striking bleeding tendency" is registered.
- The removal of "indistinct tissue" for histological analysis, which then turns out to be cerebral tissue, is tragic.
- Serious injury pattern has been induced accidentally with the shaver.
- **Symptoms:**
 - Lasting clouding of consciousness, disorientation or somnolence, and, focal neurological signs like myoclonia or headaches in recovery phase, bleeding or liquorrhea occurs.
 - Permanent "frontal brain syndrome" can occur after injury of the frontal brain with personality changes (loss of motivation, apathy), behavioral and memory disorders.
- **Diagnosis:**
 - In case of doubt, contrasted CT or MRI should be done immediately to determine the existence and extent of the damage and to exclude a pneumocephalus or bleeding requiring therapy.
 - MRI displays more subtle parenchymal damage and also the chronological sequence of a resorption of hemorrhages.
- **Management:**
 - In an acute case, emergency neurosurgical consultation has to be performed directly after imaging.



Figure 14: Postoperative axial CT-scan following seemingly uneventful routine paranasal sinus surgery. Obviously a major skull base perforation has happened and a piece of bone (red arrow) was transferred into the remote brain tissue. The surgeon noticed an increased intraoperative blood loss only.

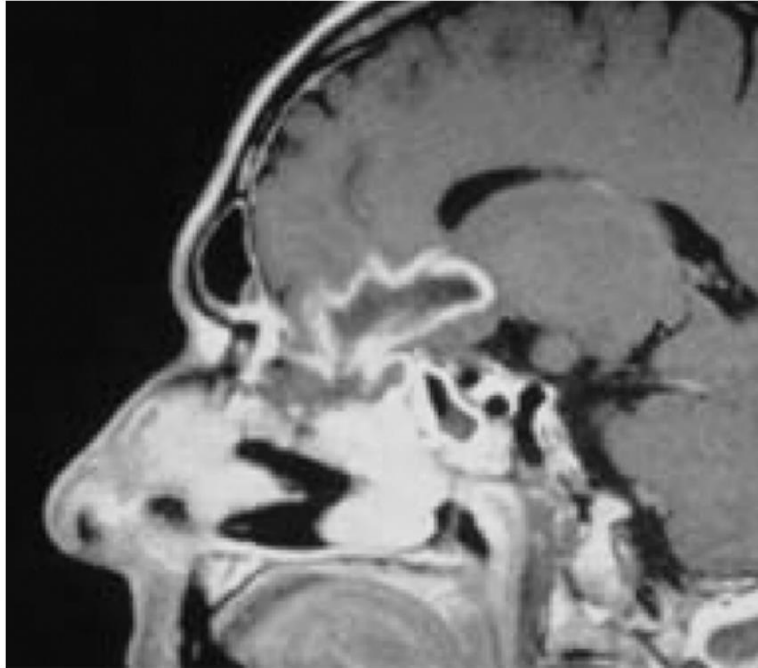


Figure 12: Postoperative sagittal MRT tomography revealing signs of a recent anterior skull base perforation by a.-p. directed force.

- **Intracranial bleeding and hematoma:**
- CSF fistulas may lead to severe complications including bleeding of intradural or subarachnoid vessels or from branches of the anterior cerebral artery or the anterior ethmoidal artery.
- This may result in an epidural, subdural or intracerebral haematoma, a localized cerebral infarction or even a traumatic aneurysm.
- After extensive reconstruction of the frontobasal region and after a large amount of CSF has been discharged, intracranial pressure may drop, which in turn can result in displacement of the graft or tension on the bridging veins causing a subdural haematoma.
- After extensive surgical procedures, a CT control must be performed on the first or second postoperative day.

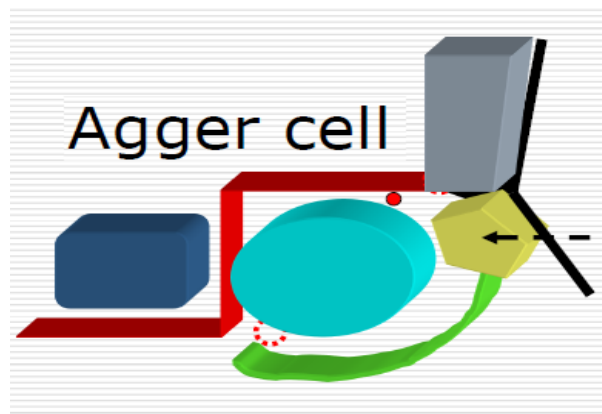
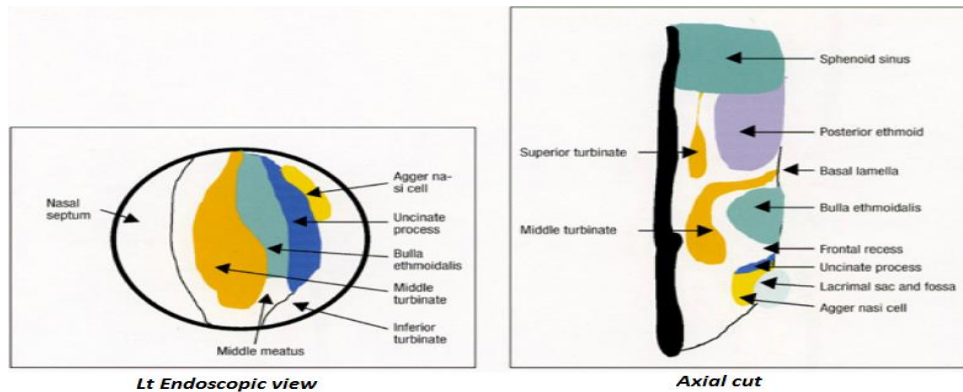
Frontal Sinus Surgery

- Endoscopic sinus surgery (ESS) has been accepted as the treatment of choice for chronic sinusitis **refractory to medical treatment**.
 - Surgery of the frontal sinus is perhaps the most technically demanding of all ESS.
 - The complex and varied anatomy, and proximity to critical structures such as: **olfactory fossa, skull base, orbit** contribute to the surgical difficulty.
 - These narrow confines enhanced for postoperative **scarring**, make surgical treatment of chronic frontal sinusitis challenging.
 - Wide spectrum of defined endonasal surgical procedures of the frontal sinus has been developed.
 - These are based on the drainage or sinusotomy classification.
- **SURGICAL ANATOMY**
- The key to successful frontal sinus surgery is understanding of its anatomy.
 - Frontal sinus doesn't present at birth "last paranasal sinus to develop".
 - At around **2 years** of age, the most anterior ethmoidal sinus invaginates into the frontal bone and continues its vertical growth trajectory at an average annual rate of 1.5 mm until the 15th year.
 - Final growth is completed before 20.
 - The sinus is compartmentalized by the intrasinus septa, which divides the sinus into halves "Usually asymmetric".
 - Between outer and table of frontal bone.
 - Ratio of table thickness between outer and inner table = 2:1
 - Adult volume: 2-3 ml at age of 12-12 years.
 - Underdeveloped in 5%.
 - Ostium located posteromedially in the floor.
 - The frontal sinus has the most complex and variable drainage of any paranasal sinus.
- Drainage depends on superior attachment of uncinate process:
- **Attaches to Lamina papyrcea (**Most common**):**
 - Drains into **Middle meatus** medial to ethmoidal infundibulum (between uncinate process and middle turbinate)
 - **Attaches to Ethmoid roof:**
 - Drains into **Ethmoidal infundibulum**.
 - **Attaches to Middle turbinate:**
 - Drains into **Ethmoidal infundibulum**.



- **Agger nasi:**

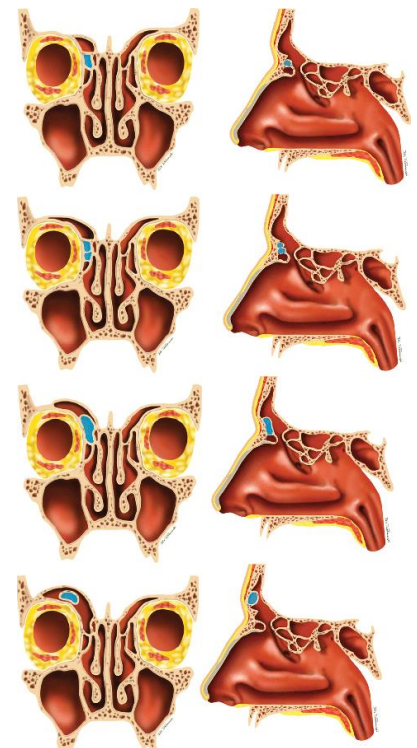
- Most anterior ethmoid cells.
- Eminence anterior & superior to the attachment of middle turbinate on lateral wall.
- Anteriorly to uncinate process.
- Medial to lacrimal sac & fossa.
- Posterior wall of Agger nasi cells forms the anterior wall of frontal recess.
- Major dimensions are correlated with frontal sinus diseases and lacrimation.



- **Frontal cells (20%):**

Anterior ethmoid cells which invade the frontal sinus or the recess.

- **Type I (37%):** Single frontal recess cell above agger nasi cell
- **Type II (20%):** Two or more cells in frontal recess above agger nasi cell
- **Type III (10%):** Single massive cell above agger nasi and pneumatizing cephalic into the frontal sinus
- **Type IV (5%):** Single isolated cell within frontal sinus.



- This classification was modified by **Wormald** who more clearly defined the frontoethmoidal cell and **re-defined type 3 and 4 cells**:
 - **Type III:** Frontal ethmoidal cell pneumatizing cephalad into the frontal sinus but extending less than 50% of the vertical height of the frontal sinus
 - **Type IV:** Frontal ethmoidal cell that extends more than 50% of the vertical height of the frontal sinus



Figure 46.2c T1 cell. One cell above the agger nasi cell. This cell does not protrude superior to the frontal beak

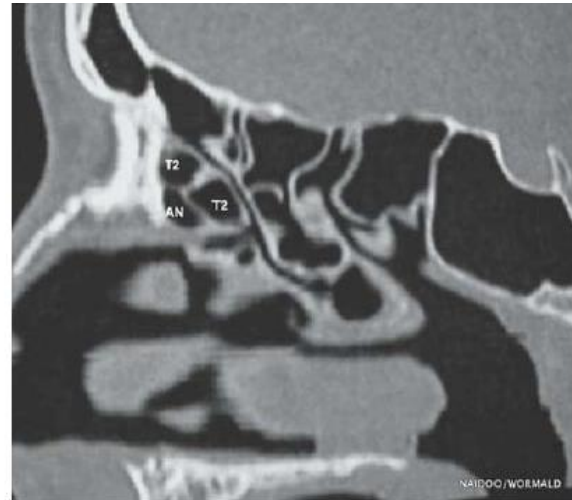
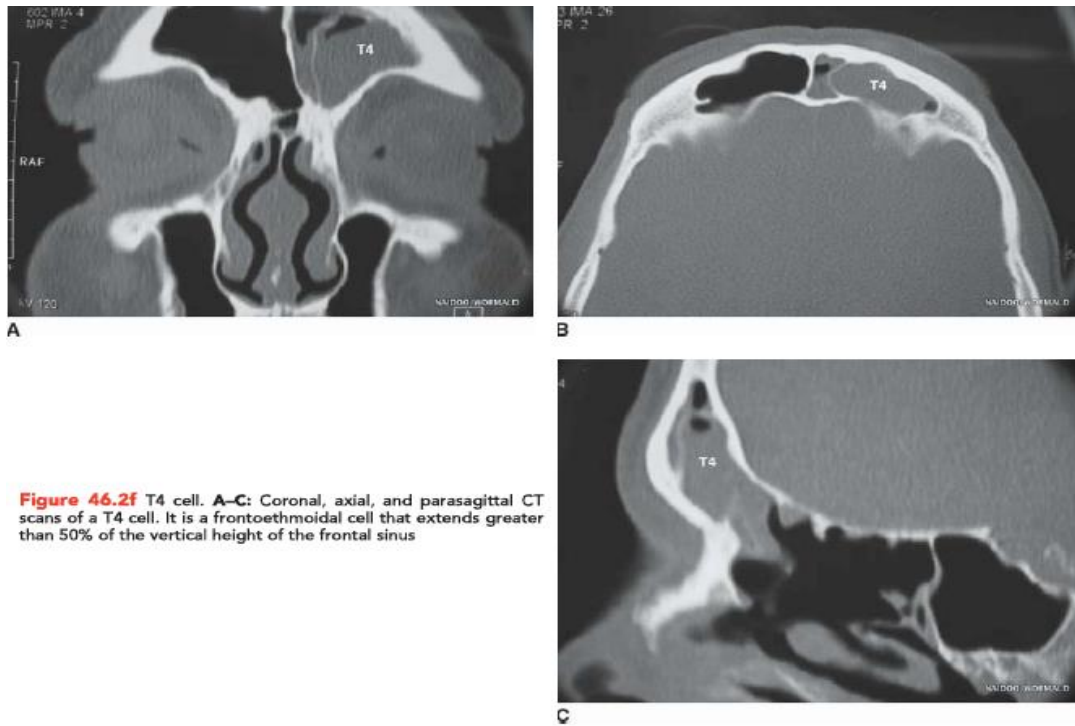


Figure 46.2d T2 cell. A tier of cells above the agger nasi cell, none of which extend superior to the frontal beak



Figure 46.2e T3 cell. A frontoethmoidal cell that extends above the frontal beak. It is limited to less than 50% of the vertical height of the frontal sinus



- **Supraorbital ethmoidal cells: (15%):**
 - Pneumatization of the orbital plate of frontal bone by ethmoid cells.
 - Located just **posterior to frontal sinus** ostium.
 - Anterior ethmoid artery present just **posterior** to cell and anterior to suprabullar lamella in fovea ethmoidalis.
 - Failure to address disease within these cells or subsequent scarring is another potential reason for persistent frontal sinus disease.

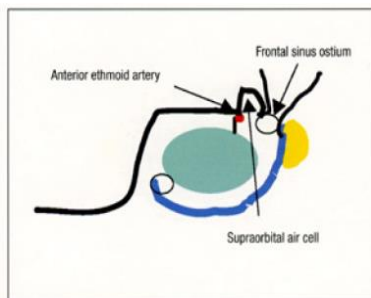
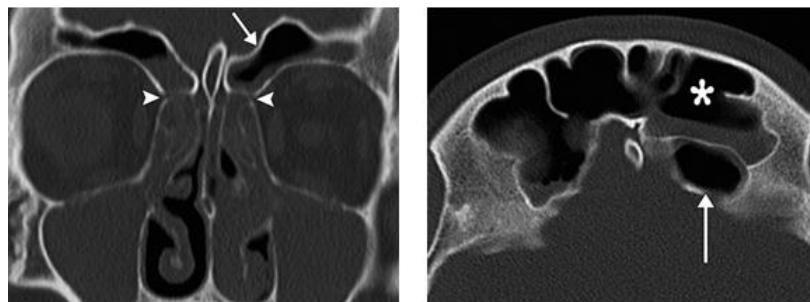
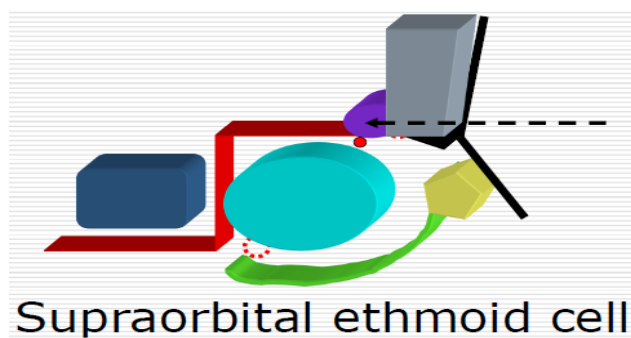


Fig. 13
Left supraorbital air cell (schematic depiction, sagittal plane).



- **Frontal recess:**

- Most anterior superior aspect of the anterior ethmoid sinus that forms connections with the frontal sinus.
- Hourglass shape; narrow waist at frontal's true os.
- Its walls are formed by the close relationship of bordering anatomic structures.
- **Bounded by:**
 - Posterior wall of agger nasi, frontal beak **anteriorly**.
 - Anterior face of the bulla ethmoidalis **posteriorly**.
 - Middle turbinate **medially**.
 - Lamina papyracea **laterally**.

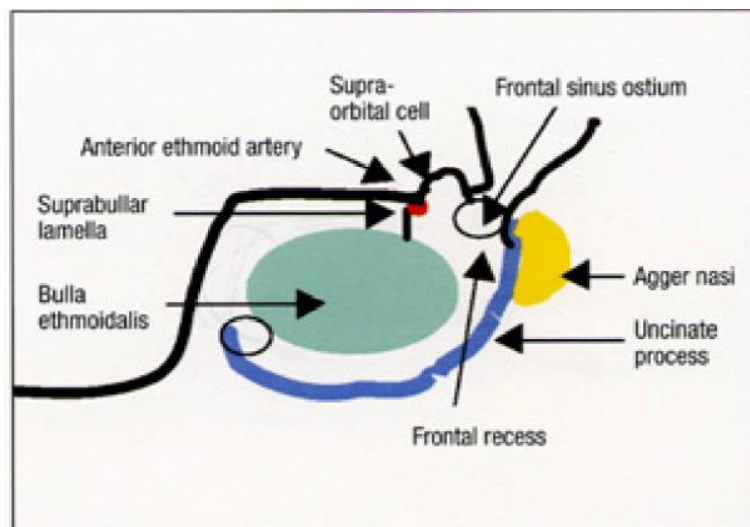
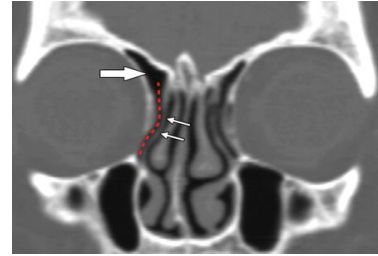


Fig. 12a
Left frontal recess (schematic depiction, sagittal plane).

- When the ethmoid bulla lamella reaches the skull base, it forms **posterior** border of the frontal recess, but when it doesn't reach the skull base, the suprabullar recess communicates directly with the frontal recess.
- Nasal communication of the frontal recess **Inferior**.
- Internal os of the frontal sinus **Superior**.

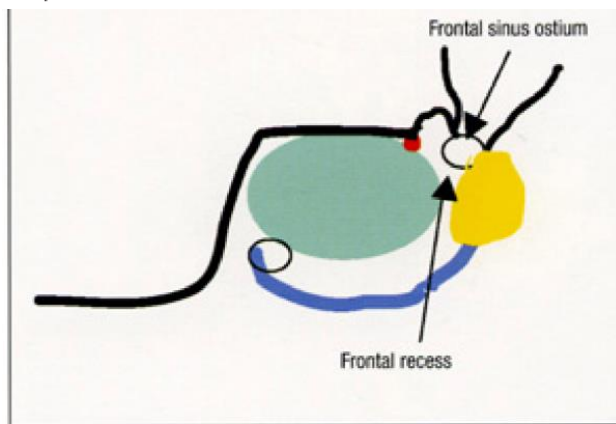
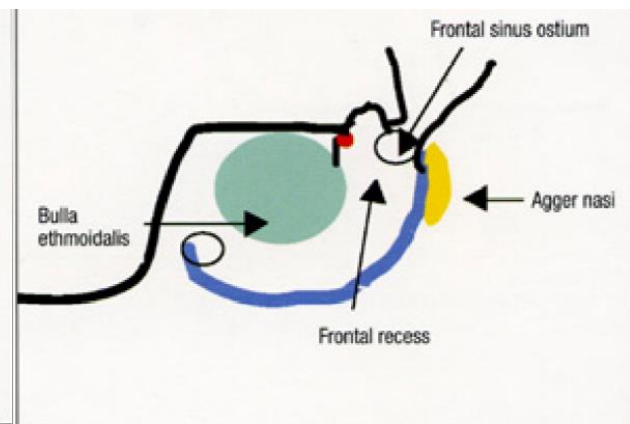
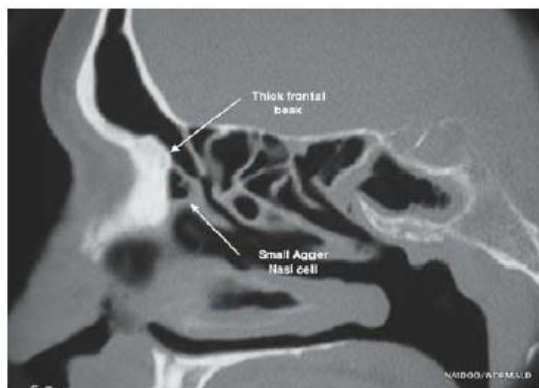
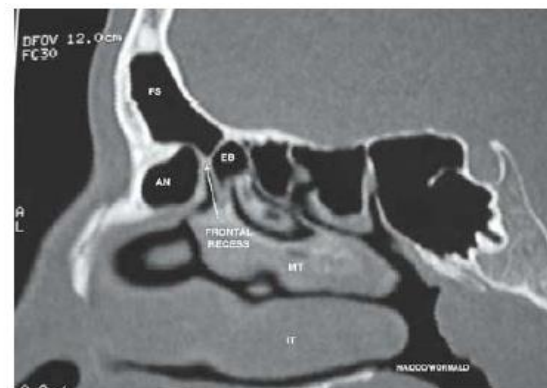
- **The patency of the frontal recess depends on:**

1. Superior attachment of the uncinate process.
2. Agger nasi cell.
3. Presence or absence of Frontal cells.
4. Presence or absence of Supraorbital cell.
5. Ethmoid bulla lamella.
6. Suprabullar cells (Along the skull base, may protrude anteriorly compressing the drainage pathway).
7. Frontal bulla cell "suprabullar cell pneumatizes along the skull base into the frontal sinus".

**TABLE
46.2****WORMALD CLASSIFICATION OF FRONTAL RECESS AND FRONTAL SINUS CELLS**

Cell	Description
Agger nasi	Cell that is either anterior to the origin of the middle turbinate or sits directly above the most anterior insertion of the middle turbinate into the lateral nasal wall (see Fig. 46.2a - Small agger and Fig. 46.2b - large agger cell)
Frontal ethmoidal	Anterior ethmoidal cell that needs to be in close proximity or touching the frontal process of the maxilla
Type 1	Single cell above agger nasi, not extending above the frontal beak (see Fig. 46.2c)
Type 2	Tier of frontal ethmoidal cells above agger nasi, not extending above the frontal beak (see Fig. 46.2d)
Type 3	Frontal ethmoidal cell pneumatizing cephalad into the frontal sinus but extending less than 50% of the vertical height of the frontal sinus (see Fig. 46.2e)
Type 4	Frontal ethmoidal cell that extends more than 50% of the vertical height of the frontal sinus (see Fig. 46.2f)
Frontal bulla	Suprabullar cell that pneumatizes along the skull base into the frontal sinus along its posterior wall (see Fig. 46.2g)
Suprabullar	Cells above the ethmoid bulla that do not extend into the frontal sinus (see Fig. 46.2h)
Intersinus septal	A medially based cell related to the frontal sinus septum which opens into the frontal recess (see Fig. 46.2i)

Adapted from the Kuhn classification.

**Fig. 12b**
Narrow left frontal recess (this is not a fronto-nasal duct) (schematic depiction, sagittal plane).**Fig. 12c**
Wide left frontal recess (schematic depiction, sagittal plane).**Figure 46.2a** Small agger cell: A small agger cell is usually associated with a prominent frontal beak, which narrows the A-P dimension of the frontal ostium**Figure 46.2b** Large agger cell: A large agger cell tends to push the frontal drainage pathway more posteriorly. In general, it is associated with a less prominent frontal beak, making the A-P diameter of the frontal ostium larger. AN, agger nasi; EB, ethmoid bulla; FS, frontal sinus; MT, middle turbinate; IT, inferior turbinate

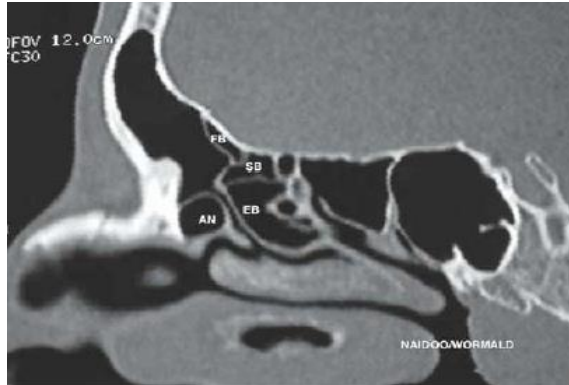
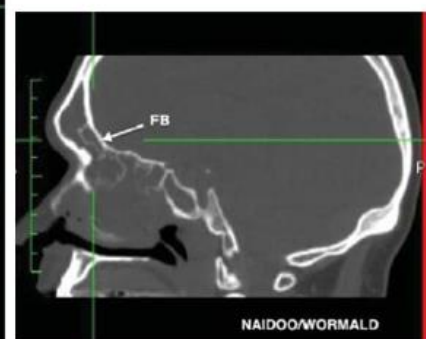
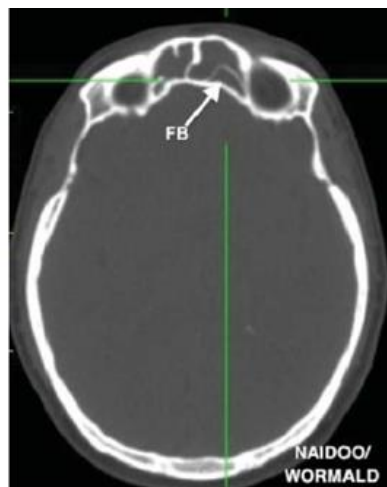
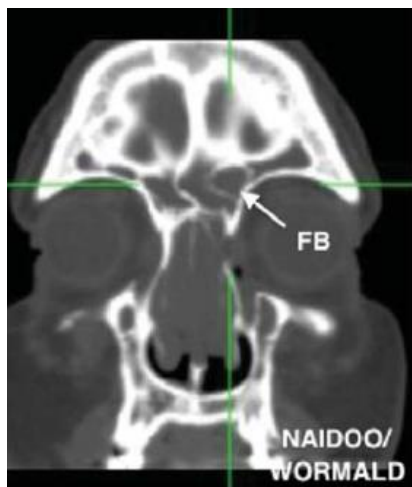
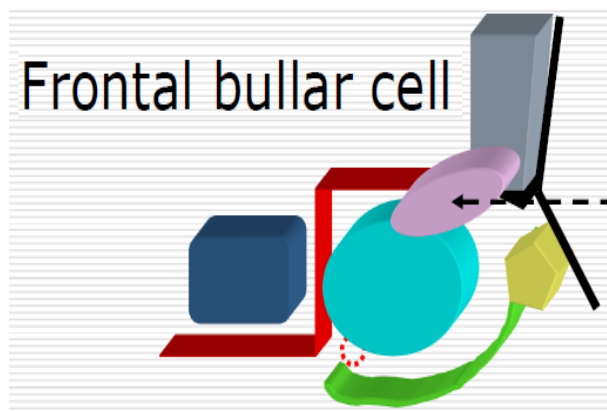
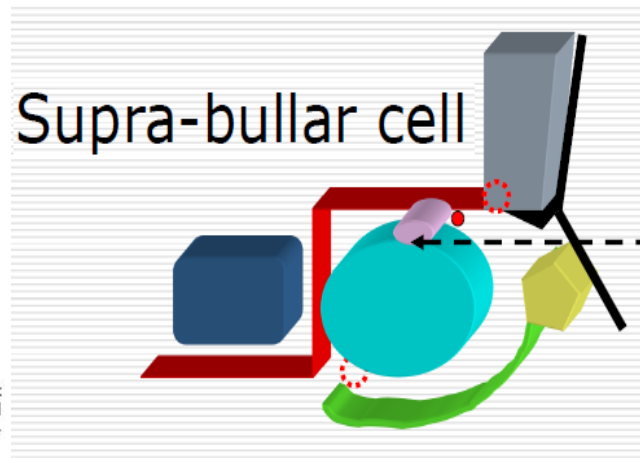
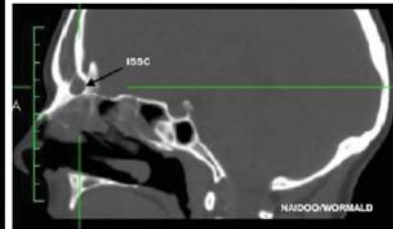
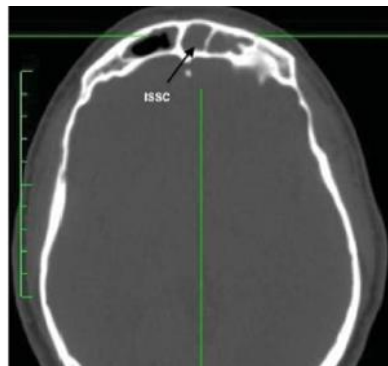
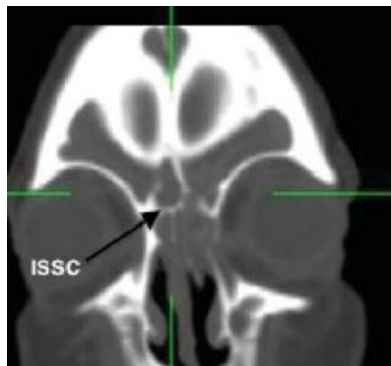
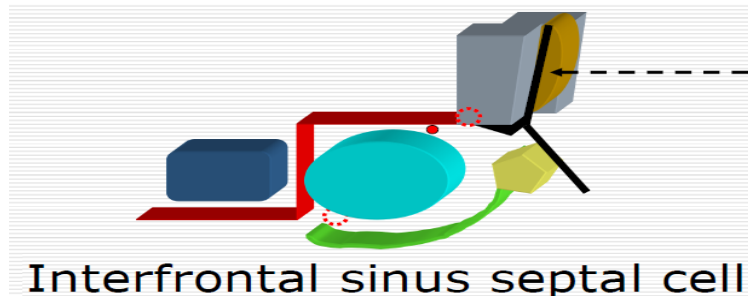


Figure 46.2g Suprabullar cell. A cell above the ethmoid bulla that pneumatizes along the skull base but does not enter into the frontal sinus. In this example, there is both a suprabullar (SB) cell and a frontal bulla (FB) cell. AN, agger nasi



frontal bulla (FB) cell.



Intersinus Septal cell (ISSC).medially based related to the frontal sinus septum, which opens into the frontal recess

- Surgeon should understand the anatomy of the frontal sinus & carefully plan the surgical approach and to identify each cell In CT to be dissected as well as to understand the drainage pathway of the frontal sinus facilitating a safe dissection.
- **Indications for Frontal Sinus Surgery**
 - Chronic Rhinosinusitis “clinical and radiologic evidence of disease in the frontal recess and sinus” that is nonresponsive to medical therapy at **least a 2-month**.
 - Course of maximal medical treatment that includes :
 1. Antibiotics for 2-3 weeks.
 2. Saline douches.
 3. Topical nasal steroids.
 4. Oral steroids.
 - Frontal pain or headaches should not be thought of as the only surgical indication for endoscopic frontal sinusotomy.
 - Mucosal disease in the frontal sinus without symptoms of headache can still contribute to the other symptoms of CRS such as “nasal obstruction, anosmia, postnasal drip, and rhinorrhea.”
 - The philosophy that frontal sinus disease should not be treated without specific frontal sinus pain being present has no scientific basis in the literature.

- **Pre-op Planning and Assessment**

- Pre-op CT scan of the paranasal sinuses performed in three planes.
- Each cell is confirmed by reference to the coronal, sagittal, and axial planes
- Assessed for surgical danger points:
 - Location and course of the **anterior and posterior ethmoidal arteries**.
 - Height and more importantly symmetry of the lateral lamella of the cribriform plate (**Keros classification**).
 - Angle and potential breaches of the skull base.
 - Dehiscence of the lamina papyracea.
 - Location and potential dehiscence of carotid artery and optic nerve
 - Building block model if necessary.
 - The superior attachment of the uncinate process:
 - If the uncinate attaches to the middle turbinate or to the fovea ethmoidalis, the frontal recess will be found draining into the infundibulum, Removal of the most superior aspect of the uncinate will access the frontal recess.
 - If the uncinate attaches to the lamina papyracea, as it usually does, the frontal recess will be found between the middle turbinate and the superior extent of the uncinate process.

- **OPERATIVE TECHNIQUE**

- **Mucosal sparing and maximizing the size of the frontal sinus** tract are the critical elements to successful surgery.
- The cells in the frontal recess are in close approximation and partial clearance predisposes the formation of **adhesions** with obstruction of the drainage pathway.
- Therefore, is an **all or nothing consideration** "the frontal sinus is either left alone or all cells are removed "so that the frontal ostium is completely visualized.

- **Approaches:**

- 1. Posterior:**

- Uncapping of the egg.
- Conservative and could be used in most cases.

- 2. Anterior:**

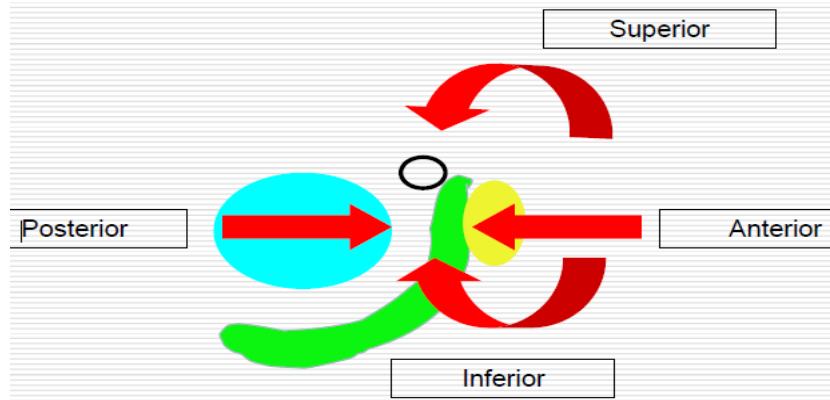
- Axillary flap.
- Direct but is reserved for difficult cases.

- 3. Inferior:**

- Intact bulla technique.
- Conservative but is reserved for limited lesions.

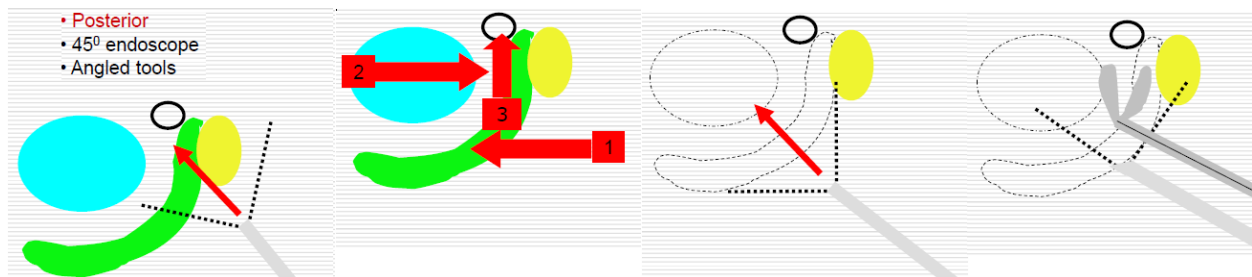
- 4. Superior:**

- Above & below technique.
- May be used as a supplementary technique in case of failure to define the site of frontal ostium.

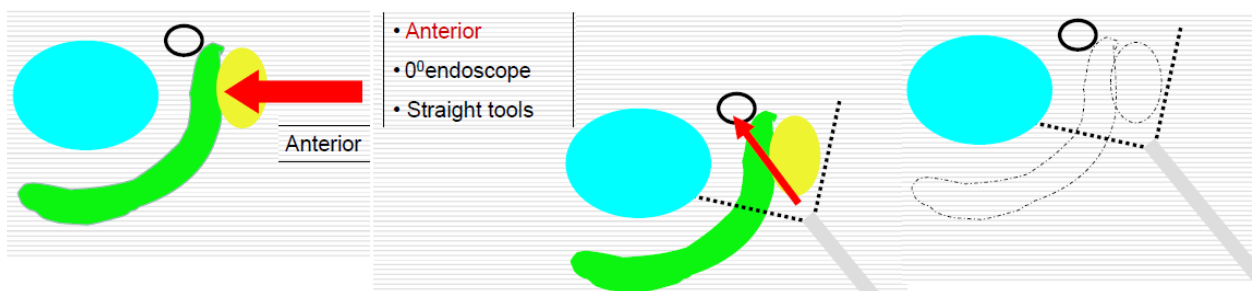


Posterior Approach	Anterior Approach (Axillary flap)
Advantages: ○ Middle turbinate stability ○ No adhesions ○ Post-operative polyps [Exposure] Disadvantages: ○ 45 degree endoscope ○ Angled instruments	Advantages: ○ Direct ○ 0 degree endoscope ○ Straight instruments Disadvantages: ○ Middle turbinate stability [lateralization] ○ Adhesions ○ Post-operative polyps [Exposure]

❖ **Posterior Approach:**



❖ **Anterior Approach (Axillary flap):**



- For accessing the frontal recess begins with:
- 1. Removal of the middle third of uncinated process.
- 2. The maxillary ostium is identified and then opened as indicated by the extent of disease involving this sinus.
- 3. The maxillary ostium allows identification of the medial orbital wall and floor of the orbit and should be the first step in all sinus surgery.
- 4. 8-mm incision superior to the axilla of the middle turbinate is extended anteriorly for 8 mm, The scalpel is turned vertically and the incision is continued inferiorly to the level of the axilla.



Figure 12.3 The incisions for the axillary flap in a left nasal cavity are outlined above the axilla of the middle turbinate.



Figure 12.4 The mucosal flap is elevated off the anterior face of the agger nasi cell (arrow) and tucked between the middle turbinate and septum.



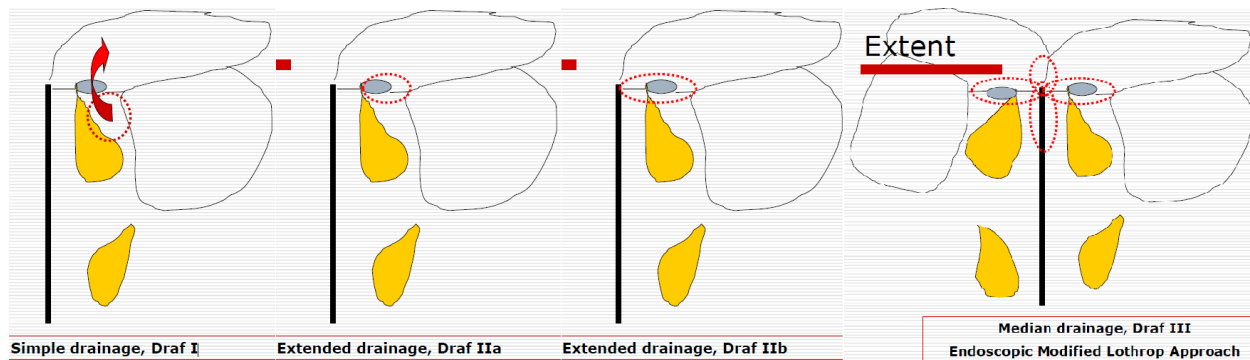
Figure 12.5 The anterior face of the agger nasi cell has been removed and the roof of the agger cell is seen (continuous arrow). The axillary flap can be seen tucked between the middle turbinate and septum (broken arrow).

- 5. Freer elevator is used to raise the flap, which is tucked medial to the middle turbinate.
- 6. Typically one or two "bites" with the rongeur are sufficient to remove the anterior face of the agger nasi cells allowing for direct visualization into the frontal recess.
- 7. A frontal sinus probe or malleable suction curette is placed behind the posterior wall of the agger nasi cell into the previously identified drainage pathway.
- 8. If the drainage pathway is posterior, cells are curetted from a posterior to anterior direction (away from the skull base) and if it is medial then curettage is medial to lateral (away from the lateral lamella of the cribriform plate).
- 9. To enlarge the frontal sinus drainage pathway, fragments of bone and soft tissue are removed with a microdebrider or an up-biting Blakesley or giraffe sinus forceps.
- 10. All the cells in the frontal recess and protruding through the frontal ostium are removed.
- 11. At the end of the dissection, the frontal ostium should be clearly visible.

- The surgical limits, therefore:
 - Frontal "beak" anteriorly
 - Skull base posteriorly
 - Middle turbinate medially
 - Lamina papyracea laterally.

Endoscopic Drainage of the Frontal Sinus [Draf]:

- **Extent of surgery depends on the extent of the pathology**
- 1. **Draf I [Simple Drainage]:**
Most Frequent.
- 2. **Draf IIa-b [Extended Drainage]:**
Extensive disease.
- 3. **Draf III , Endoscopic Modified Lothrop , frontal drillout [Median Drainage]:**
Recurrent resistant disease, failed Draf IIa-b.



**TABLE
46.1**

DRAF CLASSIFICATION OF ENDONASAL FRONTAL SINUS PROCEDURES

Type	Extent of Surgery	Indication
I	Anterior ethmoidectomy with drainage of the frontal recess by removal of obstructing disease inferior to the frontal ostium. The frontal ostium itself and any cells protruding into it are not touched	• History, endoscopy, and CT findings suggest chronic frontal sinusitis is due to sinus outflow tract obstruction at the level of the frontal recess
IIA	Removal of all ethmoidal cells protruding into the frontal sinus thereby creating an opening between the middle turbinate medially and the lamina papyracea laterally	• Complicated frontal sinusitis or failed Draf I • Recommended in frontal sinuses with a large A-P diameter (anticipated minimum diameter of frontal neo-ostium 5mm or more), hypoplastic internal nasal spine (small frontal beak), and a broad ethmoid
IIB	Removal of the frontal sinus floor between the nasal septum medially and the lamina papyracea laterally	• Complicated frontal sinusitis in frontal sinuses with a small A-P diameter, hyperplastic internal nasal spine (i.e., large frontal beak), or narrow ethmoid
III	Bilateral Draf IIB with removal of the upper part of the nasal septum and intersinus septum	• As for Draf IIB recommended over type II sinusotomy for cases with severe polyposis

- **Draf III/Endoscopic Modified Lothrop Procedure [EMLP] /Frontal Drillout:**

- Now well established.
- It is an effective intranasal approach to the treatment of frontal sinus disorders.
- The operation aims to reestablish normal or near-normal frontal sinus function.
- Frontal ostium patency rates of greater than 95%
- Significant improvement in symptoms noted in 82%.
- The overall failure rate was 13.9%.
- 1.2% of patients reported a worsening of symptoms.
- Serious complication rate of less than 1%.
- Draf III/EMLP has proved effective in the treatment of following :
 - Failed prior ESS (NOT PRIMARY)
 - Frontal sinus mucocele
 - Fractures of the frontoethmoid complex
 - Repair of CSF leaks and meningoencephaloceles
 - Failed osteoplastic flap frontal sinus obliteration
 - Resection of sinonasal neoplasms

MINI-TREPHINATION "Above & below technique":

- ✓ Frontal sinus mini-trephination is a particularly helpful technique when the drainage pathway is not seen.
- ✓ Cannula placed in the frontal sinus allows the instillation of fluorescein-stained saline.
- ✓ The stained fluid can then be followed in the frontal recess, allowing a probe to be passed into the frontal sinus, facilitating careful clearance of the frontal recess.
- ✓ facilitating irrigation of the neo-ostium postoperatively to prevent adhesion formation and stenosis



- **Complications of Frontal Sinus Surgery**

1. Orbital Complications:

- Most common.
- Potential damage to the eye, ocular muscles, optic nerve.

2. Hemorrhage:

- Anterior ethmoidal artery injury either from the nose or into the eye with proptosis.
- AEA is generally found behind the anterior face of the bulla ethmoidalis unless a suprabullar cell or recess exists.

3. CSF leak (cribriform plate, and skull base).

4. Lacrimal sac injury.

5. Iatrogenic injury with **scarring** of the frontal recess and/or incomplete removal of all cells in the frontal recess can also lead to persistence or worsening of symptoms.

POSTOPERATIVE CARE

- Careful postoperative management is crucial.

- Postoperative regimen consist usually of:

- **Oral antibiotics:**

- 10 days for frontal sinusotomy.
- 3 weeks for EMLP.
- Intraoperative cultures are checked and the antibiotic regimen changed to culture-directed antibiotics if required.

- **Saline irrigation:**

- Four to six times daily.

- **Topical and Oral steroids:**

- For 3 weeks on a tapering schedule for sinonasal polyposis.
- Where mini-trephines are placed for frontal sinusotomies, 5 mL of saline flushes every 2 hours, with every second flush, 0.5 to 1 mL of prednisolone solution is added.

- The patient is reviewed at **10 to 14 days postoperatively** "crusts removed and adhesions divided.
- A thin-curved suction is passed into the frontal sinus and the sinus cleaned, if all appear to be healing well, the patient is reviewed **4 to 6 weeks** later.

- **CONTROVERSIES IN FRONTAL SINUS SURGERY**

- The extent of frontal sinus surgery in CRS is controversial.
- Techniques such as [Balloon Sinuplasty](#) offer a minimally invasive approach to the sinuses but is not as yet supported by the literature.
- Some surgeons only performing endoscopic frontal sinusotomy when patients have symptoms of frontal headache or congestion.
- Bailey said "frontal sinus should be treated no differently to any of the other sinuses", Mucosal disease in the frontal sinus without symptoms of headache can still contribute to the other symptoms of CRS such as nasal obstruction, anosmia, postnasal drip, and rhinorrhea.
- The extent of frontal sinus surgery required for high-risk CRS patients such as Samter triad is being investigated, these patients have a higher rate of failure from standard endoscopic frontal sinusotomy and appear to do better with more extensive surgery such as the EMLP (unpublished data).

External Sinus Surgery

- The development of endoscopic endonasal techniques in sinus surgery has dramatically reduced the indications for external approaches.
- However, persistent indications for open approaches to the facial skeleton include:
 - Facial trauma surgery
 - External anterior ethmoidal artery ligation.
 - Orthognathic surgery
- In some circumstances, the open approach is combined with endoscopic endonasal techniques to treat **sinonasal pathologies and skull base lesions**.
- Therefore, even with the evolution of endoscopic endonasal surgery, it is imperative for a sinonasal surgeon to be familiar with external approaches.

MAXILLARY SINUSES:

- **Approach:** **Caldwell-Luc procedure** “described by two surgeons George Caldwell and Henri Luc”.
- Caldwell-Luc “**anterior transmaxillary approach**” which requires an incision in the superior gingivolabial sulcus and anterior maxillary antrostomy to access the maxillary sinus”.
- Remained the principal surgical intervention to treat **inflammatory maxillary sinus disease** until the advance of endoscopic endonasal techniques in the mid-1980s and the concept of a middle meatus antrostomy at the natural ostium of the maxillary sinus .
- The procedure is not performed anymore as originally describe.
- Remains a very useful procedure for treating patients with facial fractures and foreign bodies in the maxillary antrum.

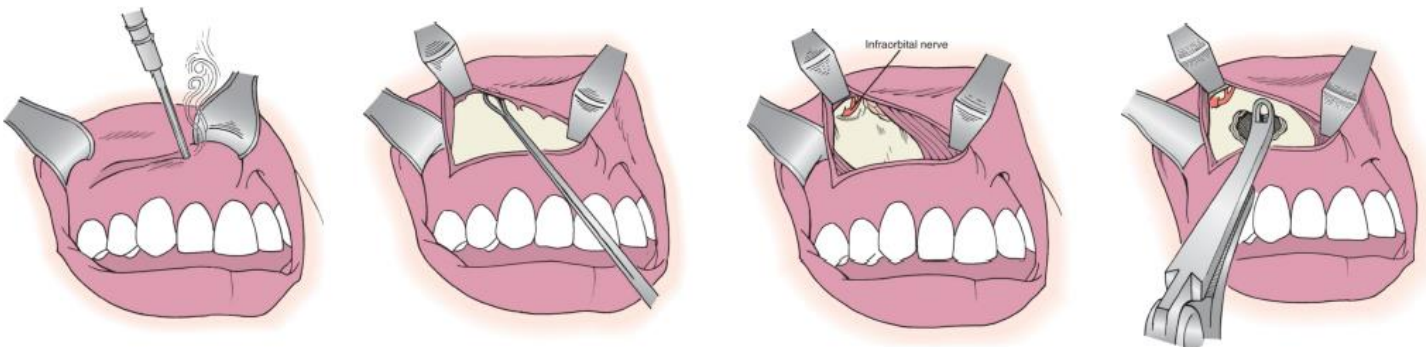
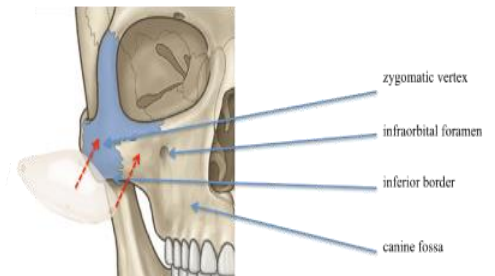
➤ **Indications**

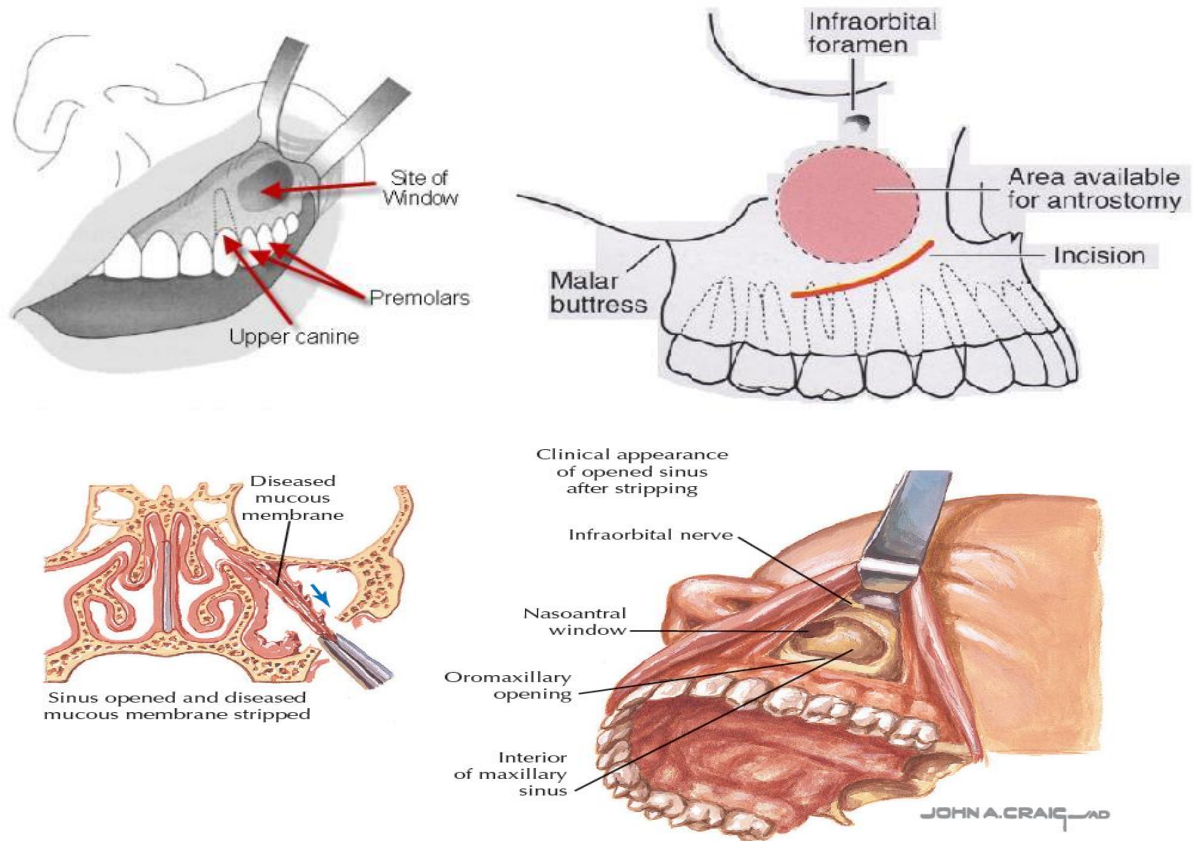
- **Adjunct to endoscopic approaches for :**
 - Sinonasal tumors (involvement of the **anterior, inferior, and posterolateral walls** of the maxillary sinus) such as:
 - Antrocoanal polyp.
 - Juvenile nasopharyngeal angiofibromas.
 - Inverted papillomas.
- **Infraorbital nerve resection:**
 - Adenoid cystic carcinoma, squamous cell carcinoma.
- **Corridor to reach:**
 - Pterygopalatine fossa.
 - Lateral recess of the sphenoid sinus (trans-antral sphenoethmoidectomy).
 - Masticator space.
 - Infratemporal fossa.

- **ORIF of comminuted orbital floor fractures.**
- **Oroantral fistula closure.**
- **Retrieval of FB.**
- **Chronic maxillary sinusitis that fails to respond to medical and conventional ESS treatment strategies "Kartagener's syndrome / Young's syndrome "**
- **Dental tumors.**
- **Orthognathic surgery.**
- **Augmentation of the maxillary alveolus "Edentulous patients with insufficient maxillary alveolar bone "**
- **Mycotic maxillary sinusitis**

➤ **Operative Technique:**

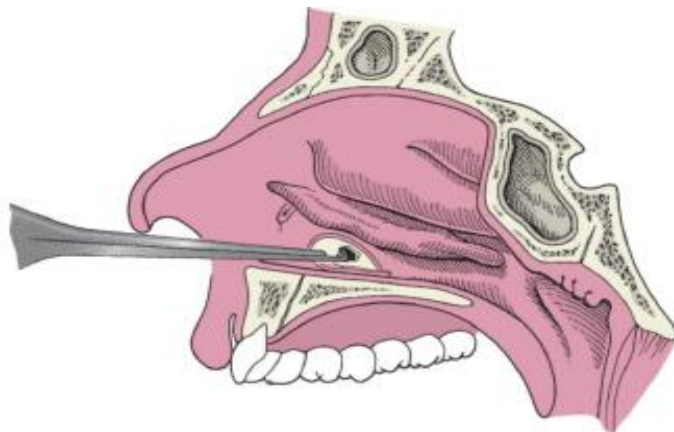
- Infiltration with local anesthetic with vasoconstrictor.
- Preserve 5 mm of gingival "to facilitate closure of the mucosa".
- Sublabial incision sited at the bucco-gingival sulcus is carried from the canine tooth to the first molar.
- Cautery till periosteum.
- Elevate periosteum up to infraorbital nerve.
- Removal of bone window in the "**canine fossa**" which is an area of thin bone just superior to the root of the canine tooth below the infraorbital nerve.
- Drill or Mallet and osteotome used to enter the sinus; enlarged with Kerrison rongeurs to the margins of the sinus (pyriform sinus, orbital rim, maxillary alveolus).
- Care is taken to avoid injury to the infraorbital nerve superiorly and dental roots inferiorly, in pediatric patients.
- At the end of the procedure, the anterior wall is not reconstructed and the incision is dosed in one layer with absorbable suture.





○ **Inferior meatal antrostomy:**

- Useful when extensive resection of mucoperiosteal lining of the maxillary sinus is performed.
- Provide drainage of secretions during the healing process.
- Can be used to irrigate maxillary sinus antrum facilitating hygiene.
- Residual hematoma following surgery can be evacuated.
- The maxillary sinus cavity can be periodically inspected for evidence of recurrence of disease through this opening.



➤ **Complications:**

- Facial edema “minimized with elevation of the head by 30 degrees and application of an ice pack”.
- Wound infection.
- Wound dehiscence with **oroantral fistula**.
- Bleeding with hematoma formation.
- Infraorbital nerve injury (transient or permanent numbness of the cheek and upper lip).
- Scar contracture of the cheek.
- Asymmetry of the facial skeleton (pediatric patients).
- Disruption of dental roots (pediatric patients).

➤ **Post-op care:**

- Packs if used should be removed within 48 hours.
- Broad spectrum antibiotics to be prescribed if packs are used.
- Regular douching of nose with saline nasal spray will prevent crust formation.

➤ **Pitfalls:**

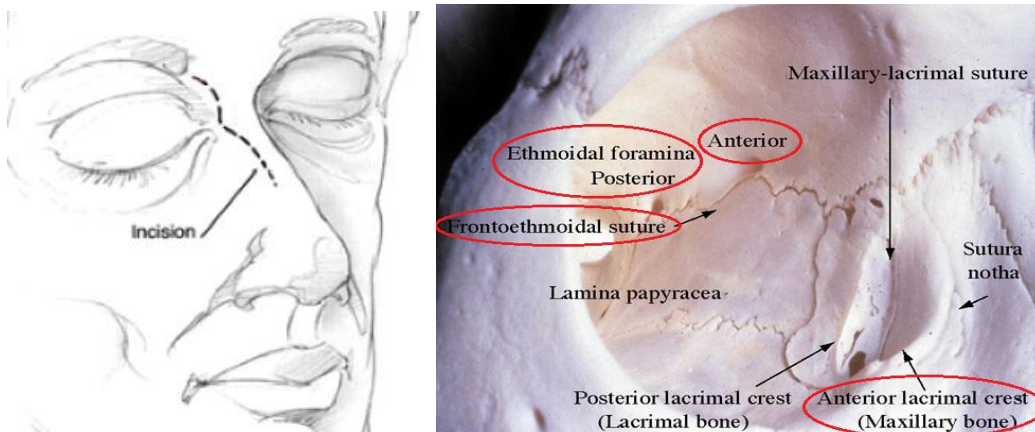
- Natural mucociliary clearance is disrupted “Regenerating maxillary antral mucosa lacks cilia”.
- Post-op CT scans are challenging to interpret because the resultant fibrosis will cause misleading artifacts.
- Difficult to perform in patients with maxillary sinus hypoplasia (commonly encountered).

ETHMOID SINUSES

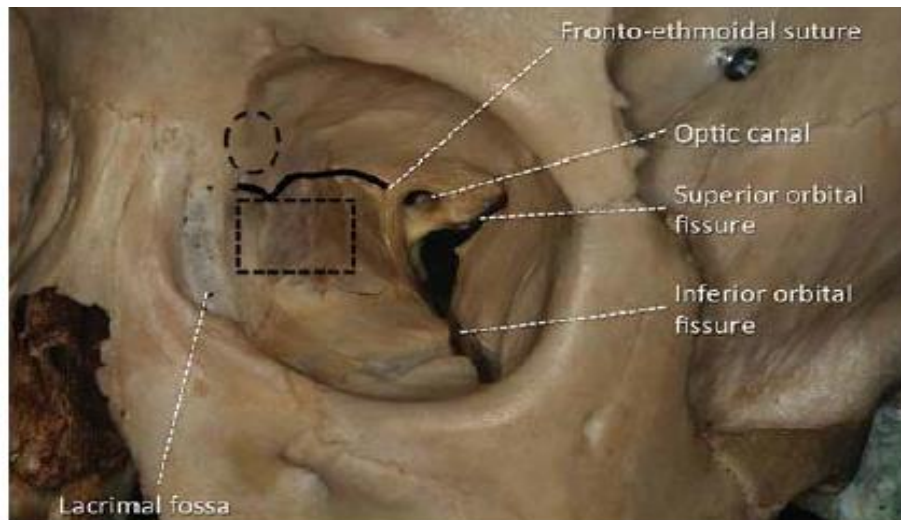
- The natural corridor to approach the ethmoid sinuses is the nasal cavity.
- Even before the advent of the endoscopes, ethmoid sinus surgeries have been performed transnasally, with or without magnification (microscope).
- Endoscopic endonasal techniques provide improved access and visualization and are generally preferred.
- External approaches (**external ethmoidectomy**) to the ethmoid sinuses are rarely indicated but can be useful in selected patients for:
 1. Resection of malignant tumors.
 2. Repair of complex midfacial fractures.
 3. External ligation of the anterior ethmoidal artery.
 4. Adjunct to frontal sinus surgery.
- External ethmoidectomy provide excellent access to:-
 - Ethmoid sinuses
 - Medial orbit
 - Cribriform plate
 - Fronto-nasal area "inferomedial aspect of the frontal sinus".

➤ **Operative Technique**

- The external approach to the ethmoid sinuses starts with a **Lynch incision**.
- The incision begins at the **inferior margin of the medial aspect of the eyebrow** and extends inferiorly along the lateral nose midway between the nasal dorsum and the medial canthus to the level of the medial canthus.
- Incision down to periosteum, Subperiosteal dissection is performed.
- Medial wall of orbit exposed along the frontoethmoidal suture line until the **anterior ethmoid artery** is seen (24 mm from anterior lacrimal crest); divide or bipolar the artery.



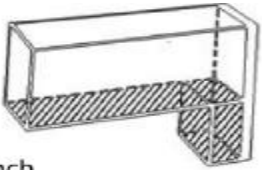
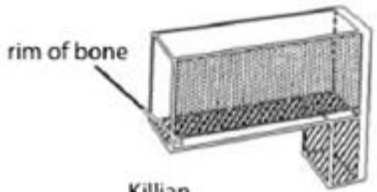
- Continue dissection until posterior ethmoid artery is seen (12 mm) posterior to the anterior artery.
- Optic nerve is 6 mm (1-8 mm range) from post ethmoid artery but dissection back here is not performed.
- Lamina papyracea is punctured and debrided with a Kerrison.
- The level of the skull base is defined by the frontoethmoidal suture line bone is removed **inferior** to this line.
- Remember that the cribriform plate is often lower than the roof of the ethmoid sinus; careful dissection is necessary to prevent injury to the lateral lamella of the cribriform and a CSF leak.
- Pack loosely and close.

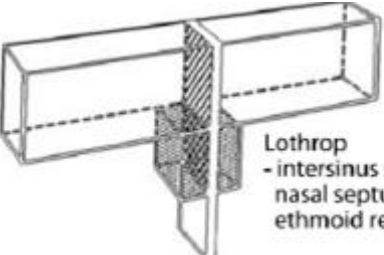


➤ **Complications:**

- 1. Epiphora** "From injury to the nasolacrimal apparatus".
- 2. Diplopia:**
 - Temporary is common and usually self-limited as edema resolves.
 - Horizontal gaze diplopia, however; may result as a consequence of trauma to the medial rectus muscle with subsequent fibrosis.
 - Blindness is a potential consequence.
- 3. Retro-bulbar hemorrhage.**
- 4. Optic nerve injury.**
- 5. CSF leak** "If the fovea ethmoidalis or cribriform plate is disrupted".
- 6. Supratrochlear nerve injury** "Anesthesia, hypesthesia, or neuralgia of the median forehead".
- 7. Hypertrophic scar formation.**

FRONTAL SINUSE

Approach	Walls Resected	Comments
Reidel	➤ Complete removal of: ○ Anterior wall frontal sinus ○ Floor of frontal sinus  Reidel anterior wall and floor removed	○ Method of frontal sinus ablation. ○ Forehead concavity. ○ <u>Performed in:</u> ○ Small frontal sinus "collapse can be mild". ○ When osteoplastic flap cannot be raised. ○ Anterior table osteomyelitis.
Lynch (External fronto-ethmoidectomy)	○ Floor of frontal sinus. ○ Portions of lamina papyracea. ○ Anterior Ethmoidectomy.  Lynch - floor and anterior ethmoid removed via medial canthal incision.	○ Removes lateral bony wall allowing the orbit to prolapse medially; does not provide easy access to the natural ostium (typically medially). ○ It does not cause any facial deformity
Killian (Modified Riedel)	➤ Complete removal of: ○ Anterior wall frontal sinus ○ Floor of frontal sinus (preservation of supraorbital ridge to reduce deformity. ○ Anterior ethmoidectomy  rim of bone Killian - ethmoidectomy performed - small rim of bone left at supraorbital rim.	○ Forehead deformity, easier reconstruction than Riedel; same problems as Lynch

Approach	Walls Resected	Comments
Osteoplast -ic Flap	Anterior wall frontal sinus based inferiorly retracted and replaced with periosteum intact.  Osteoplastic	- Large incisions, excellent exposure. - Good Eradication of disease and mucosa. - Mucosal preservation without obliteration may be possible. - Obliteration failure rates increase over time (up to 25%). - Post obliteration imaging is unreliable.
Lothrop Method	- Creates a large drainage opening into the nasal cavity by removing: Bilateral anterior ethmoids Superior nasal septum Middle turbinate Frontal septum	 Lothrop - intersinus septum, superior nasal septum and anterior ethmoid removed.

➤ **Indications**

- As an adjunct for acute frontal sinusitis
- Intracranial complications from sinusitis
- Frontal sinus trauma, fractures and CSF leaks
- Failed endoscopic frontal sinusotomy
- Intractable mucosal disease
- Lateral frontal mucocele
- Access to orbital roof for fracture repair

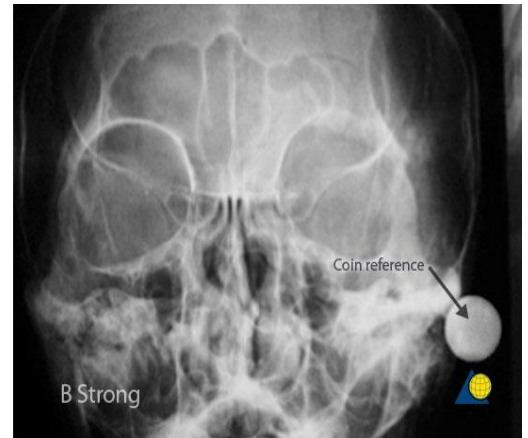
➤ **Trephination**

○ **Indications:**

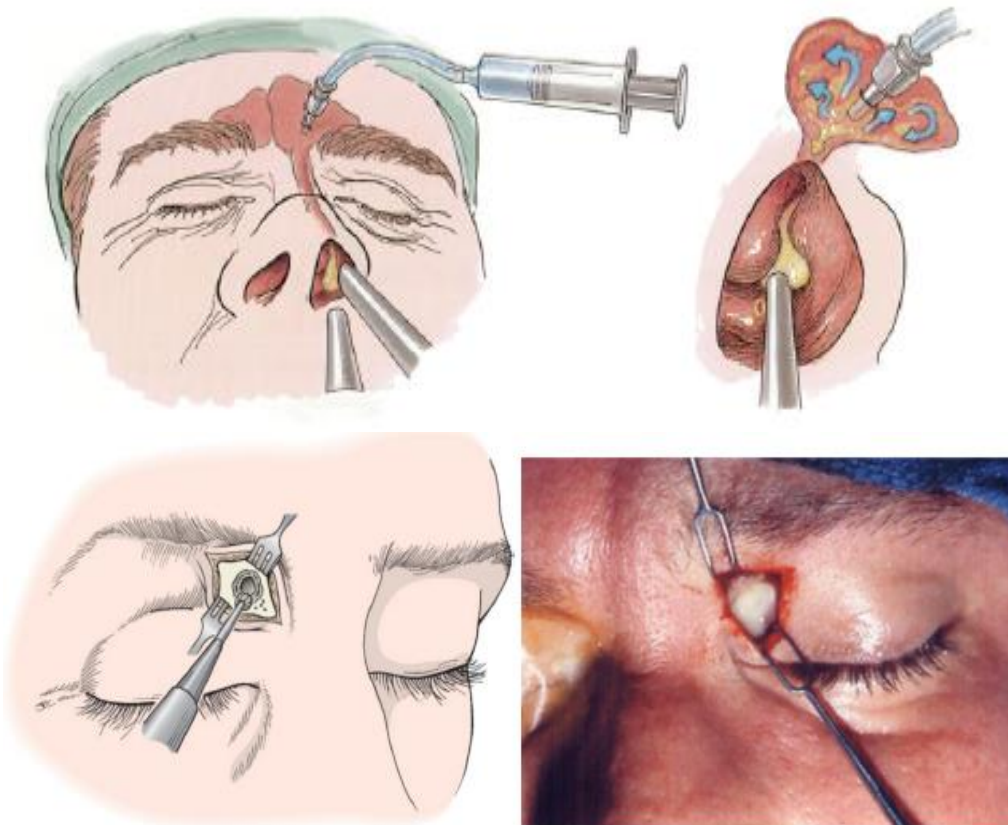
- Relieves acute frontal sinusitis that fails to resolve with medical therapy
- Mucocele
- Type 4 frontal cell
- Tumor biopsy and cultures
- when there are complications of acute frontal sinusitis
- Adjunct to ESS identify the ostium intranasally (**above and below approach**).

- **For incision planning:**

1. Image guidance (navigation).
2. 6-foot Caldwell plain film (occipitofrontal view) **with coin reference.**
3. Trans-illumination "Trephination can be made and an endoscope passed into the sinus to transilluminate its borders".



- Incision → inferomedial aspect of the eyebrow.
- A 4- to 5-mm diameter hole is drilled through the anterior table.
- Culture pus; irrigate with abx solution.
- Examine with 30 degree scope.
- 8-French red rubber catheter or other tubing may be used for 2 days for post-op irrigation.
- Catheter can be removed and the trephination allowed to close by secondary intention.
- Trephination should not be performed if pneumatization does not reach up to the superior limit of orbit.



- **Complications:**

1. Brain injury
2. Cellulitis
3. Orbital complications due to needle shift

- **Eyebrow Incision**

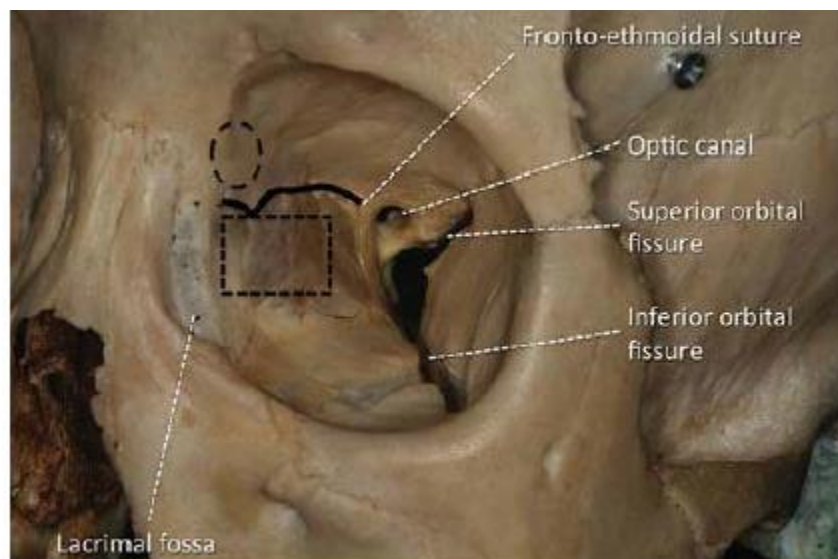
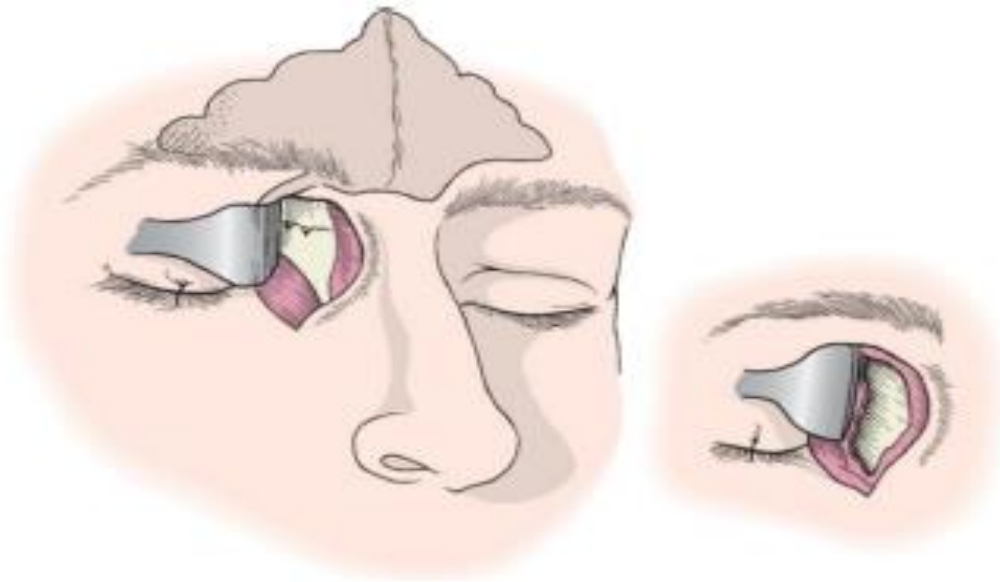
- Gives direct access to the superior orbital rim and anterior wall of the frontal sinus.
- The incision can be placed within the at the angle of the hair follicles.
- Subperiosteal plane is necessary to identify and avoid injury to the supratrochlear and supraorbital arteries and nerves.
- The eyebrow approach is useful for lesions localized **laterally** in the frontal sinus when well pneumatized.
- If the bone is not replaced, the defect is reconstructed with titanium mesh to avoid retraction of the soft tissues and a depression in the forehead.
- The use of image guidance system (navigation) to prevent inappropriate opening of the intracranial.



- **External fronto-ethmoidectomy "Lynch procedure":**

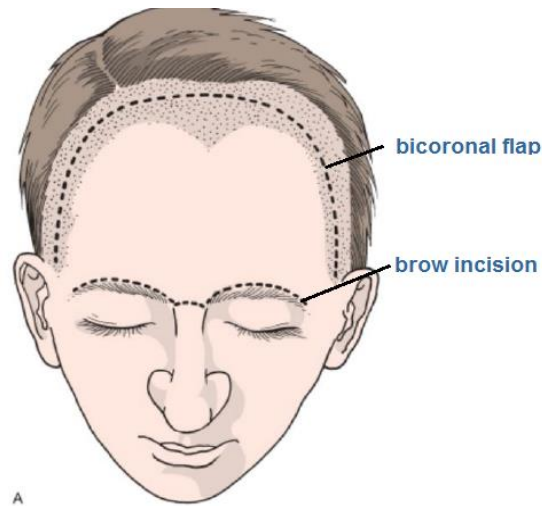
- Rarely used.
- Consists of removal of the frontal sinus floor, middle turbinate, and anterior ethmoids.
- It provides access to the:
 1. Medial orbit
 2. Ethmoid sinus
 3. Inferomedial aspect of the frontal sinus
- Easiest and quickest technique.
- Good for wide exposure.
- Risk of recurrent mucocele formation from stenosis of nasofrontal duct.
- Access same as **External ethmoidectomy** (see Operative Technique above), except incision carried into brow for frontal access.
- The frontal sinus may be accessed by (see pic below)
 - Drilling the bone of the orbital roof medially.
 - Opening the ethmoid sinus below the frontoethmoidal suture line and following the nasofrontal recess to the frontal sinus.

- Stent placement to maintain nasofrontal duct patency & prevent mucocele formation.
- Supraorbital and supra trochlear nerve is at risk.



- **Osteoplastic flap:**

- Approaches include bicoronal, brow or midforehead incisions.



- **Indications:**

- Chronic frontal sinusitis with persistent intractable symptoms, sepsis, or other complications despite previous intervention.
- Large Mucocele with Orbital or Intracranial extension.
- Osteomyelitis, subperiosteal or epidural abscesses.
- Frontal sinus tumor "lateral extension or involvement of the anterior wall will necessitate an open approach".
- Frontal sinus fracture with comminuted anterior table or displaced posterior table.
- Posttraumatic CSF leak "fractures of the posterior table are more easily repaired".
- Osteoma.

- **Contraindication:**

- Aplastic frontal sinus

- **Advantages:**

- Best view of entire frontal sinus and anterior base of skull.
- Minimal deformity.
- Direct approach.
- Safe method to eradicate frontal sinus disease (permanent and complete removal of diseased mucosa).

- **Disadvantages:**

- Technically more difficult.
- Time consuming.
- Requires hospitalization.
- Risk of mucocele formation and chronic pain.

○ **Two osteoplastic options can be considered:**

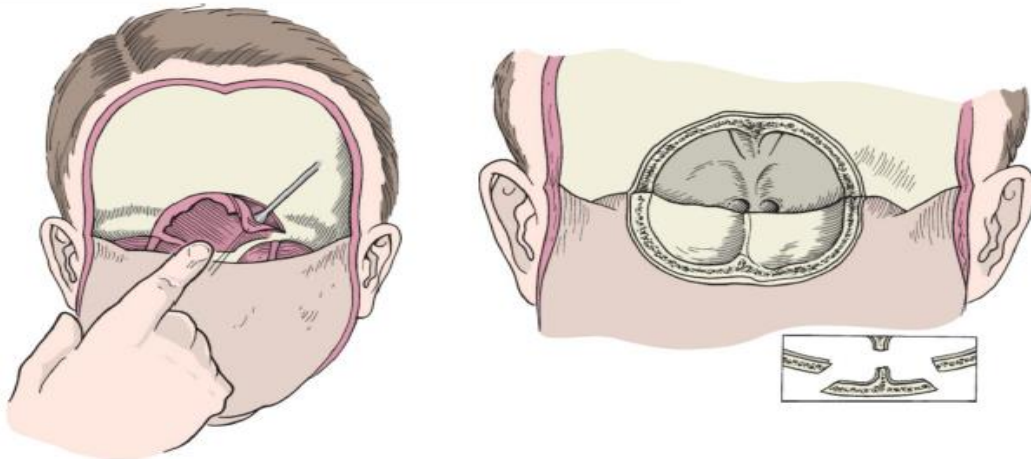
1. Obliteration of the sinus.
2. Removal of a lesion in a mucosal-sparing fashion combined with subsequent endoscopic frontal recess dissection to reestablish good drainage into the frontal sinus.
 - Advantage of permitting both endoscopic and CT evaluation for recurrent disease postoperatively.
 - Best in cases of tumors, such as inverting papilloma with frontal sinus involvement.

Frontal Sinus Obliteration:

- In general, frontal sinus obliteration should be performed rarely as a result of the ever-increasing reported success rates of endoscopic approaches.
- Considered when:
 - Frontal recess obstruction cannot be relieved by more conservative techniques "small recess".
 - When the mucosal disease is diffuse and irreversible.
- Requires careful drilling of **all the mucosa** in the frontal and supraorbital ethmoid cells to eliminate remnants of mucosa within the **foramina of Breschet** "small venules that drain the sinus mucosa into the dural veins" thus minimizing the possibility of recurrence and/or mucocoele.
- Obliteration the cavity and occluding frontonasal recess with **fat** has the advantage of being easy to remove when it became infected and re-exploration required.
- If symptoms persist, CT and MRI each only provide very limited information in the obliterated frontal sinus; obliteration with fat is generally preferable to other materials such as **bone chips, hydroxyapatite, methylmethacrylate, bioactive glass** because these are more difficult to remove when re-exploration is necessary.
- Although frontal sinus obliteration has a high success rate of 93% at 8 years, complications are relatively high, occurring in over 20% of patients.
- **Contraindications obliteration of the sinus:**
 1. Hyperpneumatized supraorbital ethmoid cells
 2. Fungal sinusitis
 3. Inverting papilloma or other sinonasal tumors that need repeated imaging to r/o recurrence "relative contraindications".
 4. Posterior frontal table or orbital roof dehiscence.

○ **Operative Technique:**

- A bicoronal flap is elevated, leaving the pericranium intact to expose the anterior table of the frontal sinuses.
- The incision extends from the tragus of one ear to the other & 1 inch posterior to the hairline.
- Alternatively, a mid-forehead or brow incision can be used.
- In either case, it is important to not elevate the pericranium while exposing the frontal sinuses because the former is needed to create the osteoplastic flap, decrease infection and protect vital structures.
- Careful dissection should be performed close to the superior orbital rim to avoid injury to the [supraorbital neurovascular bundles](#).
- The pericranium is incised around the borders of the frontal sinus.
- The inferior rim of the pericranium should be left intact because this will be the hinge of the osteoplastic flap "trapdoor access".
- Once the sinuses are opened, any diseased mucosa or tumors can be removed.
- The bone flap is then secured using a microplating system, and the periosteum is closed with an absorbable suture.
- The skin is closed in two layers over a suction drain.



○ **Cranialization "sinusectomy sinusectomy":**

- Removal of the posterior frontal sinus wall allowing the brain to come forward and obliterate the frontal sinus.
- Reserved for severe posterior frontal sinus wall fracture with significant bony loss.

- **Complications (15%)**

- Seroma
- Chronic pain
- Persistence of symptoms
- Frontal sinus mucocele
- Orbital hematoma
- Diplopia
- Abscess formation in either the frontal or abdominal wound
- Dural exposure or dural laceration (CSF leak)
- Intracranial bleed
- Nasal skin necrosis
- Anosmia
- Temporary ptosis
- Cosmetic deformity
- Hyperesthesia (supraorbital or supratrochlear)

- **Postoperative management**

- Long-term follow-up for mucocele is essential for obliteration.
-
- 93% had resolution of symptoms postoperatively.
 - Revision surgery was required in 6% of patients due to recurrent frontal sinus infection.
 - The best way to determine whether a mucocele or rhinosinusitis has recurred in symptomatic patients complaining of headaches is to obtain an **MRI with fat suppression**.
 - The amount of adipose tissue seen within the obliterated will decrease over time Because the fat is nonvascularized, it may shrink with healing and result in incomplete obliteration.

SPHENOID SINUS

- **Approach:**

- **Superior:**

- Through the planum sphenoidale is usually part of a craniofacial translocation.

- **Lateral:**

- Infratemporal access to the sphenoid sinus between the second and third divisions of the trigeminal nerve.
 - Very limited in exposure.

- **Anterior:**

1. Transantral
2. transseptal
3. external transtethmoidal

- **Indications:**

- Access to and excision of pituitary lesions
 - Invasive fungal sphenoid sinusitis
 - Access to the anterior cranial base or clivus
 - CSF leak repair
 - Mucosal disease resistant to medical and ESS management
 - tumors involving sphenoid sinus

- **Anatomy and operative technique**

- Types of pneumatization:

- Sellar 86%
 - Pre-Sellar 11%
 - Concha

- Intersinus septum is rarely in midline
 - Natural ostium located behind the inferior border of the superior turbinate.

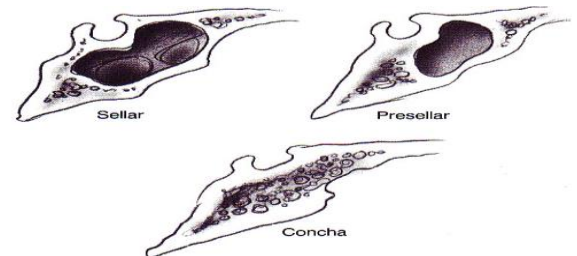
- Planum sphenoidale "Sella turcica" is above the roof of the sinus "anterior cranial fossa".

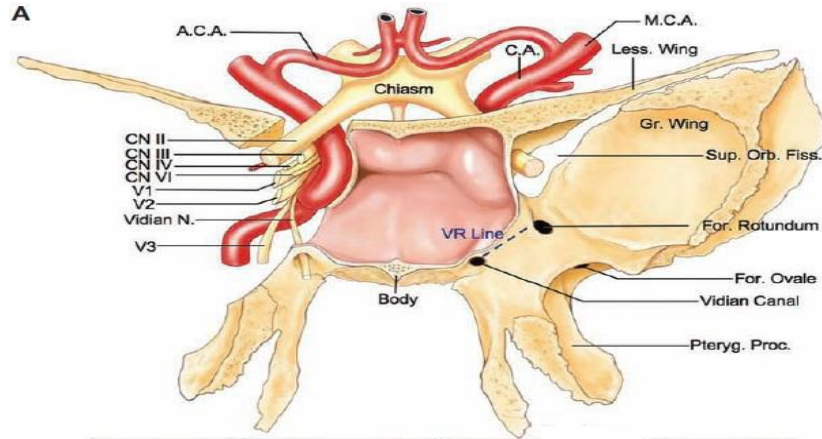
- Clivus is posterior "posterior cranial fossa".

- Cavernous sinus is lateral with ICA and CN III-VI; "middle cranial fossa".

- 4% of cases, ICA canal is dehiscant within the sinus.

- Vidian canal lies below and lateral.



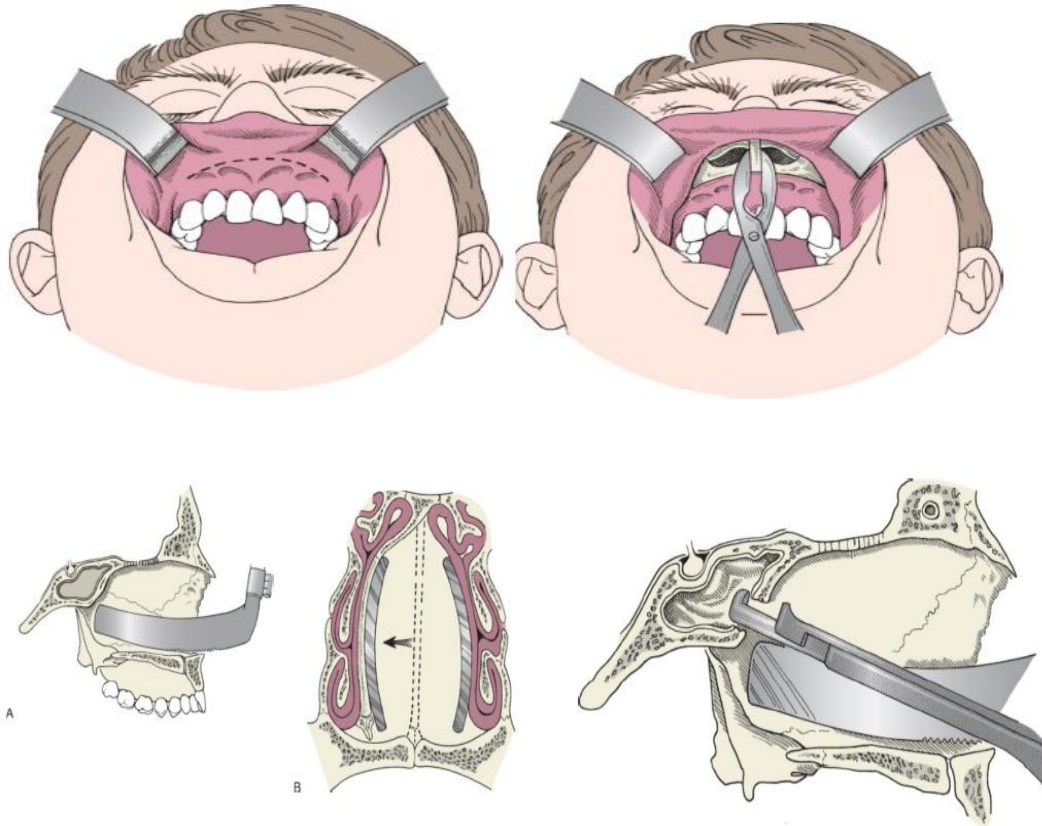


- **Trans-septal Approaches**

- Trans-septal good for smaller lesions.
- Can be accessed through:
 1. Trans-nasal (Hemitransfixion or External rhinoplasty)
 2. Sub-labial
 3. External rhinoplasty transeptal approach
 4. Columellar flap modification approach

- ✓ **Sublabial transseptal approach**

- Commonest trans-septal approach.
- Incision is made 5-10mm superior to the gingiva and is carried down to the bone of premaxilla.
- The periosteum is elevated up to the inferior margin of pyriform aperture.
- Anterior nasal spine is exposed "It can be fractured for exposure but left attached to the septum".
- Anterior & inferior tunnels are created over nasal septum by elevating mucoperichondrium performed until the rostrum and anterior sphenoid
- Cartilaginous portion of nasal septum is dislocated from the floor and pushed to one side
- Perpendicular plate of ethmoid and maxillary crest displaced to one side.
- Inferior turbinate can be out fractured for creating more space.
- Transsphenoidal speculum inserted for exposure.
- Operating microscope or endoscope "posterior septum, rostrum, and anterior sphenoid wall is removed with a drill".
- Sinus entered in the midline; intersinus septa often insert into the ICA canal.



○ Advantage:

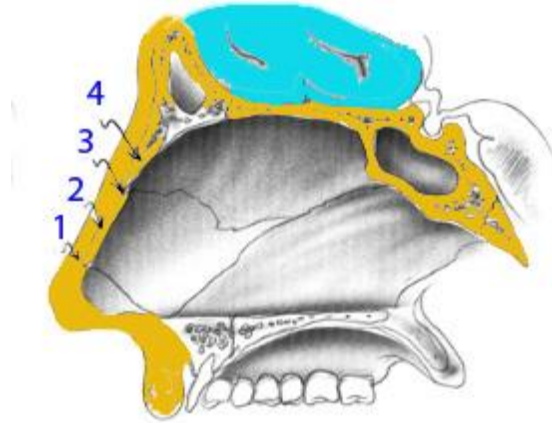
- Easy procedure
- Scarless
- Use of midline speculum increases visibility
- Minimal post op nasal deformity
- Suited for nasal cavity of any size

○ Disadvantage:

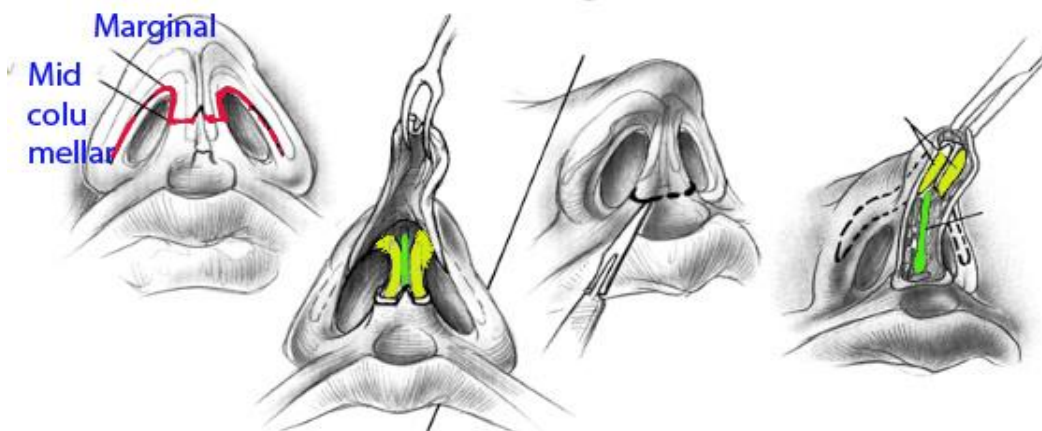
- Oral contamination of wound
- Incisions may cause problems with dentures
- Dental complications like devitalization of teeth is a possibility

✓ **Transnasal transseptal approach:-**

- This approach is without sublabial incision.
- Avoiding the morbidity of the sublabial dissection.
- Allows direct access to the rostrum of sphenoid.
- Incisions used include:
 1. Hemitransfixation
 2. Killian's
 3. Vertical
 4. Bony cartilaginous junction .
- This approach may not be suitable for small noses because of difficulties faced in inserting the speculum



- Advantage:
 - Oral cavity contamination is avoided
 - Scarless
 - Septal incisions can be placed anteriorly / posteriorly
 - Posterior incisions are useful in septal reoperations
 - Disadvantage:
 - High risk of nasal disfigurement
 - Requires meticulous post op wound care
 - Ideally suited only for large nasal cavities
 - Columellar incision scar may be visible in some patients
- ✓ **External rhinoplasty transseptal approach**
- Medial crura are separated after incising the intercrural ligaments.
 - Exposure of caudal edge of quadrilateral cartilage



- Advantage:
 - Exposure is excellent
 - Midline approach
 - Oral cavity contamination is avoided
 - Nasal deformities present preoperatively can also be corrected
 - Can be used in noses of any size

✓ **Columellar flap modification:**

- No separation of medial crura
- Columellar flap contains the medial crura

• **Complications**

- Synechia
- Saddle nose deformity
- Tip deformities
- Septal perforations
- Rhinosinusitis
- Chronic rhinitis with crusting
- ICA injury
- Optic nerve injury
- CSF leak
- Brain herniation
- Meningitis
- Pneumocephalus
- Ophthalmic artery injury

SinoNasal Neoplasms:

- Difficult to differentiate between nasal and paranasal tumors except in early stages.
- Unilateral nasal obstruction is the most common symptom in patients with benign or malignant sinonasal tumors.
 - o Should be assessed with endoscopy, imaging studies, histologic examination to establish an accurate diagnosis.

TABLE 129.1 DIAGNOSIS PARANASAL TUMORS TECHNIQUE	
History and Physical	Risk Factors/Cranial Nerve Deficits, Nasal Mass
Imaging	
CT scanning	Evaluation of bony boundaries of PNS; not cost-effective
MRI	Evaluation of soft tissue invasion and perineural spread; differentiate retained secretions from tumor
PET	Routine evaluation for recurrent disease after primary treatment and distant metastasis; useful for SCCA; cost-effectiveness unknown
Biopsy	
Sinus lavage/cytology	
Fine needle aspiration	
Transnasal biopsy	Direct or endoscopic. Preferred modality

- Benign tumors:
 - o Smooth, localized and covered with mucous membrane.
- Malignant tumors:
 - o Friable, have a granular surface and tend to bleed easily.

TABLE 129.2 TUMORS OF THE SINONASAL TRACT	
Benign	Malignant
Epithelial	Epithelial
Exophytic papilloma	Squamous cell carcinoma
Inverted papilloma	Transitional cell carcinoma
Columnar papilloma	Adenocarcinoma
Adenoma	Adenoid cystic carcinoma
	Melanoma
Nonepithelial	Olfactory neuroblastoma
Fibroma	Undifferentiated carcinoma
Chondroma	
Osteoma	Nonepithelial
Neurilemmoma	Soft tissue sarcoma
Neurofibroma	Rhabdomyosarcoma
Hemangioma	Leiomyosarcoma
	Fibrosarcoma
	Liposarcoma
	Angiosarcoma
	Myxosarcoma
	Hemangiopericytoma
	Connective tissue sarcoma
	Chondrosarcoma
	Osteosarcoma
	Lymphoreticular tumors
	Lymphoma
	Plasmacytoma
	Giant cell tumor
	Metastatic carcinoma

Benign SinoNasal Neoplasms:

- **Osteoma:**
 - Most common benign sinonasal neoplasm.
 - Non-epithelial tumor.
- **Inverted papilloma:**
 - 2nd most common benign sinonasal neoplasm.
 - Most common indication of surgery in benign sinonasal neoplasm.
 - Epithelial tumor.
- **Juvenile Angiofibroma:**
 - 3rd most common benign sinonasal neoplasm.
 - Non-epithelial tumor.

TABLE 48-1. Distribution of Benign Tumors of the Sinonasal Tract by Histology and Surgical Approach

Tumor Histology	Surgical Approach			Total
	Endoscopic	Combined	External	
Inverted papilloma	403	41	12	456
Osteoma	109	39	11	159
Juvenile angiofibroma	120	1	34	155
Lobular capillary hemangioma	50	1	—	51
Fibrous dysplasia	26	3	1	30
Cavernous hemangioma	11	1	—	12
Schwannoma	7	3	1	11
Ossifying fibroma	8	—	1	9
Hamartoma	9	—	—	9
Glioma	5	1	1	7
Pleomorphic adenoma	5	—	—	5
Miscellaneous	20	7	—	27
Total	773	97	61	931

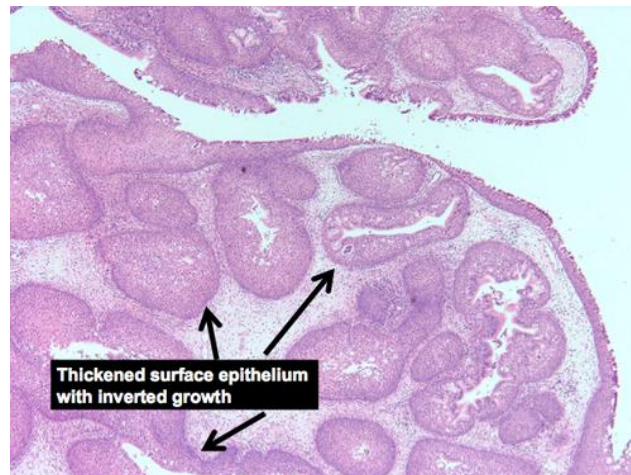
Data from a series of 931 patients treated at the University Hospitals of Brescia and Varese (Italy) from 1994 to 2013.

Benign Epithelial SinoNasal Neoplasms:

- **Squamous (Schneiderian) Papilloma:**
- Also called **Keratotic Papilloma**.
- Benign epithelial lesions arise from squamous or schneiderian (nasal) epithelium.
- **Subtypes:**
 - 1. Inverted:**
 - Originates from lateral nasal wall.
 - **Most common subtype (50%).**
 - **Highest recurrence rate (27-73%)**
 - Associated with **HPV-6, HPV-11 and EBV.**
 - **Association with malignant transformation (SCC) in 10% of patients.**
 - 2. Fungiform (Exophytic):**
 - Originates from nasal septum.
 - 2nd most common subtype (47%).
 - Recurrence rate 25-50%
 - Associated with **HPV-6 and HPV-11.**
 - Low association with malignant transformation (SCC) in 5% of patients.
 - 3. Cylindrical (Oncocytic):**
 - Originates from lateral nasal wall.
 - Least common (3%).
 - Recurrence rate 25-35%
 - Not associated with HPV.
 - **Association with malignant transformation (SCC) in 10% of patients.**
- **Clinical picture:**
 - Verrucous lesion.
 - Commonly on Nasal vestibule.
 - Often multiple and painless.
- **Diagnosis:**
 - Anterior Rhinoscopy
 - Nasal Endoscopy
 - Biopsy confirms the diagnosis.
- **Management:**
 - Simple excision or Laser ablation.
 - In septal keratotic papillomas, a cuff of normal mucoperichondrium should be taken with lesion to avoid recurrence.



- **Inverted Papilloma:**
- 2nd most common benign sinonasal neoplasm (after osteomas).
- Most common indication of surgery in benign sinonasal neoplasm.
- Most commonly diagnosed in white males (above 50 years).
- **Histopathology:**
 - Arise from proliferation of squamous epithelium through fingerlike projections into underlying stroma (rather than on the surface).
 - Associated most commonly with **HPV-6, HPV-11 and EBV.**
- **Benign pathology with locally aggressive behavior.**
 - Extends beyond its site of origin and destroy bone into sinuses, orbit and skull base.
 - Recur if not excised completely (Recurrence rate 27-73%)
 - 15% Associated with Malignancy (SCC).



- **Clinical picture:**
 - Unilateral polyp originates from **Lateral Nasal wall.**
 - Red or grey masses.
 - Nasal obstruction.
 - Epistaxis.
 - Rhinosinusitis.
 - Symptoms of complication and extension:
 - Diplopia.
 - Proptosis
 - Epiphora
 - Persistent headache
 - CSF leak
 - Often misdiagnosed as a simple nasal polyp:
 - More translucent.
 - Bilateral
 - Less vascular.

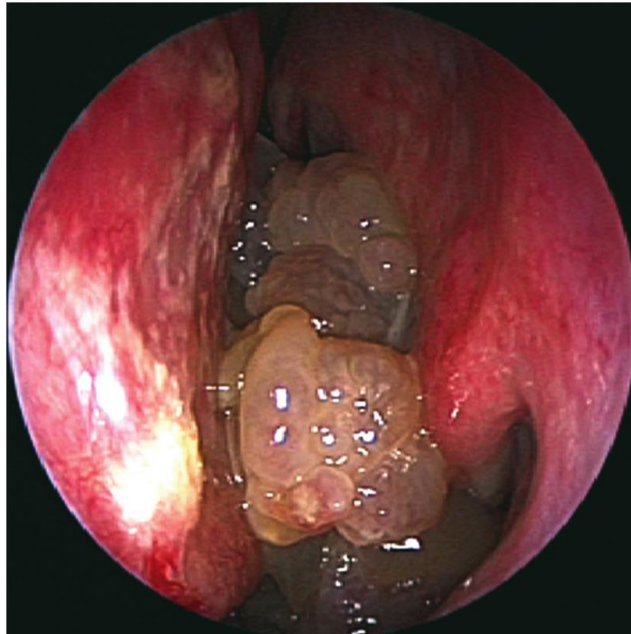
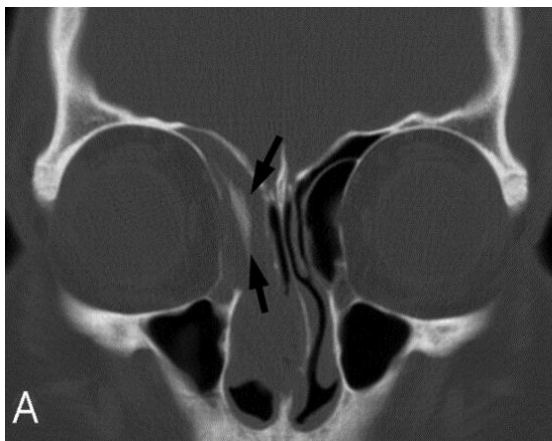


FIGURE 48-1. Typical endoscopic appearance of an inverted papilloma. A polypoid lesion with a pale, papillary surface protrudes from the middle meatus and extensively fills the left nasal cavity.

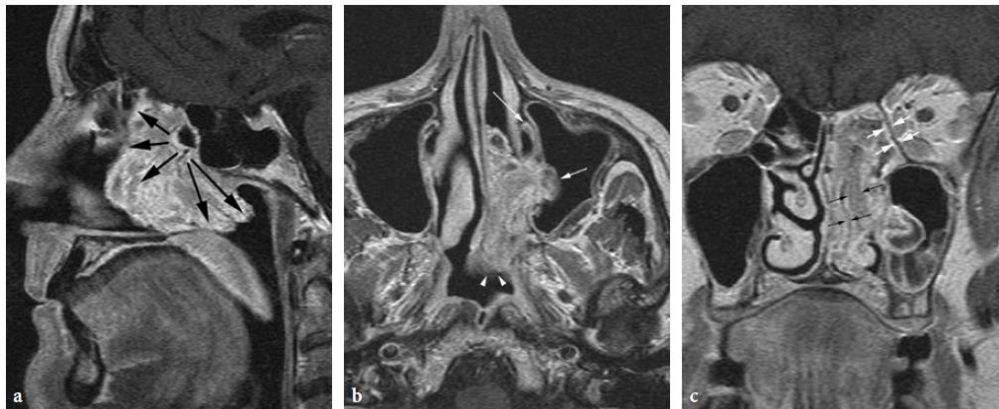
- Diagnosis:

○ **CT Scan:**

- Unilateral soft tissue density mass with some enhancement.
- Erosion into lateral nasal wall or extension into maxillary or ethmoid sinuses.
- May reveal calcifications.
- **Focal hyperostosis** indicates site of tumor origin.



- **MRI:**
 - Gold standard for evaluation of sinonasal tumors.
 - **T1:**
 - Isointense to muscle.
 - **T1 with Gad:**
 - Heterogeneous enhancement.
 - **Convoluted Cerebriform Pattern:**
 - Alternating roughly parallel lines of high and low signal.
 - Present in 50-80% of cases.
 - **T2 :**
 - HYPER-intense to muscle
 - **Convoluted Cerebriform Pattern:**
 - Alternating roughly parallel lines of high and low signal.
 - Present in 50-80% of cases.



MRI T1 with contrast



FIGURE 48-2. Inverted papilloma on an axial contrast-enhanced, T1-weighted, spin-echo magnetic resonance image. The maxillary sinus is occupied by a solid mass that protrudes into the nasal fossa through an accessory ostium. The lesion exhibits a cerebriform-columnar pattern, which is typically seen in inverted papilloma (arrows).

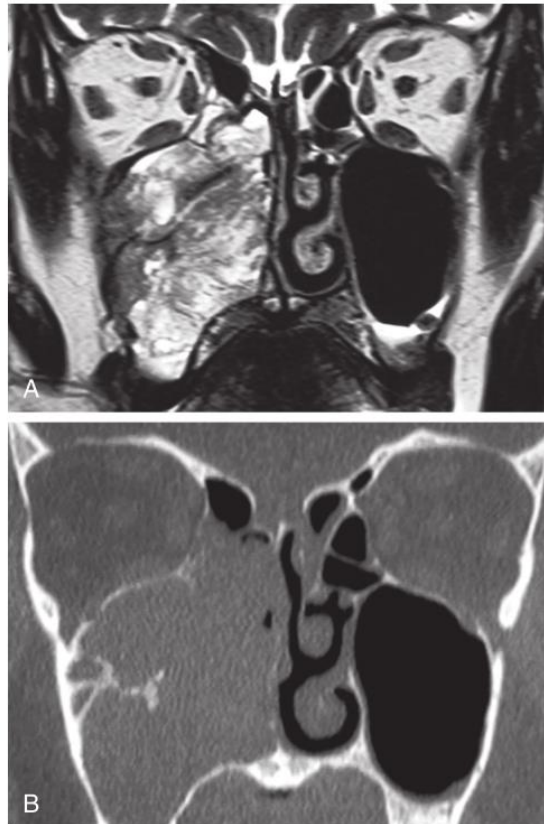


FIGURE 48-3. Inverted papilloma on a magnetic resonance image (MRI) and a computed tomography (CT) scan, both in the coronal plane. The maxillary sinus is completely occupied by an expansile lesion that destroyed the medial wall and invaded the right nasal fossa. **A**, The T2-weighted spin-echo MRI demonstrates the cerebriform-columnar pattern of the lesion and permits visualization of the bony spur along the lateral maxillary sinus wall, where the lesion originates. **B**, The CT scan does not provide a good characterization of soft tissue density opacification but gives a superior view of the sclerotic bony spur.

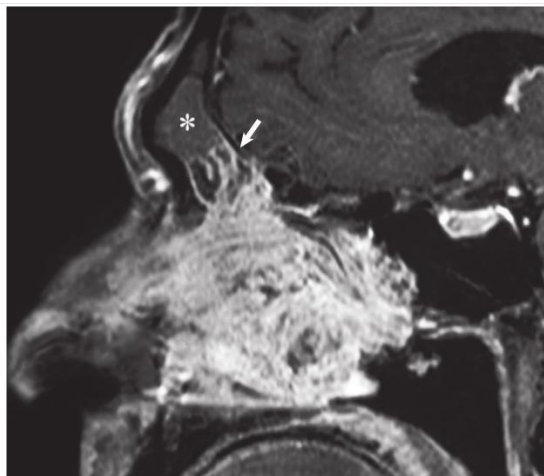


FIGURE 48-5. Inverted papilloma on a sagittal contrast-enhanced magnetic resonance image. The advantage of this acquisition plane is the excellent assessment of tumor growth into the most caudal part of the frontal sinus. The upper part of the sinus is filled with inflammatory secretions (*asterisk*). The sagittal plane also permits accurate evaluation of the relationships between the tumor and the floor of the anterior cranial fossa, which is not invaded (*arrow*).

- **Biopsy:**
 - Confirms the diagnosis.
 - Imaging should be done before biopsy to avoid possible vascular or brain injury.
 - Histopathology:
 - Endophytic growth of epithelium.
 - Cristae-laden senescent mitochondria, inflammatory cells throughout epithelium.
- Krouse staging system (No standard staging system):
 - **T1:**
 - Disease limited to nasal cavity only.
 - **T2:**
 - Disease limited to ethmoid sinuses and medial or superior aspects of maxillary sinuses.
 - **T3:**
 - Disease involves lateral or inferior aspects of maxillary sinus or extension into frontal or sphenoid sinuses.
 - **T4:**
 - Disease with extra-sinonasal extension or presence of malignancy.
- Management:
 - **Endoscopic surgical resection:**
 - Reliable alternative to traditional external approach for most cases.
 - Equal recurrence rates as traditional external resection.
 - Surgery goals:
 - Dissect involved mucosa along subperiosteal plane.
 - Drill the underlying bone.
 - Creation of large marsupialized cavity for post-op for endoscopic inspection.
 - Types of endoscopic resection:
 - **Type 1: Simple ESS:**
 - Indicated for disease involving middle meatus, ethmoid, superior meatus, sphenoid or combination of these structures.
 - **Type 2: Medial Maxillectomy:**
 - Indicated for disease involving maxillary sinus mucosa (but not the lateral or the anterior walls).
 - **Type 3: Sturman-Canfield operation/Endoscopic Denker operation:**
 - Includes removal of medial portion of anterior maxillary wall to enable access to all the antrum walls.
 - Indicated for disease involving anterior or lateral walls of maxillary sinus mucosa.

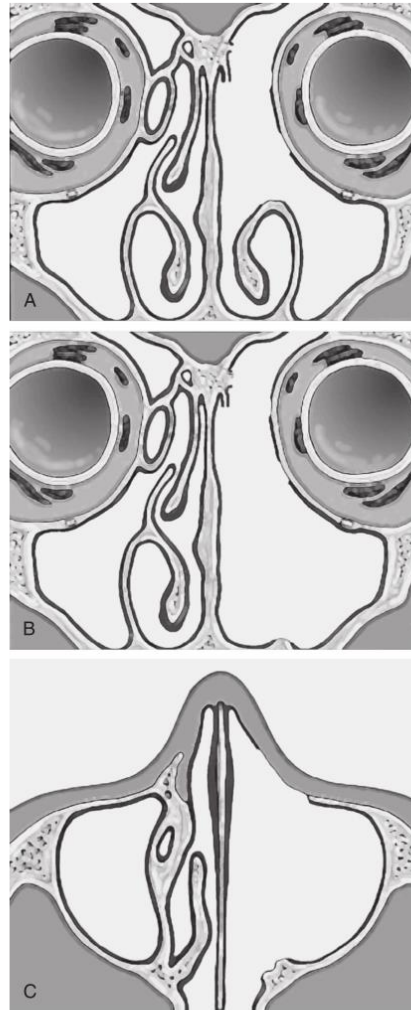


FIGURE 48-4. Schematic drawings summarize the different endoscopic resections used for inverted papillomas of the nasoethmoid-maxillary region. **A**, Type I resection. **B**, Type II resection. **C**, Type III resection.

- Frontal sinus approaches:
 - Depends on extent of frontal mucosa involvement which can be definitively assessed intra-op only.
 - Surgical approach may involve Draf IIB or III endoscopic sinusotomy or a combination of endoscopic and external approaches through osteoplastic flap.
- Contraindications of exclusive endoscopic approach:
 - Trans-orbital extension.
 - Massive frontal sinus and/or supraorbital cell involvement.
 - Malignancy that involves critical areas
 - Significant scarring and anatomic distortion from previous surgery.

- **External surgical resection:**

- Open En bloc resection with Medial maxillectomy through lateral rhinotomy or midfacial degloving approaches.



- **Radiation:**

- Indications:

- Patients with SCC
- Multiple recurrent disease
- Incompletely resectable disease.

- Post-operative follow up:

- Endoscopic inspection should done periodically for early evaluation of residual or recurrent lesions.
- High rate of recurrence rate 27-73%.
- Indications of imaging (MRI/CT):
 - Inaccessible sinus cavity for exploration
 - Symptomatic patient
 - Histological evidence of residual or recurrent lesion
- Risk factors for recurrence:
 - Incomplete resection
 - High-stage disease
 - Smoking

- Adenoma:

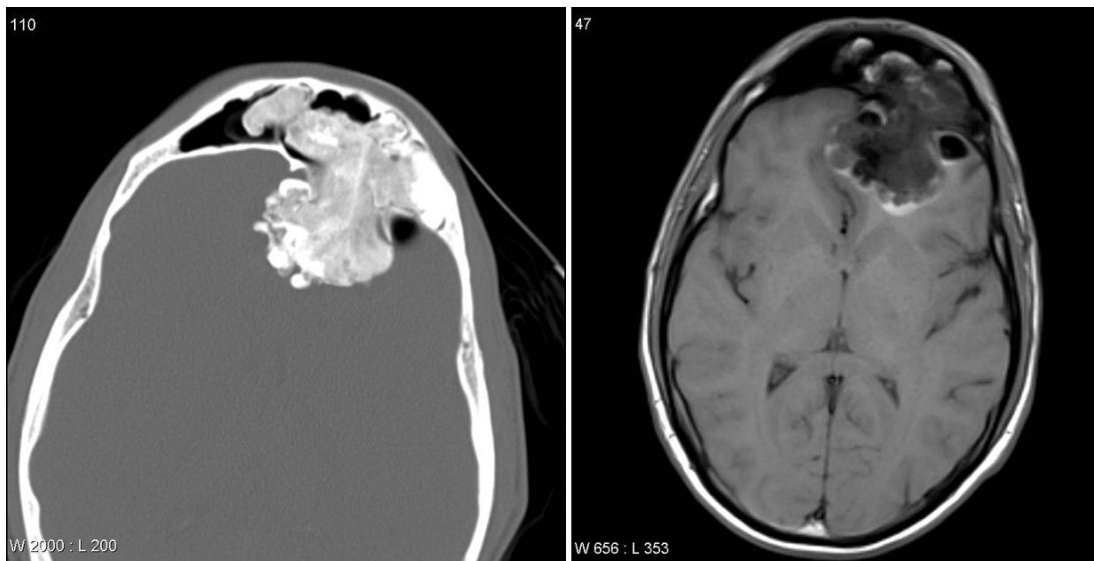
- Benign epithelial lesions arise most commonly from Nasal septum.
- 4th-7th decades
- M=F
- Treatment is wide surgical excision.
- Recurrence rate after removal is 10%.

Benign Non-Epithelial SinoNasal Neoplasms:

- **Osteomas:**
 - Most common benign sinonasal neoplasm.
 - Slow-growing osteoblastic lesion.
 - Found in 3% of patients undergoing CT for sinus symptoms.
 - Diagnosed between 2nd-5th decades of life.
 - Male preponderance commonly reported.
- **Pathophysiology:**
 - **Embryologic theory:**
 - Osteoma develops at junction between the embryonic cartilaginous ethmoid and the membranous frontal bone.
 - **Traumatic theory:**
 - Osteoma develops at site of a previous trauma.
 - **Infective theory:**
 - Osteoma develops at site of local inflammation due to osteitis and activation of osteogenesis.
- **Histological classification:**
 - **Ivory (Eburnated) osteoma (Most common):**
 - Made of compact dense bone.
 - Contains minimal amount of fibrous tissue without evidence of haversian canals.
 - **Mature (Spongiosum) osteoma:**
 - Made of spongy mature bone (resembles normal bone).
 - Bony trabeculae are divided by large amount of fibrous tissue.
 - Contains fibroblasts, collagen fibers and haversian canals (containing small vessels).
 - **Mixed osteoma:**
 - Findings typical for both ivory and mature osteoma.
- **Clinical picture:**
 - **Asymptomatic (Most common):**
 - Incidental findings during imaging for unrelated reasons.
 - **Sinusitis:**
 - Osteoma interfering with sinus drainage and causing frontal sinusitis or mucocele formation.
 - **Cosmetic deformity:**
 - Osteoma involving outer table of frontal bone.
 - **Proptosis or diplopia:**
 - Osteoma growing toward the orbit.
 - **CSF rhinorrhea:**
 - Osteoma growing into the anterior skull base.
 - **Gardner syndrome:**
 - Colon polyps, skull osteomas, PTC, epidermoids, fibromas and desmoid tumors.

- Diagnosis:

- **Endoscopic examination:**
 - Usually normal as the lesion is deeply located.
 - Rarely, osteoma grows toward nasal cavity and visualized as firm mass covered by normal or atrophic mucosa.
- **CT scan:**
 - Gold standard imaging.
 - Not requiring contrast.
 - Most common locations:
 - Frontal sinus **(80%)**.
 - Ethmoid sinuses (15%).
 - Maxillary sinus (5%).
 - Osteomas exhibit a different density depending on the amount of mineralized bone within the lesion:
 - Ivory osteomas are uniformly very dense.
 - Mature osteomas are having gradually decreasing density with marrow space sometimes visible.
- **MRI:**
 - Indicated for lesions with intra-orbital or intra-cranial extension.
 - Delineate the relationship between osteoma and adjacent soft tissues (dura, brain, orbital content).
 - Predominantly HYPO-intense in all sequences.



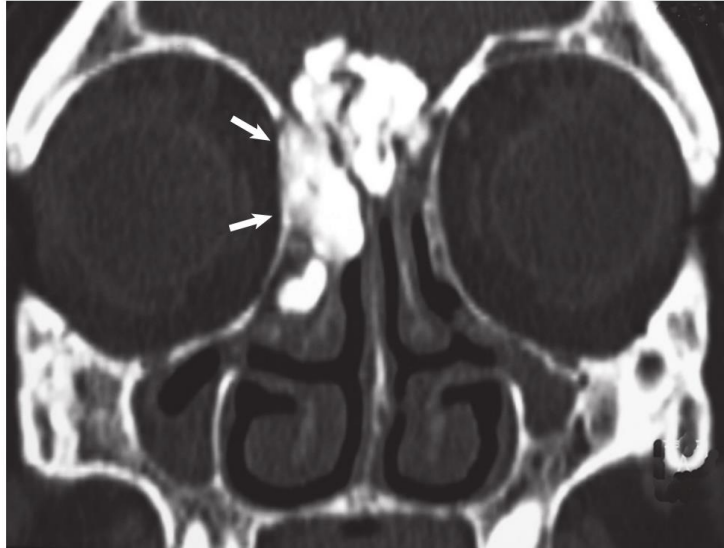


FIGURE 48-10. Osteoma on a coronal computed tomography scan. A huge ivory osteoma is shown in the right ethmoid, encroaching on the crista galli and the cribriform plate and invading the anterior cranial fossa. The right orbit is not invaded, and the thin lamina papyracea can still be detected (*arrows*).



FIGURE 48-11. Coronal computed tomography (CT) scan shows a fronto-ethmoid osteoma that exhibits a mixed CT pattern that is ivory in the anterior ethmoid and spongiosum into the frontal sinus.



FIGURE 48-12. Coronal computed tomography scan shows a right ethmoid osteoma that displays a ground-glass density typically seen in the spongiosum variant. The lesion is adjacent to the frontal recess, which is unobstructed (arrows).

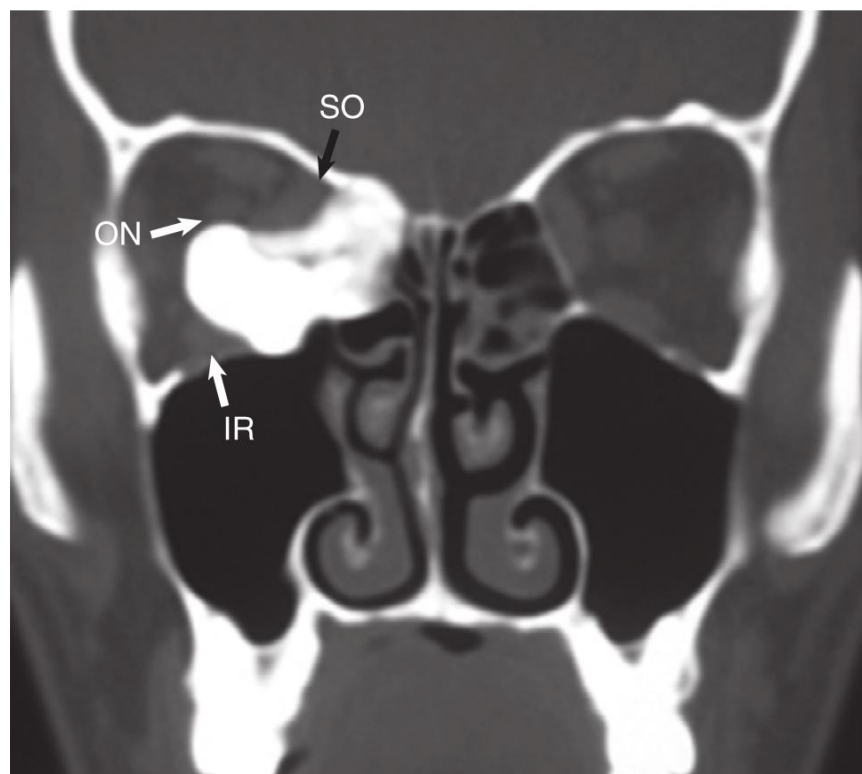


FIGURE 48-13. On a coronal computed tomography scan, a large ethmoid osteoma can be seen invading the orbit and displacing the optic nerve (ON), the superior oblique muscle (SO), and the inferior rectus muscle (IR). The high and quite homogeneous density is characteristic of the ivory variant. The lesion was resected via an endoscopic approach.

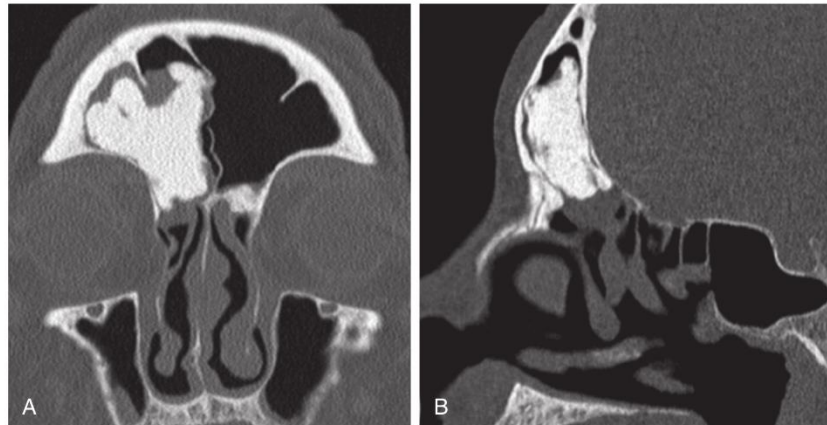


FIGURE 48-14. Axial (A) and coronal (B) computed tomography shows a large right frontal osteoma. In view of a large anteroposterior diameter and the presence of minimal residual space between the lesion and the surrounding wall, complete removal could be achieved through a Draf III procedure.

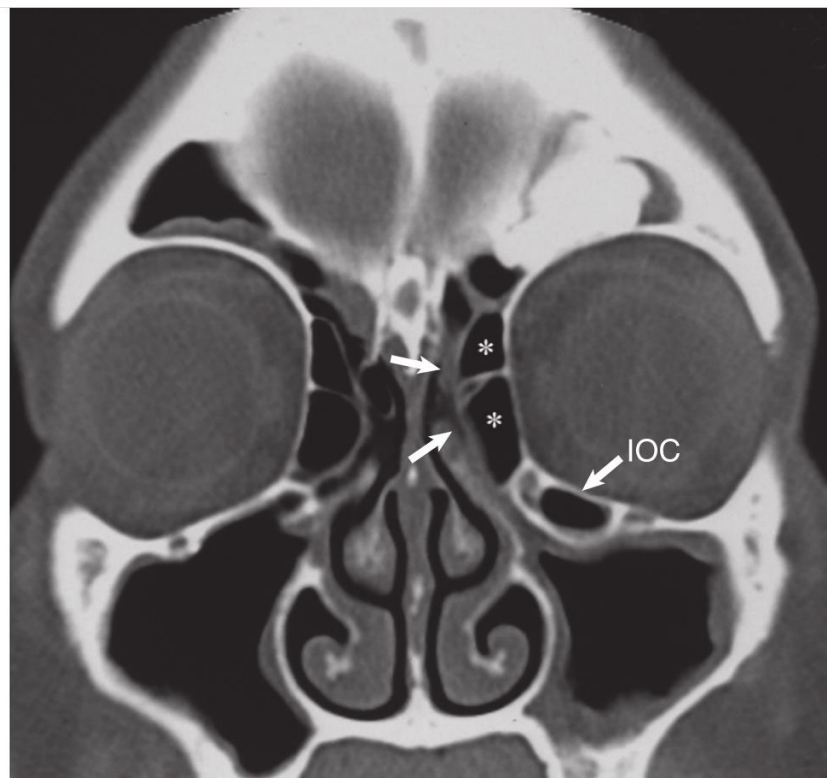
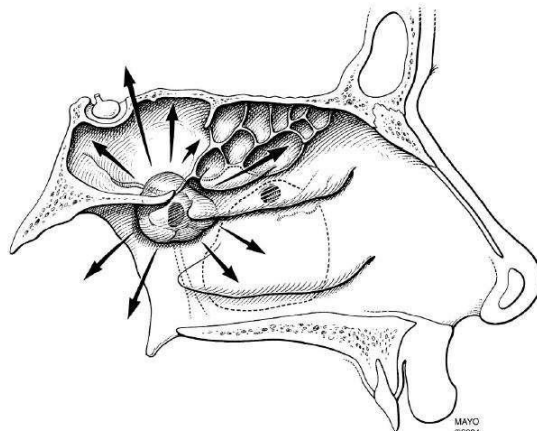


FIGURE 48-15. An ivory osteoma is shown in the most lateral aspect of the left frontal sinus in this coronal computed tomography scan. The lesion is rather distant from the frontal recess (*arrows*), which is narrowed by large agger nasi cells (*asterisks*). The osteoma was managed with a combination of an endoscopic approach with an osteoplastic frontal sinusotomy. IOC, infra-orbital cell.

- Management:
 - **Observation (wait and see):**
 - Advisable to monitor the osteoma with periodic imaging.
 - Indicated in asymptomatic patients with:
 - No intra-cranial or intra-orbital invasion.
 - Not encroaching upon critical structures (optic nerve).
 - **Surgical resection:**
 - For patients with symptomatic lesions.
 - Endoscopic approach:
 - Used for most osteomas.
 - Not helpful with far lateral osteomas in narrow frontal sinus.
 - Cavitation is a surgical trick helps resect large osteomas; core of the lesion is drilled with a cutting or diamond bur, leaving very thin shell of bone that can be easily fractured and dissected from adjacent tissues.
 - External approach:
 - Used in combination with endoscopic approach for lateral osteomas in narrow frontal sinus.
 - Frontal trephination
 - Osteoplastic flap
- Because osteoma recurrences are very rare, routine periodic postoperative surveillance by CT is not justified.

- **Juvenile Angiofibroma (JA):**
- 3rd most common benign sinonasal neoplasm.
- Most common vascular mass in the nose.
- Most common of benign nasopharyngeal neoplasms.
- Exclusive to adolescent males (14-25 years).
 - o May have hormonal element (Androgen dependent).
- **Etiology:**
 - o Originates from embryologic chondrocartilage during the development of cranial bones.
 - o Originated at pterygopalatine fossa at **superior margin of sphenopalatine foramen**, formed by junction of:
 - Palatine bone
 - Vomer
 - Root of pterygoid
- **Histology:**
 - o Non-encapsulated fibrovascular tumor.
 - o Vascular endothelium-lined spaces embedded in fibrous stroma.
 - o Lack the surrounding smooth muscle of normal blood vessels:
 - Profusely bleed when surgically manipulated (Lose the ability to contract, and cannot be controlled by application of adrenaline).
- **Pathophysiology:**
 - o Benign slow growing tumor.
 - o Locally invasive tumor with extensive bone destruction and extensive invasion of adjacent structures.
 - o Does not metastasize.
 - o Spread typically follows skull base foramina and fissures:
 - **Sphenopalatine foramen** into nasal cavity and nasopharynx.
 - **Vidian canal** into sphenoid sinus.
 - **Pterygomaxillary fissure** into infratemporal fossa.
 - **Inferior and superior orbital fissures** into orbit and intracranially.
 - **Foramen rotundum** along maxillary nerve to the parasellar region.
 - **Anteriorly** pushing posterior maxillary wall forward.



- Intracranial extension found in 10-35% of all cases, mainly from the following routes:
 - Direct extension through foramen rotundum, ovale or lacerum.
 - From infratemporal fossa directly into middle cranial fossa.
 - From pterygomaxillary fissure through inferior and superior orbital fissures into middle cranial fossa.
 - From ethmoid cells through cribriform plate into anterior cranial fossa.
 - From roof of sphenoid into the sella and medial to the cavernous sinus.
- JA receive feeding vessels from the following:
 - **External carotid artery (Most common):**
 1. Internal maxillary artery (**Mainly**).
 2. Ascending pharyngeal artery
 3. Palatine arteries
 - **Internal carotid artery (Large tumors with intracranial involvement):**
 1. Ophthalmic artery
- Clinical picture:
 - **Unilateral nasal obstruction and recurrent epistaxis:**
 - Involvement of nasal cavity and nasopharynx.
 - Small to intermediate size JA.
 - OME and CHL:
 - Blockage of the Eustachian tube.
 - Swelling of cheek:
 - Involvement of infratemporal fossa.
 - Diplopia, visual loss and proptosis:
 - Involvement of orbit.
 - Headache:
 - Involvement of cranial fossa.
- Diagnosis:
 - **Nasal Endoscopy:**
 - Smooth hyper-vascularized lesion.
 - Located in lateral nasal wall behind the middle turbinate, which is laterally displaced against the lateral wall.

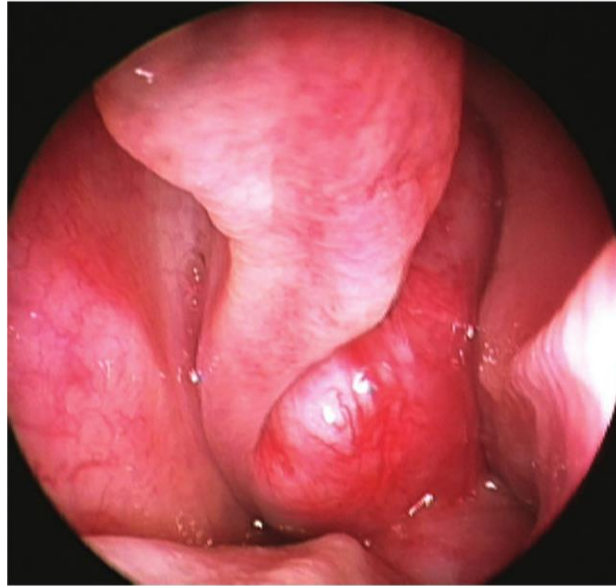
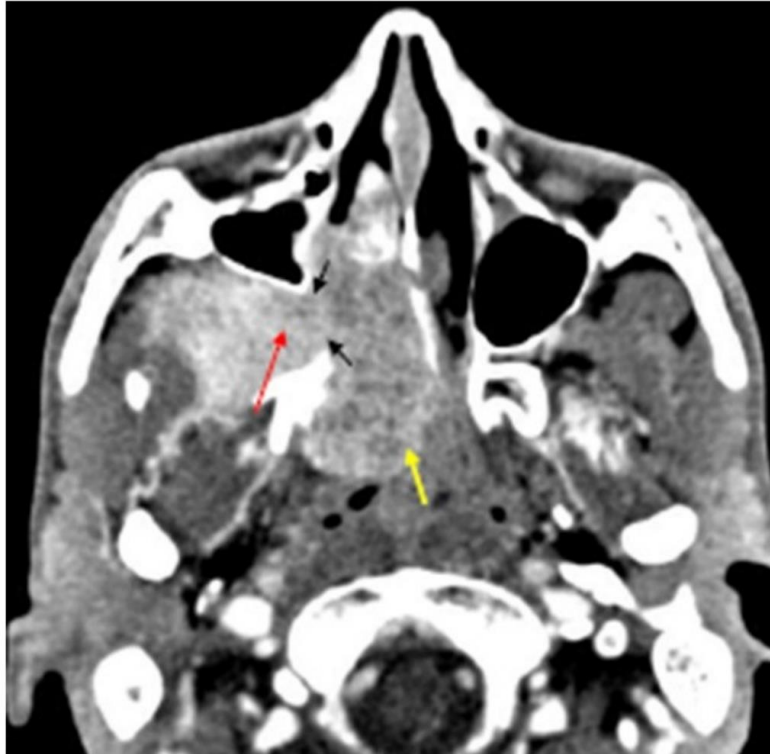


FIGURE 48-7. Juvenile angiofibroma. On endoscopy, the lesion typically appears as a polypoid hypervascularized mass that bulges from the lateral wall behind the middle turbinate, which is laterally compressed. The choana is completely obstructed.

○ **Imaging:**

- Confirm the diagnosis **without** the need for histopathology confirmation.
- Important features:
 - Originated at level of pterygopalatine fossa.
 - Prominent contrast enhancement.
 - Widened sphenopalatine foramen.
 - **Holman-Miller sign:**
 - Bowing of posterior maxillary wall anteriorly.
- CT with contrast:
 - Provides bony anatomy and degree of erosion and remodeling of sphenopalatine area and skull base.
- MRI with Gad:
 - Differentiate between tumor and surrounding soft tissues and intra-cranial structures.
 - Differentiate between tumor and sinus disease.
 - Presence of several signal voids (salt and pepper appearance) indicate major intra-lesional vessels.
 - **T1:** Intermediate signal.
 - **T1 C+ (Gd):** HYPER-intensity.
 - **T2:** Heterogeneous signal with flow voids.



Homogeneously enhancing mass lesion centered at sphenopalatine foramen (widened by the mass, shown by black arrows), extending into nasopharynx (yellow arrow) and pterygopalatine fossa (red arrow). The mass also fills ipsilateral nasal cavity, pushing the septum to left.



FIGURE 48-9. Juvenile angiofibroma on axial T2-weighted, spin-echo magnetic resonance image. The lesion invades the left orbital apex (OA), the lateral wall of the left sphenoid sinus (*arrows*) is completely destroyed, and the internal carotid artery (ICA) is encased. On the right side, a bony barrier (*arrowheads*) still separates the lesion from the internal carotid artery.

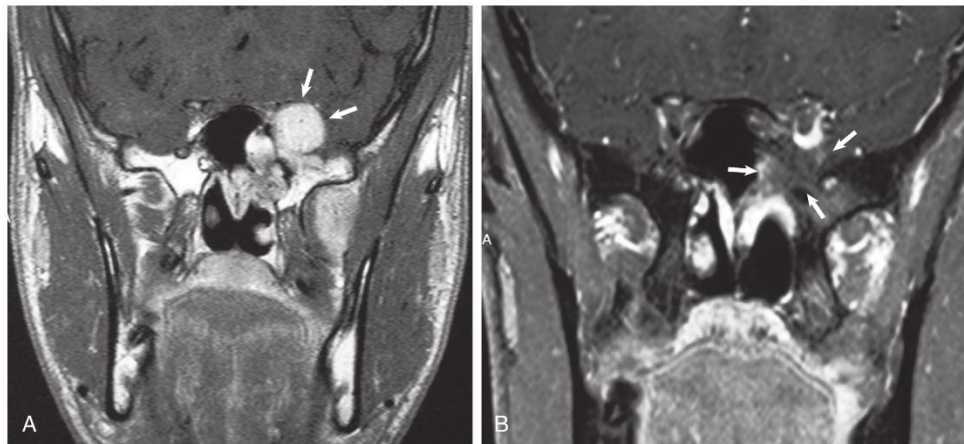


FIGURE 48-8. Juvenile angiofibroma on coronal contrast-enhanced magnetic resonance images (MRIs) obtained before and after endoscopic resection. **A,** Pretreatment image shows encroachment of both the floor and lateral wall of the left sphenoid sinus and infratemporal fossa. Intracranial extension is demonstrated in close proximity to the superior orbital fissure (*arrows*). **B,** MRI obtained after surgical resection shows solid tissue (*arrows*) along the lateral sphenoid sinus wall; the lack of contrast enhancement suggests residual scar tissue.

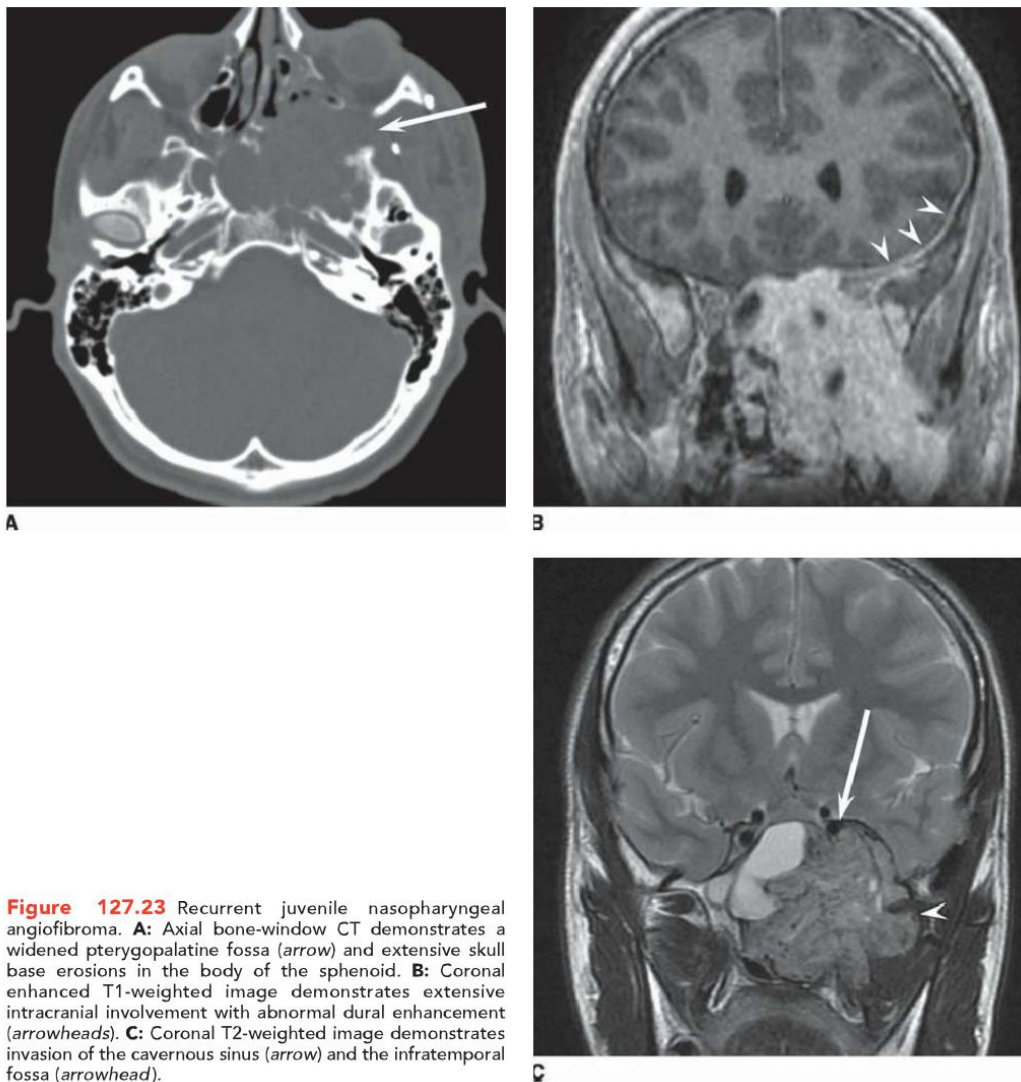


Figure 127.23 Recurrent juvenile nasopharyngeal angiofibroma. **A:** Axial bone-window CT demonstrates a widened pterygopalatine fossa (*arrow*) and extensive skull base erosions in the body of the sphenoid. **B:** Coronal enhanced T1-weighted image demonstrates extensive intracranial involvement with abnormal dural enhancement (*arrowheads*). **C:** Coronal T2-weighted image demonstrates invasion of the cavernous sinus (*arrow*) and the infratemporal fossa (*arrowhead*).

- **Biopsy:**
 - **Not required** to confirm the diagnoses in teenage boy with hyper-vascularized lesion originates behind the middle turbinate with radiological confirmation of JA due to high risk of bleeding.
 - If needed, it should be done under GA in OR with all arrangements to control bleeding and transfuse blood.
- **Staging:**
 - Multiple staging systems have been elaborated for JA and no classification system has been universally accepted.

Stages of the Fisch classification

Stage	Description
I	Tumors limited to the nasal cavity nasopharynx with no bony destruction.
II	Tumors invading the pterygomaxillary fossa, paranasal sinuses with bony destruction.
III	Tumors invading the infratemporal fossa, orbit and parasellar region remaining lateral to the cavernous sinus.
IV	Tumors with invasion to the cavernous sinus, optic chiasmal region and pituitary fossa.

**TABLE
127.7****RADOWSKI CLASSIFICATION
OF JNA**

Stage	Tumor Extent
Ia	Limited to the posterior nares and/or nasopharyngeal vault
Ib	Involving the posterior nares and/or nasopharyngeal vault with involvement of at least one paranasal sinus
IIa	Minimal lateral extension to the pterygomaxillary fossa
IIb	Full occupation of the pterygomaxillary fossa with or without erosion of orbital bones
IIc	Extension into the infratemporal fossa or extension posterior to the pterygoid plates
IIIa	Erosion of the base of skull (middle cranial fossa/base of pterygoids)—minimal intracranial extension
IIIb	Extensive intracranial extension with or without extension into the cavernous sinus.

- Differential diagnosis:
 - Nasal lesions having similar enhancement pattern on imaging includes:
 - Lobular capillary hemangioma
 - Hemangiopericytoma
 - Schwannoma
 - However, it do not involve the pterygopalatine fossa and occur in different age group.
- Treatment:
 - **Surgical Excision:**
 - Primary treatment modality for treating JA.
 - Recurrence is highly related to incomplete removal in 10-40% of surgeries.
 - Pre-excision preparations:
 - **Angiography and embolization:**
 - Performed 48 hours before surgery.
 - Goals:
 - Identifying the feeding vessel.
 - Pre-op embolization will facilitate complete resection of tumor by decreasing intra-op bleeding and tumor size.
 - Performing balloon occlusion test if internal carotid is involved to facilitate preoperative counseling.
 - Complications:
 - CVA
 - Blindness
 - Skin and soft-tissue necrosis
 - **Hormonal therapy (Not used):**
 - Hypothesized to decrease the size and vascularity to JA via estrogen therapy or anti-androgens (flutamide).
 - Not used due to:
 - Lack of efficacy.
 - Physical and psychological side effects

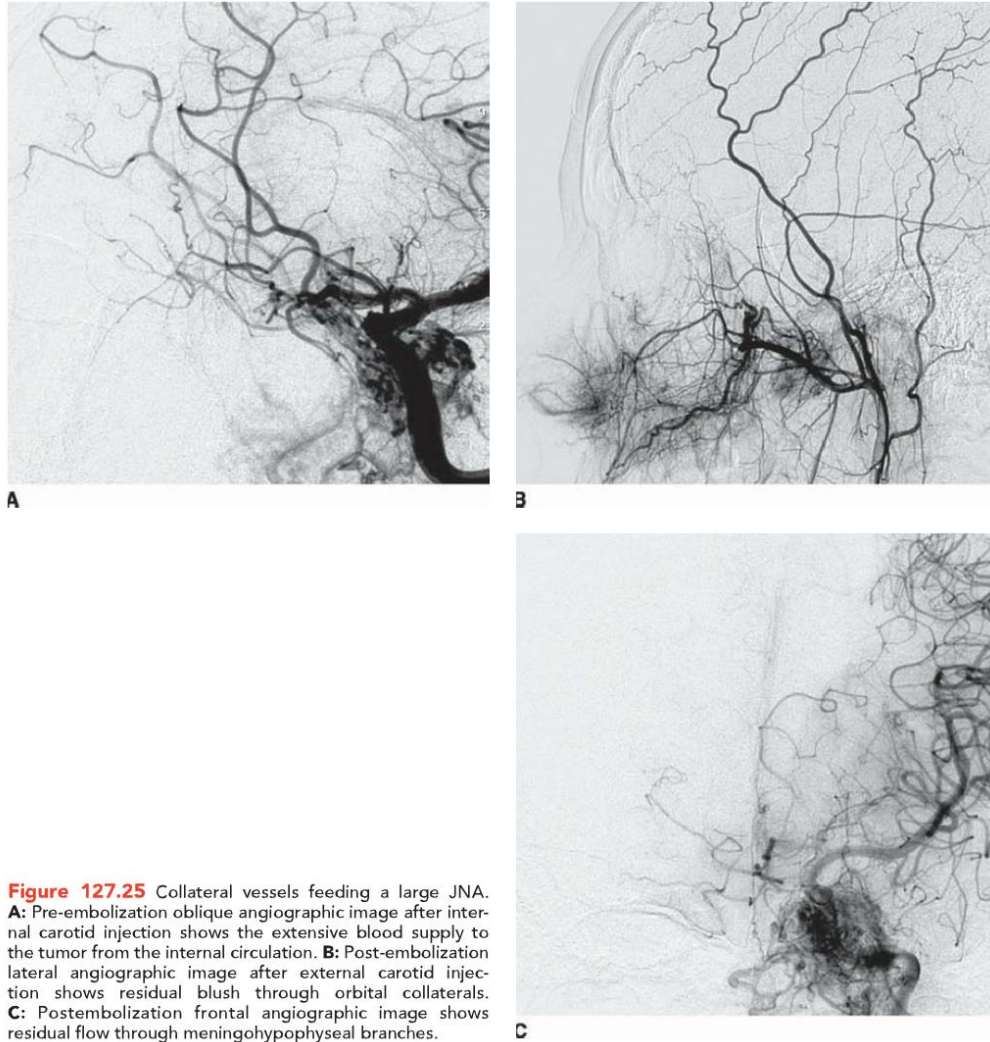


Figure 127.25 Collateral vessels feeding a large JNA. **A:** Pre-embolization oblique angiographic image after internal carotid injection shows the extensive blood supply to the tumor from the internal circulation. **B:** Post-embolization lateral angiographic image after external carotid injection shows residual blush through orbital collaterals. **C:** Postembolization frontal angiographic image shows residual flow through meningohipophyseal branches.

- Surgical approaches:
 - **Endoscopic:**
 - Considered for small to intermediate tumors limited to:
 - Sinonasal cavity
 - Pterygomaxillary fossa
 - Limited infratemporal fossa involvement
 - Limited intracranial extension
 - Limitations of endoscopic approach:
 - Extensive intracranial extension
 - Extensive infratemporal fossa involvement
 - Lateral extension to cavernous sinus
 - Posterior extension to pterygoid plates.

- **Open/Combined:**

- For large tumors with extensive involvement.
- Choice of approach depends on tumor extent and facility of the surgical team.
- Open approaches will achieve 80% complete resection avoiding tumor recurrence even in advanced and salvage cases.

TABLE 127.8	OPEN APPROACHES FOR JNA SURGICAL RESECTION
Open anterior approaches Intraoral	Degloving-Le fort-I osteotomy and downward palatal translocation Degloving, transantral approach Degloving modified transantral approach with medial maxillectomy and lateral buttress removal Degloving, maxillary/zygomatic removal and re-insertion Transpalatal and modified transpalatal approach
Open anterior approaches Facial incision	Weber-Ferguson and extended WF incision with facial translocation (removal and reinsertion of zygomatic bone including orbit floor and lateral wall)
Open lateral approaches	Fisch type D transtemporal approach with facial nerve mobilization Preauricular infratemporal fossa approach with zygomatic arch displacement and pterional craniectomy extradural approach

- **Radiation:**

- Effective at controlling JA with control rate of over 80%.
- Conventional radiation therapy of 30-35 Gy can achieve these results.
- Other modalities include:
 - Intensity-modulated radiation therapy (IMRT)
 - Stereotactic radiation (Gamma Knife radiosurgery and Cyber knife system)
- Indications:
 - Recurrent tumors
 - Extensive tumors
 - Non-operative candidates

- Post-op follow up:

- Based on periodic endoscopic and imaging examination for minimum of 3 years.
- Recurrence rate 10-40%.
- Most residual lesions are diagnosed within 1 year post-op.
- Risk factors for recurrence:
 - Invasion of sphenoid or pterygoid process
 - Infratemporal fossa involvement
 - Foramen lacerum involvement
 - Cavernous sinus involvement

- **Lobular Capillary Hemangioma:**
- Rapidly growing lesion.
- Characterized by proliferation of capillaries arranged in lobules and separated by a loose connective tissue stroma.
- Known previously as **Pyogenic Granulomas** (misnomer).
 - o Not caused by bacterial infection
 - o Not true granuloma
- **Epidemiology:**
 - o Occur mainly in boys (82%).
 - o Occurs in female patients in 3rd decade of life during pregnancy.
- **Etiology:**
 - o **Trauma:**
 - Habitual nose picking
 - Nasal packing
 - o **Hormonal influences:**
 - Pregnancy
- **Clinical picture:**
 - o **Symptoms:**
 - Unilateral epistaxis and nasal obstruction.
 - o **Signs:**
 - Red to purple mass not larger than 1 cm.
 - o **Location:**
 - Anterior septum at Little area.
 - Head of inferior and middle turbinates.
- **Treatment:**
 - o Resolves spontaneously after delivery in pregnant patients.
 - o Surgical Excision with a cuff of surrounding mucoperichondrium is the treatment of choice.



- **Fibrous dysplasia:**
- Genetically based tumor-like condition (not a true neoplasm).
- Normal medullary bone is replaced with fibro-osseous tissue.
- Commonly diagnosed within the first two decades of life.
- Self-limiting process and ceasing when affected bone reaches maximum growth and maturation.
- Reported cases of sarcomatous transformation.
- **Juvenile variant:**
 - o Rapidly destructive, aggressive, destroys teeth, refractory to treatment.
- **Types:**
 - 1. Monostotic:**
 - One bone involved.
 - Most common form.
 - 2. Polyostotic:**
 - More than one bone involved.
 - 3. Disseminated (McCune-Albright Syndrome):**
 - Polyostotic fibrous dysplasia, precocious puberty, abnormal skin pigmented lesions (café-au-lait).
- **Clinical picture:**
 - o Slow-growing painless bony hard swelling.
 - o Progressive facial distortion (unilateral and not crossing midline).
 - o Mostly involves Maxillary sinus.
 - o Displacement of the eye and teeth.
 - o Nasal obstruction.
 - o Cranial nerve dysfunction via compression (Optic nerve).
 - o Endoscopic findings are usually unremarkable.
 - o **Juvenile variant:**
 - Rapidly destructive, aggressive, destroys teeth, refractory to treatment.

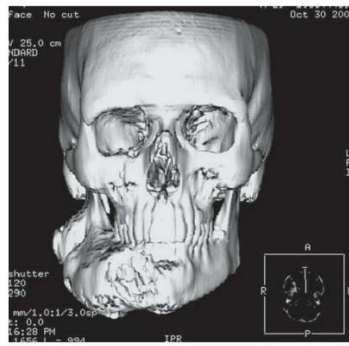


- Diagnosis:
 - **CT scan:**
 - Findings depends on degree of tissue mineralization:
 - Early stage:
 - High density of fibrous tissue.
 - Radiolucent or lytic appearance.
 - Difficult to differentiate from simple bone cyst.
 - Late stage:
 - High density of bony tissue.
 - **Sclerotic appearance/Ground-glass/Chinese writing:**
 - Irregular margins
 - Multiloculated
 - Expanded cortex (eggshell-thin)
 - NOT displacing inferior alveolar canal.



FIGURE 48-17. Coronal computed tomography scan shows fibrous dysplasia. An expansile lesion can be seen in the right ethmoid with a mixed density pattern that includes calcifications, a ground-glass appearance, and low-density areas. This pattern reflects the different degrees of mineralization of the fibrous tissue that replaces normal bone. The lamina papyracea is interrupted (*arrowheads*), but the orbit is not invaded. The cribriform plate is thickened and displays a ground-glass pattern (*arrows*) suggestive of fibrous dysplasia.

- Treatment:
 - **Observation:**
 - For asymptomatic patients.
 - **Medical:**
 - Bisphosphonates (Pamidronate) inhibit osteoclastic activity.
 - Used with success in patients with extensive lesions.
 - **Surgical excision and curettage of involved bone:**
 - Surgery is performed after maturation of affected bone.
 - Higher recurrence rate if surgery performed during active growth period.
 - Re-contouring of affected facial bone is performed rather than resection.
 - Indications of surgery:
 - Facial disfiguring
 - Cranial nerve dysfunction
- Radiotherapy is contraindicated because of the risk of malignant transformation and the side effects on facial skeleton growth in young patients.



- **Ossifying Fibroma**
- True benign neoplasm.
- Occurs in 3rd-4th decades of life.
- Commonly in black women.
- Space-occupying lesion evident in the nasal cavity at endoscopic examination.
- CT scan:
 - Well-defined, multiloculated lesion bordered by a peripheral eggshell-like dense rim.
- Treatment:
 - Radical surgical resection.
 - Recurrence is high (40%).



- **Schwannomas:**

- Neurogenic tumor that arises from Schwann cells of the sheath of myelinated nerves.
- Sinonasal involvement occurs in only 4% of all head and neck Schwannomas.
- Most common site of origin is ophthalmic and maxillary divisions of trigeminal nerve.

- **Histopathology:**

- Unlike other regions, Sinonasal schwannomas lack tumor encapsulation.
- Types
 - **Antoni Type A:**
 - Histologically parallel nuclei, uniform spindle cells, compact cells.
 - **Antoni Type B:**
 - Histologically less uniform, may have fatty or hyaline degeneration, less cellular.

- **Clinical picture:**

- Polypoid mass covered with network of capillaries on its surface.

- **CT scan:**

- Mild enhancement with contrast.

- **MRI:**

- Intermediate signal on T1-T2 in Antoni type A.
- HYPER-intense on T2 in Antoni type B.

- **Treatment:**

- Surgical resection.

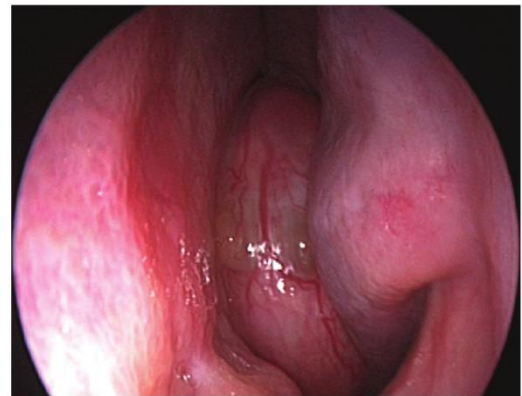


FIGURE 48-18. This benign schwannoma appears on endoscopy as a large polypoid mass that entirely fills the left nasal cavity. A network of capillaries on the surface of the lesion may suggest a diagnosis of hypervascularized tumor.

- **Hemangiopericytoma:**

- Rare highly vascular tumor.
- Arises from pericytes of Zimmerman that are found spiraling around capillaries and postcapillary venules.
- Clinical course may vary from indolent, with low rate of local recurrence, to aggressive, with rapid development of metastases.
- 10% malignant degeneration.

- **CT scan:**

- Enhancing lesion with contrast.

- **Treatment:**

- Preoperative embolization.
- Wide surgical resection is the only curative modality.
- No neck dissection needed as lymphatic spread is rare.
- Adjuvant Radiation therapy for recurrence.

Malignant SinoNasal Neoplasms:

Risk Factors:

1. Toxin exposure:
 - Nickel
 - Chromium
 - Mustard gas
 - Hydrocarbons
 - Radium
2. Woodworkers:
 - Softwood: Increases risk of **SCC**.
 - Hardwood: Increases risk of **Adenocarcinoma**.
3. Chronic infection
4. Previous radiation

Clinical Picture:

- Unilateral persistent Sinusitis, Nasal obstruction, or Epistaxis.
 - Nasal lesions (exophytic, ulcerative, septal perforation).
 - Orbital symptoms (Proptosis, Diplopia, Vision loss).
 - Anosmia and Taste disturbances.
 - Palatal fistula.
 - CN Palsies
 - Headache.
 - Facial Paresthesias and pain.
- **SCC** is the most common type of Sinonasal cancer.
 - **Maxillary Sinus** is the most common site of Paranasal cancer.
 - Followed by Ethmoid, Frontal and Sphenoid sinuses.
 - **Nasal Cavity** is the 2nd most common site of Sinonasal cancer.

Malignant Epithelial SinoNasal Neoplasms:

SCC:

- Most common of all Sinonasal tumors (80%).
- Most common in elderly white males (5th-6th decade).
- Aggressive.
- High rate of Mets.
- Majority are moderately differentiated keratinizing type.
- Prognosis is related to extent of tumor and site of origin.

Adenocarcinoma:

- 4-8% of all Sinonasal tumors.
- Most Common in Ethmoid and Nasal cavity.
- Associated with Hardwood and Leather workers.
- Approaches:
 - Anterior Craniofacial Resection.
 - Lateral Rhinotomy
 - Endonasal with or without Radiotherapy.

- **Adenoid cystic carcinoma (ACC):**
 - 15-20% of all H&N ACC.
 - Minor salivary gland tumor.
 - Early Neurovascular and Submucosal spread.
 - High rate of Local Recurrence (50-75%) and Mets.
 - Treatment:
 - Surgery and Post-op Radiation.
- **Melanoma:**
 - Either Primary or Metastatic (More common).
 - Less than 1% of Melanoma are Sinonasal melanoma.
 - Grey or bluish-black polypoid mass.
 - Most commonly found in Nasal cavity (Anterior Nasal Septum).
 - Then Maxillary, Ethmoid and Frontal sinuses.
 - Early vascular and lymphatic invasion.
 - High incidence of Local recurrence after surgical excision.
 - Post-op Radiation therapy may be beneficial.
 - 5-year overall survival is 23%.
 - Most important factor in predicting survival is Metastatic disease.
- **Olfactory Neuroblastoma:**
 - Rare.
 - Arising in Olfactory epithelium.
 - Pink or brown friable Nasal mass.
 - Present with Nasal obstruction and Epistaxis.
 - Bimodal frequency at 10-20 and 50-60 years of age.
 - Males = Females.
 - **Kadish Staging System:**
 - **Stage A:** Tumor confined to Nasal cavity
 - **Stage B:** Extending to Sinuses.
 - **Stage C:** Extending to Orbit, Base of Skull, Cranial cavity or with Cervical/Distant metastasis
 - **UCLA Staging System:**
 - **T1:** Tumor involving Nasal cavity and/or Paranasal sinuses (Excluding sphenoid), sparing Most Superior Ethmoidal air cells.
 - **T2:** Tumor involving Nasal cavity and/or Paranasal sinuses (Including Sphenoid) with Extension to or Erosion of Cribriform plate.
 - **T3:** Tumor Extending into Orbit or Protruding into Anterior Cranial Fossa.
 - **T4:** Tumor involving Brain
 - Management:
 - Open or Endoscopic Craniofacial resection with Post-op Radiation to Primary site and Neck.

- **Sinonasal Undifferentiated Carcinoma (SNUC):**
 - Extremely Poor prognosis.
 - Rapidly progressing tumor.
 - Extensive tissue destruction.
 - Treatment is Trimodal (Chemotherapy + Radiation + Surgery).
- **Small Cell Carcinoma:**
 - Extremely Poor prognosis.
 - Treatment is Nonsurgical (Chemotherapy and Radiation).
- **Malignant Non-epithelial SinoNasal Neoplasms:**
- **Rhabdomyosarcomas:**
 - 10% of patients with H&N Rhabdomyosarcomas are originate in Paranasal sinuses.
 - Classification:
 - Embryonal: 1st decade of life.
 - Alveolar: Adolescents.
 - Pleomorphic: Adults.
 - Management:
 - Adults: Wide surgical excision and post-op Radiotherapy.
 - Children: Chemotherapy and Radiotherapy.
- **Neurogenic Sarcomas:**
 - Rare.
 - Commonly associated with Neurofibromatosis..
 - Locally aggressive.
 - Frequently present with Distant metastases.
 - Management:
 - Surgery plays a primary role.
 - Radiation and Chemotherapy are reserved for:
 - Incomplete removal.
 - Inoperable cases.
 - Recurrences.
 - 5-year survival rate is 60%.
 - Sarcomas associated with Neurofibromatosis behave more aggressively, yielding a 5-year survival rate of 30%.
- **Hemangiopericytoma:**
 - Rare tumor.
 - Highly vascular.
 - Arises from pericytes of Zimmermann (located outside of endothelial cells, capable of changing the caliber of the capillaries).
 - Presents as a lobulated mass and epistaxis.
 - May require preoperative embolization prior to excision.

- **Leiomyosarcoma:**
 - Smooth muscle tumor.
 - Poor prognosis.
- **Fibrosarcoma:**
 - Tumors of fibroblasts.
 - Associated with trauma and radiation.
- **Angiosarcomas:**
 - Slow growing but locally aggressive with indistinct margins.
- **Osteogenic Sarcomas and Chondrosarcomas:**
 - Aggressive.
 - Poor prognosis (10–20% 5-year survival).
 - Arise from the facial skeleton, more common in the mandible than in the maxilla.
- **Lymphomas:**
 - 0.17% of all lymphoma.
 - Typically non-Hodgkin's.
 - Maxillary sinus most common then Nasal Cavity.
- **Metastatic Tumors:**
 - Renal cell carcinoma most common.
- **Chordoma:**
 - Malignant.
 - Slow growing.
 - From notochord remnant.
 - Arise from Clivus/skull base.
 - Key pathologic finding is physaliferous cells.

- **Maxillary Sinus Ca:**
- Most common site of origin for malignancies of the sinonasal tract.
- SCCA is usually diagnosed at advanced stages.
 - o Remain silent for a long time giving only vague symptoms of "Sinusitis".
- Spreads to destroy the bony confines of the maxillary sinus and invades the surrounding structures.
- Most Antral malignancies are Antroethmoidal in nature.

- Clinical Picture:
- Early features:
 - o Nasal stuffiness.
 - o Blood-stained nasal discharge
 - o Facial paraesthesias or pain
 - o Epiphora.
- Late features depend on direction of spread:
 - o Medial spread:
 - Nasal obstruction.
 - Discharge and Epistaxis.
 - o Anterior spread:
 - Cheek swelling
 - Invasion of facial skin.
 - o Inferior spread:
 - Expansion of alveolus with dental pain.
 - Loosening of teeth
 - Ulceration of gingiva
 - Swelling in hard palate.
 - o Superior spread:
 - Proptosis
 - Diplopia
 - Ocular pain.
 - Epiphora.
 - o Posterior spread:
 - Trismus.
 - o Intracranial spread:
 - Through ethmoids, cribriform plate or foramen lacerum.
 - o Lymphatic spread:
 - Uncommon.
 - Only in late stages of disease into Retropharyngeal, Submandibular and upper jugular LN.
 - o Systemic mets:
 - Rare.
 - Lungs (Most common)

- **Staging of Maxillary Sinus Tumors:**

- **T1:**
 - Tumor is limited to Maxillary sinus mucosa.
 - No erosion or destruction of bone.
- **T2:**
 - Tumor is causing bone erosion or destruction, including extension into Hard palate and/or Middle nasal meatus, **EXCEPT** extension to Posterior wall of the maxillary sinus and pterygoid plates.
- **T3:**
 - Tumor invades any of the following:
 - Bone of the posterior wall of the maxillary sinus, subcutaneous tissues, floor, or medial wall of the orbit, pterygoid fossa, or ethmoid sinuses.
- **T4a:**
 - Tumor invades anterior orbital contents, skin of cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses.
- **T4b:**
 - Tumor invades any of the following:
 - Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve (V2), nasopharynx, or clivus.

- **Management:**

- Early-stage SCC:
 - Treated by surgery or radiation.
- Advanced stages:
 - Combination therapy.

- **Prognosis:**

- Typically presents in advanced stage (90% invades more than one wall).
- Maxillary tumors in general have 5-year survival rates between 20–50%.

○ **Ohngren's Line:**

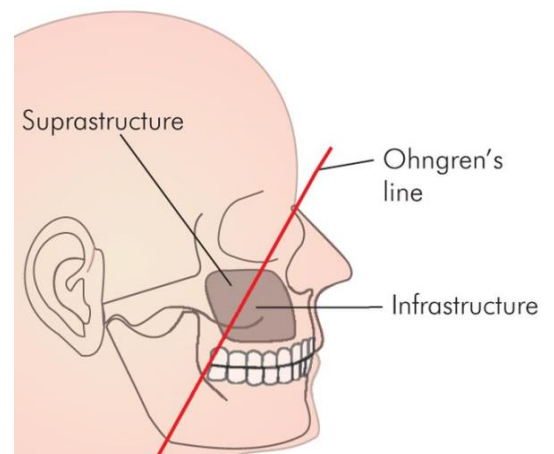
- Imaginary line between Medial canthus of Eye and Angle of mandible.

▪ **Poor Prognosis:**

- Tumor Situated above this line (Suprastructural).
- Higher risk of skull base invasion and perineural spread.

▪ **Good Prognosis:**

- Tumor Situated below this line (Intrastructural).



- **Nasal Cavity Ca:**
- 2nd most common site of Sinonasal cancer.
- SCCA is the most common type.
- Most commonly arises from Lateral nasal wall.
- Sometimes arises from mucocutaneous junction in the septum causes burning and soreness in the nose (Nose-picker's cancer).
- Tumors arising at floor, lateral wall, and septum are all amenable to surgical excision.
- Prognosis:
 - o <10% regional mets potential.
 - o 60% 5-year survival.
 - o Worse prognosis for (Required post-op Radiation):
 - Larger tumors > 2cm.
 - Extension outside the nasal cavity.
 - Bony involvement.
 - Regional mets.
- **Ethmoid Sinus Ca:**
- Tumors originating in ethmoid sinuses produce symptoms at early stages, leading to earlier diagnosis and better prognosis.
- SCCA is the most common type.
- Early features:
 - o Nasal obstruction
 - o Blood-stained nasal discharge
 - o Retro-orbital pain.
- Late features:
 - o Broadening of nasal root
 - o Lateral displacement of eyeball and diplopia
 - o Extension through cribriform plate may cause meningitis.
- Nodal involvement is not common.
- Endoscopic surgical resection is indicated in:
 - o Small tumor.
 - o T1 or T2
- Combined therapy with radiation is recommended for:
 - o Advanced stages.
 - o Positive surgical margins
 - o Recurrent tumors.
- Prognosis:
 - o The overall 5-year survival rate is 50% to 60%.

- **Staging of Nasal Cavity and Ethmoid Sinus Tumors:**
 - **T1:**
 - Tumor is limited to Ethmoid sinus with or without bone erosion.
 - **T2:**
 - Tumor invades two subsites in a single region or extends to involve an adjacent region within the nasoethmoidal complex, with or without bony invasion.
 - **T3:**
 - Tumor extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribriform plate.
 - **T4a:**
 - Tumor invades any of the following:
 - Anterior orbital contents, skin of nose or cheek, minimal extension to anterior cranial fossa, pterygoid plates, sphenoid or frontal sinuses.
 - **T4b:**
 - Tumor invades any of the following:
 - Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve (V2), nasopharynx, or clivus.
- **Frontal Sinus Ca:**
 - Rare.
 - Pain and swelling of the frontal region.
 - Growth may erode through the floor of frontal sinus and present as a swelling above the medial canthus.
 - Growths of frontal sinus may extend through ethmoids into the orbit.
 - Dura of anterior cranial fossa may be involved if growth penetrates the posterior wall of the sinus.
 - The anatomical site is amenable to surgical resection.
 - Managed similarly to ethmoidal tumors.
- **Sphenoid Sinus Ca:**
 - Extremely rare (<1% of malignant sinonasal tumor).
 - Sphenoid sinus has a higher instance of Neurological sequelae
 - CN Palsies.
 - Orbital symptoms.
 - Headache.
 - En bloc surgical resection is technically difficult due to the anatomical relationships of the sphenoid sinus.
 - Gross total removal and postoperative radiation therapy is the treatment of choice.
- **Prognosis:**
 - The overall prognosis at 5-year follow-up varies from 5% to 25%.

- **Treatment Principles of Sinonasal Ca:**

1. Surgery:

- Biopsy:
 - Endoscopic (preferred) or Open (Caldwell-Luc, External Ethmoidectomy; Rhinotomy).
- Drainage/debridement:
 - Nasoantral window is indicated for:
 - Patients with secondary bacterial sinusitis.
 - Patients who will require radiation therapy as primary treatment.
- Resection:
 - Surgical Resection for cure.
 - Palliative surgical resection is indicated in:
 - Intractable pain.
 - Rapid decompression of vital structures.
 - Social embarrassment (massive lesion).
 - New endoscopic approaches are becoming more favorable.
 - Surgery as a single treatment modality for malignant tumors of the sinonasal tract has yielded 5-year survival rates from 19% to 86%.

- **Most common open surgical approaches:**

1. Medial Maxillectomy Lateral Rhinotomy:

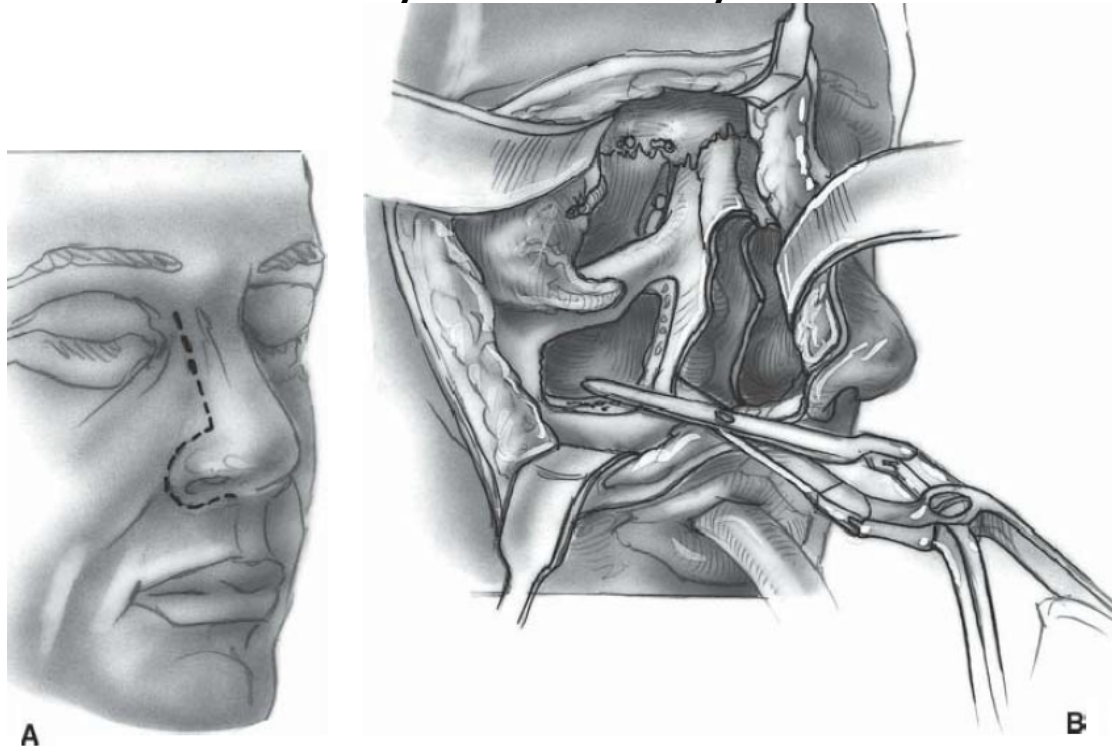
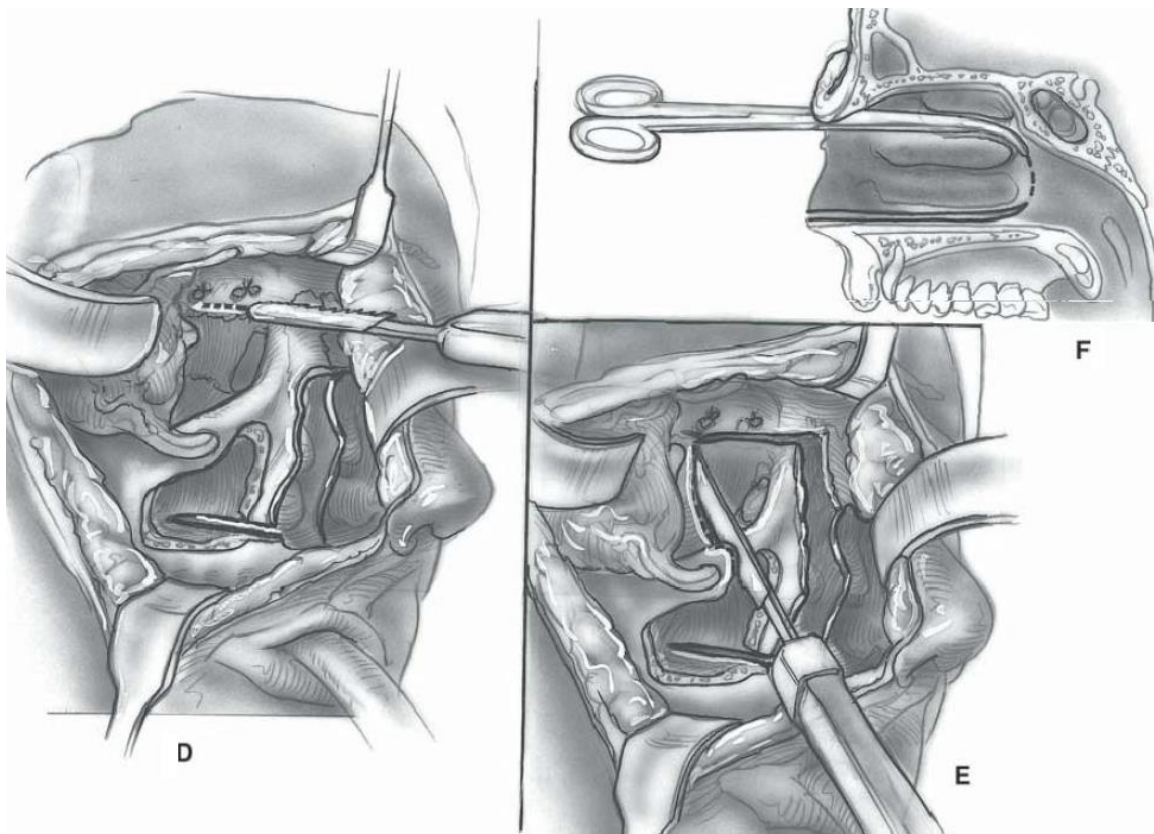
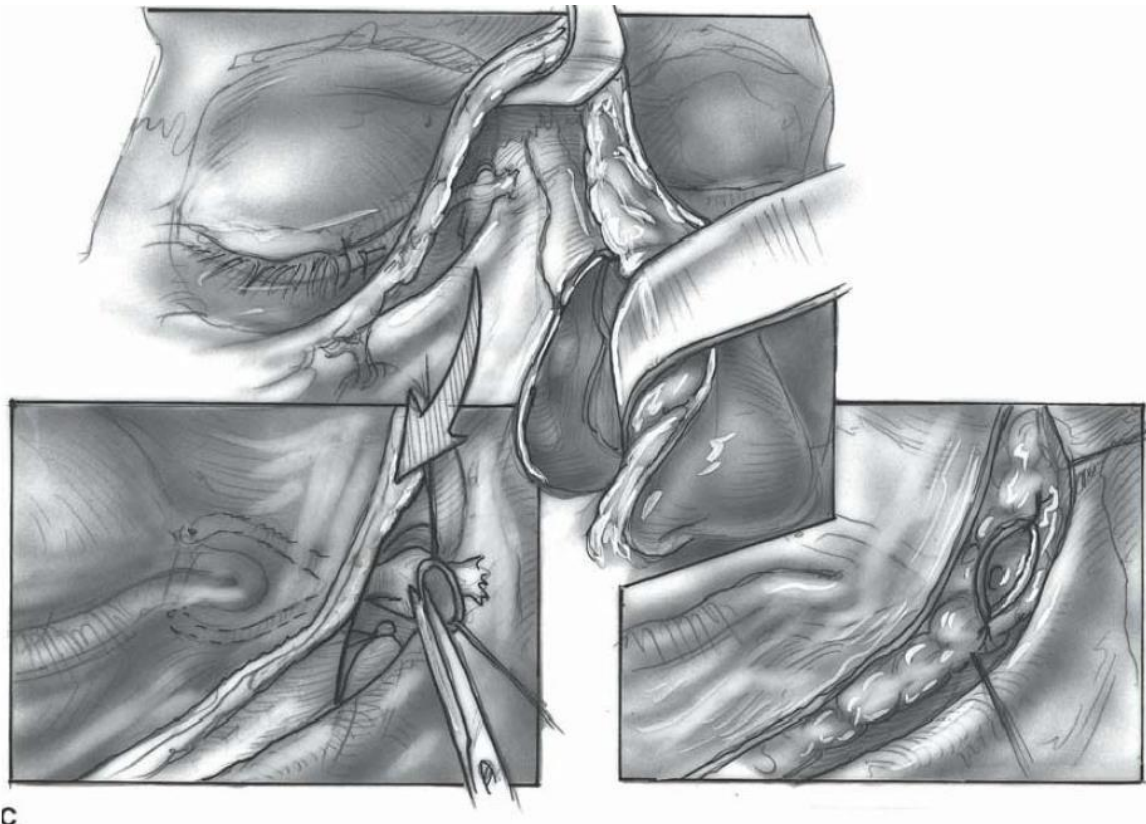


Figure 129.1 Medial maxillectomy. Lateral rhinotomy: **A:** The skin incision begins beneath the medial aspect of the eyebrow and continues 4 to 5 mm anterior to the medial canthus and over the nasal bone along the deepest portion of the nasomaxillary groove and following the alar crease. A lip-splitting extension of the incision is not necessary. To expose the surgical area, the cheek flap is elevated subperiosteally over the maxilla and around the infraorbital nerve. The periorbital is elevated over the lamina papyracea, and the frontoethmoid suture is identified and followed posteriorly until the anterior and posterior ethmoid arteries are identified. The anterior wall of the antrum is penetrated at the canine fossa using a 4-mm chisel. The antrostomy is enlarged with a Kerrison rongeur around the infraorbital nerve and superiorly toward the inferior orbital rim. **B:** Bone is removed across the orbital rim, including the lacrimal fossa. The nasolacrimal duct is divided and the lacrimal sac is opened and marsupialized (**C**). **D:** Osteotomies and removal of the specimen. The first osteotomy involved in the actual removal extends through the piriform aperture at the level of the nasal floor, directed posteriorly until the osteotomy perforates the posterior wall of the antrum. The orbit is retracted laterally, and a second osteotomy is performed at the frontoethmoid suture, extending posteriorly to a point 2 to 3 mm posterior to the posterior ethmoid artery (i.e., anterior to the optic foramen). **E:** The thin bone of the medial floor of the orbit is sawed following a line that joins the lacrimal fossa with the superior osteotomy. The final bone cut involves three steps. First, a 2-mm osteotome is introduced through the anterior antrostomy and directed through the medial posterior antral wall. The osteotome is advanced superiorly to reach the level of the superior osteotomy and is then pushed medially. Second, a wide osteotome, introduced through the nose, is impacted into the anterior wall of the sphenoid sinus, and then pushed laterally. Heavy right-angle scissors (e.g., upper-lateral-cartilage scissors) are guided through the inferior osteotomy with one blade in the nose and the other in the antrum to start the posterior cut, behind the turbinates. **F:** Heavy curved scissors are then introduced with one blade in the nasal cavity and the other in the superior osteotomy, directed through or along the posterior attachments of the turbinates. The specimen is removed by anterior and inferior traction. Hemostasis is achieved by direct clamping or cautery. The bony edges are smoothed with a rongeur. Residual ethmoid mucosa is removed with ethmoid forceps, and a wide sphenoidotomy is opened with Kerrison rongeurs. The cavity is covered with Gelfoam for hemostasis. The medial canthal tendon is sutured to the periosteum of the nasal bones. The wound is closed using a meticulous layered closure. AEF, anterior ethmoidal foramen; OF, optic foramen; PEF, posterior ethmoidal foramen.



2. Mid-Face Degloving:

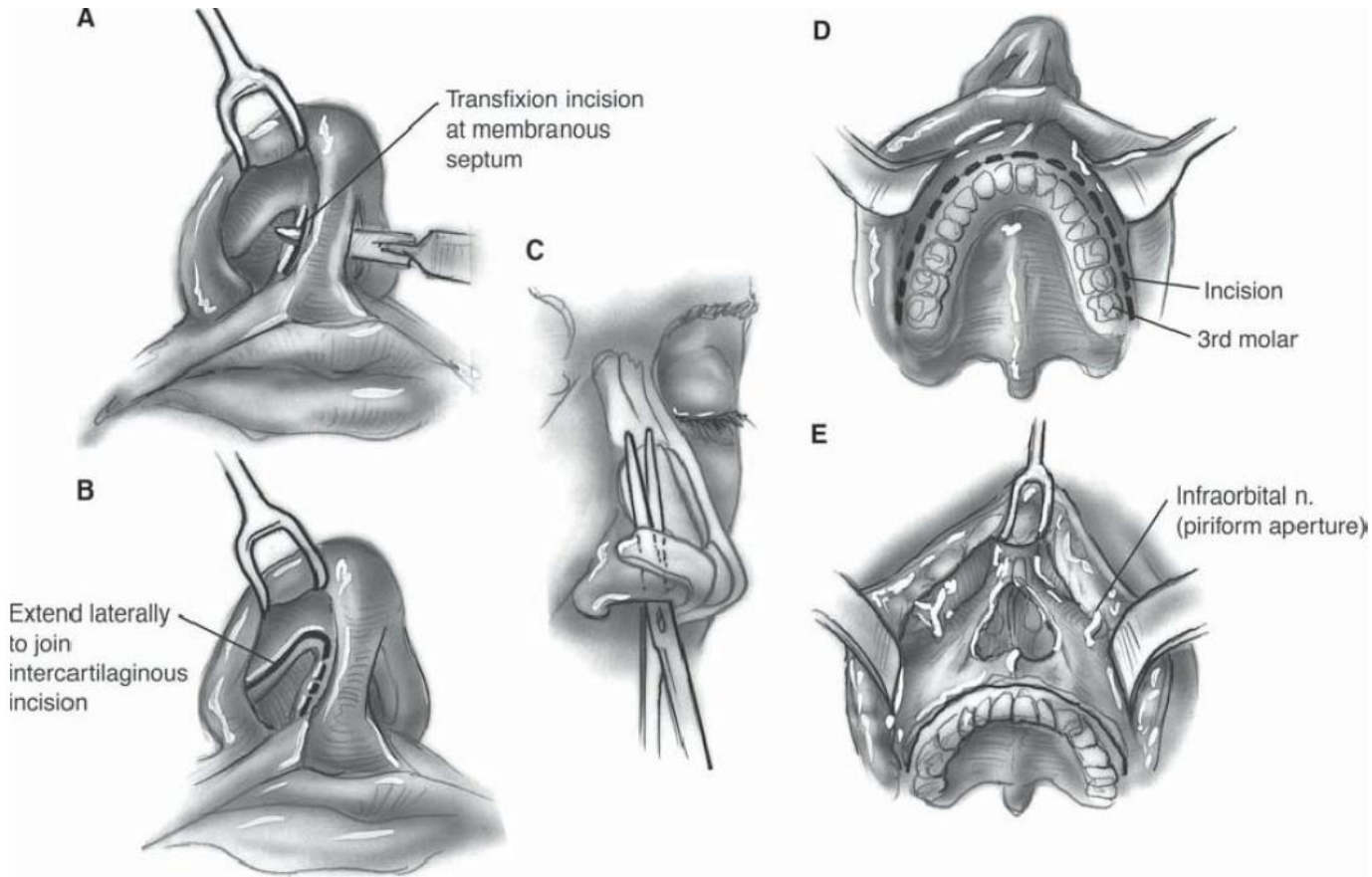


Figure 129.2 A,B: Mid-face degloving. A transfixion incision is performed at the membranous septum and is extended laterally to join a transcartilaginous incision. **C:** A gingivobuccal incision is performed, extending laterally from the midline to the maxillary tuberosities. **D:** In exposing the surgical area, tenotomy scissors are introduced through the transcartilaginous incisions to dissect the skin from the nasal skeleton. The dissection over the maxilla is carried out in a subperiosteal plane. This dissection joins the nasal degloving using sharp dissection over the piriform aperture attachments. The dissection is extended superiorly exposing the mid-face skeleton. **E:** The exposure is limited by the infraorbital neurovascular bundles. The osteotomies, removal of the specimen, reconstruction, and closure are performed as described for the lateral rhinotomy.

3. Total Maxillectomy:

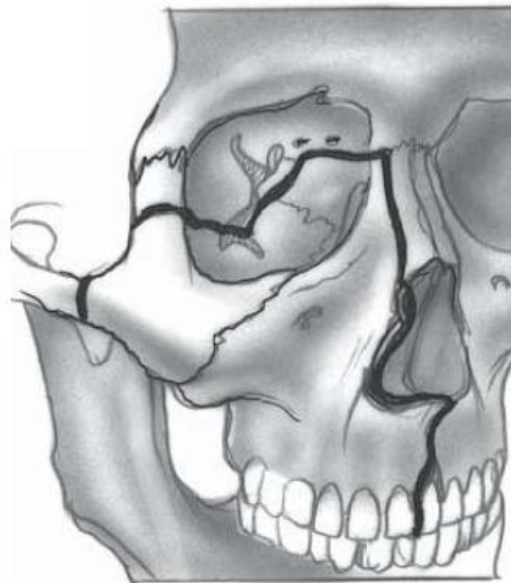
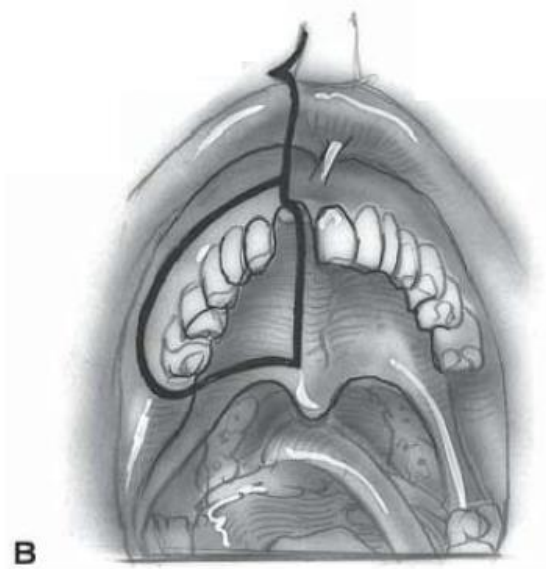
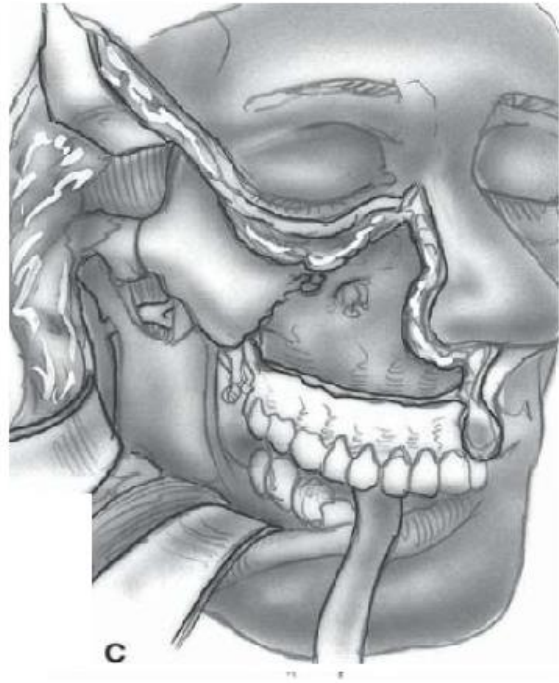
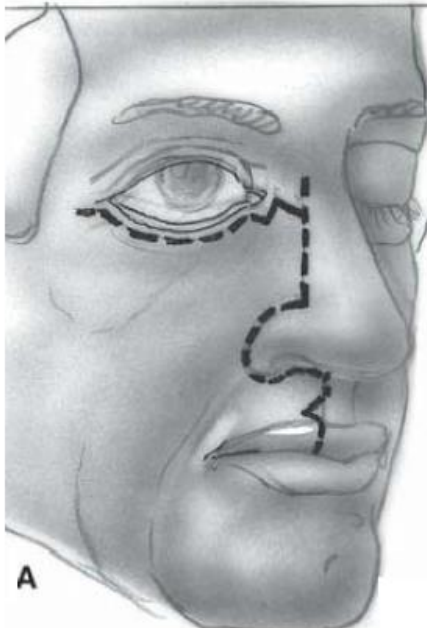
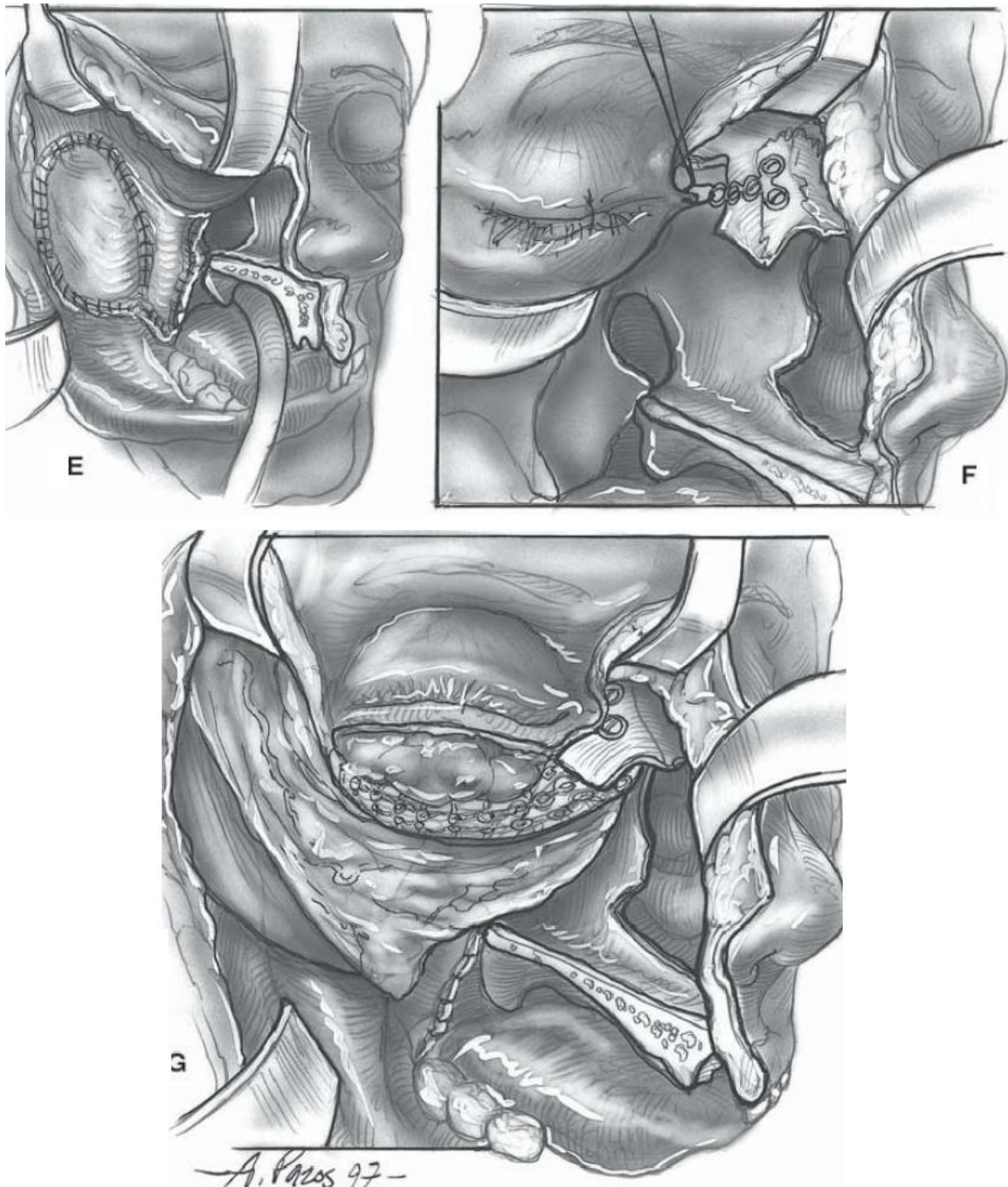


Figure 129.3 Total maxillectomy. A total maxillectomy with preservation of the orbit can be performed using incisions identical to lateral rhinotomy incisions with a lip-splitting extension. Alternatively, a lateral rhinotomy incision may be combined with an ipsilateral degloving approach. **A:** If increased exposure is necessary, the facial incisions may be modified. The superior incision begins at the lateral canthus and extends medially, passing 3 to 4 mm below the ciliary line. The eye is protected by a temporary tarsorrhaphy stitch. This incision may be substituted by a transconjunctival incision. The subciliary limb is joined to a lateral rhinotomy incision. The orbicularis oculi muscle is incised with an inferiorly directed slant, exposing the orbital septum. The gingivobuccal incision is extended laterally to the ipsilateral maxillary tuberosity. **B:** The soft palate is incised at the junction with the hard palate, and its attachments are sharply transected. The mucoperiosteum of the hard palate is incised following a paramedian line ipsilateral to the lesion. The paramedian strip of mucosa will be later imbricated over the bony edge of the hard palate to facilitate the fitting of a prosthesis. **C:** The orbital contents are dissected from the medial inferior and lateral walls, exposing the lacrimal sac, the anterior and posterior ethmoid arteries, and the infraorbital fissure. These are managed as described for a medial maxillectomy (Fig. 129.1). Osteotomies and removal of the specimen. **D:** The body and frontal process of the zygoma are divided with the saw. The maxilla is severed from the nasal bones with the saw, and the osteotomy is extended superiorly to the frontoethmoidal suture. A superior osteotomy is carried out posteriorly to a point 3 to 4 mm posterior to the posterior ethmoid artery. An osteotomy is performed connecting the lateral and medial wall osteotomies across the inferior orbital fissure. The hard palate is transected with a Gigli or sagittal saw. The maxilla is detached from the skull by tapping a chisel placed into the pterygomaxillary fissure in a posterosuperior direction. The superior attachments of the turbinates are sharply severed, as for a medial maxillectomy (Fig. 129.1). The specimen is removed by anteroinferior traction. Remnants of ethmoid sinus mucosa are removed in a piecemeal fashion. The coronoid process of the mandible is removed to avoid displacement of the prosthesis when opening the mandible. (*Continued*)

Figure 129.3 (*Continued*) **E:** The exposed facial and pterygoid muscles and periorbital are lined with a split-thickness skin graft that is 0.35 to 0.45 mm thick. The obturator or denture is wired to the remaining dentition or suspended from the zygomatic arch and piriform aperture or leg screwed to the remaining hard palate. **F:** A medial canthopexy, using a Y-shape titanium plate fixed to the nasal bones. A figure-8 nonabsorbable suture is used to medialize the medial canthal tendon. **G:** The floor and medial walls of the orbit are reconstructed with titanium mesh. This is then covered by a vertically split temporalis muscle flap. The anterior half is used for the reconstruction, and the posterior half is transposed to the anterior temporal fossa to obliterate the defect.



4. Orbital Exenteration:

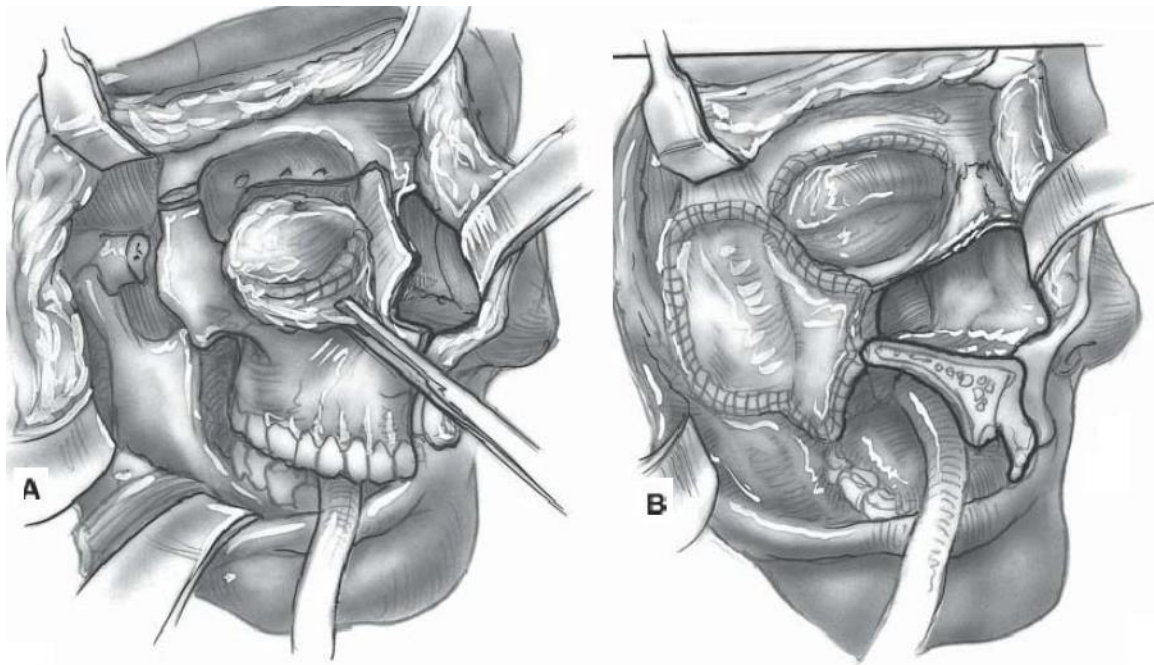


Figure 129.4 Orbital exenteration. **A:** Incisions for orbital exenteration include those described in Figure 129.3 and a supraciliary incision along the upper eyelid. These incisions allow the preservation of the lids, which can be used to line the remaining orbital cavity. If the eyelids are involved by the tumor, the incisions are modified to include their resection en bloc. The exposure proceeds as previously described (Fig. 129.3), omitting the dissection of the orbit from the inferior wall. The upper eyelid is retracted superiorly, and the periorbita is incised over the superior orbital rim. The orbit is dissected from the superior wall, identifying optic foramen and superior orbital fissure. These are infiltrated with lidocaine to block the autonomic innervation and prevent cardiac arrhythmias. The neural and vascular structures traveling through these foramina are transected after hemostasis with bipolar cautery or clamping. Inferior traction on the globe allows further visualization of the orbital floor and inferior orbital fissure. Lateral and medial wall osteotomies are connected as in Figure 129.3C. Other osteotomies are identical to Figure 129.3C. The roof of the orbit may be lined with a skin graft or left to granulate and mucosalize. Alternatively, the cavity may be filled with a temporalis muscle flap. It should be remembered that squamous epithelium tolerates trauma (e.g., prosthesis) better than mucosa. **B:** In exposing the surgical area, the facial flap is elevated subperiosteally. In the case of extension through the anterior wall of the antrum, the facial flap may be elevated in a subcutaneous plane, including the facial musculature in the specimen. The skin may also be resected en bloc, to address direct invasion by the tumor. The dissection is carried out along the lateral wall of the maxilla and the internal maxillary artery is identified at the pterygomaxillary fissure and clipped.

5. Inferior Maxillectomy:

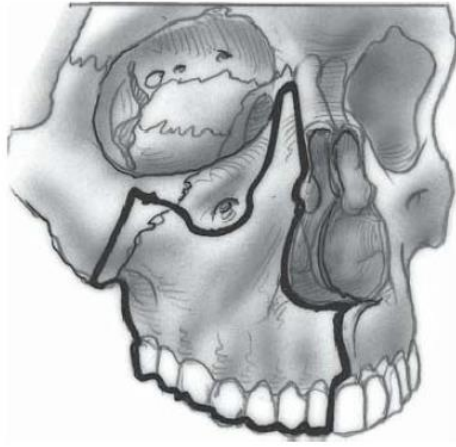


Figure 129.5 Inferior maxillectomy. Tumors confined to the floor of the antrum may be managed by partial maxillectomy. It differs from total maxillectomy on the preservation of the orbital floor and, in selected cases of the infraorbital nerve. Bilateral maxillectomy. The procedure is performed bilaterally as in Figure 129.4. The nasal septum may be sacrificed or preserved for suspension of the prosthesis or reconstructive flaps.

- Treatment Principles of Sinonasal Ca (Cont.):

2. Rehabilitation

- Goals of post-op Rehabilitation:
 1. wound healing
 2. Preservation/reconstruction of facial contour
 3. Restoration of oronasal separation (speech/swallowing)
- Function more important than cosmetics.
- Rehabilitation can be achieved with prostheses or flaps of various kinds.
- A total maxillectomy defect should not be obliterated at the initial operation.
 - An open cavity facilitates cleansing and direct visual inspection during the follow-up period.
- Patients requiring a craniofacial resection, needs immediate separation of the cranial cavity from the upper aerodigestive tract and support of the brain.
 - A pericranial flap and transfer of a temporalis muscle flap achieves these goals.
 - The temporalis muscle, however, is often devascularized after an infratemporal fossa dissection or its bulk may be inadequate to obliterate the dead space.
 - Under these circumstances, the maxillectomy cavity may be obliterated with a free microvascular flap, offering immediate palliation and oronasal separation without the need for a prosthesis.

3. Radiation Therapy:

- Response of sinonasal tract tumors to radiation varies with the stage and histology of the tumor.
- Radiation may be used as a single modality, as an adjunct to surgery, or as palliative therapy.
- Postoperative radiation improves local control but not cause specific or absolute survival.
- Radiation therapy is the primary treatment for:
 - Lymphoreticular tumor.
 - Patients poor surgical candidates..
 - Patients who refuse surgery.

4. Chemotherapy:

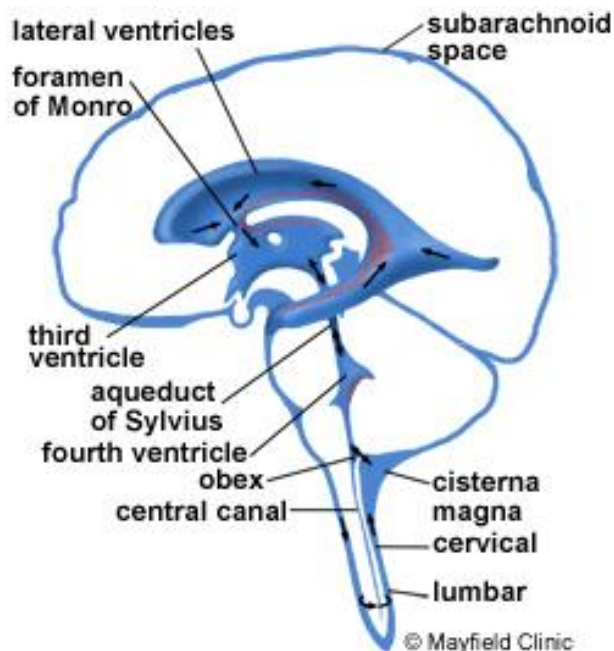
- Role of chemotherapy is usually palliative to relieve pain, obstruction, or to debulk a massive external lesion.

CSF Rhinorrhea:

- Cerebrospinal fluid rhinorrhea is result of bony defect at skull base with disruption of arachnoid, dura mater and sinonasal mucosa with a resultant pressure gradient that leads to active CSF leak and flow of clear fluid from the nose.

CSF Physiology:

- **Function:**
 1. Acts as a cushion for the brain.
 2. Transportation of metabolite.
- **Production:**
 1. Choroid plexus of ventricular system **(70%)**
 2. Capillary ultra-filtration **(18%)**
 3. Metabolic water production **(12%)**
- **Circulation:**
 - **Lateral ventricles**
 - Foramen of Monro
 - **Third Ventricle**
 - Aqueduct of Sylvius
 - **Fourth Ventricle**
 - Foramina of Magendie and Luschka
 - **Subarachnoid space** over brain and spinal cord
 - Reabsorption into venous sinus blood via Arachnoid villi.



- **Consistency:**

- Nearly iso-osmotic to plasma.
- Ions: Na^+ , K^+ , Mg^{2+} , Ca^{2+} , Cl^- , and HCO_3^-
- Glucose (70% of blood glucose)
- Water
- Proteins
- Very few cells (polymorphonuclear and mononuclear cells)

Table 4. Normal CSF characteristics and main pathological alterations

Characteristics	Normal	Meningitis		
		Acute bacterial	Viral	Chronic
Pressure (mm/H ₂ O)	100-200	N or ↑	N or ↑	N or ↑
Aspect	Clear	Turbid/purulent	clear/cloudy	Clear/cloudy
Color	Clear	white	Clear	Clear/white
Cytology (mm ³)	Until 4	> 1,000	500-1,000	<500
Cell type	Lymphocytes	Neutrophils	Lymphocytes	Lymphocytes
Protein (mg/dL)	V 5-10 SO 10-25 L 15-45	↑↑	Normal or ↑	Normal or ↑
Glucose (mg/dL)	2/3 from serum	↓	Normal	Normal or ↓
Lactic acid (mmol/L)	<3.5	>3.5	<3.5	<3.5

- **Amount:**

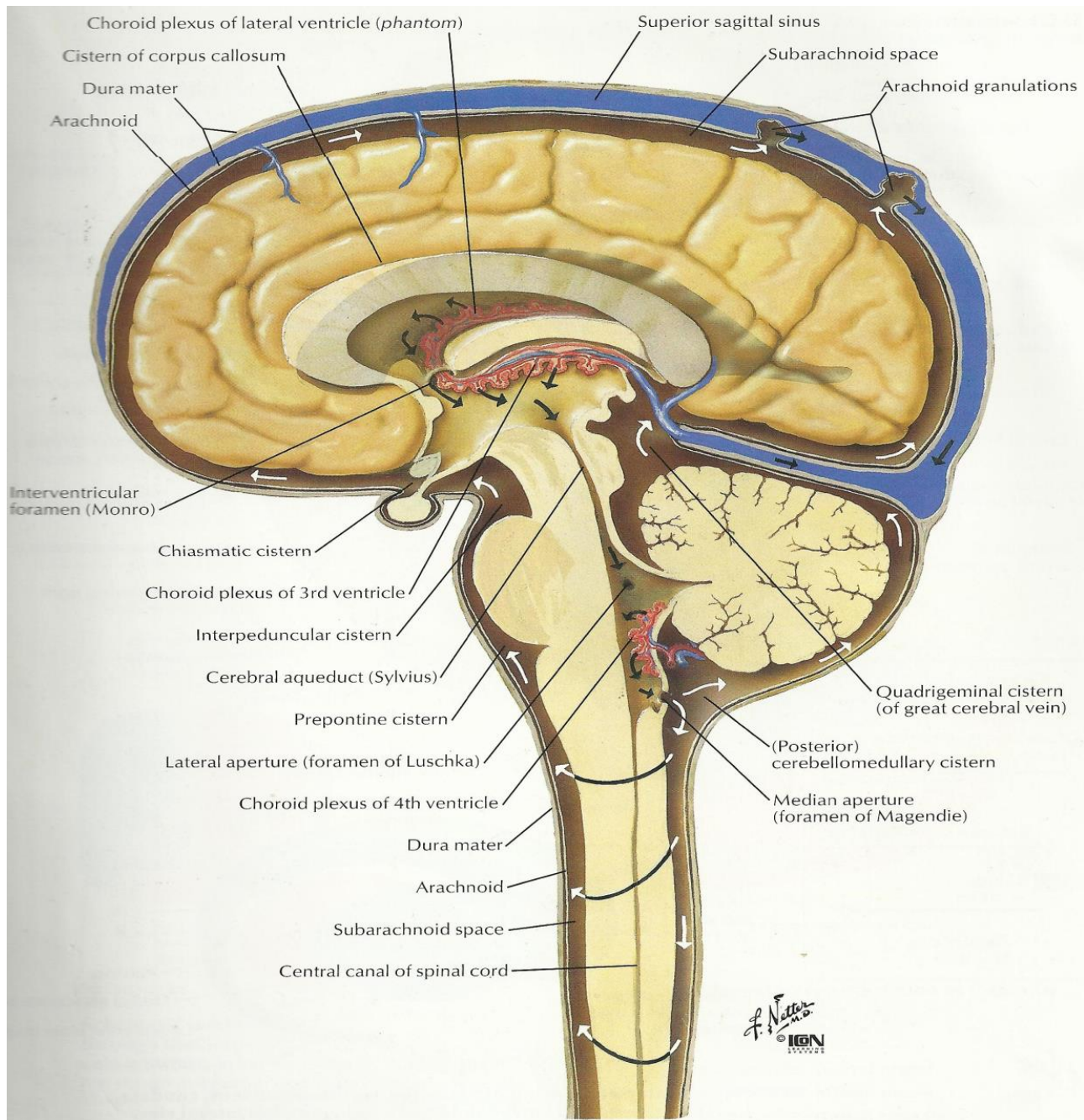
- Total CSF produces is **500 ml/day**.
- Production rate of **20 ml/hour**.
- Total CSF volume is 140-150 ml.
 - 30 ml in ventricles.
 - 80 ml in subarachnoid space of the brain.
 - 30 ml in subarachnoid space of spine

- **Intracranial Pressure (ICP)::**

- Normal pressure in a supine position **5-15 cm/H₂O**.
- Activities (Change in position, straining, and sneezing) transiently increasing ICP to 40 cm/H₂O.
- ICP is maintained by balance between CSF secretion from the choroid plexus and its resorption by the arachnoid villi.
- Because CSF secretion occurs at a steady rate, the rate of CSF resorption plays the major role in determining CSF pressure.
- Processes disrupt CSF resorption will lead to increased ICP.

Table 1: Causes of Increased Intracranial Pressure

Aneurysm rupture	Cerebral edema
Encephalitis	Epidural hematoma
Head injury	Hydrocephalus
Intraventricular hemorrhage	Meningitis
Status epilepticus	Stroke/Intracerebral hemorrhage
Subdural hematoma	Tumor



Box 52-1. CEREBROSPINAL FLUID RHINORRHEA CLASSIFICATION

Traumatic

Accidental

Immediate

Delayed

Surgical

Complication of neurosurgical procedures:

- Transsphenoidal hypophysectomy
- Frontal craniotomy
- Other skull base procedures

Complication of rhinologic procedures:

- Sinus surgery
- Septoplasty
- Other combined skull base procedures

Nontraumatic

Elevated Intracranial Pressure

Intracranial neoplasm

Hydrocephalus:

- Noncommunicating
- Obstructive

Benign intracranial hypertension

Normal Intracranial Pressure

Congenital anomaly

Skull base neoplasm:

- Nasopharyngeal carcinoma
- Sinonasal malignancy

Skull base erosive process:

- Sinus mucocele
- Osteomyelitis

Idiopathic

- **Causes of CSF Rhinorrhea:**

1. Acquired:

- Traumatic (95%):
 - Accidental Head Trauma (80%).
 - Iatrogenic Surgical Trauma (15%).
- Non-Traumatic (5%):
 - Normal ICP.
 - High ICP.

2. Congenital.

- **Acquired Traumatic Causes of CSF Rhinorrhea:**

- Most common site is Anterior cranial fossa.
 - Dura is tightly adherent to the thin skull base and easily torn with injury to the skull base.
- **Head Trauma (Accidental):**
 - **Most common cause of CSF leak.**
 - 2-4% of patients with blunt head injury.
 - Most CSF fistulas that result from accidental head trauma resolve spontaneously or with conservative management.
 - Presentation:
 - 1. Immediate:**
 - Most common presentation (80%).
 - CSF leak occurs within 48 hours after trauma.
 - 2. Delayed:**
 - CSF leak occurs within 3 months.
 - Due to:
 - Delayed elevation of ICP.
 - Lysis of clots in the region of injury.
 - Resolution of edema.
 - Loss of vascularity with resultant necrosis of soft tissue and bone.
- Most common locations of skull base fractures:
 - 1.** Ethmoid-cribriform plate region.
 - 2.** Posterior table of frontal sinus
 - 3.** Orbital roof.
 - 4.** Sphenoid sinus.

- **Surgical Trauma (Iatrogenic):**

- 2nd most common cause of CSF leaks.
- Most common surgical procedures:

- 1. FESS:**

- **Most common iatrogenic cause.**
 - Risk of CSF leak is 0.5%.
 - 70% of the leak manifested intra-op.
 - 30% of the leak manifested post-op.
 - Most common side of injury is Right side.
 - Especially in right-handed surgeons, given tendency to drift medially during ethmoid dissection.
 - Most common location of injury:
 - Lateral lamella.
 - Posterior ethmoid roof.
 - At risk during high entry into basal lamella.
 - Posterior aspect of frontal recess.
 - Anatomical risk factors:
 - Asymmetrical Skull base.
 - Low-lying skull base.

- 2. Endoscopic Neurosurgical Surgeries:**

- Examples:
 - Trans-cribriiform approach to the anterior cranial fossa.
 - Trans-ethmoid approaches to anterior cranial fossa.
 - Trans-clival approach to posterior fossa.
 - The iatrogenic high-flow CSF leaks in these cases are expected sequelae of the surgical procedure and addressed as an integral component of the Surgery.

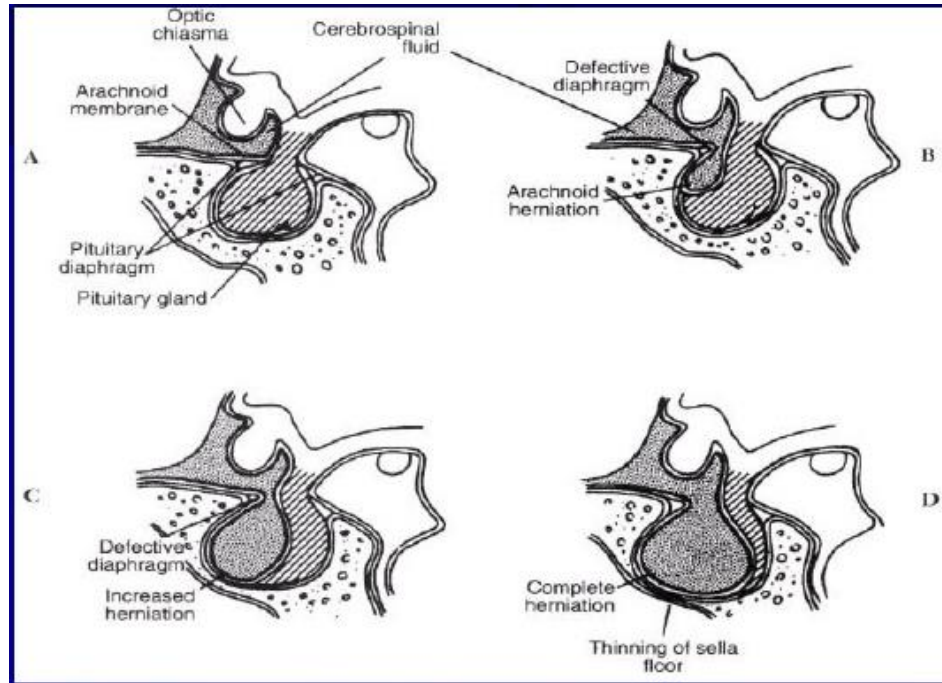
- **Acquired Non-Traumatic Causes of CSF Rhinorrhea:**

1. High ICP:

- Sustained high ICP lead to remodeling and thinning of skull base and formation of a skull base defects.
- CSF leak may serve as a release valve that decompresses the elevated ICP.
- Majority of these patients exhibit clinical and radiographic signs of elevated ICP.
- Causes:
 - Benign Intracranial Hypertension (BIH)
 - Hydrocephalus
 - Intracranial tumors

- **Benign Intracranial Hypertension/Pseudotumor Cerebri:**

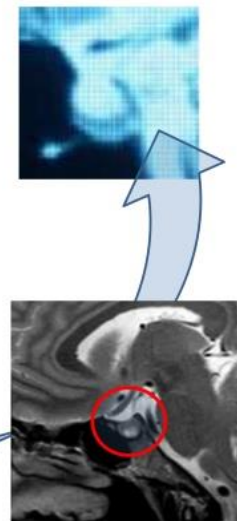
- Idiopathic increased ICP in the absence of specific causes such as intra-cranial masses, hydrocephalus, and dural sinus thrombosis.
- Most patients are middle-aged obese women.
- Clinical manifestations:
 - Headache
 - Vertigo
 - Pulsatile tinnitus
 - Papilledema
 - Visual disturbance
 - CN-6 palsy
 - Encephalocele formation
- Diagnosis:
 - Lumbar puncture opening pressure:
 - High ICP >25 cmH₂O.
 - If the patient is actively leaking CSF at the time of lumbar puncture, the opening pressure may be normal because the leak has decompressed the ICP.
 - Increased ICP may only be apparent after a site of leakage has been surgically repaired.
 - CT imaging may demonstrate a broadly attenuated skull base or arachnoid pit formation.
 - MRI imaging may show partially or completely empty sella (pituitary fossa is largely empty of tissue and replaced by CSF).



- Modified Dandy criteria to diagnose BIH:
 1. Patient is awake and alert
 2. Symptoms of raised ICP
 3. No localizing signs except for CN-6 palsy.
 4. Normal CT/MRI except for empty sella turcica.
 5. LP opening pressure of >25 cmH₂O and normal biochemical and cytological composition of CSF.



PATIENT HAD AN EMPTY SELLA
DETECTED ON MRI BRAIN



NORMAL SELLA ON MRI
BRAIN

2. **Normal ICP:**

- Examples:
 - **Extracranial tumors:**
 - Nasopharyngeal carcinoma.
 - Angiofibroma.
 - Inverting papilloma.
 - **Skull base erosive process:**
 - Sinus mucocele
 - Fungal sinusitis
 - Osteomyelitis

- **Congenital Causes of CSF Rhinorrhea:**

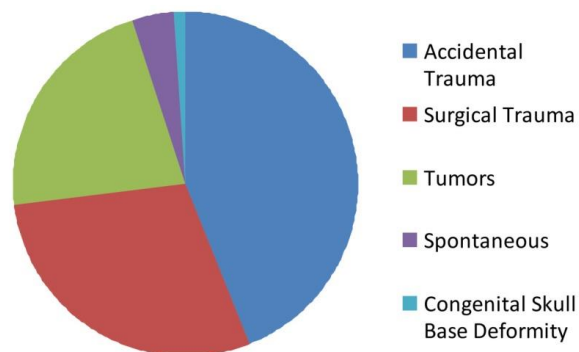
- May have either increased ICP or normal ICP.
- Causes:

1. Failure of closure of Anterior Neuropore:

- Leads to Herniation of meninges (Encephaloceles).
 - Typically involves Foramen cecum and fonticulus frontalis.
- Endoscopic techniques has facilitated minimally invasive resection of encephaloceles with layered skull base reconstruction and avoidance of external incisions with high success rate.
- In select cases, surgical management may need to be deferred till age of 2 to 3 years to allow for adequate facial growth to accommodate an endoscopic approach.

2. Persistent Craniopharyngeal Canal (Sternberg's Canal):

- Vertical defect connecting Middle cranial fossa to lateral side sphenoid sinus.
- Associated with spontaneous CSF-rhinorrhea and meningoencephaloceles in lateral recess of sphenoid sinus.
- Its existent is still a controversial topic.



- **Sites of CSF Leakage:**

- CSF from Anterior cranial fossa reaches the nose by way of:
 - Cribriform plate.
 - Ethmoid air cells.
 - Frontal sinus.
- CSF from Middle cranial fossa reaches nose via Sphenoid sinus.
- Injuries of temporal bone result in leakage of CSF into Middle ear and then via Eustachian tube into the nose (OtoRhinorrhoea).

- **Clinical Picture:**

- History:

- Unilateral clear, watery discharge on bending or straining which can't be sniffed back.
 - CSF rhinorrhoea should be differentiated from nasal discharge of allergic rhinitis.
- Postnasal drip increased in supine position.
- Salty taste in patient mouth.
- Headache resolves when CSF leak occurs.
- History of Sinonasal or neurosurgical procedure.
- History of Head trauma.
- History of Meningitis.
- History of intracranial or skull base tumors.

Table 29-1. Differences between CSF and nasal secretions

Features	CSF fluid	Nasal secretion
History	Nasal or sinus surgery, head injury or intracranial tumour	Sneezing, nasal stuffiness, itching in the nose or lacrimation
Flow of discharge	A few drops or a stream of fluid gushes down when bending forward or straining; cannot be sniffed back	Continuous, No effect of bending forward or straining. Can be sniffed back
Character of discharge	Thin, watery and clear	Slimy (mucus) or clear (tears)
Taste	Sweet	Salty
Sugar content	More than 30 mg/dl (Compare with sugar in CSF after lumbar puncture as sugar is less in CSF in meningitis)	Less than 10 mg/dl
Presence of β_2 transferrin	Always present. It is specific for CSF	Always absent

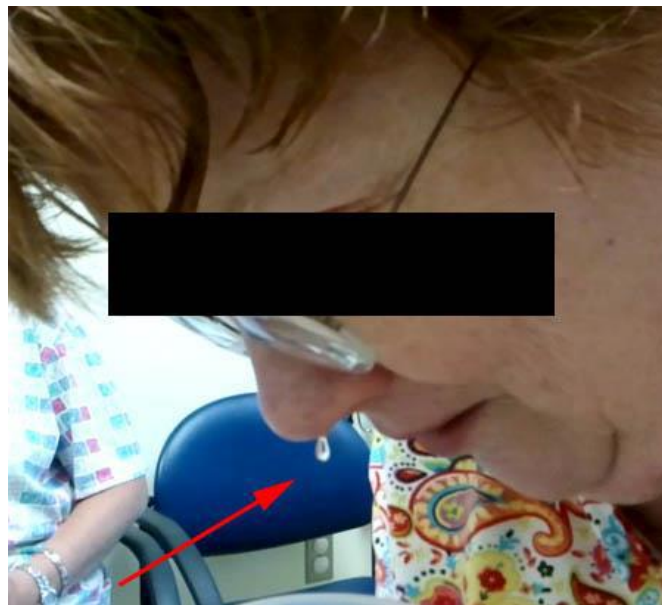
- Physical Exam:

○ **CSF Rhinorrhea:**

- When collected into a test-tube and allowed to stand, it remains clear.
- If trauma has occurred, it may be mixed with blood.
- Maneuvers to elicit a CSF leak ($\uparrow$ ICP):
 1. Chine over the chest for 1 minute with straining (**Reservoir sign / Teapot sign**).
 2. Compression of both jugular veins.

○ **Nasal endoscope:**

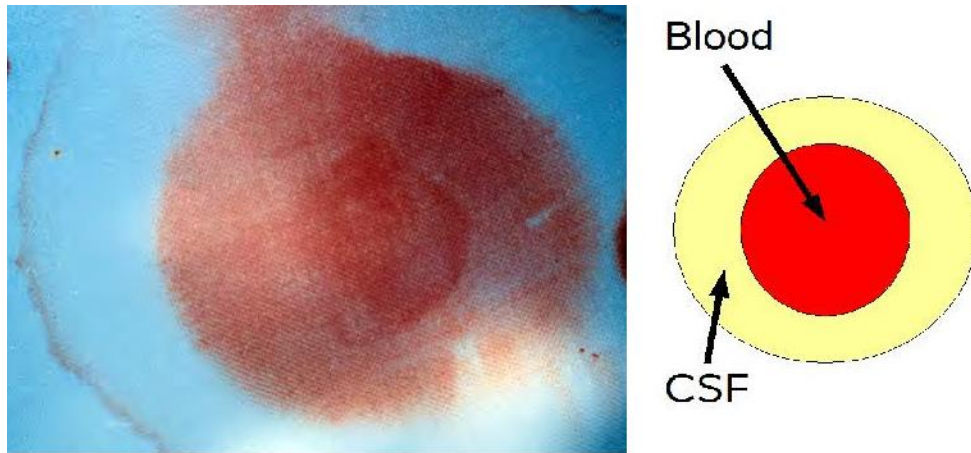
- Unremarkable in most cases.
 - Glistening moist nasal mucosa may be identified on the side of the CSF leak.
 - Stream of clear fluid may herald an active CSF leak.
 - Meningoceles and meningoencephalocele:
 - Develop from the constant pressure exerted by the dura over a weakened portion of the skull base.
 - Over time, a portion of dura may protrude through the bony defect into the paranasal sinuses and nose.
 - If the defect is large enough, and sufficient time has elapsed, a meningoencephalocele may eventually develop.
 - Compression of ipsilateral jugular vein leads to increase the size of the encephalocele (Furstenberg test).
- **Signs of High ICP:**
- Papilledema.
 - CN-6 palsy.



- **Diagnosis of CSF Rhinorrhea:**

1. Ring (Halo) sign:

- Mixing CSF with blood and placing it onto a piece of filter paper will give a Ring sign.
 - Central blood with clear ring of CSF.
- **Not specific:**
 - Occurs when mixing blood with water, saline, and other mucus.



2. Glucose Level:

- A rapid but highly unreliable test.
- Detect glucose level with use of glucose oxidase paper.
 - This paper changes color with glucose concentrations as low as 5 mg/dl.
- Not recommended as a screening or confirmatory test to detect the presence of CSF in the nasal cavity due to:
 - **False Positive Results:**
 - Presence of blood increase glucose readings.
 - Lacrimal secretions and nasal mucus contain enough glucose to change the color of Glucose oxidase paper.
 - **False Negative Results:**
 - Presence of meningitis or other intracranial infections lower glucose readings.

3. Beta-Trace Protein:

- Prostaglandin D synthase.
- Synthesized primarily in arachnoid cells, oligodendrocytes, and the choroids plexus within the CNS.
- Found in:
 1. CSF
 2. Heart
 3. Serum
- Rapid and cost-effective test and widely available.
- More sensitive and specific compared to Beta-2-Transferrin.
- Beta trace protein level in CSF should ALWAYS be compared with blood level.
 - **Cut-off point for positive CSF in nasal secretion is $\geq 0.35\text{mg/L}$.**
- **False positive results:**
 - Renal insufficiency
 - Multiple sclerosis
- **False negative results:**
 - Bacterial meningitis

4. Beta-2-Transferrin:

- **Single best laboratory test for identifying the presence of CSF in sinonasal fluid:**
 - Highly specific for CSF.
 - Not found in blood, mucus or tears.
- Found in:
 1. CSF
 2. Aqueous and vitreous humor of the eye
 3. Inner ear perilymph
- Test requires 0.5 ml of fluid.
- Specimens should be refrigerated.
 - If refrigerated, can last for 3 days.
 - If not, protein will become unstable at room temperature within 4 hours.
- **False positive results:**
 - Liver failure

- **Localization of CSF Leak:**

1. Imaging:

- Next step in diagnosing CSF rhinorrhea.

1. High Resolution CT Scan:

- Initial imaging study of choice.
- Should have 1mm cuts with axial, sagittal and coronal views.
- Sensitivity and specificity > 90%
- Advantages:
 - Detects large bony defects
 - Detects pneumocephalus
 - Detects soft tissue masses
 - Detects hydrocephalus
- Disadvantages:
 - Doesn't differentiate CSF from other soft tissue.
 - Cannot firmly establish location of CSF leak because defects may not be actively leaking and defects may be present without leak.



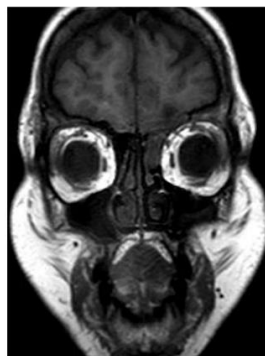
2. **CT Cisternography:**

- Helps for diagnosing occult site of Active CSF leak.
- Sensitivity of 92% with active leaks.
- Sensitivity of 40% with inactive leaks.
- **Advantages:**
 - Confirm the presence of active leak.
 - Establish the exact location of the skull base defect
 - Helpful for identifying frontal and sphenoid sinus leaks because these act as reservoir for contrast material.
- **Disadvantages:**
 - Invasive and requires injection of intrathecal contrast dye (iopendylate) prior to obtain CT scan.
 - May miss intermittent and inactive leaks.



3. **MRI:**

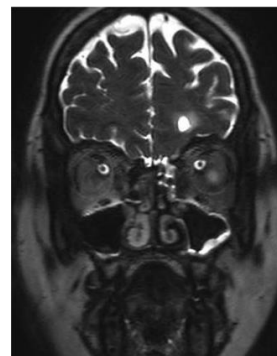
- Gold standard tool for evaluating for CSF leak.
- **T2 MRI:**
 - Differentiate between CSF and secretions vs. nasal polyposis.
 - CSF and secretions will be HYPER-intense.
 - Nasal polyposis will be HYPO-intense signal.
- **FLAIR MRI:**
 - T2 with Fluid Suppression.
 - Differentiate between CSF vs. Encephalocel and secretions.
 - Encephalocel and secretions will remain HYPER-intense.
 - CSF will be HYPO-intense signal.
- **MRI Cisternography:**
 - Non-invasive protocol.
 - Heavily T2 with Fat Suppression.
 - CSF will be HYPER-intense signal.
 - All adjacent tissues will be HYPO-intense.
 - Some machines do gray-scale reversal in which the CSF is HYPO-intense.
- **Advantages:**
 - Differentiate between Nasal secretions vs. CSF leak vs herniated intracranial contents in an opacified cell adjacent to the skull base defect on CT.
 - Detects associated intracranial abnormalities, such as aberrant vessels (**MRA**) or adjacent areas of encephalomalacia.
 - Detects presence of partially or completely empty sella in cases of spontaneous CSF leak.
- **Disadvantage:**
 - Not as good as CT when it comes to detecting bony defects.



T1 MRI



T2 MRI



T2 MRI + FAT SUPP.

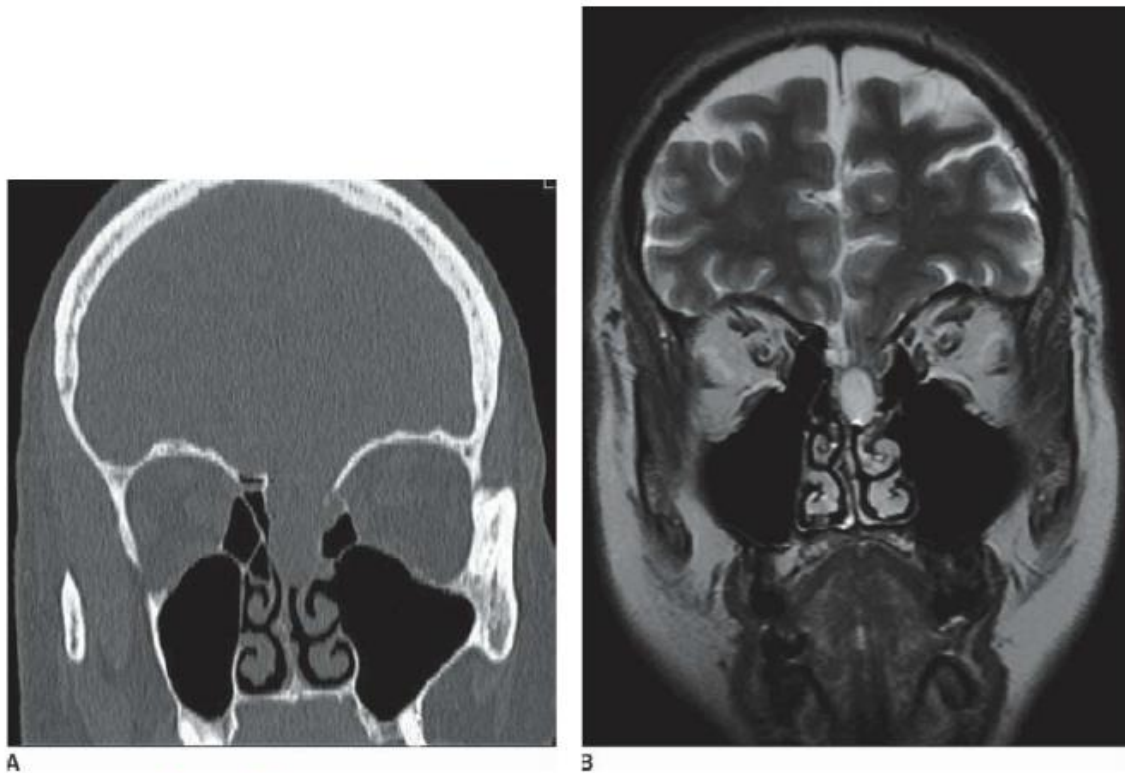
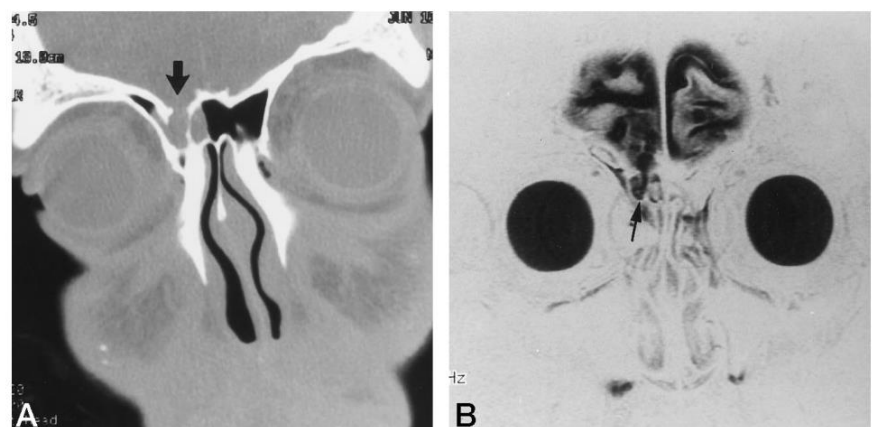


Figure 45.4 **A:** Corresponding reformatted coronal CT scan in patient from Fig. 45.3 demonstrates an extensive cribriform plate/ethmoid roof defect with resultant meningoencephalocele. **B:** Coronal T2-weighted MR shows that the mass contains both CSF and brain parenchyma.

FIG 1. Fistula detection (positive MR cisternography) in a patient with posttraumatic rhinorrhea after a motor vehicle accident.

A, Thin-section coronal CT scan shows bone defect in roof of right anterior ethmoidal sinus (*arrow*).

B, Coronal MR cisternogram shows CSF-intensity fistulous tract (*arrow*) at site of bone defect seen in **A** caused by post-traumatic fistula, confirmed at surgery.



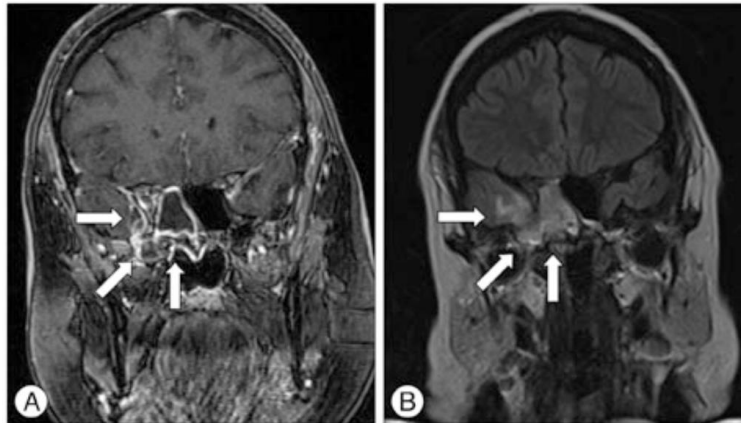


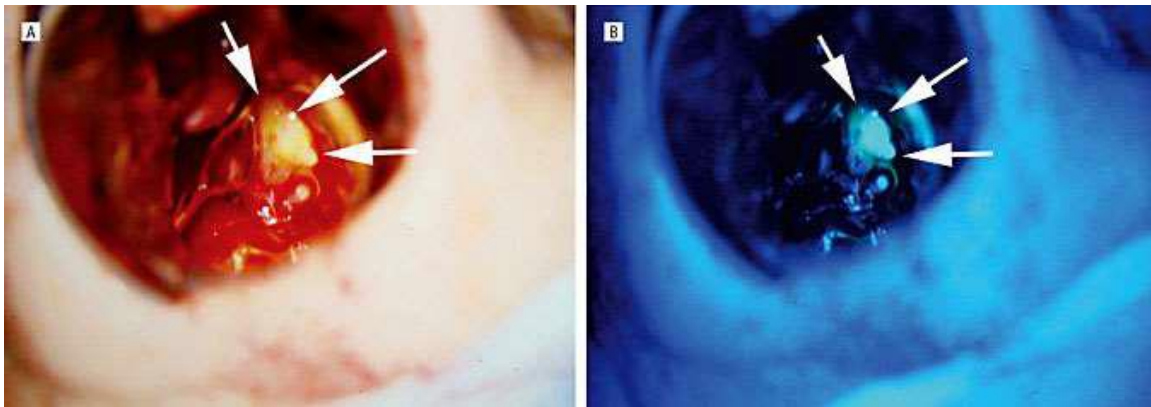
Fig. 2. A : Coronal T1-weighted magnetic resonance imaging scan of the head showing a disruption of the skull base in the area of the roof of the right sphenoid sinus where the encephalocele penetrated. A contrast enhancement in terms of an inflammatory area reaching to the fronto-mesial temporal lobe is detectable (arrows). B : Coronal T2-weighted/FLAIR magnetic resonance imaging scan of the head showing a high signal intensity of the right temporal lobe reconcilable with an encephalitis (arrows). FLAIR : fluid attenuated inversion recovery.

4. **Radionuclide Cisternography:**

- Intrathecal injection of radioactive tracers.
 - Technetium-99, I-131, Indium 111.
- Pledgets placed at areas suspected of leak and scintigrams of the skull are obtained.
- Pledgets are removed after 12-24 hours and measured for radioactive tracer.
- Pledges location:
 - Olfactory slit → Cribriform plate
 - Middle meatus → Frontal or ethmoid sinuses
 - Sphenoethmoidal recess → Sphenoid sinus
 - Inferior meatus near the eustachian tube → Temporal bone
- Advantages:
 - Useful for very slow or intermittent leak since the pledgets are left in place for many hours and can continuously collect CSP.
- Drawbacks:
 - Low sensitivity 60-70%.
 - Poor localization in most cases.

2. Intra-thecal injection of Fluorescein dye:

- Good for pre-op and intra-op localization of active CSF leaks to ensure watertight closure.
- Indications:
 - Multiple skull base defects
 - Abnormal skull base anatomy from trauma.
- Lumbar drain is done and 15 ml of patient CSF is withdrawn.
- Mixing of 0.1 ml of 10% Fluorescein dye with 10 ml of Patient CSF is performed then injected intrathecally over 10 mins and then flushed with 5 ml of patient CSF.
- Examination done with nasal endoscope 30-60 minutes later.
- In most cases, Dye can be seen without filters.
- Smaller defects may require **BLUE** light filter.
- Fluoresceine is not U.S FDA approved for intrathecal injection because seizures, encephalitis and neurotoxicity have been reported with using higher concentrations or rapid injections.
 - Arrhythmia that resolved within 24h was the only complication observed over 15 years of intra-thecal Fluoresceine use in this concentration.
- Topical intranasal application of 5% fluorescein can be used intra-op to evaluate presence of active CSF leak.
 - CSF will change the fluorescein from yellow to green.



- **Management of CSF Leak:**

- Management of CSF leak is of extreme importance as presence of a CSF leak increases the risk of meningitis 10-fold.
- Generally:
 - Traumatic leaks respond well to conservative management.
 - Spontaneous leaks tend to require surgical correction.

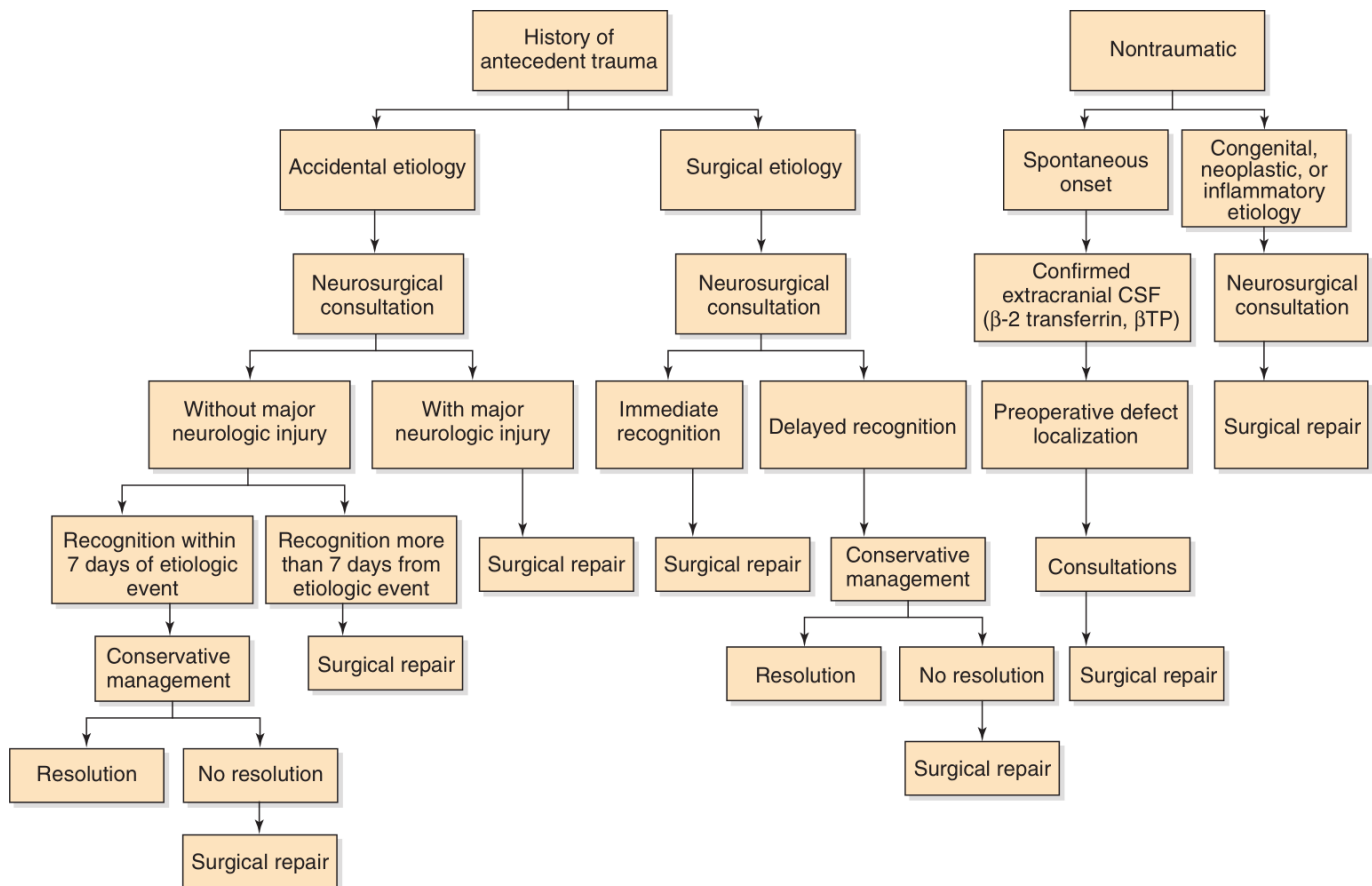


FIGURE 52-9. Management strategy for cerebrospinal fluid (CSF) rhinorrhea. β TP, β -trace protein.

- **Conservative Management:**

- **Indications:**
 - Early (not immediate) recognition of traumatic leak (surgical and non-surgical).
 - Spontaneous Leaks (unlikely to be successful).
- The goal of conservative management is to reduce the CSF leak flow by decompressing the intracranial pressure; in this way, healing at the defect site may seal the leak without surgical intervention.
- 70% of traumatic CSF leak will spontaneously resolve with conservative measures.
 - Dura does not regenerate in these cases and skull base only be covered by a thin monolayer of nasal mucosa or fibrous tissue which break down over time by sinonasal inflammatory processes or by pulsatile effect of the brain.
 - 30% incidence of meningitis with long-term follow-up of CSF leak that are managed conservatively.
- **Consist of:**
 - 1. Bed Rest (5-7 days):**
 - Head of bed elevated 30 degrees.
 - Maintain normal BP.
 - 2. Avoid (6-8 weeks):**
 - Sneezing (Anti-histamine).
 - Coughing (Anti-tussives).
 - Vomiting (Anti-emetics).
 - Straining (Stool softeners).
 - Nose blowing.
 - Heavy lifting (> 10lbs = 4.5kg).
 - 3. Antibiotics:**
 - Controversial.
 - Used to prevent intracranial infection (meningitis).
 - Studies have shown no difference in preventing intracranial infection with or without their use.
 - Risk of developing more virulent bacterial strains.
 - 3rd generation cephalosporin with blood-brain barrier penetration is used mainly in presence of Lumbar drain.
 - 4. Diuretics:**
 - Indicated in patients with high ICP (benign intracranial hypertension).
 - Acetazolamide (Diamox) is used to decrease ICP.
 - Decrease CSF leak recurrence by decrease CSF production up to 50%.
 - Dose of 500mg BID.

5. Lumbar Drain (5-7 days):

- Indicated if CSF leak fails to respond after 5-7 days of conservative management.
- Function to lower ICP and reduce flow through defect.
- Allows for measuring of ICP.
- Continuous drainage at rate of 10-15 ml/hr.
 - Higher drainage rates produce a greater decompression of ICP leading to severe headache.
 - Low ICP also may cause pneumocephalus because air is drawn through the skull base defect.
- Patient should be covered with antibiotics cross the brain blood barrier (3rd generation cephalosporin)
- Complications:
 - Meningitis
 - Pneumocephalus
 - Headache
 - Paresthesias
 - Cellulites

- Surgical Management:

- Indications:
 1. Failed conservative management
 2. Intra-op recognition of CSF leak
 3. Large defects/leaks
 - Especially in association with pneumocephalus.
 4. Idiopathic (spontaneous) leaks.
 5. Open traumatic head wounds with CSF leakage
- Basic Surgical Principles:
 - Precise identification of CSF leak site is an absolute requisite to attaining good closure.
 - This may be accomplished through use of preoperative CT imaging or intraoperative surgical navigation, along with intraoperative intra-theal fluorescein injection.
 - Intra-op iatrogenic injury with immediate recognition/onset CSF leaks should be repaired at the time that they are recognized.
 - Surgical approach differs based on location and size of skull base defect.

- **Open Transcranial Approach:**

○ Indications:

1. Comminuted skull fractures with displaced fragments requiring reduction.
2. Extensive skull base fractures.
3. Fractures associated with intracranial hemorrhages or contusions that require craniotomy for treatment.

- After craniotomy, the defect site is identified, and then a tissue graft is placed to close the defect.
- Access to the cribriform plate region and roof of the ethmoid requires a frontal craniotomy.
- Extended craniotomy and skull base techniques with even greater brain compression provides access to the sphenoid sinus defects.

○ Advantages

- Direct visualization of defect
- Inspection of adjacent cerebral cortex
- Better chance of patching a defect in the face of increased ICP.

○ Disadvantages:

- Increased morbidity
- Increased hospital time
- Injury to brain from retraction (hematoma, seizures, cognitive dysfunction, risk of permanent anosmia)
- Not good for visualization of sphenoid sinus
- Despite direct access to the skull base defect, failure rates are quite high exceeding 25%.

- **Endoscopic Transnasal Approach:**

- Success rates of more than 90% in primary repair and 95% in secondary repair.
- Good visualization and exposure is the key.
- Highest failure rates occur in spontaneous leak and intracranial hypertension.
 - Decreasing the underlying intracranial hypertension is essential for increasing success rate in patients with spontaneous CSF leak.
 - Medical management with acetazolamide is the best option at this time.
- Sites of failure:
 - Sphenoid 50%, Ethmoid 40%, Frontal 15%.
- Advantages:
 - Better magnified visualization
 - Angled visualization
 - No external incisions
 - Minimizes intranasal mucosal injuries
- If Encephalocele is present:
 - Cauterize encephaloceles stalk with bipolar cautery to avoid retracting the sac into the cranial cavity and subsequent intracranial hemorrhage and stroke.
 - Monopolar devices should be avoided as the depth of heat penetration cannot be controlled and may result in dural or brain parenchymal injury.
- For good results:
 - All mucosa at the defect should be removed circumferentially for 5 mm to decrease risk of mucus lifting the graft from the recipient bed and risk of mucocele formation.
 - Gentle drilling of the bone adjacent to the defect may stimulate osteoneogenesis and further facilitate the closure.

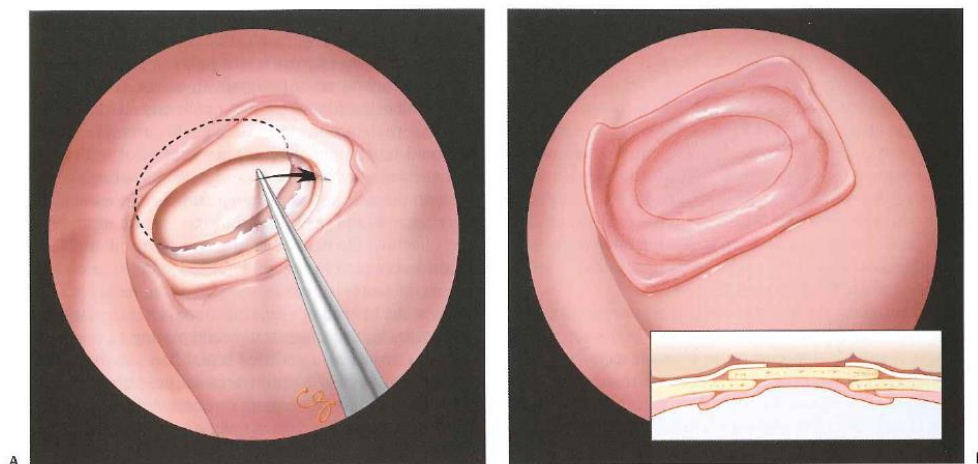
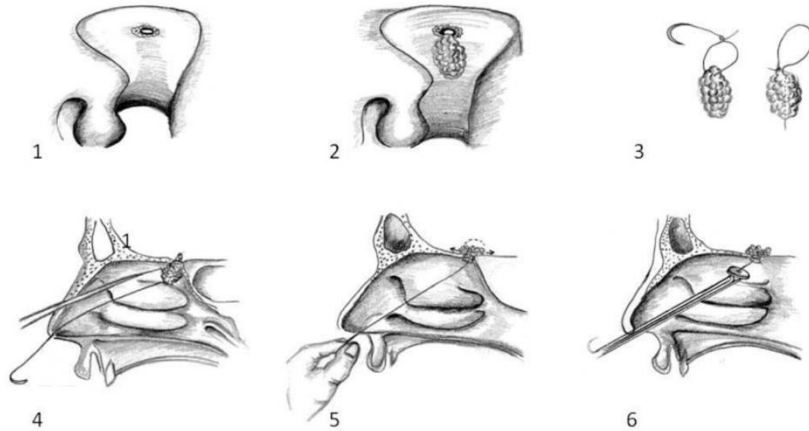


Fig. 44.5 Illustrations demonstrating the proper technique in the placement of an underlay bone graft (A) and an overlay mucosal graft (B).

- Type of grafting material:
 - **Fat bath plug:**
 - Abdominal wall or ear lobule fat.
 - Used for repairing anterior skull base defects.
 - A fat plug, three times the width of the defect, is positioned in underlay in the epidural or sub-dural space.
 - It overcomes the increased ICP by preventing the high pressure from displacing the graft.
 - In one series, 90% of repairs by the bath plug technique were successful.
 - **Dural substitutes:**
 - Xenogeneic collagen dural substitutes (Durepair, Dura-Gen, Dura-Guard).
 - Provides a scaffold for the native fibroblasts to produce a collagen layer that blends and eventually replaces the implant.
 - **Fascia:**
 - Autograft (Temporalis, Fascia lata).
 - Irradiated cadaveric Tutoplast (Fascia lata).
 - **Cartilage (Septum).**
 - **Bone (Septum, Middle turbinate):**
 - Not recommended following skull base tumor resection if the patient will undergo postoperative radiation due to the risk of osteroradionecrosis.
 - **Medpor implant:**
 - Biocompatible porous polyethylene implants for craniofacial reconstruction and augmentation.
 - **Mucosa:**
 - Nasal floor.
 - Middle turbinate mucosa.
 - Should be 30% larger than defect to account for shrinkage.
 - **Pedicled Middle Turbinate Flap:**
 - Based on branches of sphenopalatine artery.
 - **Pedicled Septonasal flap:**
 - Based on posterior septal branch of sphenopalatine artery.
 - Used mainly in patients with large dural high-flow defects (in the setting of endoscopic skull surgery) or if there is history of prior radiation.
 - Tend to tent, fold and contract, so not as good as free tissue use.

Fig. 1. The bath plug technique for repair of anterior skull base defects. (1) Mucosal stripping around the defect. (2) Harvesting an oblong fat plug three times the width of the defect. (3) A knot is tightened at the top of the fat plug. The suture is passed through the whole length of the plug. (4) The fat plug is gently manipulated through the defect while the suture is still attached to it. (5) Traction on the suture to expand the plug. (6) Reinforcement of the reconstruction with overlay mucosal graft ascended along the suture.



○ Grafting Techniques:

- Size and location of the defect play a role in the grafting technique utilized in CSF leak repair.

1. Underlay/Inlay:

- Place graft between dura and bony defect.
- Used with overlay grafts in multilayered reconstruction.
- Materials:
 - Rigid grafts (bone, cartilage or medpor implants).
 - Soft tissue grafts (fat or fascia graft).
 - Mucosal grafts should never be placed intracranially to avoid risk of intracranial mucocele.

2. Overlay/Onlay:

- Place graft directly over defect.
- Indications (as only reconstruction method):
 - Small skull base defects (<5mm).
 - Cribriform plate defects.
- Materials:
 - Soft tissue graft (Fascia, free mucosal graft or pedicled mucosal flap).

3. Multilayered:

- Utilized both underlay and overlay grafts together.
- Indications (as only reconstruction method):
 - Large skull base defects (>5mm).
 - High flow CSF leak.
 - Patients with high ICP.
- Gasket seal method:
 - Watertight closure of skull base defect.
 - Fascia is placed underlay with Medpor implant or bone graft as a buttress.



- **Multilayered skull base Repair:**

A: Dura

B: Fascial autograft (**Underlay**)

C: Bone or Cartilage autograft (**Underlay**)

D: Fascial autograft (**Underlay**)

E: Mucosal free autograft (**Overlay**)

F: Surgical sealant

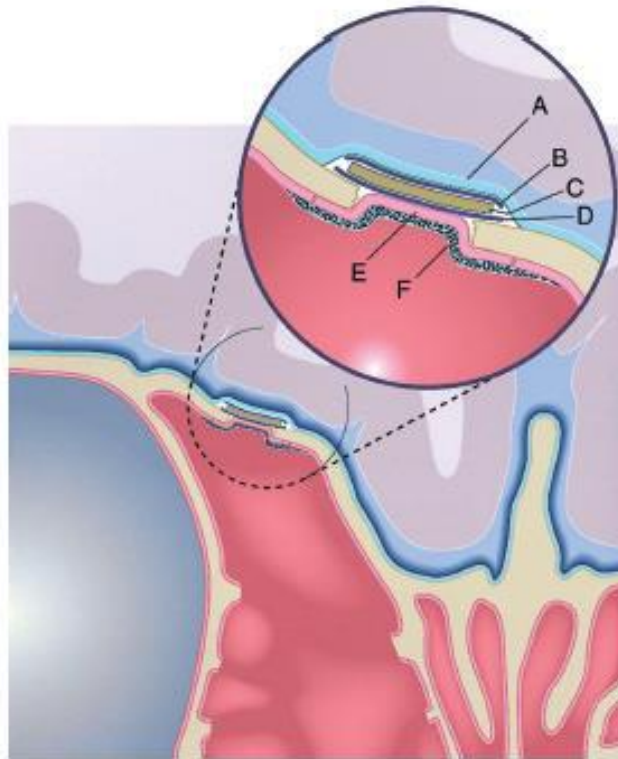


Fig. 1. Correct placement of layers for reconstruction of ethmoid roof and cribriform plate defects is illustrated in schematic coronal image. The acellular dermal allograft and bone/cartilage autograft is placed intracranially. (A) Dura; (B) acellular dermal allograft; (C) bone and/or cartilage; (D) acellular dermal allograft; (E) mucosal graft; (F) fibrin glue.

○ **Steps to improve the seal and stability of the reconstruction:**

- Fibrin glue
- Gelfoam packing over the seal.
- The entire reconstruction is further supported by non-absorbable nasal pack.

Site-Specific Repair Strategy:

- **Cribriform Plate/Ethmoid Roof:**
 - Amenable to repair by a purely endoscopic approach.
 - Maxillary antrostomy, total ethmoidectomy, frontal sinusotomy and sphenoidotomy are required to:
 - Minimize risk of scarring and mucocele formation.
 - Enhance exposure to anterior and posterior ethmoid roof.
 - **One exception to this approach is small cribriform plate encephaloceles that resides strictly in the olfactory cleft.**
 - Middle turbinate can be lateralized and the CSF leak can be addressed via transnasal approach to olfactory cleft.
 - Middle turbinate can be partially resected to increase exposure, if necessary.
 - Bone at Ethmoid roof is thick; and reconstruction with a bone graft can be placed in underlay fashion.
 - Bone of cribriform plate is thin and any attempts to place underlay grafts can further disrupt the bone and substantially increase the size of the skull base defect.
 - Ablating the mucosa by bipolar cauterization and placing overlay graft or pedicled flap should suffice for these olfactory cleft meningoceles.

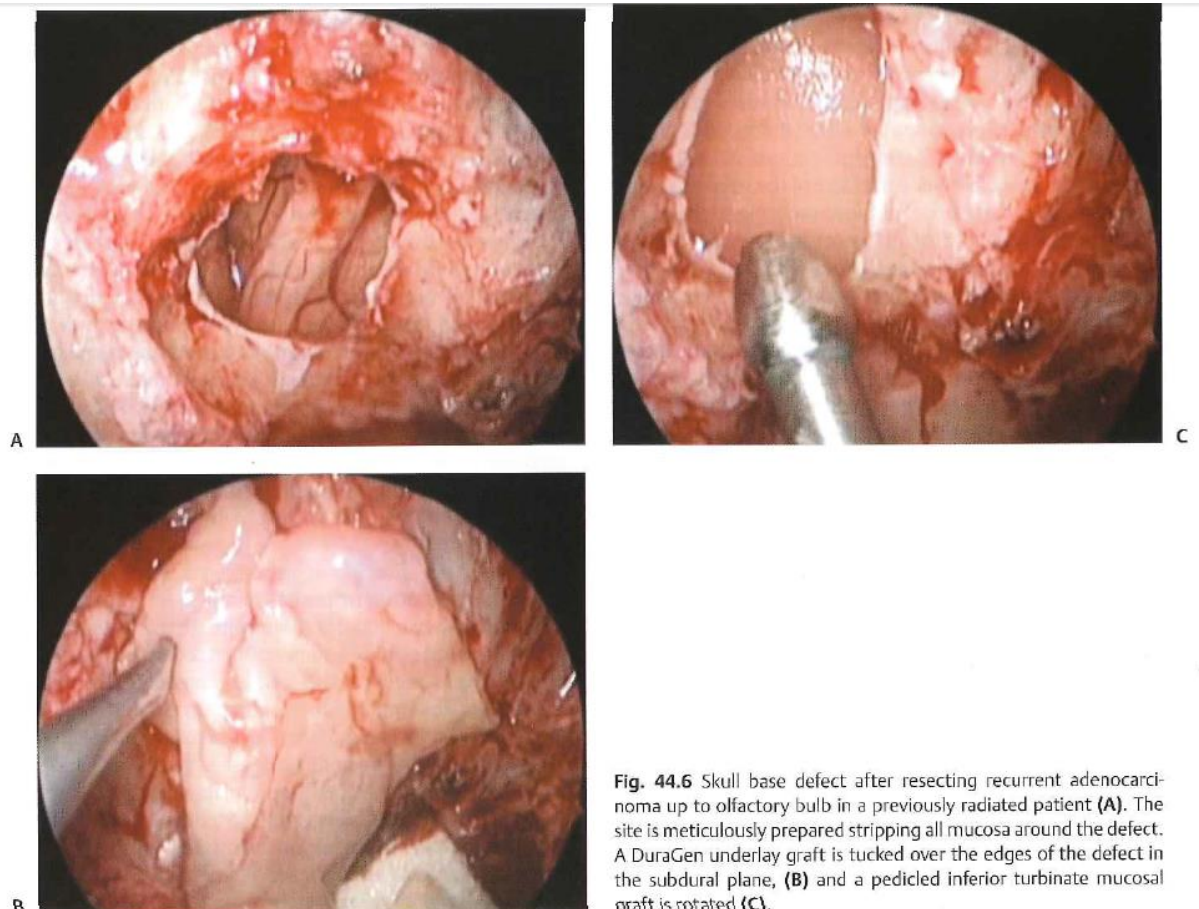


Fig. 44.6 Skull base defect after resecting recurrent adenocarcinoma up to olfactory bulb in a previously radiated patient (A). The site is meticulously prepared stripping all mucosa around the defect. A DuraGen underlay graft is tucked over the edges of the defect in the subdural plane, (B) and a pedicled inferior turbinate mucosal graft is rotated (C).

- **Sphenoid Sinus:**

- Central sphenoid leaks:
 - Managed via an endoscopic transnasal or transtethmoid approach.
 - Wide sphenoidotomy extending from the planum sphenoidale to floor of sphenoid and from lateral sphenoid wall to intersphenoid septum is essential to gaining wide exposure.
 - Great care must be exercised during surgical maneuvers inside the sphenoid given close proximity of internal carotid artery (ICA) and optic nerve.
 - Careful fulguration of herniated dura and brain and circumferential removal of the mucosa around the defect remain key steps.
 - Defects of planum may be repaired with use of epidural bone grafts, along with other tissue arrays.
 - Nasoseptal flap may be used for skull base reconstruction of the central sphenoid.
 - Fat obliteration of sphenoid sinus after removal of all sphenoid sinus mucosa can be used as well.
 - Complete removal of mucosa is difficult and may pose risk to the ICA and optic nerve.
 - Any residual mucosa may result in mucocele formation.

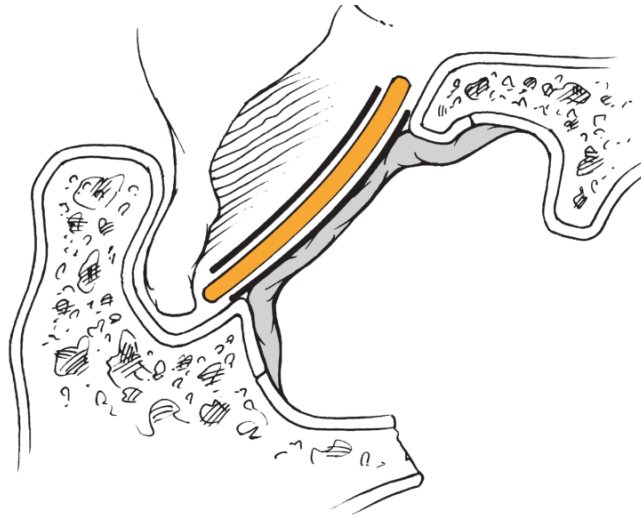


FIGURE 52-12. Layered reconstruction also provides an optimal barrier for sphenoid cerebrospinal fluid leaks, as shown in the illustration of such a repair for a midline sphenoid defect (after transsphenoidal hypophysectomy). Layers are as indicated in [Figure 52-11](#). (Modified from Lorenz RR, Dean RL, Hurley DB, et al: Endoscopic reconstruction of anterior and middle cranial fossa defects using acellular dermal allograft. *Laryngoscope* 113:496–501, 2003.)

- Lateral recess leaks:
 - Surgical challenge.
 - Not amenable to standard transnasal or transtethmoid approaches.
 - Transpterygoid approach affords access to lateral sphenoid sinus.
 - Wide maxillary antrostomy, ethmoidectomy, and sphenoidotomy are performed to establish initial landmarks.
 - Posterior maxillary sinus wall is removed to expose Pterygomaxillary fossa (PMF).
 - Dissection in the PMF will expose internal maxillary artery, which is either divided or reflected inferiorly.
 - Great care must be taken to preserve sphenopalatine ganglion, along with vidian, greater and lesser palatine, and infraorbital nerves.
 - Pterygoid process that forms anterior wall of lateral recess is gently drilled to gain exposure.
 - Skull base defect is then managed as per standard technique.
 - Since spontaneous CSF rhinorrhea form majority of the leaks in this region, placement of a bone graft in the epidural space is preferred as a component of the definitive reconstruction.



Figure 45.6 **A:** Reformatted coronal bone window CT shows a soft tissue mass in the left sphenoid sinus with a planum sphenoidale defect. **B:** Postoperative reformatted coronal bone window CT demonstrates septal bone graft (arrow) in the epidural space. The associated soft tissue density in the sphenoid sinus represents packing and mucus in the early postop period.

- **Frontal Sinus:**

- Surgical challenge.
- Narrow anatomic confines of the frontal recess and proximity to critical structures, including the skull base and orbit.
- Two important goals:
 1. Successful repair of skull base defect
 2. Maintaining patency of frontal sinus or successfully obliterating the sinus with meticulous removal of all mucosa.
- Great care must be taken to preserve uninvolved mucosa to minimize risk of frontal sinus stenosis and mucocele formation.
- Two most common locations of frontal leaks include:
 - Frontal recess:
 - Repaired via an endoscopic frontal sinusotomy or Draf IIB approach if greater exposure is required.
 - Posterior table of frontal sinus:
 - Repaired via an endoscopic modified Lothrop procedure (Draf III) providing broad exposure of frontal sinuses orbit to orbit.
- Nasoseptal flap is useful for repairing skull base defects involving the frontal sinus and can cover up to 3 cm of the posterior table.
- Defects in an extreme superior or lateral frontal sinus location restrict the ability to address these endoscopically.
 - An adjunct endoscopic frontal trephination or osteoplastic flap may be required to address these leaks.



Fig. 44.7 A 70-degree nasal endoscopic view of a right frontal sinus posterior table cerebrospinal fluid leak repair with a nasoseptal flap at 1 year postoperatively. The flap extends 1 cm up the posterior table here, but can reach 3 cm for certain defects.

- **Nasoseptal Flap:**
- Based on posterior septal branch of sphenopalatine artery.
- Used mainly in patients with large dural high-flow defects (in the setting of endoscopic skull surgery) or if there is history of prior radiation.

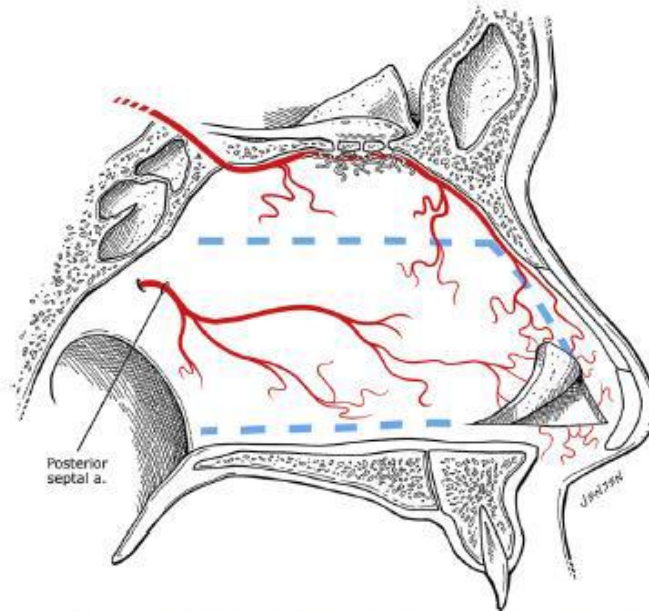


Fig. 5. Axial view of the nasal septum demonstrating the posterior septal branch of the sphenopalatine artery.

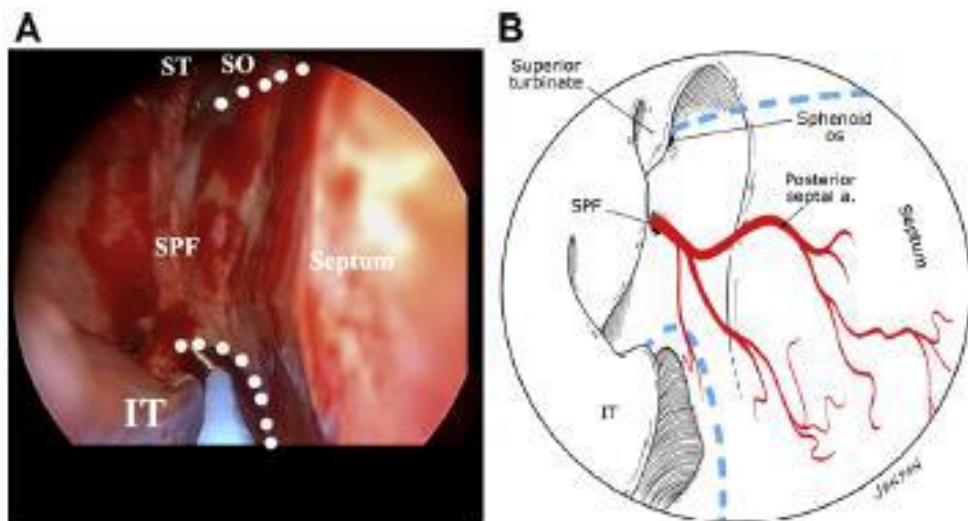


Fig. 4. Endoscopic view of the posterior nasal cavity (A) with an artist's representation of the same view (B). Dashed lines mark the location of cuts for the posteriorly based nasal septal flap. IT, inferior turbinate; SO, sphenoid os; SPF, sphenopalatine fossa; ST, superior turbinate.

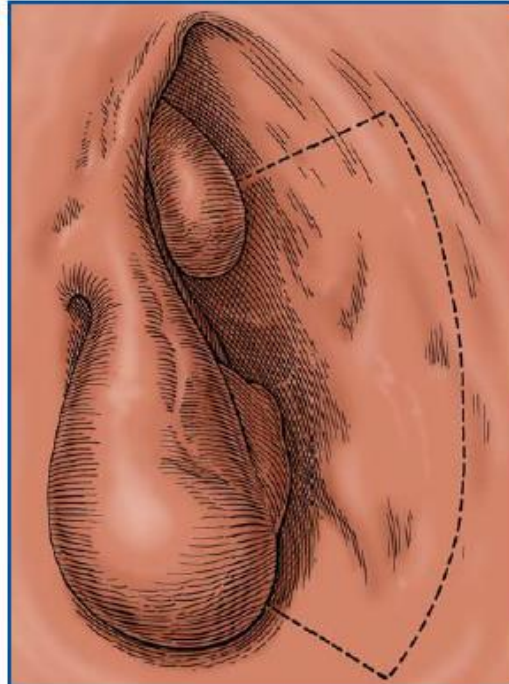


FIGURE 1. The nasoseptal flap incisions at the anterior nasal cavity. Two parallel incisions are joined by a vertical incision anterior to the inferior turbinate.

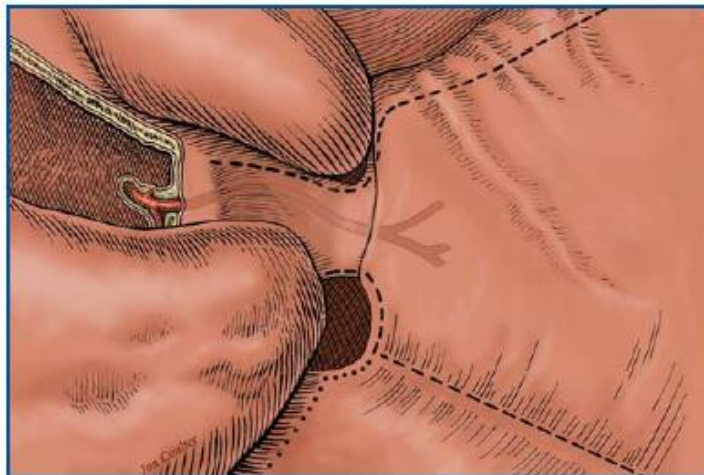


FIGURE 2. The nasoseptal flap incisions at the right posterior nasal cavity. Two parallel incisions (dashed lines), one following the maxillary crest and the other 1 to 2 cm inferior to the olfactory cleft, are extended to reach the lateral nasal wall. The inferior incision follows the free edge of the posterior septum and then crosses the posterior choana. The inferior incision may be designed to include the mucoperiosteum of the floor of the nose. The superior incision extends laterally to cross the rostrum of the sphenoid sinus at the level of its natural ostium. A large middle antrostomy and exposure of the terminal internal maxillary artery are illustrated. This is only necessary if the flap will be stored in the maxillary sinus so as to approach the clival and paraclival areas.

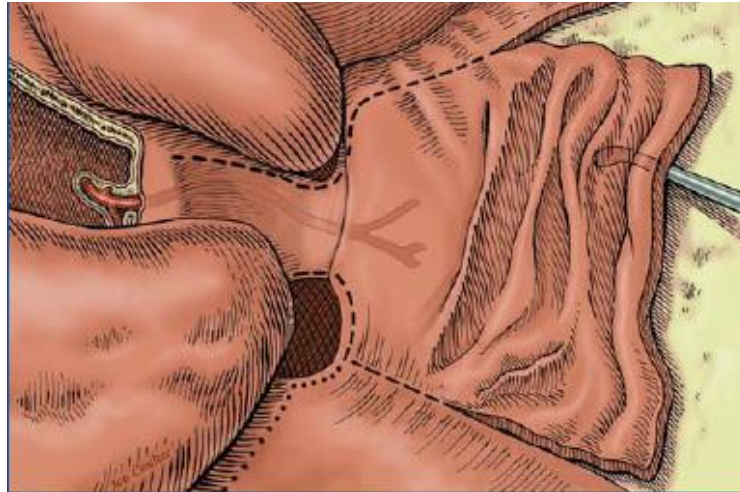
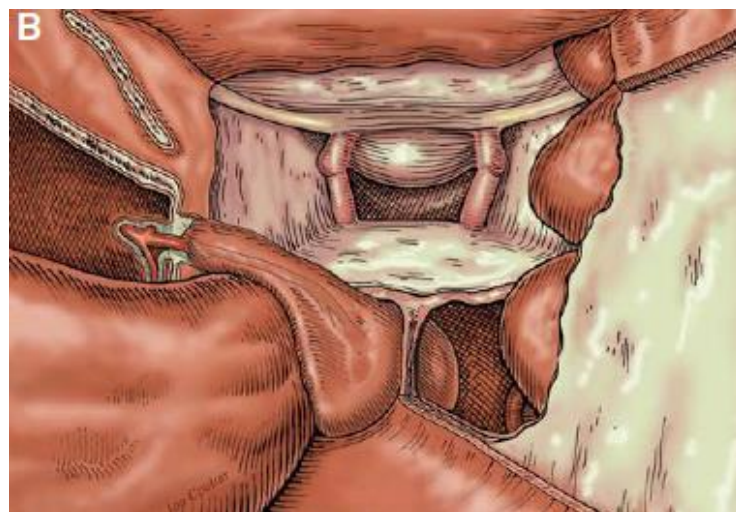


FIGURE 3. Elevation of the nasoseptal flap following a subperichondrial and subperiosteal plane.



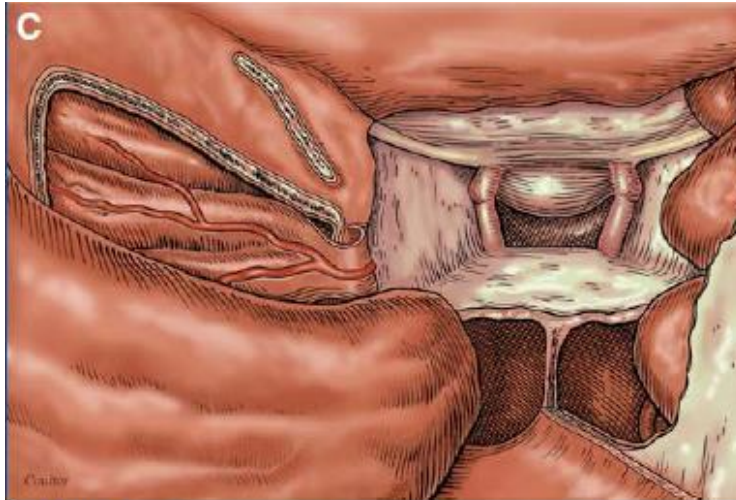


FIGURE 4. *A, the nasoseptal flap is mobilized posteriorly after a posterior septectomy. B, the nasoseptal flap is "stored" at the nasopharynx. C, the nasoseptal flap is "stored" inside the maxillary sinus.*

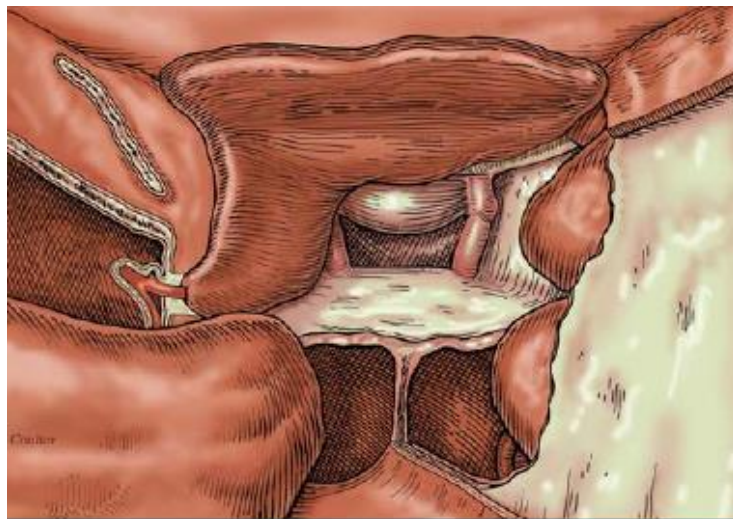


FIGURE 5. *The nasoseptal flap is shown covering a defect at the planum sphenoidale.*

- **Post-Operative Management:**

1. Bed Rest (5-7 days):

- Head of bed elevated 30 degrees.
- Maintain normal BP.

2. Avoid (6-8 weeks):

- Sneezing (Anti-histamine).
- Coughing (Anti-tussives).
- Vomiting (Anti-emetics).
- Straining (Stool softeners).
- Nose blowing.
- Heavy lifting (> 10lbs = 4.5kg).

3. Nasal Pack:

- Kept for 5 days.
- Patient should be covered with antibiotics.

4. Antibiotics:

- Used to prevent intracranial infection (meningitis).
- 3rd generation cephalosporins with blood-brain barrier penetration is used mainly in presence of nasal pack or Lumbar drain.

5. Diuretics:

- Indicated in patients with high ICP (benign intracranial hypertension).
- Acetazolamide (Diamox) is used to decrease ICP.
- Decrease CSF leak recurrence by decrease CSF production up to 50%.
- Dose of 500mg BID.
- Important side effect is development of hyperchloremic metabolic acidosis with hypokalemia.

6. Lumbar Drain (5-7 days):

- Function to lower ICP and allowing for better apposition of the graft to the recipient site and attenuating sudden ICP changes.
- The recommendation is against the routine use of lumbar drain in all cases of skull base reconstruction based on the available evidence.
- Indicated mainly:
 - High-pressure or high-flow leak
 - Problematic skull base reconstruction
 - Persistent postoperative CSF Rhinorrhea.
- Continuous drainage at rate of 10-15 ml/hr.
 - Prevent side effects such as headaches, nausea, and emesis.
- The LD is then clamped and patient is carefully observed for any signs of CSF leakage in the bed rest position.
- Patients may slowly be elevated to a sitting position and then to a standing position as long as there is no sign of CSF leak.
- Patient should be covered with antibiotics.

7. Surgical diversion:

- Ventriculoperitoneal or lumbar peritoneal shunting.
- Considered for patient with benign intracranial hypertension to control the high ICP and prevent recurrence of CSF leak.

- Long-Term Management:

- Patients are kept on saline sprays.
- Postoperative debridement is conservative at the 1st visit post-op and limited to suctioning of mucus or debris in the dependent sinuses to maintain patency.
- Debridement of the skull base reconstruction commences 4 to 6 weeks after surgery and continues until all healing has taken place.
- Patients are instructed to avoid any vigorous activity for 6-8 weeks.
- Patients are followed long-term by serial endoscopy to ensure the success of the reconstruction and continued patency of the adjacent sinuses.

Facial Trauma

- Injuries of face may involve soft tissues, bones or both.
- Facial injuries are caused by:
 - ✓ Automobile accidents (most common).
 - ✓ Sports
 - ✓ Assaults and fights.
- Management of facial trauma can be divided into:
 - General Approach.
 - Soft tissue injuries and their management.
 - Bone injuries and their management.
- ❖ General Approach for Facial trauma:
 - **Resuscitation (ABC) + Secondary survey.**
 - **Head and Neck History:**
 - **Nature of Injury.**
 - **When:** elapsed time of injury.
 - **Where:** motor vehicle, pedestrian, etc.
 - **What** was the mechanism of injury: assault (fists, bat, etc), penetrating trauma (gunshot wound, stab wound), blunt injury (motor vehicle accident), vector of force (anterior versus lateral impacts), and fall.
 - **Motor Vehicle Accidents:** speed of injury, restrained, air bag deployed.
 - **Gunshot Wound:** caliber of gun, distance fired, number of gunshots.
 - **Review of Systems**
 - **Ocular:** change in vision, diplopia.
 - **Otological:** dizziness, change in hearing, otalgia.
 - **Nasal:** nasal obstruction, epistaxis, rhinorrhea.
 - **Laryngeal:** hoarseness, difficulty breathing, odynophagia.
 - **Oral:** malocclusion, pre-injury missing teeth, dysarthria.
 - **Neurological:** loss of consciousness, numbness, weakness.
 - **General:** facial pain, shortness of breath, chest tightness.
 - **Head and Neck Physical Exam:**
 - **Otologic:**
 - **Inspection** (otoscopic exam) foreign bodies, hemotympanum, CSF otorrhea, tympanic membrane perforation, Battles' sign (mastoid ecchymosis).
 - **Palpation:** auricular hematoma.
 - **Assess Function:** hearing, balance.

- **Nasal:**
 - **Inspection:** epistaxis, septal hematoma, septal dislocations, mucosal tears, external deformities (depressed, angulation), edema, ecchymosis, open fractures.
 - **Palpation:** palpate for nasal fractures (crepitus, tenderness, step-offs, mobility).
 - **Assess Function:** CSF leak (**halo sign**, peripheral clear zone with a central pigmented spot appears when fluid placed on a gauze).
- **Mandible and Oral Cavity:**
 - **Inspection:** floor of mouth hematoma, dental exam (missing teeth, condition of dentition, dental fractures), palatal fracture, intraoral and lip lacerations.
 - **Palpation:** palpate for mandible fractures (tenderness, step-offs, mobility).
 - **Assess Function:** hypesthesia of lower lip (inferior alveolar nerve) and chin (mental nerve), occlusion.
- **Face and Facial Skeleton:**
 - **Inspection:** lacerations, hematomas, ecchymosis.
 - **Palpation:** bimanual palpation for facial fractures (crepitus, tenderness, step-offs, mobility), mobility of maxilla.
 - **Assess Function:** cranial nerve exam, sensory defects, salivary gland and ductal injuries (Parotid tissue, if exposed, is repaired by suturing. Injuries of parotid duct are more serious. Both ends of the duct are identified and sutured over a polyethylene tube, with fine suture, if severed, the **facial nerve** is exposed by superficial parotidectomy and cut ends are approximated by silk).
- **Larynx and Neck:**
 - **Inspection:** lacerations, hematomas, ecchymosis, vocal fold mobility.
 - **Palpation:** crepitus, tenderness.
 - **Assess Function:** hoarseness, stridor (auscultate), airway obstruction.

- **Ocular:**
 - **Inspection:** (fundoscopic exam) enophthalmos, proptosis, ptosis, extraocular range of motion, "raccoon eyes" suspect basilar skull fracture (cribriform fracture), eyelid lacerations, telecanthus.
 - **Palpation:** palpate bony rim (crepitus, tenderness, step-offs, mobility), periorbital soft tissue, assess medial and lateral canthus (**lid torsion test**, tests for medial canthal ligament by palpating ligament while pulling lid laterally)
 - **Assess Function:** visual acuity, nystagmus, hypesthesia of infraorbital and supraorbital nerves, assess lacrimal system (epiphora), pupil response (equal, reactive, consensual response, Marcus Gunn pupil).

- **Head and Neck Radiologic Exams:**

- **Facial Plain Films:**
 - Largely been replaced by CT (except for the mandible).
 - European Fifth (Hyperextended Base View): views posterior table of frontal sinus
 - Water's: views orbital rim, nasal bones, zygoma, and medial and lateral maxillary wall
 - Lateral Face: views frontal sinus
 - Caldwell: views orbital wall and frontal sinus
 - Submental Vertex: views zygomatic arch
- **Computed Tomography (CT):**
 - Most informative radiographic exam for head and neck trauma.
 - Axial and coronal facial CT with bone and soft tissue windows, 2–3 mm sections.
 - Inspect Areas of Concern: skull base, orbit (walls, floor, retrobulbar hematoma, muscle entrapment), vertical buttresses, horizontal beams, zygomatic arch, bony palate, mandible (condyles), soft tissue (fluid in sinuses, subcutaneous air, hematomas, etc).
- **Special Radiologic Exams:**
 - **Angiography:** indicated if risk of vascular injury.
 - **MRI:** may be considered for intracranial injury and complications.
 - **Modified Barium Swallow and Esophagram:** evaluate esophageal tears and aspiration

❖ **Bone injuries and their management**

Fracture of the face		
Upper third Above the level of supraorbital ridge	Middle third Between the supraorbital ridge and the upper teeth	Lower third Mandible and the lower teeth
1. Frontal sinuses 2. Supraorbital ridge 3. Frontal bone	1. Nasal bones and septum 2. Naso-orbital area 3. Zygoma 4. Zygomatic arch 5. Orbital floor 6. Maxilla: ○ Le Fort I (transverse) ○ Le Fort II (pyramidal)- ○ Le Fort III (craniofacial dysjunction)	1. Alveolar process 2. Symphysis 3. Body 4. Angle 5. Ascending ramus 6. Condyle 7. Temporomandibular joint

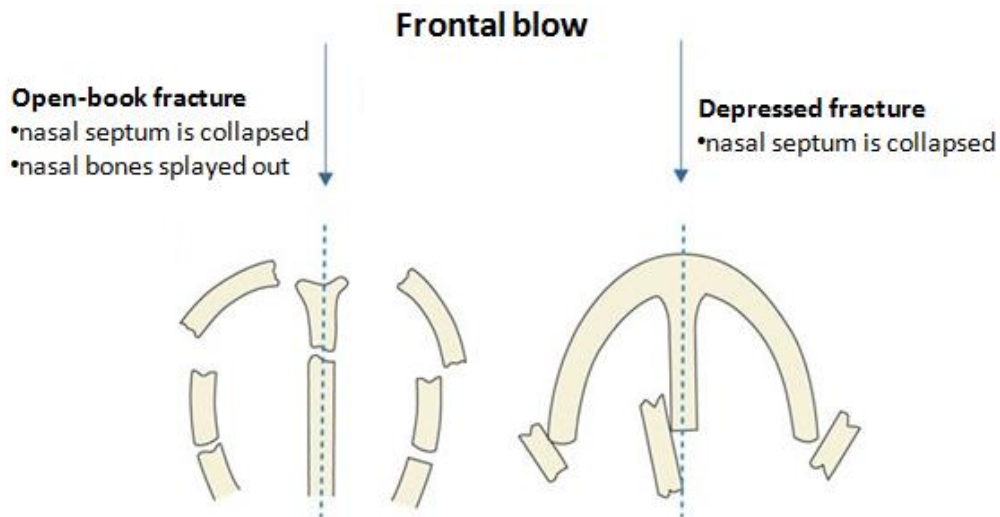
• **Nasal Bones and Septum:**

- Fractures of nasal bones are the **most common** facial bone fracture because of the projection of nose on the face.
- Traumatic forces may act from:
 - **Front** (anterior impacts): may result in tip fractures, flattened nasal dorsum, splayed nasal bones, and septal deformities.
 - **Side** (lateral impacts): may result in a depression of the lateral nasal bone, "C-" or "S-shaped" nasal dorsum deformities, medial maxillary wall fractures, and septal deformities (**Dislocated** quadrangular cartilage inferiorly or **buckled** to "C-shaped" deformity superiorly , or **fractured into several pieces** are common septal abnormalities from trauma)
- Magnitude of force will determine the depth of injury.
- **Dislocated or greenstick fractures** of septal more common in children (More flexible) usually have and have a **higher risk of septal hematomas**.
- **Fractures with/out comminution** of septal are more common in adults.

- **Types of Nasal Fractures**

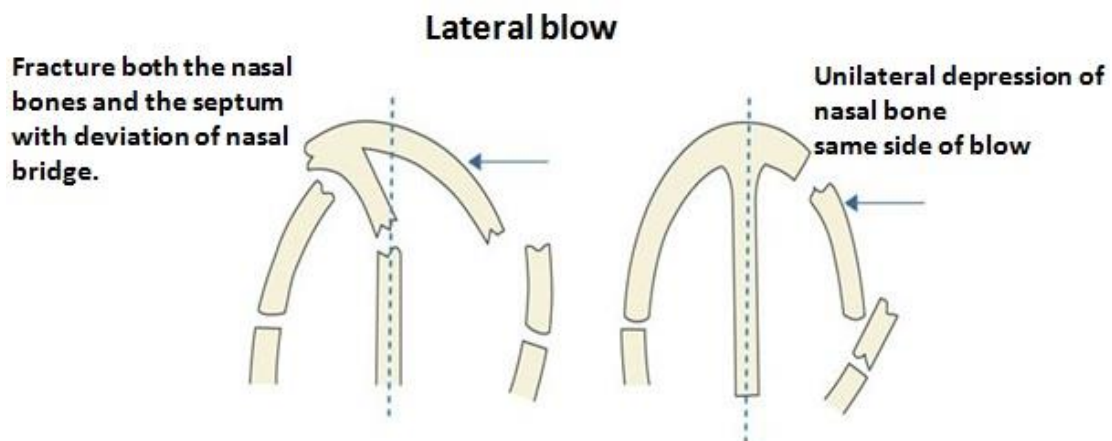
1. **Depressed:**

- They are due to frontal blow.
- Lower part of nasal bones which is thinner, easily gives way.
- A severe frontal blow will cause:-
 - **Open-book fracture.**
 - **Comminution** of nasal bones and even the frontal processes of maxillae with flattening and widening of nasal dorsum.



2. **Angulated:**

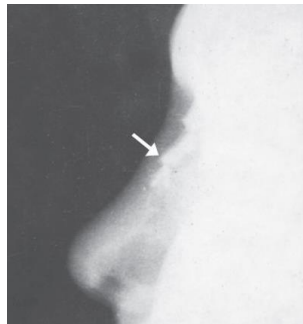
- Due to lateral blow.



Other Nasal fracture classification:

- I: Unilateral, simple
- II: Bilateral, simple
- III: Comminuted: Uni- ,, Bi- ,,Frontal
- IV a: Associated septal hematoma
- IV b: Open #
- V: N.O.E.

- **Clinical picture:**
 1. **Swelling of nose.** Appears within few hours and may obscure details of examination.
 2. **Periorbital ecchymosis.**
 3. **Tenderness.**
 4. **Nasal deformity.** Depressed from the front or side OR deviated to one side.
 5. **Crepitus and mobility of fractured fragments.**
 6. **Epistaxis.**
 7. **Nasal obstruction** due to septal injury or haematoma.
 8. **Lacerations** of nasal skin with exposure of nasal bones and cartilage seen in compound #.
- **Diagnosis:**
 - Physical examination.
 - X-rays:
 - May or may not show fracture.
 - Should include Waters' view, right and left lateral views and occlusal view.
 - CT :
 - May be considered if additional fractures are suspected.



- **Complications of Nasal Fractures:**
 - Medial canthal ligament injury,
 - Lacrimal duct injury,
 - Cribriform plate fracture (CSF leak, anosmia),
 - Septal hematoma may cause saddle nose deformity if untreated.
 - Persistent deformity.
 - Epistaxis

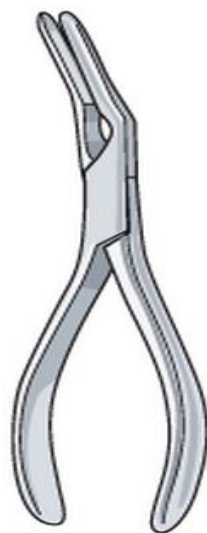
TABLE 71A.3  COMPLICATIONS NASAL FRACTURES	
Early, Temporary	Delayed
Edema	Airway obstruction
Ecchymosis	Fibrosis, contracture
Epistaxis	Secondary deformity
Hematoma	Synechiae
Infection	Saddle nose
Cerebrospinal fluid leak	Septal perforation

- **Treatment:**

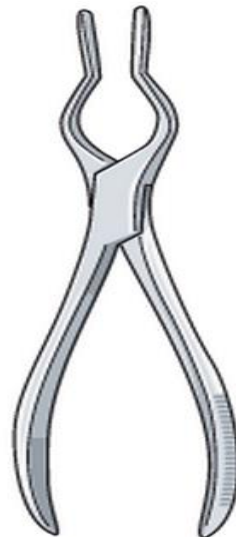
- Preoperative photography/x-ray may be considered for medicolegal documentation.
- Septal hematomas requires immediate evacuation of hematoma, nasal packing, and antibiotic prophylaxis.
- Open fractures must be cleaned then given antibiotics.
- Simple fractures without displacement need no treatment others may require closed or open reduction.

- **Closed Reduction:**

- Presence of edema interferes with accurate reduction by closed methods.
- Therefore, the best time to reduce a fracture is before the appearance of edema (**within 30 min - 3 hours**), or after it has subsided, which is usually in **5-7 days**.
- It is difficult to reduce a nasal fracture after **2 weeks** because it heals by that time.
- Pediatric Nasal Fractures is faster in healing: generally should be treated early & conservatively to avoid damage to growth centers
- Indicated as an initial trial for most nasal fractures.
- Should generally be avoided for cribriform plate fractures with CSF leaks (to avoid nasal packing).
- Anesthetize (general, or topical and injection); sedate as needed
- Reduce fracture before closing lacerations in open fractures



Asch septum forceps
use for disimpact &
reducing # of nasal
bone

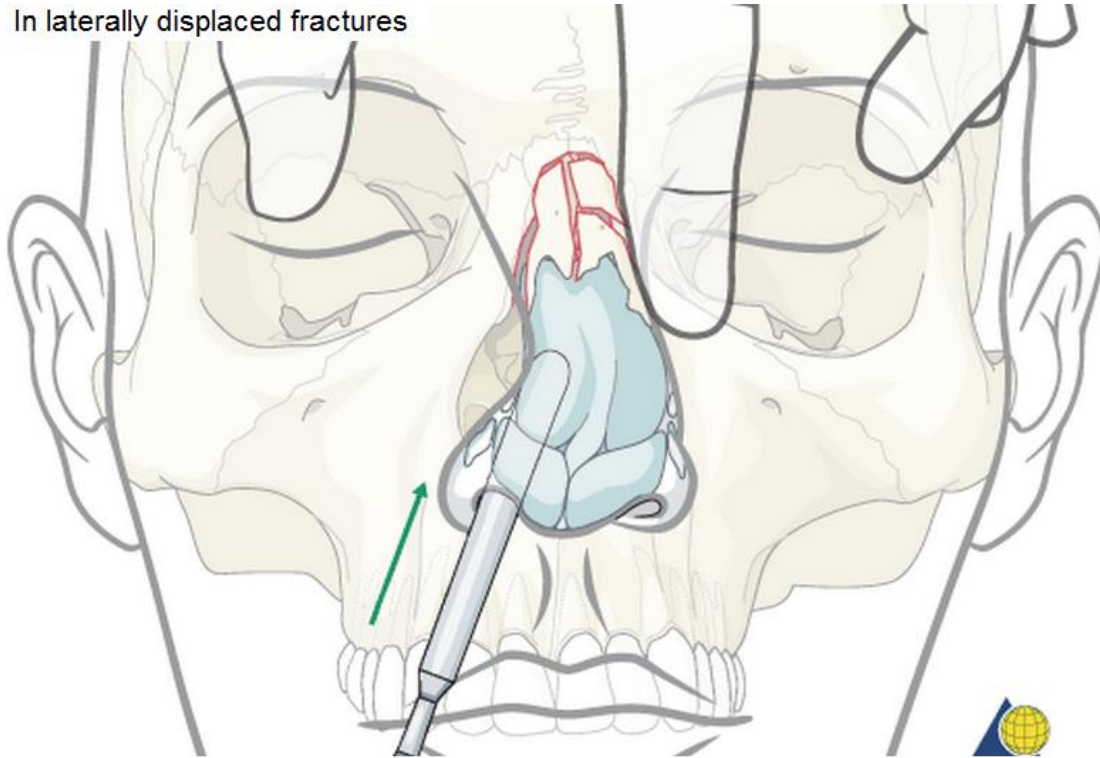


Walsham forceps
use for reducing
of nasal
septum

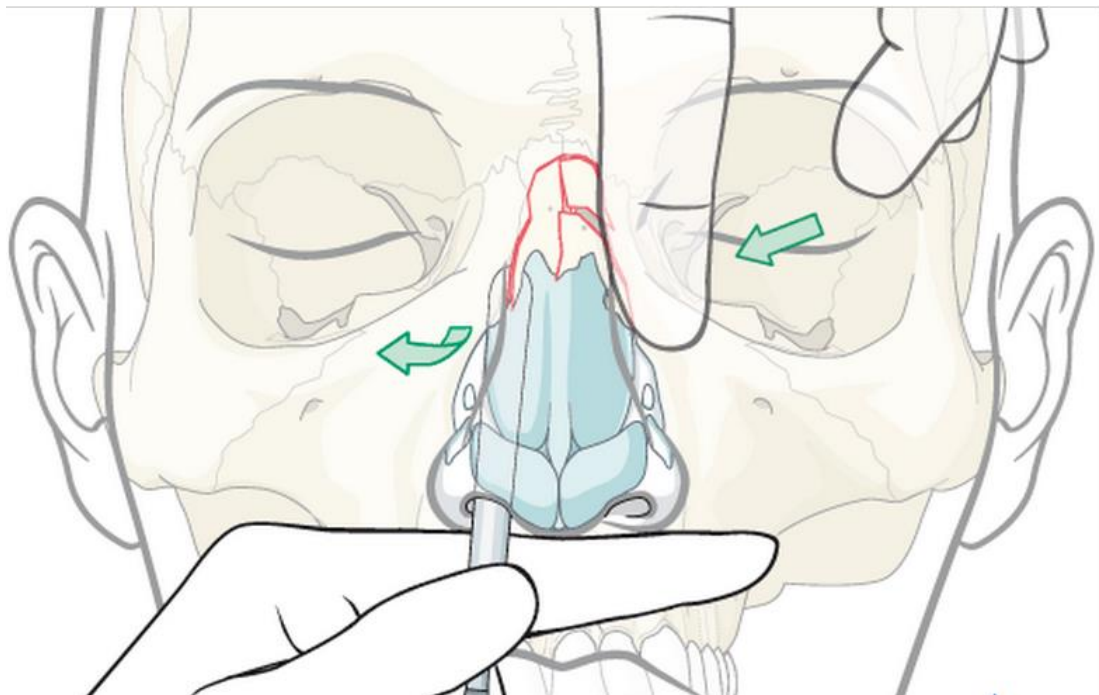


Boies nasal
fracture elevator

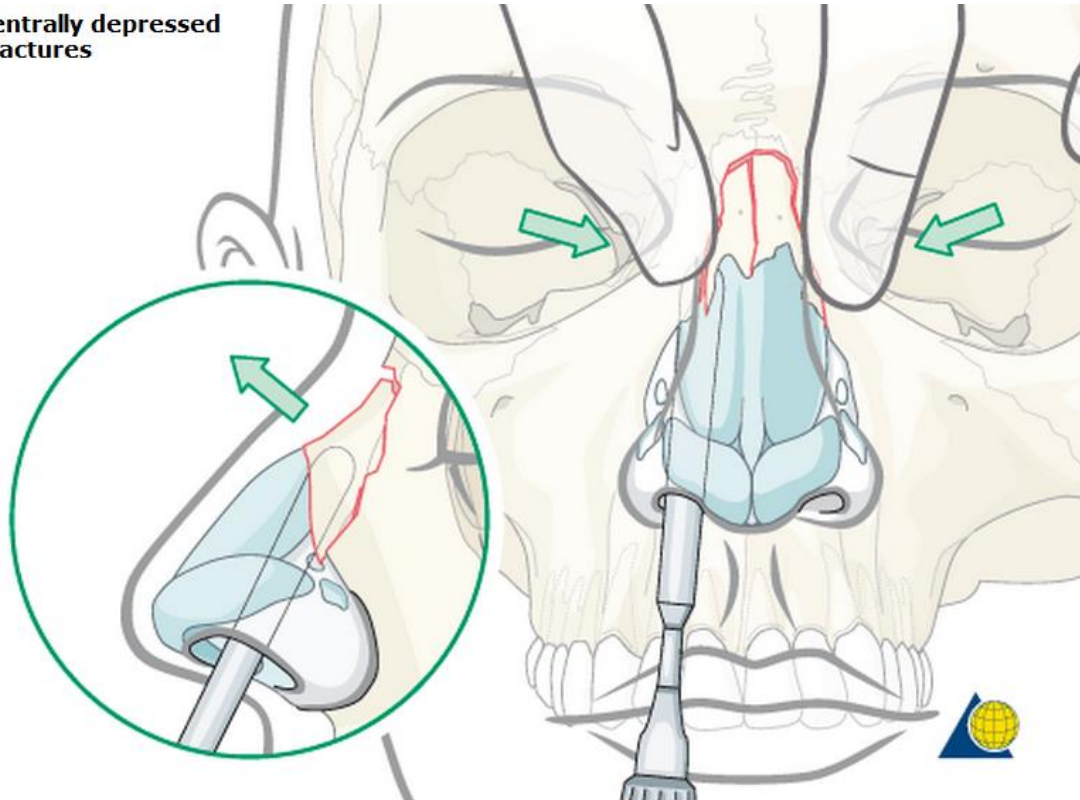
In laterally displaced fractures



- Place an instrument (eg, Boies elevator) in the **depressed side** along the lateral wall of the nose to a point 1 cm below the nasal frontal angle.
- Place a finger along the lateral side of the nose above the depressed area.; elevate instrument **ant/lat**; **medialize** contra bone if displaced laterally by finger



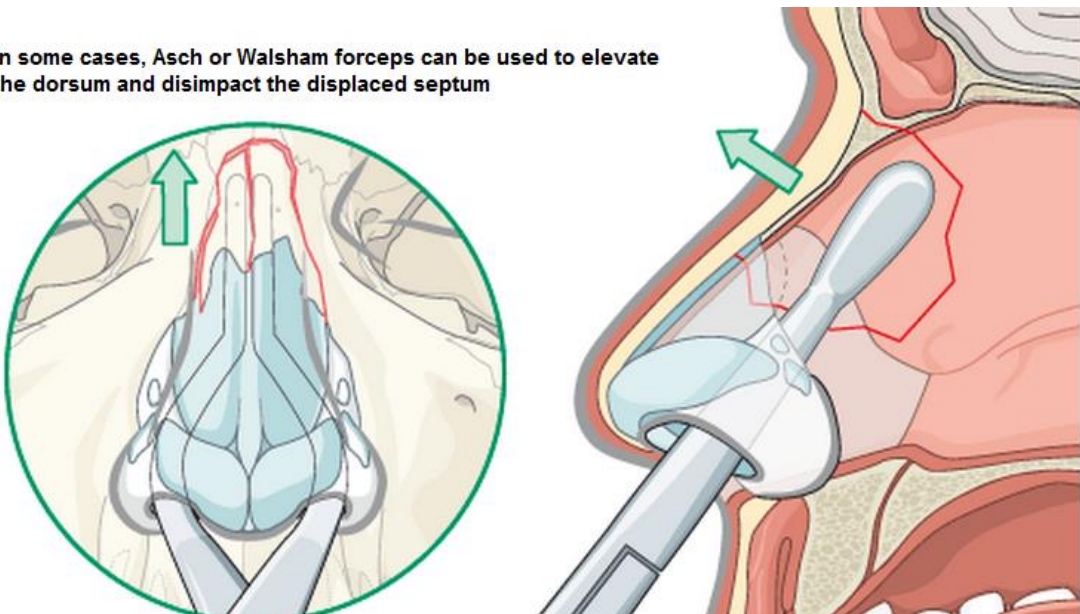
centrally depressed fractures



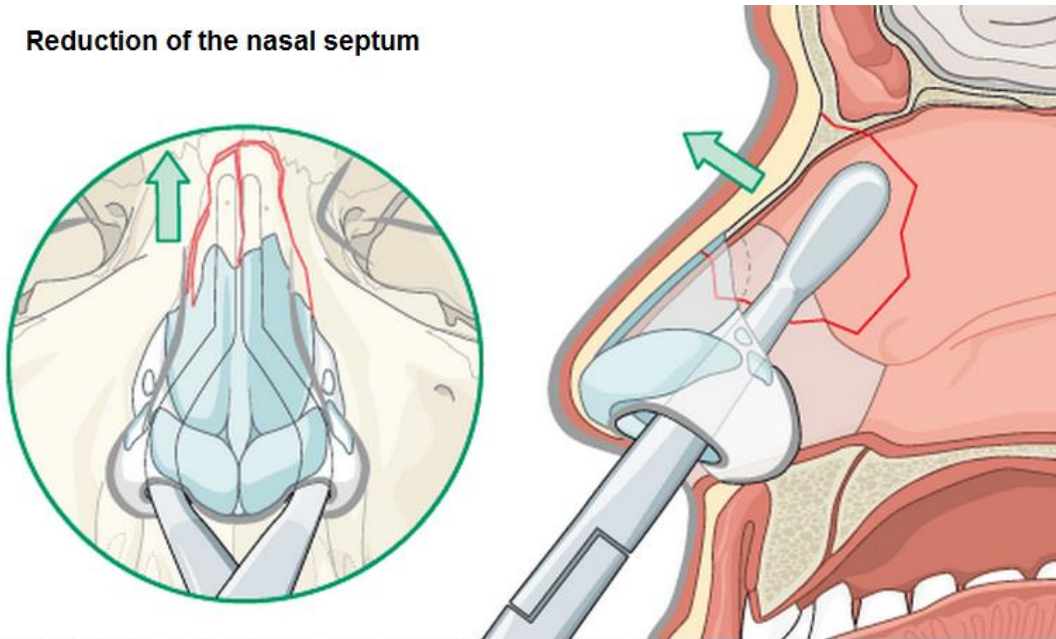
Sometimes the frontal processes of the maxillae are displaced laterally with the nasal bones impacted inside them.

The elevator must not be inserted too far into the nasal cavity and lifts the nasal dorsum **anteriorly**, while simultaneously the **thumb and index finger** put medial pressure on the displaced frontal processes of the maxillae **medially**.

In some cases, Asch or Walsham forceps can be used to elevate the dorsum and disimpact the displaced septum



Reduction of the nasal septum



The Asch or Walsham septum forceps are used to **straighten** the nasal septum.

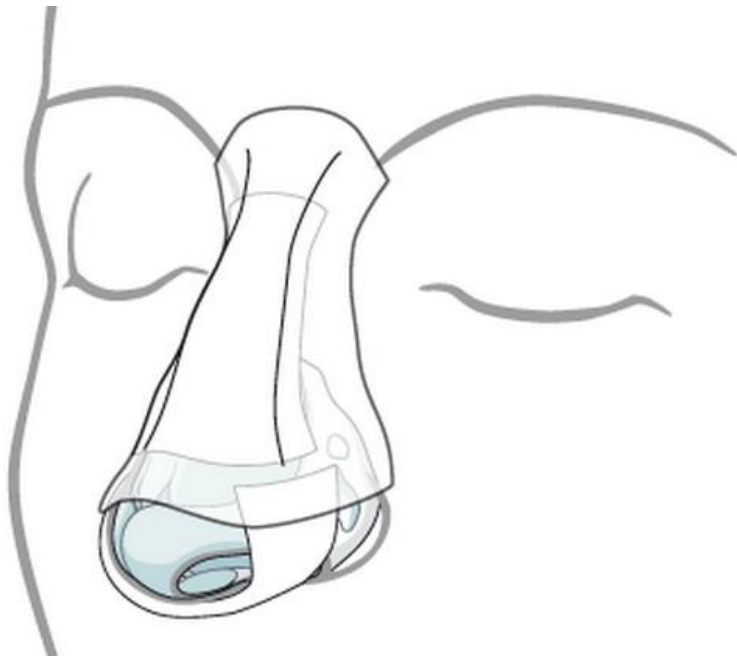
Grasp the nasal septum with the blades of the instrument and gently manipulate the septum into **proper alignment**

Centrally depressed fractures require posterior to anterior elevation.

Always splint dorsum

Simple fractures may not require intranasal packing.

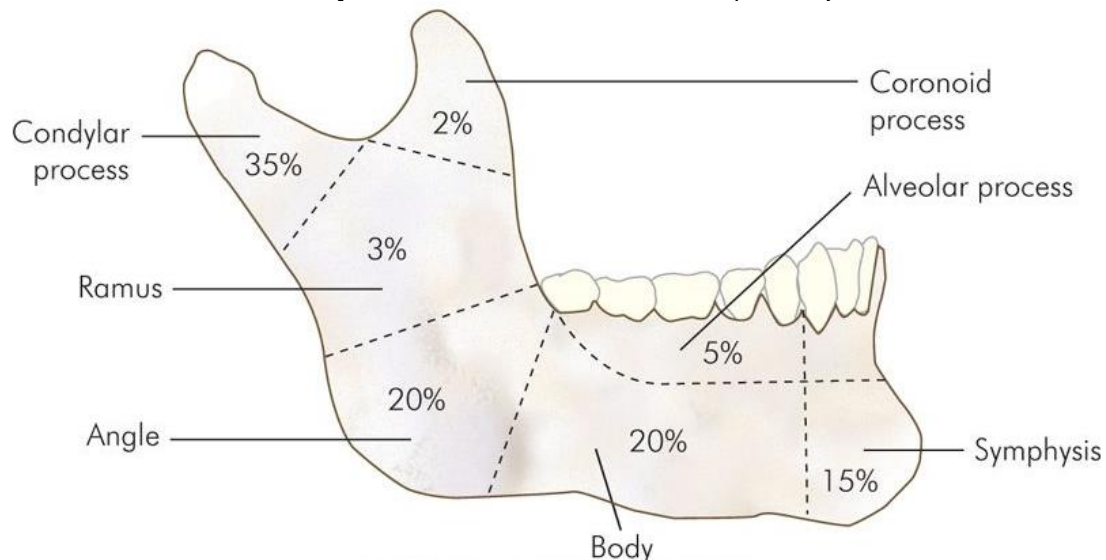
Unstable fractures (comminuted or loose) require intranasal packing and external splintage.



- **Indication for close reduction:**
 - ✓ **Depressed fractures of nasal bones** sustained by either frontal or lateral blow, can be reduced by a straight blunt elevator guided by digital manipulation from outside.
 - ✓ **Laterally displaced nasal bridge** can be reduced by firm digital pressure in the opposite direction.
 - ✓ **Impacted fragments** sometimes require disimpaction with Walsham or Asche's forceps before realignment.
 - ✓ **Septal fractures** are also reduced by Asche's forceps.
- **Open reduction (Septorhinoplasty):**
 - Early open reduction in nasal fractures is rarely required.
 - Indications of Open reduction:
 1. Extensive fracture/dislocation of the nasal bones and septum
 2. Nasal pyramid deviation > ½ the width of the nasal bridge
 3. Fracture/dislocation of the caudal septum
 4. Open septal fractures
 5. Persistent deformity after closed reduction
 6. Healed nasal deformities resulting from nasal trauma
 7. Pediatric/uncooperative patients (GA +/- open)
 - Surgical Complications and Associated Injuries:
 - **Persistent Deformity:** Failed closed reduction should be addressed with an open rhinoplasty approach after healing (3–6 months).
 - **Nasal Obstruction:** Due to synechiae, scar contracture, or structural deformity obstructing the nasal vault.
 - **Septal Hematoma.**
 - **Septal Perforations and Deviations:** May result from open septorhinoplasty techniques.
 - **Cribriform Plate Fracture:** May result in anosmia or CSF leak.
 - **Emphysema** of the face, neck.
- **Postoperative Care:**
 - Ice for 24 hours
 - Prophylactic antibiotics
 - Analgesics,
 - Nasal splints
 - Nasal packing

❖ **Fractures of Mandible**

- Fractures of mandible have been classified according to their location (mnemonic CABS are common).
 - Condylar # are the most common (weakest point).
 - Angle # (especially with an unerupted third molar).
 - Body #.
 - Symphysis #.
 - Fractures of the **ramus** (protected by pterygoid and masseter muscles), **coronoid**(protected by zygoma) and **alveolar processes** are uncommon, rarely fractured.

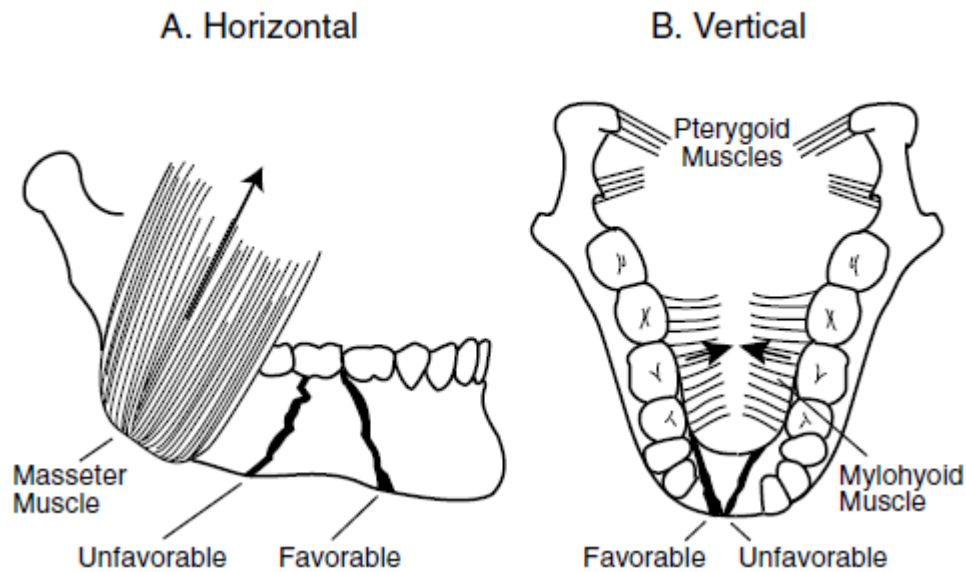


- Most of the mandibular fractures are the result of direct trauma
- Most common in young males (ages 18–30).
- Condylar fractures are caused by indirect trauma to the chin or opposite side of the body of mandible.
- Displacement of mandibular fractures is determined by:
 1. The pull of muscles attached to the fragments.
 2. Direction of fracture line.
 3. Bevel (oblique) of the fracture.
- **Risks:** Impacted teeth, osteoporosis, edentulous areas (lack teeth), pathologic lytic lesions.

○ **Classification:**

• **Classification by Favorability:**

- Favorability is determined by direction of fracture line and result of muscle force on the mandibular segments.
- **Favorable:** Vector forces from muscles pull fragments together.
- **Unfavorable:** Vector force muscles pulls fragments apart.
- **Horizontally Unfavorable:** Vector force of masseter and temporalis muscles pulls fragments apart (**angle fracture**).
- **Vertically Unfavorable:** Vector force of anterior muscles and pterygoid muscles pulls fragments apart (**body and symphyseal fractures**).



Anterior Muscles:

- ✓ Weaker force.
- ✓ Muscle action depresses and retracts (opens mandible)
- ✓ Mylohyoid, geniohyoid, genioglossus, platysma, anterior digastric muscles

Posterior Muscles:

- ✓ Stronger force
- ✓ **Temporalis Muscle**: raises and retracts mandible
- ✓ **Masseter Muscle**: raises and retracts mandible
- ✓ **Medial Pterygoid Muscle**: attaches to medial ramus, raises mandible
- ✓ **Lateral Pterygoid Muscle**: attaches to condylar neck, **protrudes** and depresses mandible

- **Classification by Type of Fracture:**

- Open versus Closed
- Fracture Pattern: comminuted, oblique, transverse, spiral, greenstick (incomplete fracture through only one cortical surface).
- Pathologic: secondary to bone disease (osteogenic tumors, osteoporosis).

- **Dental Classification:**

- ✓ **Class I**: teeth on both sides of fracture line, dentulous
- ✓ **Class II**: fracture has teeth on one side (partially dentulous)
- ✓ **Class III**: completely edentulous

- **Angle's Classification:**

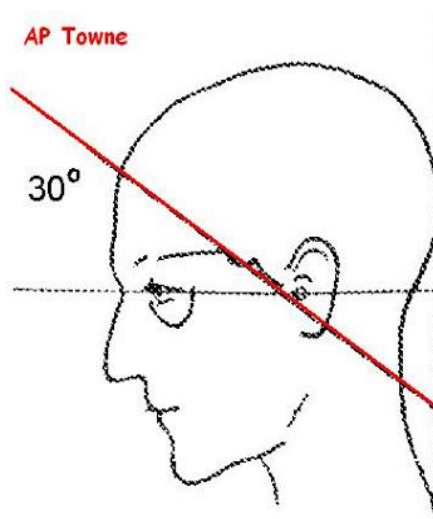
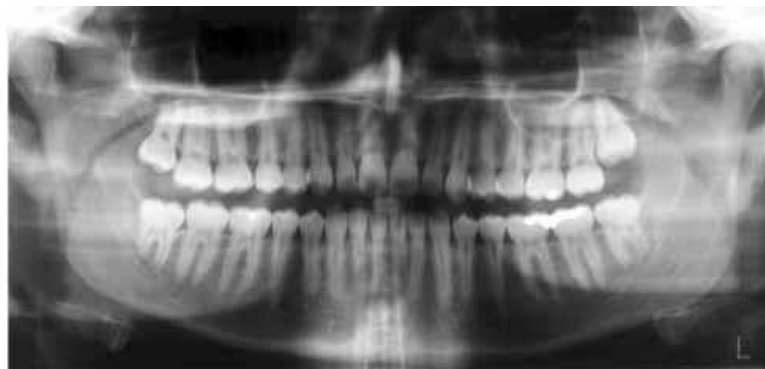
- ✓ **Class I**: normal bite, cusp of the maxillary first molar lines the buccal groove of mandibular first molar
- ✓ **Class II**: retrognathic (**over-bite**)
- ✓ **Class III**: orthognathic (**under-bite**)

Clinical Features

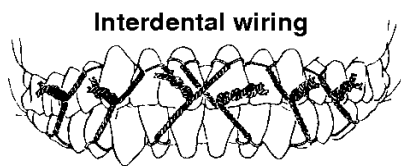
- **Fractures of condyle:**
 - If fragments are not displaced:
 - Pain, trismus and tenderness is elicited at the site of fracture.
 - If fragments are displaced:
 - Same as above in addition to malocclusion of teeth and deviation of jaw to the opposite side on opening the mouth.
- **Fractures of angle, body and symphysis:**
 - Most of the can be diagnosed by intraoral and extraoral palpation.
 - Step-deformity.
 - Malocclusion of teeth.
 - Ecchymosis of oral mucosa.
 - Tenderness at the site of fracture and crepitus may be seen.

Diagnosis

- **Plain X-ray Film Mandible Series and Panorex:**
 - Posterior Anterior Projection: views symphyseal region.
 - Lateral Oblique: views ramus, body, and angle.
 - Towne's: views condyle and coronoid process.
 - Panorex (panoramic): **most informative mandibular film**, requires patient to be able to sit erect, poor view of symphyseal region and the lingual surface of the mandible.



- **Treatment:**
- Both closed and open methods are used for reduction and fixation of the mandibular fractures.
- **Goals:** restore occlusion, establish bony union, return of function, avoid TMJ ankyloses.
- **Closed methods (Maxillo-Mandibular Fixation/MMF):**
 1. interdental wiring
 2. intermaxillary fixation.
 3. External pin fixation can also be used.
 - ✓ Requires an **intact maxilla**.
 - ✓ **Complications:** airway compromise, dental injury, weight loss, loss of fixation (malunion, nonunion), TMJ dysfunction, aspiration



- **Open methods (Open Reduction Internal Fixation/ORIF):**
- Fracture site is exposed and fragments fixed by direct **interosseous wiring** OR **compression plates**.
- With open method, prolonged immobilisation and intermaxillary fixation can be avoided.
- **Surgical Complications:**
 - **Chin and Lip Hypesthesia (most common):** inferior alveolar or mental nerve injury ; spontaneous regeneration occurs in the majority of cases.
 - **Osteomyelitis:** increased risk from **poor oral hygiene, devitalized teeth, infected teeth within fracture line**, improperly repaired fracture (nonunion).
 - **Malunion:** healing of bone in malalignment caused by inadequate immobilization, inaccurate reduction, infection, gross loss of bone, compromised blood supply, malnutrition, , or osteopetrosis.
 - **Nonunion:** failure of bone to produce osteogenic tissue, nonosteogenic matrix produced (**fibrous union**).
 - **Plate Exposure:** similar causes as malunion.
 - **Marginal Mandibular Nerve Injury:** increased risk with external approaches; Rx: direct anastomosis if noticed intraoperatively.
 - **Necrosis of Condylar Head (Aseptic Necrosis):** compromised vasculature to the condylar head with damage to the condylar neck; Rx: debridement of necrotic bone with reconstruction.

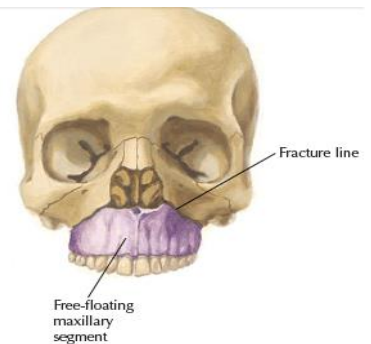
- **TMJ Ankylosis** (stiffness of a joint): defined as inability to open jaw beyond 5 mm between incisors, In children may cause facial deformities from growth disorders occurring at joint; **Rx:** if **fibrotic ankylosis** has occurred treat with passive jaw opening exercises, if **skeletal ankylosis** occurs must resect with reconstruction of the TMJ.
- **Dental Injury:** may be secondary to improper placement of arch bars and MMF.
- **Condylar fractures are treated by:-**
 - **Close method** by intermaxillary fixation with arch bars and rubber bands.
 - **Open reduction** and interosseous wiring may be required in adult edentulous (Lacking teeth) patients with bilateral condylar fractures or in fractures of children.
 - Immobilization of mandible beyond **three weeks**, in condylar fractures, can cause ankylosis of temporomandibular joints.
 - Therefore, intermaxillary wires are removed and jaw exercises started.
 - If occlusion is still disturbed, intermaxillary wires are reapplied for another week and the process repeated till the bite and jaw movements are normal.
- **Postoperative Care:**
 - Antibiotics (penicillin, **clindamycin (excrete into saliva)**),
 - Analgesics
 - Oral hygiene (hydrogen peroxide rinses),
 - Soft diet

❖ **MAXILLARY FRACTURES**

- **Causes:** assault, motor vehicle accidents, sports, falls, and gunshot wounds.
- **Matrix of the maxilla** absorbs energy with impact, thereby protecting the orbit, intracranial contents, and nose from total destruction.
- Severe maxillary fractures are associated with high incidence of intracranial and orbital injuries.
- **Sinusitis** is a potential complication and is generally avoided with prophylactic antibiotics and early removal of nasal packing; persistent sinusitis may require surgical intervention to address structural abnormalities.
- **Le Fort Classification** classified into 3 types.

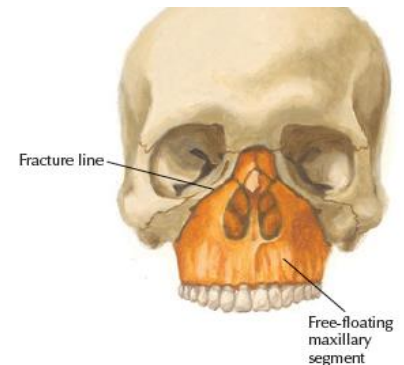
(1) Le Fort I (Transverse, low maxillary):

- Fracture runs above and parallel to the palate.
- It crosses **lower part of nasal septum, maxillary antra and the pterygoid plates** (upper alveolus becomes separated from upper maxilla).
- Typically caused by a low anterior-to-posterior force.



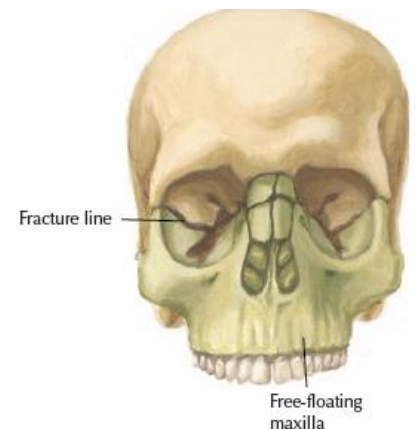
(2) Le Fort II (Pyramidal):

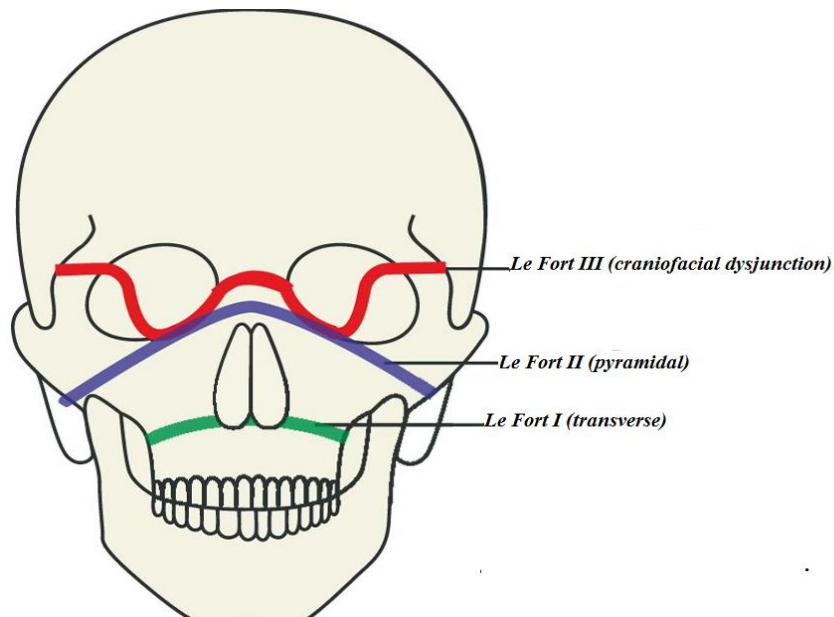
- Fracture passes through the **root of nose, lacrimal bone, floor & rim of orbit, upper part of maxillary sinus, lamina papyracea and pterygoid plates**.
- This fracture has some features common with the zygomatic fractures.
- Typically from a superiorly directed force against the maxilla or an anterior to posterior blow along the Frankfort plane (auriculo-orbital plane).



(3) Le Fort III (Craniofacial dysjunction):

- There is complete separation of facial bones from the cranial bones (base of skull).
- The fracture line passes through **root of nose, nasal septum, ethmoidal junction, superior orbital fissure, lateral wall of orbit, frontozygomatic and temporozygomatic sutures and the upper part of pterygoid plates**.
- Typically caused by high velocity impacts, motor vehicle accidents, oblique forces.

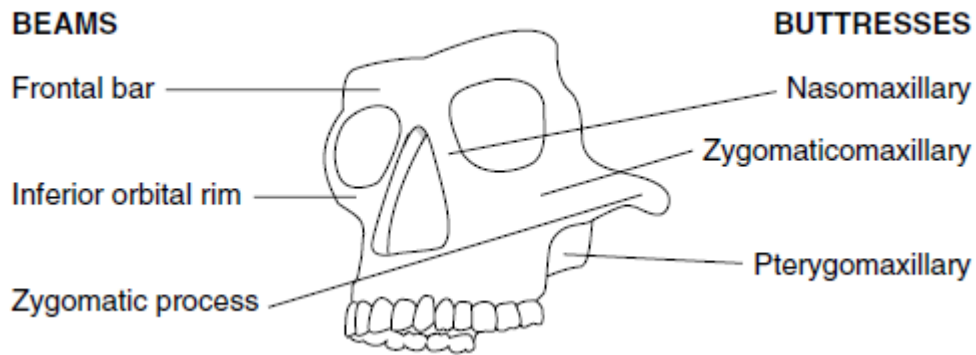




- Le Fort fractures may present in many combinations or on one side (hemi-Le Fort).

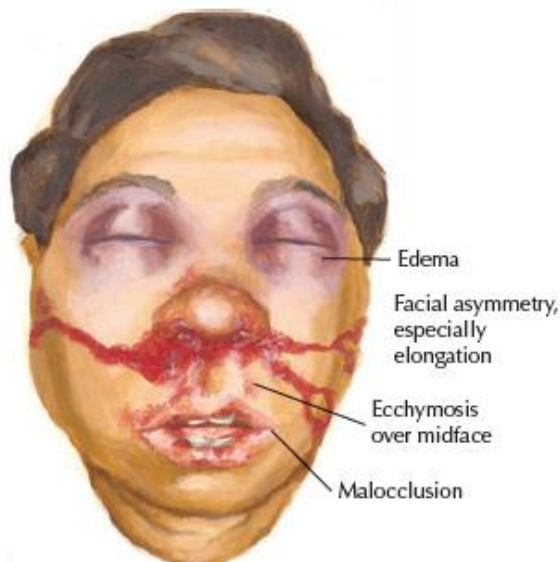
- **Buttress System**

- **Vertical Buttresses:** steady, developed to resist load from mastication.
 1. **Naso-Maxillary (NM)**
 2. **Zygomatico-Maxillary (ZM):** bears the strongest load, begins above
 3. maxillary first molars
 4. **Pterygo-Maxillary (PM):** projects into skull base
 5. **Nasal Septum:** midline support
- **Horizontal Buttresses (Beams):** weaker, reinforces vertical buttresses and provides width and projection of the face.
 1. **Frontal Bar:** essential support, made from superior orbital rim
 2. and glabellar area, suspends the naso-maxillary and zygomaticomaxillary
 3. struts
 4. **Inferior Orbital Rims**
 5. **Maxillary Alveolus and Palate**
 6. **Zygomatic Process**
 7. **Greater Wing of the Sphenoid**
 8. **Medial and Lateral Pterygoid Plates**
 9. **Mandible**

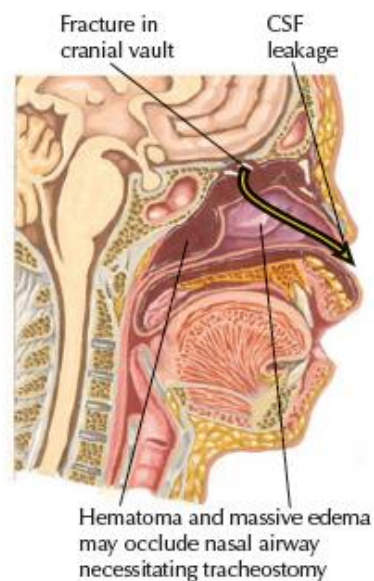


Clinical Features:

- Malocclusion of teeth with anterior open bite.
- Elongation of midface.
- Mobility in the maxilla.
- CSF rhinorrhoea. Cribriform plate is injured in Le Fort II and Le Fort III fractures.



Craniofacial dysjunction in Le Fort III fracture distorts facial symmetry



Diagnosis

- X-rays (Waters' view, posteroanterior view, lateral view)
- CT scan: Review of CT should include:
 - The buttress system
 - Zygomatic arches
 - Orbital volume and orbital contents

- **Treatment**

- Treatment of maxillary fractures is complex.
- ABC: Immediate restore the **airway** and stop severe **hemorrhage** from maxillary artery or its branches.
- The goal of treatment must be the exact anatomic restoration of the midfacial skeletal unit rather than approximate repositioning of its component parts.
- Most favorable operative time is within first 24–48 hours prior to significant facial edema.
- For good cosmetic and functional results, fractures should be treated as early as the patient's condition permits, it is necessary to prevent **soft tissue contraction**, scarring and bone resorption.
- Associated intracranial and cervical spine injuries may delay specific treatment.
- Direct fixation techniques can allow up to 2 weeks for delayed repair

Fixation of maxillary fractures can be achieved by:

- **Closed methods (Maxillo-Mandibular Fixation/MMF):**

1. Interdental wiring.
2. Intermaxillary wiring using arch bars.
 - Should be done first before the ORIF to ensure occlusion.
 - If the **mandible** zygomatic and **palatal** also fractured, it must first be stabilized.

- **Open methods (Open Reduction Internal Fixation/ORIF):**

1. Open reduction and interosseous wiring.
2. **Plate Fixation (Miniplates)**
3. Wire slings from frontal bone, zygoma or infraorbital rim to the teeth or arch bars.
4. **Bone Grafts:** indicated for significant bone loss

- **Management by Le Fort Classification**

- **Le Fort I:** generally may be reduced (digitally or with disimpaction forceps) and placed in MMF; may consider fixation of ZM buttress; edentulous patients may require splints, circummandibular wires, or circumzygomatic fixation.
- **Le Fort II:** stabilization of the ZM buttress with fixation essential after placing in MMF, may also consider fixation of the nasofrontal process or inferior orbital rim.
- **Le Fort III:** usually requires coronal flap for adequate exposure for exploration and miniplate fixation.

Postoperative Care:

- Antibiotics, analgesics, soft diet

Surgical Complications

- ✓ **Malunion** (Asymmetry)
- ✓ **Nonunion**
- ✓ **Plate Exposure:** address similar to mandible fractures (as above)
- ✓ **Forehead or Cheek Hypesthesia:** injury to supraorbital or infraorbital nerves; spontaneous regeneration occurs in the majority of cases
- ✓ **Osteomyelitis:** address similar to mandible fractures (as above)
- ✓ **Dental Injury:** may be secondary to reduction techniques or improper placement of arch bars and MMF

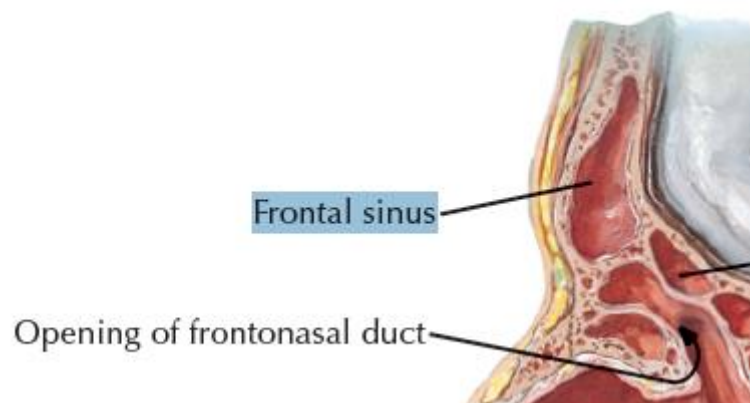
❖ FRACTURES OF UPPER THIRD OF FACE

- 1. Frontal Sinus**
- 2. Supraorbital Ridge**
- 3. Fractures of Frontal Bone**

○ Frontal sinus fractures:

- 1. Anterior wall**
- 2. Posterior wall**
- 3. Nasofrontal duct**

- **SSx:**
- Laceration of the forehead, forehead swelling (may be confused with a subgaleal hematoma), palpable frontal defect, frontal pain,
- Frontal sinus fractures are rare in children (frontal sinus does not appear until age 5–6 years old).
- Frontal sinus anterior wall is thick, posterior wall and floor are thin.
- **The frontal sinus is absent in 4% to 10% of adults**
- **Forehead Hypesthesia:** from injury to the supraorbital nerves.
- **Forehead Paralysis:** from injury of the frontal branch of the facial nerve, return of function may occur.



- **Anterior frontal wall fractures:**

- Defect is mainly cosmetic.
- Linear, Depressed or comminuted.
- Sinus is approached through a wound in the skin if that is present, or through a brow incision.
- The bone fragments are elevated,
- The interior of the sinus is always inspected to rule out fracture of the posterior wall, may place interosseous wire.
- **If <1 cm** of total frontal bone fragments are missing may consider **skin** covering only.
- **If >1 cm** fragments are missing may use a split calvarial bone graft or mesh (eg,titanium) to reconstruct.

- **Posterior frontal wall fractures:**

- Isolated Nondisplaced Posterior wall Fracture: **rare**.
- Conservative (observation) & ABx.
- If sinus fluid does not clear within 6 weeks may consider exploration.
- May be accompanied by:
 - Dural tears (can be covered by temporalis fascia)
 - Brain injury
 - CSF rhinorrhoea.
 - Pneumocephalus (raises the potential for meningitis)
- They may require neurosurgical consultation.
- Small sinuses can be obliterated with fat.

- **Injury to nasofrontal duct:**

- Cause obstruction to sinus drainage.
- May later be complicated by a mucocele (accumulation of mucus), chronic sinusitis.
- In such cases, make a large communication between the sinus and the nose.
- Small sinuses can be obliterated with fat after removing the sinus mucosa completely.

TABLE 71B.1  **COMPLICATIONS**
FRONTAL SINUS FRACTURES

Frontal sinus
Headache, fullness
Sinusitis
Mucocoele

Intracranial
Cerebrospinal fluid leakage
Meningitis
Brain abscess
Seizures

Cosmetic
Scar
Numbness
Wound infection
Forehead depression

Ophthalmic
Diplopia
Eye pulsations

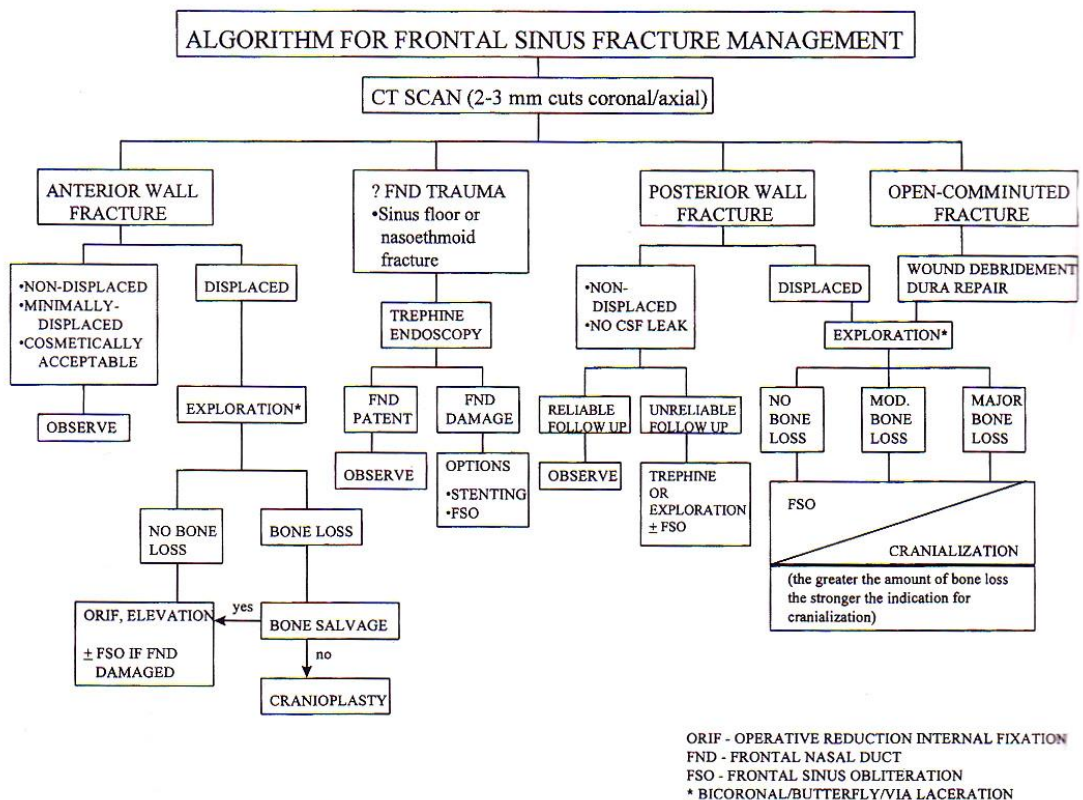


Figure 71B.1 Algorithm for management of fractures of the frontal sinus.

- **Supraorbital Ridge**

- ✓ Ridge fractures often cause:
 - Periorbital ecchymosis
 - Flattening of the eyebrow
 - Proptosis or downward displacement of eye.
- ✓ Fragment of bone may also be pushed into the orbit and get impacted.
- ✓ Ridge fractures require **open reduction** through an incision in the brow or transverse skin line of the forehead.

- **Fractures of Frontal Bone :**

- ✓ Depressed or linear.
- ✓ With or without separation.
- ✓ They often extend into the orbit.
- ✓ Brain injury and cerebral edema are commonly associated with each other and require neurosurgical consultation.

❖ **FRACTURES OF MIDDLE THIRD OF FACE**

1. Nasal Bones and Septum
2. Naso-orbital Fractures
3. Fractures of Zygoma (Tripod Fracture)
4. Fractures of Zygomatic Arch
5. Fractures of Orbital Floor
6. Fractures of Maxilla

Naso-orbital Fractures (Naso-orbitoethmoid (NOE) Fractures):

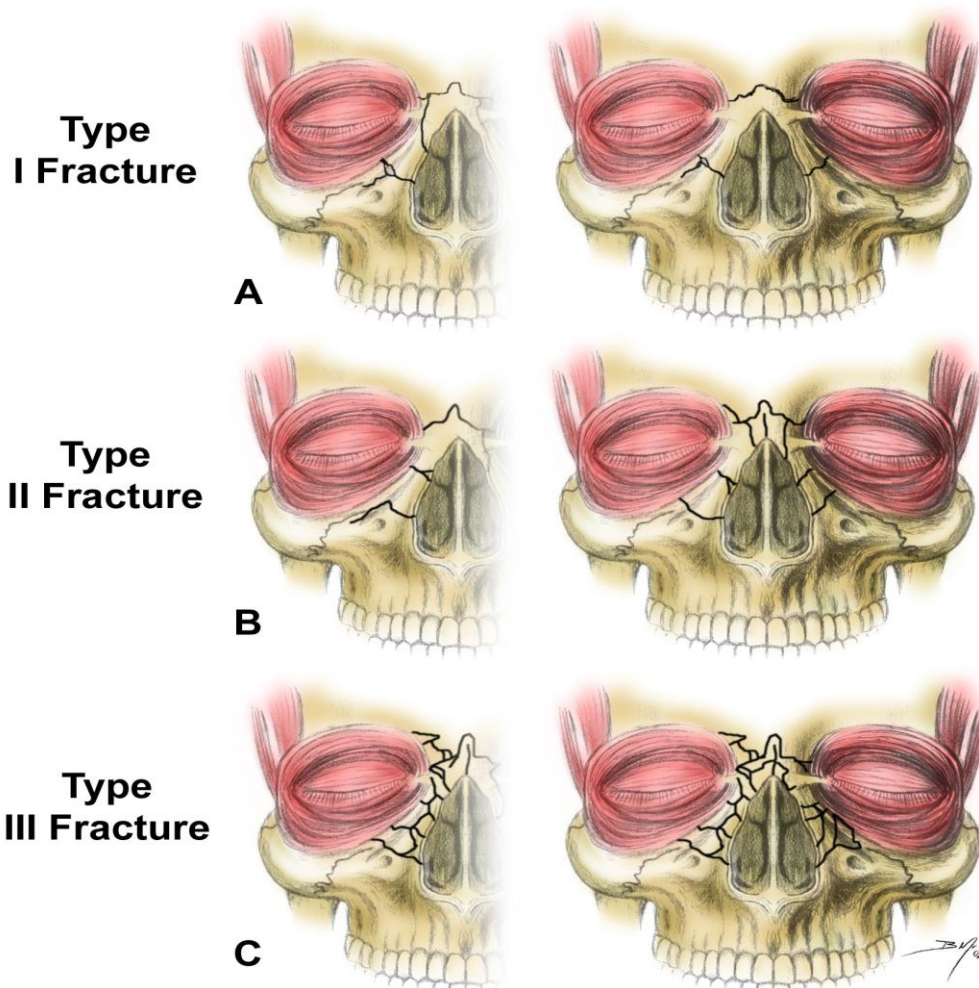
- Direct force over the **nasion** fractures nasal bones and displaces them posteriorly.
- **Perpendicular plate of ethmoid, ethmoidal air cells and medial orbital wall** are fractured and driven posteriorly.
- Injury may involve **cribriform plate, frontal sinus, frontonasal duct, extraocular muscles, eyeball and the lacrimal apparatus.**
- **Medial canthal ligament** (extension of the tarsal plates which attaches to the medial orbital wall) may be avulsed.

- **Clinical Features**

- **Telecanthus** (increased distance between the medial canthi of the eyelids), due to lateral displacement of medial orbital wall.
- **Pug nose.** Bridge of nose is depressed and tip turned up.
- **Periorbital ecchymosis.**
- **Orbital haematoma** due to bleeding from anterior and posterior ethmoidal arteries.
- **CSF leakage** due to fracture of cribriform plate and dura.
- **Displacement of eyeball.**
- **Bowstring sign** tests the integrity of the medial canthal ligament, grasp medial eyelid near lash line and pull laterally, normally should snap back

○

- **Diagnosis:**
- **CT scan** is more useful.
- **Fracture classification:**
- Each fracture type is sub-classified as either **unilateral** or **bilateral**
- **Type I fractures** represent a single non-comminuted central fragment without medial canthal tendon disruption
- **Type II fractures** involve comminution of the central fragment, but the medial canthal tendon remains firmly attached to a definable segment of bone
- **Type III fractures** are uncommon and result in severe central fragment comminution with disruption of the medial canthal tendon insertion



- **Treatment:**

- **Closed reduction:**

- In uncomplicated cases.
- Fracture is reduced with **Asche's forceps**.
- Stabilized by a wire passed through fractured bony fragments and septum and then tied over the lead plates.
- Intranasal packing is given.
- Splinting is kept for 10 days

- **Open reduction**

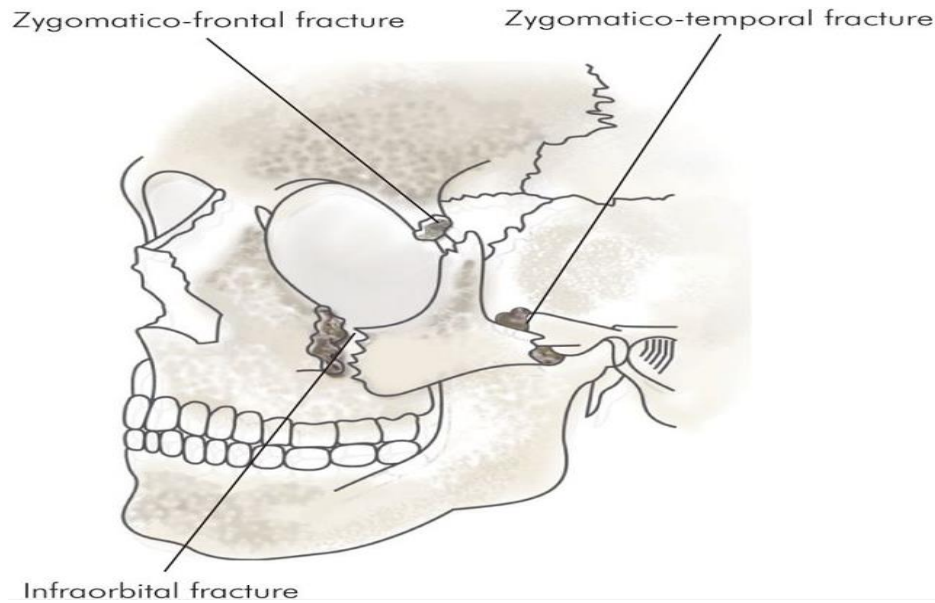
- This is required in cases with **extensive comminution** of **nasal and orbital bones, and those complicated by other injuries to lacrimal apparatus, medial canthal ligaments, frontal sinus.**
- **H-type incision** gives adequate exposure of the fractured area.
- Nasal bones are reduced under vision and bridge height is achieved.
- Medial orbital walls can be reduced.
- Medial canthal ligaments, if avulsed, are restored with a through and through wire.
- Intranasal packing may be required to restore the contour.
- When bone comminution is severe: Restoration of **medial canthal ligaments** and **lacrimal apparatus** should receive **preference** over reconstruction of nasal contour.

Fractures of Zygoma (Tripod Fracture)

- After nasal bones, zygoma is the **second most** frequently fractured bone.
- Direct trauma most common cause.
- Lower segment of zygoma is pushed medially and posteriorly resulting zygoma is separated at its three processes.
- Fracture line passes through zygomaticofrontal suture, orbital floor, infraorbital margin and foramen, anterior wall of maxillary sinus and the zygomaticotemporal suture.

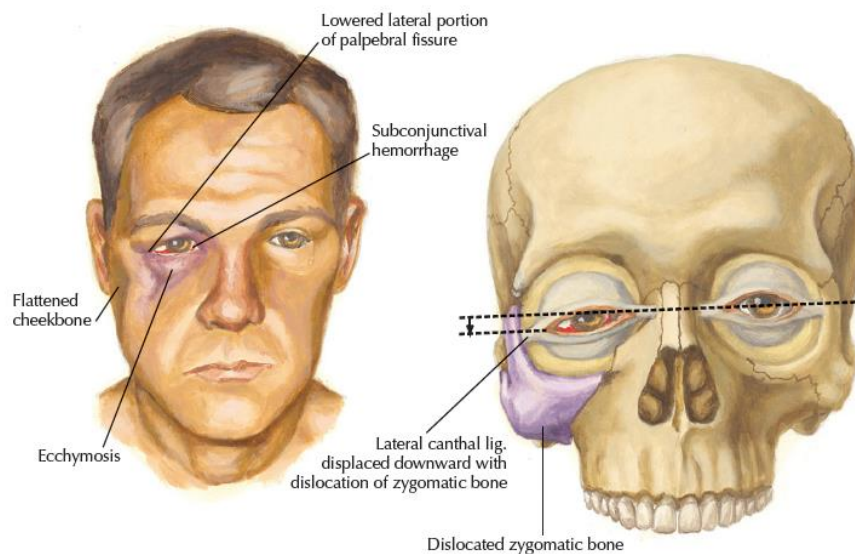
- **Four Sutures Involved in Zygomaticomaxillary Complex Fractures:**

- 1. Zygomaticofrontal Suture:** usually fractures cleanly
 - 2. Zygomaticomaxillary Suture:** at the face of the maxilla, high risk of comminution
 - 3. Zygomaticotemporal Suture:** zygomatic arch, typically fractures at midpoint or produces a double fracture
 - 4. Zygomaticosphenoid:** broad suture line
- Orbital contents may herniate into the maxillary sinus.



Clinical Features:

1. **Flattening** of malar prominence.
2. **Step-deformity** of infraorbital margin.
3. **Anaesthesia** in the distribution of **infraorbital nerve**.
4. **Trismus**, due to depression of zygoma on the underlying coronoid process.
5. **Oblique palpebral fissure**, due to the **displacement of lateral palpebral ligament**.
6. **Restricted ocular movements**, due to entrapment of **inferior rectus muscle**. It may cause **diplopia**.
7. **Periorbital emphysema**, due to escape of air from the maxillary sinus on nose-blowing.
8. **Subconjunctival, periorbital ecchymosis, eyelid edema and epistaxis.**

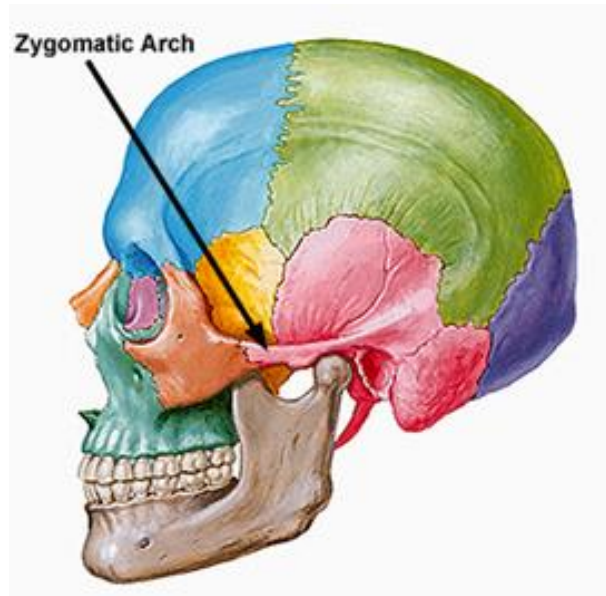


- **Diagnosis:**
- ✓ **X-rays**
 - Waters' view shows the fracture and displacement the best.
 - Maxillary sinus may show clouding due to the presence of blood.
 - Comminution with depression of orbital floor and herniation of orbital contents cannot be seen on plain X-rays.
- ✓ **CT scan:**
 - Orbital CT will be more useful.
- **Treatment:**
 - Only displaced fractures require treatment.
 - **Open reduction** and internal wire fixation gives **best** results.
 - Stabilization of the zygomaticomaxillary complex requires a minimum of 2-point fixation (usually ZF and infraorbital rim).
 - Fracture is exposed at the **frontozygomatic suture** through **lateral brow incision** and reduced by passing an elevator behind the zygoma.
 - Wire fixation is done at **frontozygomatic suture** and **infraorbital margin** ([exposed by a separate incision in the lower lid](#)).
 - Fracture of orbital floor can also be repaired through this incision ([lower lid](#)).
- ✓ **Transantral approach is less favorable:**
 - Antrum is exposed as in **Caldwell-Luc operation**.
 - Blood is aspirated.
 - Fracture reduced then stabilized by a **pack** in the antrum.
 - Fractures of orbital floor can also be reduced.
 - Antral pack is removed in about 10 days through the buccal incision, which is left open at the end of operation, **or** through the intranasal antrostomy route.



Fractures of Zygomatic Arch:

- Zygomatic arch generally breaks into two fragments which get depressed.
- There are **three fracture lines** (one at each end and third in the centre of the arch).



Clinical Features

- Depression in the area of zygomatic arch.
- Local pain aggravated by talking and chewing.
- Trismus or limitation of the movements of mandible due to impingement of fragments on the condyle or coronoid process.

Diagnosis:

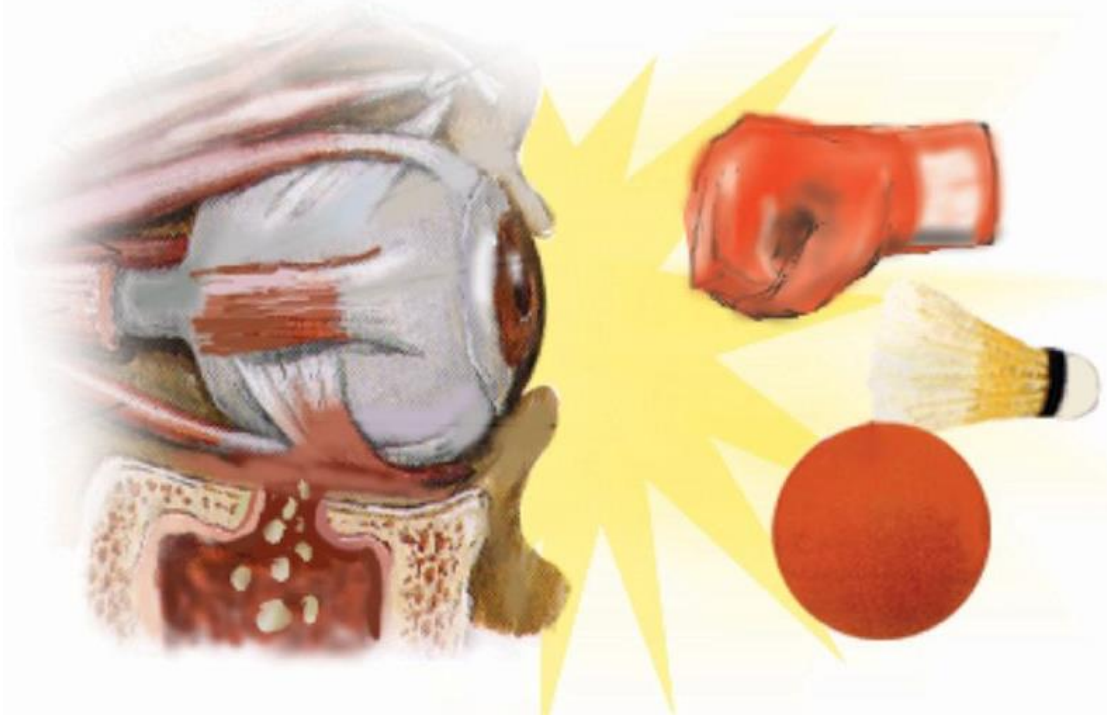
- ✓ **X-ray:** Arch fractures are best seen on submentovertical view of the skull. Waters' view is also taken.
- ✓ **CT Scan.**

Treatment:

- ✓ **Vertical incision** is made in the hair-bearing area above or in front of the ear.
- ✓ Cutting through temporal fascia.
- ✓ Elevator is passed deep to temporal fascia and carried under the depressed bony fragments which are then reduced.
- ✓ Fixation is usually not required as the fragments remain stable.

Fractures of Orbital Floor

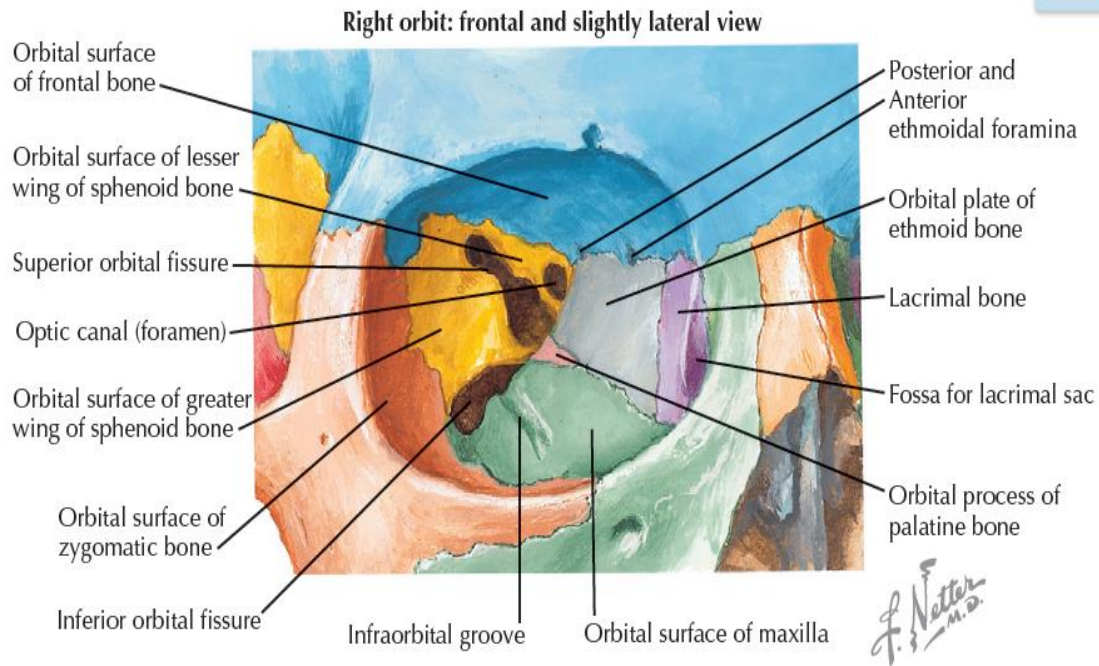
- Zygomatic and **Le Fort II** maxillary fractures are always accompanied by fractures of orbital floor.
- Isolated fractures of orbital floor, when a **large blunt object strikes the globes**, are called "**blow out fractures**"
- Orbital contents may herniate into the antrum



- **Seven Orbital Bones:**

1. Frontal,
2. Lacrimal
3. Ethmoid (lamina papyracea weakest portion)
4. Maxilla,
5. Zygoma,
6. Sphenoid
7. Palatine bones

- ✓ **Optic Canal Contents:** optic nerve (CN II), ophthalmic artery
- ✓ **Superior Orbital Fissure Contents:** trochlear nerve (CN IV), lacrimal and frontal divisions of V1, **supraorbital vein**, oculomotor and abducens (CN III and CN VI), nasociliary division of V1 (3.4.5.6)
- ✓ **Inferior Orbital Fissure Contents:** zygomaticofacial and zygomaticotemporal divisions of V2, **inferior ophthalmic vein**



- **Clinical Features:**

1. **Ecchymosis** of lid, conjunctiva and sclera.
2. **Enophthalmos** with inferior displacement of the eyeball. This becomes apparent when oedema subsides.
3. **Diplopia**, which may be due to **displacement of the eyeball or entrapment of inferior rectus and inferior oblique muscles**.
4. **Hypoaesthesia** or anaesthesia of cheek and upper lip (**infraorbital nerve**)

- **Theories of Orbital Floor Injury**

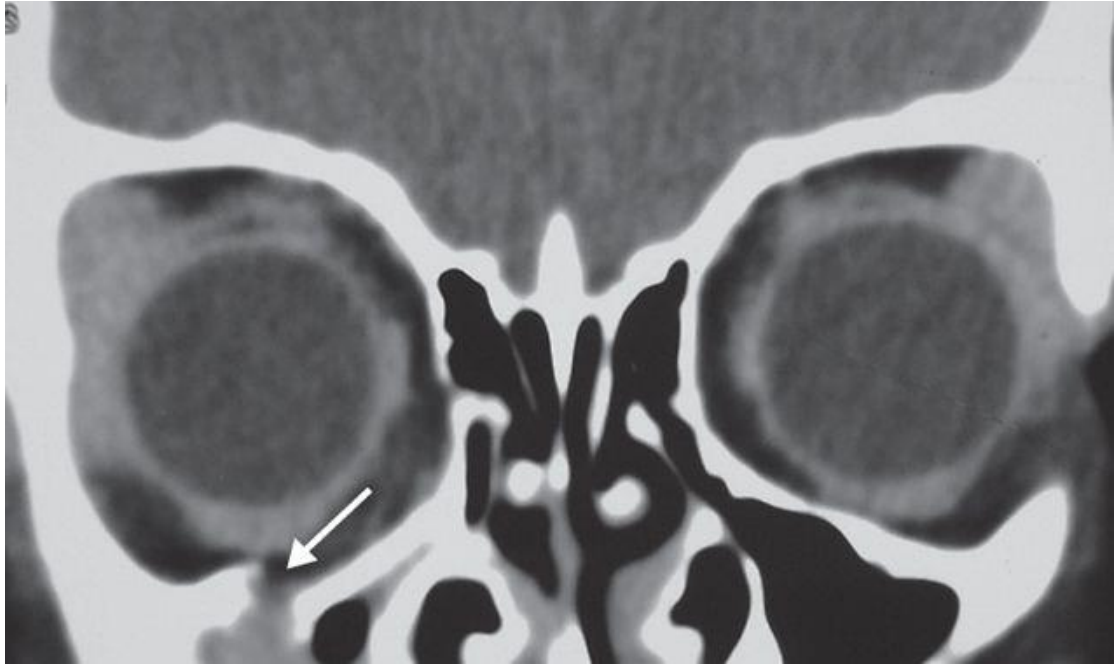
- **Hydraulic Theory:** force to orbital region → intraocular pressure → fractures floor.
- **Buckling Theory:** force on inferior rim → directly fractures floor.

- **Types:**

1. **Pure:** involves the central area of a wall or floor only
2. **Impure:** extension of the fracture to include an orbital rim
3. **Junctional:** involves floor and medial wall

- **Diagnosis:**

- **X-ray** :Waters' view shows a convex opacity bulging into the antrum from above (tear-drop opacity).
- CT scans may confirm the diagnosis.
- Entrapment of **inferior rectus and inferior oblique muscles** is diagnosed by asking the patient to **look up and down**, or by the **traction test** (performed by grasping the globe and passively rotating it to check for restriction of its movements.)



Treatment

- Indications for surgery include:-
 - Enophthalmos
 - Persistent diplopia due to entrapment of muscle.
- Orbital floor fractures can be satisfactorily reduced by:-
 - **Trans-antral approach**: finger passed into the antrum, pack can be kept in the antrum to support the fragments.
 - **Infra-orbital approach**: through a skin crease of the lower lid, can also be used either alone or in combination with transantral approach.
- ✓ Badly comminuted fractures of orbital floor can be repaired by a **bone graft** from the **iliac crest, nasal septum or the anterior wall of the antrum**.
- ✓ Silicon or teflon sheets have also been used to reconstruct the orbital floor but autogenous grafts are preferable.

Granulomatous and Systemic Diseases Affecting the Nose:

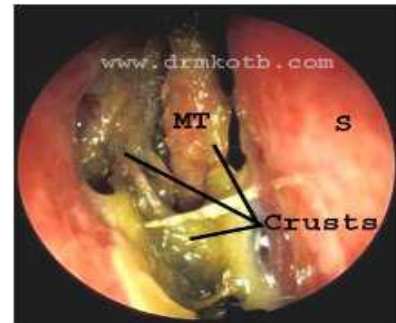
GRANULOMATOUS DISEASES	SYSTEMIC DISEASES
1- Infectious ○ Bacterial ○ Rhinoscleroma (Klebsiella) ○ Syphilis (Treponema pallidum) ○ TB ○ Lupus Vulgaris ○ Leprosy ○ Fungal ○ Rhinosporidiosis ○ Sporotrichosis ○ Blastomycosis (dermatiditis) ○ Coccidiomycosis (immitis) 2- Autoimmune/vasculitis ○ Wegeners Granulomatous ○ Churg-Strauss syndrome ○ Relapsing Polychondritis ○ Systemic lupus Erythematosus ○ Sjogrens Syndrome 3- Idiopathic ○ Sarcoidosis 4- Neoplastic ○ T-Cell lymphoma ○ Pyogenic granuloma (Lobular capillary hemangioma) 5- Foreign Body Reactions ○ Cocaine-induced midline granuloma	1- IMMUNODEFICIENCY DISEASES ○ SINUSITIS IN THE IMMUNOCOMPROMISED PATIENT ○ AIDS AND THE NASAL AIRWAY 2- CUTANEOUS DISEASES ○ PEMPHIGUS VULGARIS ○ PEMPHIGOID ○ SCLERODERMA ○ BEHÇET DISEASE 3- MUCOCILIARY DISEASES ○ PRIMARY CILIARY DYSKINESIA ○ CYSTIC FIBROSIS 4- HEMATOLOGIC DISEASES ○ HEREDITARY HEMORRHAGIC TELANGIECTASIA

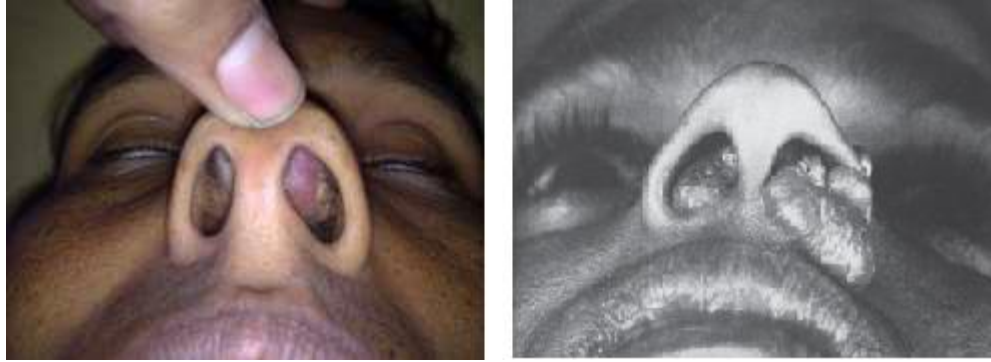
- Various granulomatous lesions involving the nose.
- They are the result of **bacterial** or **fungal** infections, **Autoimmune** or due to **causes not yet clear "idiopathic"**.
- Many of these lesions may be manifestations of **systemic disease**
- Biopsy of the lesion is also **essential**, not only to establish the correct diagnosis of granulomatous disease but also to exclude a **neoplasm**, which many of these diseases may clinically simulate.

❖ **Bacterial Infectious**

➤ **Rhinoscleroma (Klebsiella)**

- ✓ It is **noncontagious** a chronic granulomatous disease caused by Gram-negative bacillus called [Klebsiella rhinoscleromatis \(Frisch bacillus\)](#).
- ✓ Sometimes called as "Balkan leprosy"
- ✓ **Pathology**
 - The disease **starts in the nose** and extends to
 - **Nasopharynx**
 - **Oropharynx**
 - **Larynx (mostly subglottic region)**
 - **Trachea and bronchi.**
 - Mode of infection is Air-borne transmission.
 - Both sexes of any age may be affected.
 - Endemic in central and South America, Central Europe, East Africa, and the Indian subcontinent and is highly associated with poor hygiene and overcrowded conditions
- ✓ **Clinical features**
 - The disease runs through the following **3 stages** (stages may last years):
 - **(a) Atrophic stage (Catarrhal)**
 - It resembles **atrophic rhinitis** and is characterized by
 1. Foul smelling
 2. Purulent nasal discharge for weeks/months
 3. Honeycomb color crusting
 - **(b) Granulomatous stage (Proliferative)-Mikulicz cells**
 - Granulomatous nodules form in nasal mucosa ,,adherent to Nasal septum
 - Subdermal infiltration of lower part of external nose and upper lip.
 - Nodules are painless , friable and non-ulcerative → Epistaxis and obstruction
 - Cartilage destruction
 - Larynx: glottic, subglottic granulomas
 - **(c) Fibrosis stage (Cicatricial ,,Sclerotic).**
 - Lesion heals with extensive scarring (dense fibrotic)
 - This causes stenosis of nares, up to complete stenosis
 - Distortion of upper lip,
 - Adhesions in the nose extend to nasopharynx and oropharynx & may be subglottic stenosis with respiratory distress.





✓ **Diagnosis**

✓ **Biopsy**

- **Histopathology:**
 - **Mikulicz's cell** (large Foamy histiocytes (macrophage) with numerous vacuoles containing viable or non-viable causative bacteria),
 - **Russell bodies** (eosinophilic structures within the cytoplasm of bloated plasma cells.)
 - **Pseudoepitheliomatous hyperplasia**
- **Cultured**
 - The causative organisms from the biopsy material.
 - **MacConkey agar**
 - Only 50% of lesions will be culture positive

✓ **Treatment**

- **Abx:-**
 - 3/12 of ABx (Streptomycin plus Tetracycline)
 - Treatment is stopped only when two consecutive cultures from the biopsy material are negative.
- **Steroids** can be combined to reduce fibrosis.
- **Surgical treatment** may be required to debridement & establish the airway and correct nasal deformity.
- Long term follow-up; high relapse rates

➤ **Syphilis**

- ✓ Chronic infection caused from the spirochete **Treponema pallidum**.
- ✓ Nasal syphilis is of two types: **acquired and congenital**.

1. **Acquired**: Usually as a sexually transmitted disease it occurs as 3 stage:

- **(a) Primary**.
- It manifests as primary **chancre of the vestibule of nose**. It is rare.

○ **(b) Secondary:**

- Highly contagious, It manifests as **simple rhinitis with crusting and fissuring in the nasal vestibule**.
- Diagnosis is suggested by the presence of:

1. Mucous patches in the pharynx,
2. Skin rash,
3. Fever & general malaise
4. Generalized lymphadenitis
5. Superficial "snail-track" ulcers of the tonsils and soft palate
6. Arthralgia
7. Hepatosplenomegally
8. Genital condyloma lata
9. Nephrotic syndrome

- It is rarely recognized.

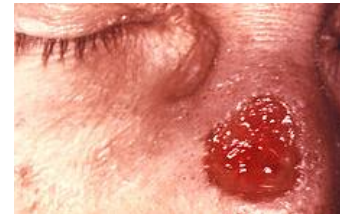


○ **(c) Late Stages**

- **Latent:** (1/3) asymptomatic phase (may have return to mucocutaneous lesions)

- **Tertiary:** (1/3)

- ✓ Noninfectious stage, slowly progressive, may occur years after initial infection.
- ✓ Nose is commonly involved:
- ✓ Typical manifestation is the **formation of a gumma on the nose**.
- ✓ The septum is destroyed both in its **bony and cartilaginous** parts.
- ✓ Offensive nasal discharge with crusts.
- ✓ Bony or cartilaginous sequestra may be seen.
- ✓ Perforation may also appear in the hard palate.
- ✓ Bridge of the nose collapses causing a **saddle nose deformity**.



- **Remission:** (1/3) resolution

○ **H&N Symptoms of syphilis:-**

- **Lymphadenopathy:** generalized cervical adenopathy
- **Laryngeal:** laryngitis with mild edema of the larynx, vocal fold paralysis,
- **Dysphagia :** pharyngitis, tonsillitis
- **Oral Cavity:** chancre Leukoplakia , , granulomatous infiltration of tongue, palate
- **Otologic:** abrupt profound SNHL, Ménière's symptoms, interstitial keratitis, TM perforation, gummas of the temporal bone , facial palsy (unilateral , bilateral)
- **Nasal:** see above
- **Other:** patchy alopecia, cranial nerve involvement

2. Congenital:

- ✓ From maternal transmission , often fatal
- ✓ It occurs in two forms: **early and late**.

a) Early form: It is seen in the first 3 months of life.

- ✓ Manifests as **rhinitis (runny nose)**, Soon the nasal discharge becomes purulent.
- ✓ Associated with:
 - Fissuring and excoriation of the nasal vestibule and of the upper lip.
 - Hepatosplenomegally
 - Jaundice
 - Lymphadenopathy

b) Late form:

- ✓ Usually manifests around **puberty**.
- ✓ Clinical picture is **similar to that seen in tertiary stage of acquired syphilis**.
- ✓ Other stigmata of syphilis:
 - **Neurosyphilis**
 - **Mulberry molars** "dental condition characterized by multiple rounded rudimentary"
 - **Frontal bossing**
 - **Mental retardation**
 - **Saber chins**
 - **Hutchinson's Triad:** abnormal central incisors" Notched incisors" (Hutchinson's teeth), interstitial keratitis, deafness



- **Diagnosis**

- ✓ **Serological tests**

- **Nonspecific** RPR or VDRL (Venereal Disease Research Laboratories)
- **Specific** FTA-ABS (Fluorescent Treponemal Antibody- Absorption Test)

- ✓ **Biopsy** of the tissue with special stains to demonstrate *Trep. pallidum* by dark field microscopy, **Warthin-Starry tissue staining**

- ✓ **False positive VDRL is associated with what diseases?**

- Viruses & infections – Measles, Malaria, EBV, Smallpox, HSV, HIV, Hepatitis
- Rheumatic fever, Rheumatoid arthritis
- Leprosy, Lupus



- **Treatment**

- ✓ Penicillin , ampicillin, tetracycline, erythromycin
- ✓ Steroids for otologic involvement
- ✓ Nasal crusts are removed by irrigation with alkaline solution.
- ✓ Bony and cartilaginous sequestra should also be removed.
- ✓ Cosmetic deformity is corrected after disease becomes inactive.

➤ TB

- ✓ Primary tuberculosis of nose is **rare**.
- ✓ More often it is secondary to lung tuberculosis (inhalation of acid-fast bacilli, Mycobacterium Tuberculosis).
- ✓ Hematogenous /lymphatic spread
- ✓ **Anterior part of nasal septum and anterior end of inferior turbinate** are the sites commonly involved.
- ✓ First, there is nodular infiltration followed later by ulceration and perforation of nasal septum in its **cartilaginous part**.
- ✓ **Risks factor** : immunocompromised (50% of HIV population), health care workers, immigrants, elderly, poor
- ✓ **Diagnosis**:
 - i. skin test positive if :-
 - any reaction in HIV patients,
 - >5 mm in household contacts
 - >10 mm in health care workers,
 - >15 mm in low risk population),
 - ii. sputum culture &AFB
 - iii. chest x-ray
 - iv. biopsy for acid fast bacilli & culture
- ✓ **Treatment** (RIPE)
 - Isoniazid (INH) and rifampin for active disease (for drug resistance consider adding ethambutol, pyrazinamide)
 - Prophylaxis INH for immunosuppressed patients with exposure or positive PPD
- **H&N manifestations usually secondary from pulmonary source :**
 - ✓ **Cervical Lymphadenopathy**: bilateral, anterior and posterior, firm,nontender, most common H&N manifestation
 - ✓ **Larynx**: granulation and ulcerative tissue in **posterior glottis** (posterior interarytenoids, laryngeal surface of epiglottis, vocal folds)
 - ✓ **Otologic**: painless, odorless, watery otorrhea, multiple TM perforations
 - ✓ **Salivary Glands**: diffuse glandular involvement
 - ✓ **Oral Cavity**: ulcerative lesions
 - ✓ **Ocular**: conjunctivitis, uveitis

➤ **Lupus Vulgaris**

- ✓ It is a **low-grade tuberculous infection** commonly affecting nasal vestibule or the skin of nose and face presents as:
 - Chronic vestibulitis
 - Face skin lesions brown, gelatinous nodules called "apple-jelly" nodules.
 - Perforation may in the cartilaginous part of nasal septum.



- ✓ **Dx:** Biopsy of the lesion is useful to make the diagnosis.
- ✓ **Treatment** : same as for tuberculosis of nose

➤ **Leprosy** (Hansen's Disease)

- ✓ It is caused by *Mycobacterium leprae*. highly infective, but low virulence "3-10 yr incubation"
- ✓ **Tissues affected:** Schwann cells or peripheral nerves; small vessels; monocytes
- ✓ The nose is involved as a part of systemic disease (cutaneous lesions)
- ✓ **Most common route of spread is via nasal discharge**
- ✓ Infection starts in the **anterior part of nasal septum and anterior end of inferior turbinate** (like TB)
- ✓ May present with ulcerative lesions in the larynx and oral mucosa, lymphadenopathy
- ✓ **Initially**, there is excessive nasal discharge with red and swollen mucosa.
- ✓ **Later**,
 - Crusting and bleeding.
 - Nodular lesions on the septum may ulcerate and cause perforation.
 - Atrophic rhinitis
 - Depression of bridge of nose
 - Destruction of anterior nasal spine with retrusion of the columella (nasal collapse)



- ✓ **Diagnosis:**
 - Nasal mucosa and biopsy (Acid-fast lepra bacilli can be seen in the foamy appearing histiocytes called lepra cells (lipid-laden histiocytes)).
- ✓ **Treatment:**
 - Dapsone ((Diaminodiphenylsulfone), rifampin and isoniazid & steroid
 - Reconstruction procedures are required when disease is inactive

❖ Fungal Infectious

➤ **Rhinosporidiosis**

- ✓ It is a fungal granuloma caused by *Rhinosporidium seeberi*.
- ✓ Endemic to Africa, Pakistan, India, Sri Lanka, spread from contaminated water (public bathing)
- ✓ The disease mostly affects **nose and nasopharynx**
- ✓ **Other sites** such as : lip, palate, conjunctiva, epiglottis, larynx, trachea, bronchi, external ear canal, and the parotid duct ,skin, vulva, vagina may also be affected.
- ✓ **In the nose:** the disease presents as:-
 - **Friable**, polypoidal mass, "**strawberry**" red (vascular) in colour
 - Attached to nasal septum or lateral wall.
 - Sometimes, it extends into the nasopharynx and may hang behind the soft palate.
 - The mass is very vascular and bleeds easily on touch.
 - Its surface is studded with white dots representing the (spore of fungus).
 - Patient may complain of nasal discharge , epistaxis



- ✓ **Diagnosis** is made on biopsy.
 - It shows several spore "thick-walled giant sporangia containing up to 10,000 sporangiospore"
 - It has **not** been possible to culture the organism
- ✓ **Treatment :-**
 - Complete excision of the mass with diathermy knife and cauterisation of its base..
 - Oral antifungal agents, corticosteroid injections, may consider dapsone
 - Recurrence may occur after surgical excision

Other rare Fungal Infections of nose such as:-

- ✓ Candidiasis,
- ✓ Histoplasmosis
- ✓ Blastomycosis
- ✓ Sporotrichosis
- ✓ Blastomycosis (dermatiditis)
- ✓ Coccidiomycosis (immitis)

❖ Autoimmune/Vasculitis

➤ **Wegeners Granulomatous** "Granulomatosis with Polyangiitis"

✓ **Aetiology** :- idiopathic necrotizing granulomatosis small vessel vasculitis

✓ **Involving:**

- Mainly the upper airways, lower airways (lungs)
- Kidneys (glomerulonephritis)
- Skin.

✓ It should be differentiated from T-cell lymphoma (non-healing midline granuloma) because the treatment of the two is quite different.

✓ Most patients present in 5th decade, mostly white, M=F

✓ **3 Types:**

✓ **Type 1**:- limited disease, characterized by :-

- Upper airway symptoms and few systemic findings.
- Several weeks of symptoms similar to those of an upper respiratory tract infection but that are unresponsive to antibiotics
- Nasal pain, serosanguinous rhinorrhea, and crusting.

✓ **Type 2** disease are sicker, characterized by :-

- Seen initially with systemic features, not severe
- The initial presentation similar to that of **type 1** + nasal ulcerations, cough, hemoptysis, and cavitory lesions on chest radiography.

✓ **Type 3** widely disseminated form of the systemic features are more profound characterized by :-

- Similar to that of type 2 + cutaneous lesions, and **progressive renal involvement**.

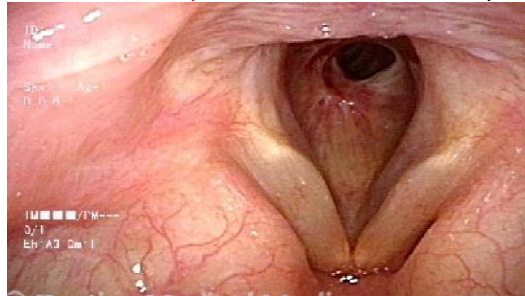
✓ **Clinical Features**

○ General systemic symptoms include anaemia, fatigue, night sweats and migratory arthralgias.

○ **Nasal findings include** (most common H&N symptom) :-

- Early symptoms of Wegener's granulomatosis include clear or blood-stained nasal discharge which later becomes purulent.
- Recurrent and chronic sinusitis (most common)
- Crusting
- Epistaxis
- Granulations
- Septal perforation
- Nasal airway stenosis.
- Saddle nose.
- Nasal endoscopy typically reveals mucosal cobblestoning, edema, and crusting. diagnosis and treatment Because nonspecific nature of many of the symptoms

- **Laryngeal:** **subglottic stenosis**, ulcerative lesions ,



- **Otologic:** CHL, serous chronic otitis media
- **Oral:** Gingival hyperplasia, gingivitis
- **Ocular:** Uveitis, keratitis
- **lung:** Manifested by cough and sometimes haemoptysis. X-ray chest may show a single or multiple **cavity lesions**.
- **kidneys:** Urine examination will show red cells, casts and albumin. Serum creatinine level is raised. Renal failure is the usual cause of death in these patients.
- **Skin:** Ulcers of distal extremities, Wart-like lesions around the elbows, Pyoderma gangrenosum like lesions ,Petechia ,Crusted plaques

✓ **Diagnosis**

- **cANCA (antineutrophil cytoplasmic autoantibodies):**
 - Suggests **active disease**" monitor disease activity"
 - Highly sensitive for WG, but a negative result does not exclude the diagnosis.
 - (cANCA + antibodies against proteinase-3 = **86%** specific)
 - False positives have been seen in amebiasis, leprosy, and infectious endocarditis
- **Biopsy** from the nose is diagnostic (necrotising vasculitis involving **small arteries** or **veins** "with multinucleated giant Cells").
- **ESR is raised.**
- **Renal profile**

TABLE 31.3	THE AMERICAN COLLEGE OF RHEUMATOLOGY DIAGNOSTIC CRITERIA OF WG (TWO OF FOUR REQUIRED)
-----------------------	--

The American College of Rheumatology Diagnostic Criteria of WG

Oral ulcers and nasal discharge
 Nodules, fixed infiltrate, or cavities on chest film
 Nephritis urinary sediment (red cell cast or >5 red blood cells per high power field)
 Granulomatous inflammation on biopsy

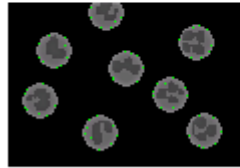
- ✓ **Treatment** It consists of :-
- Systemic steroids "prednisone 0.5 to 1.0 mg/kg/day up to a maximum of 80 mg/day. tapering may begin after 1 month with the goal to discontinue the agent completely within 6 to 9 months"
 - Cytotoxic drugs.(Cyclophosphamide and azathioprine OR methotrexate)
 - Local therapy nasal hypertonic saline irrigations and nasal debridement
 - Surgical reconstruction may be used to restore function once the disease is in remission, and it includes:-
 - Correction of saddle nose deformity
 - Septal perforation repair
 - FESS may benefit selected patients with chronic nasal crusting with Saline irrigations

Wegener's Granulomatosis

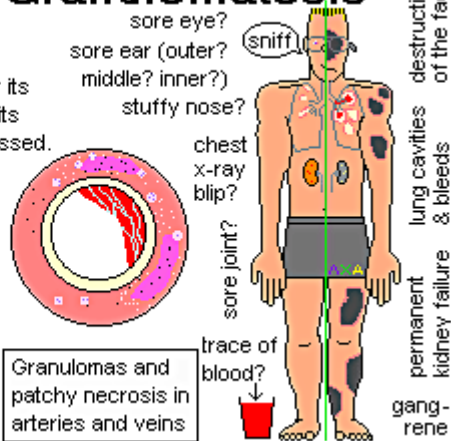
Easy to diagnose and treat -- if you think of it.

Wegener's is infamous for its subtle presentation, and its lethality if (and only if) missed.

Caused by autoantibodies against proteinase 3.



Positive anti-neutrophil cytoplasm test (c-ANCA).



- **Churg-Strauss syndrome** "Allergic Granulomatosis Angiitis"
- ✓ **Pathophysiology:** rare unknown etiology, small to medium vessel **necrotizing vasculitis** causes **angiitis** and **allergic granulomatosis**
- ✓ **Clinical Features**
 - Triad:-
 - Asthma,
 - Eosinophilia (>10%)
 - Systemic vasculitis
 - Allergic rhinosinusitis, nasal polyposis nasal obstruction, septal lesions
 - Pulmonary infiltrates" eosinophilic pneumonia"
 - Mononeuritis or polyneuropathy
 - Lung lesions
 - Myocardial infarction (secondary to coronary arteritis),
 - Sensorineural and conductive hearing loss
 - Fever
 - Weight loss

TABLE 31.5	THE AMERICAN COLLEGE OF RHEUMATOLOGY CRITERIA FOR DIAGNOSIS OF CSS (AT LEAST FOUR REQUIRED FOR DIAGNOSIS)
	The American College of Rheumatology Criteria for Diagnosis of CSS History of asthma Eosinophilia > 10% Neuropathy Nonfixed pulmonary infiltrates Paranasal sinus abnormalities Extravascular eosinophils on biopsy

- ✓ **Diagnosis :**
 - Presence of principal symptoms,
 - Nerve or muscle biopsy
 - Histopathologically, is characterized by:-
 - Necrotizing vasculitis of small and medium-sized vessels.
 - Necrotizing extravascular granulomas
 - Eosinophilia of the vessels and perivascular tissue is prominent.
 - Serum IgE and eosinophils
 - CXR
 - CT of paranasal sinuses
- ✓ **Treatment:**
 - Corticosteroids
 - Cytotoxic agents (cylophosphamide) reserved for life-threatening conditions,
 - Symptomatic medications for nasal symptoms, polypectomy or sinus surgery as needed
 - Consultation (rheumatologist , medical pulmonologists, cardiologists, nephrologists)

➤ Relapsing Polychondritis

✓ **Pathophysiology:**

- Rare autoimmune disease of unknown etiology,
- Inflammation of elastic cartilaginous tissue with high concentration of glycosaminoglycans "Hyaline cartilage"
- Typically occurs in the fourth decade
- Affects men and women equally

✓ **Clinical Features** (episodic and progressive)

- Auricular chondritis "lobule-sparing",
- Cochlear and vestibular injury (vertigo, hearing loss)
- Respiratory chondritis (laryngeal collapse)
- Nasal chondritis (saddle-nose deformity ,septal perforation , crusting, rhinorrhea, epistaxis)
- Polyarthritits (nonerosive, migratory)
- Cardiac valve insufficiency
- Ophthalmic structures "ulcerative keratitis, uveitis"
- The cause of death in the majority of these patients is secondary to respiratory tract or cardiovascular involvement.



✓ **Diagnosis :**

- McAdam "described diagnostic criteria" require three or more criteria in combination with histologic confirmation at two or more locations

TABLE 31.4	McADAM CRITERIA FOR DIAGNOSIS OF RELAPSING POLYCHONDritis
-----------------------	--

McAdam Criteria for Diagnosis of Relapsing Polychondritis

- Bilateral auricular chondritis - Nonerosive, seronegative inflammatory polyarthritits - Nasal chondritis - Ocular inflammation - Respiratory tract chondritis - Cochlear/vestibular dysfunction
--

- H&P :- Perichondrial inflammation, fibrous tissue Replacement
- Elevated **nonspecific** immunologic markers (eg, ESR, IgG , Antinuclear antibodies)
- Elevated antibodies to type II and type IV collagen (must differentiate from rheumatoid arthritis, gout, and other connective tissue diseases)
- CT, and MRI may be helpful in determining the diagnosis and extent of the disease.

✓ **Treatment:**

- NSAIDs
- Corticosteroids for severe attacks
- Cytotoxic medications

➤ **Systemic lupus Erythematous**

- ✓ Late stage cause ulcerated nasal septum (nasal perforation)
- ✓ See premalignant lesion lecture

➤ **Sjogrens Syndrome**

- ✓ See salivary gland lesion lecture
- ✓ Sjögren syndrome may come to medical attention with nasal dryness that leads to crusting , hyposmia and epistaxis.
- ✓ Recurrent sinusitis were rare
- ✓ Examination finding was dry mucosa. with septal ulceration and crusting occurring less commonly
- ✓ Nasal saline sprays/irrigations and increasing environmental humidification are recommended.

➤ **Idiopathic**

➤ **Sarcoidosis**

✓ **Pathophysiology:**

- ✓ It is a chronic granulomatous disease of **unknown aetiology**
- ✓ Resembling tuberculosis on history but with the **absence of caseation**
- ✓ May resolve spontaneously within 2 years or progress to death
- ✓ Mononuclear cells accumulate in affected organs followed by **formation of granulomas, may lead to irreversible fibrosis of tissue**
- ✓ More common in **African-American women & Scandinavian individuals**

✓ **Clinical Features:**

- ✓ It is a systemic disorder capable of involving almost any organ in the body
- ✓ Symptoms may refer to involvement of:

- Lungs
- Lymphatic system,
- Liver
- Spleen
- Eyes
- Skin.
- Bones
- Epithelium of the upper respiratory tract

- ✓ 90% pulmonary involvement
- ✓ 40% of patients with sarcoidosis have granulomatous changes in extrapulmonary organs 40% asymptomatic (incidental chest x-ray finding)

- ✓ In the **nose** (1-4% of sarcoid patient), it presents with:-

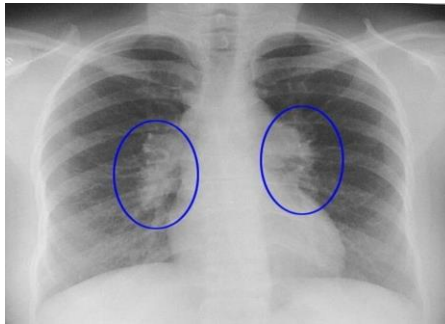
- Submucosal yellowish nodules involving septum or the inferior turbinate (cobblestoning of sinonasal mucosa)
- Nasal obstruction
- Nasal pain
- Crusting
- Epistaxis
- Septal perforation
- Epiphora
- More advanced disease, irregular polypoid mucosa is seen "friable and bleeds readily"
- Bony lesions of the nasal bones "The suture lines may disappear, but no periosteal reaction is seen"



- ✓ Skin nodules "Lupus pernio" may form in the nasal vestibule or skin of face



- ✓ Pulmonary:
 - Primary organ affected (cough, hilar adenopathy, dyspnea 90%)
 - X-ray chest shows diffuse pulmonary infiltrate "fibrosis " with hilar adenopathy.



- ✓ Cervical adenopathy (25–50%, most common H&N presentation)
- ✓ Salivary Glands: parotid mass, uveoparotid fever or Heerfordt's disease

Uveoparotid Fever (Heerfordt's Syndrome)

- More common in women
- Self-limited uveitis, parotid enlargement, SNHL, facial palsy, malaise, fever
- Rx: Corticosteroid

- ✓ Laryngeal: Supraglottic submucosal mass (epiglottis most common), TVF paralysis
- ✓ Rheumatologic : arthralgia, myalgia

- ✓ **Diagnosis :**
- ✓ Combination of "histologic ,radiographic, immunologic, biochemical data"
- ✓ **Biochemical**
 - Serum and urinary calcium levels are raised.
 - Serum angiotensin-converting enzyme (ACE) elevations occur in 83%with active sarcoidosis "useful for the diagnosis of sarcoidosis and for monitoring for relapse"
 - Nonspecific :- CRP , ESR
 - ANCA should be measured to rule out Wegner
- ✓ **Radiology :-**
 - CXR classified as follows:-
 1. stage 0 :normal CXR
 2. stage 1 : isolated intrathoracic adenopathy
 3. stage 2 : intrathoracic adenopathy and parenchymal disease
 4. stage 3 :parenchymal disease
 5. stage 4 :pulmonaiy fibrosis
 - Sinus CT findings are abnormal in most cases of nasal sarcoidosis "Mucous membrane thickening or opacification".
 - Radioactive gallium uptake may be increased in the nasal mucosa in sarcoidosis.
- ✓ **Histology**
 - Biopsy of the lesions helps to establish the diagnosis (noncaseating granulomas)
 - Negative stains for fungus and acid-fast bacilli help to support the diagnosis.
- ✓ **Complications:**
 - Progressive interstitial lung disease
 - Blindness (progression of uveitis)
 - Airway obstruction (rare)
- ✓ **Treatment :-**
 - Systemic steroids "prednisone in doses of 10 to 40 mg daily" Six to nine months
 - NSAIDs
 - For nasal symptoms, steroids can be used locally as nasal spray & saline irrigations.
 - Do not treat asymptomatic lesions
 - Methotrexate has been used to treat nasal sarcoidosis successfully
 - May require surgical for symptomatic nasal obstruction or chronic sinusitis

❖ **Neoplastic**

➤ **T-Cell lymphoma**

- ✓ Previously known as midline **malignant reticulosis , Midline Destructive Syndromes or polymorphic reticulosis**
- ✓ The most notable neoplastic, systemic disease with nasal manifestation is nasal **T-cell lymphoma**.
- ✓ **Leukemia and B-cell lymphomas** may also have nasal manifestations
- ✓ **B-cell lymphomas**: manifest as unilateral nasal obstruction by an enlarged nasal or nasopharyngeal mass.
- ✓ **Acute leukemia** :-manifest as symptoms of an upper respiratory infection or as epistaxis secondary to friable mucosa in the anterior nose
- ✓ Nasal T-cell lymphomas differ phenotypically from lymphomas of the paranasal sinuses and in the Waldeyer ring, which tend to be of B-cell origin

✓ **Pathophysiology:**

- Originates from **T-cells**, form of **extranodal Non-Hodgkin's Lymphoma** forms perivascular infiltrates (angiocentric, **not a true vasculitis**)
- Can lead to vessel occlusion and local tissue infarction
- Association of Epstein-Barr virus (EBV) and nasal T-cell lymphoma has been frequently reported

✓ **SSx:**

1. Pansinusitis and nasal obstruction , purulent rhinorrhea (most common initial finding)
2. Typically, unilateral involvement of midface (paranasal, oral, orbital) usually unilateral mucosal ulceration with extension into the palate, maxillary sinus, and upper lip, "nasal septal perforations & Oronasal fistulae often occur"
3. Cutaneous involvement (maculopapular rash, ulcerative cutaneous lesions)
4. Unlike Wegener's granulomatosis, it is rapidly destructive, and usually localized to the head and neck & devoid of systemic involvement
- 5- Can progress to **autorhinectomy**



✓ **Stages:**

- 1. **Prodromal**: clear rhinorrhea, nasal obstruction
- 2. **Ulcerative (Active)**: purulent rhinorrhea, ulceration and septal perforations (epistaxis), destruction of osteocartilaginous tissue
- 3. **Terminal**: malaise, fever, sloughed tissues, death

- ✓ • **Dx**: Biopsy, CT/MRI of paranasal sinuses



- ✓ **Histopathology**: sheets of **atypical** polymorphonuclear cells, no granuloma, no palisading histiocytes
- ✓ **Rx**: radiation therapy for local disease, chemotherapy for disseminated disease or relapses

➤ **Pyogenic granuloma (Lobular capillary hemangioma)**

✓ **Introduction:-**

- Pyogenic granuloma also known as
 1. "Lobular capillary hemangioma"
 2. "Eruptive hemangioma"
 3. "Granuloma gravidarum"
 4. "Pregnancy tumor"
 5. "Tumor of pregnancy"
- Pyogenic granuloma is **benign** tumorlike tissue overgrowth of tissue
- Most pyogenic granuloma of the head and neck arise in the **oral cavity, and fewer in the nasal cavity**.
- Its also found in skin
- The name for pyogenic granuloma is misleading because it is not a true granuloma.
- In actuality, it is a **capillary hemangioma** of lobular subtype which is the reason they are often quite prone to **bleeding**.
- It is also not truly "pyogenic," as the origin is traumatic and not infectious

✓ **Etiology:-**

- Etiology is unclear
 1. Trauma (nose picking , intubation)
 2. Chronic irritation
 3. Infection
 4. Hormonal factors
- Have been suggested to play a role
- Most likely to occur in children and younger adults
- More females affected than men.
- In pregnant women, it is most likely to occur in the first trimester with an increasing incidence up until the seventh month

✓ **Clinical presentation:-**

- Pyogenic granulomas mostly involves anterior gingivae (75% of cases)
 - Often maxillary more than mandibular jaw.
 - Anterior areas are more often affected than posterior areas.

✓ **It can also be found on the other sites :-**

- Anterior nasal septum and the tip of the turbinate (source of frequent nose bleeds)
- Tongue
- Buccal mucosa
- Lips





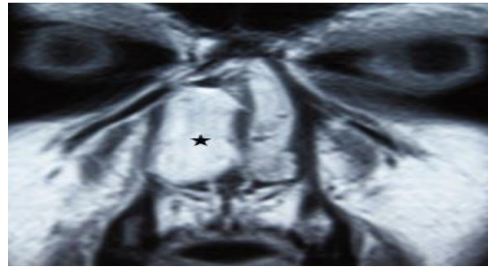
- Appearance of pyogenic granuloma can be smooth or lobulated.
- Color ranging from red/pink to purple
- Size ranges from a few millimeters to centimeters.
- It can be painful, especially if located in an area of the body where it is constantly disturbed.
- Pyogenic granulomas friable , will often bleed profusely with little or no trauma.
- Presents with :-
 - Epistaxis
 - Unilateral obstruction
 - Nasal discharge
 - Epiphora
 - Rarely with facial pain, alteration of smell, and headache
 - Rarely present as a mass of considerable size and entirely fill the nasal cavity

✓ **Dx :-**

- **Endoscopy** : the lesion is usually seen as a red to purple single hypervascularized mass, less than 10 mm in diameter, with a predilection for the anterior portion of the nasal septum.



- **Radiology :-**
- CT:
- Unilateral mass with soft tissue density,
- MRI:
 - T2 hyperintensity and spontaneous T1 hypointensity; enhancement with contrast medium.
 - Determining any intracranial extension or connection for big lesions or lesions originating from the roof of the nasal cavity

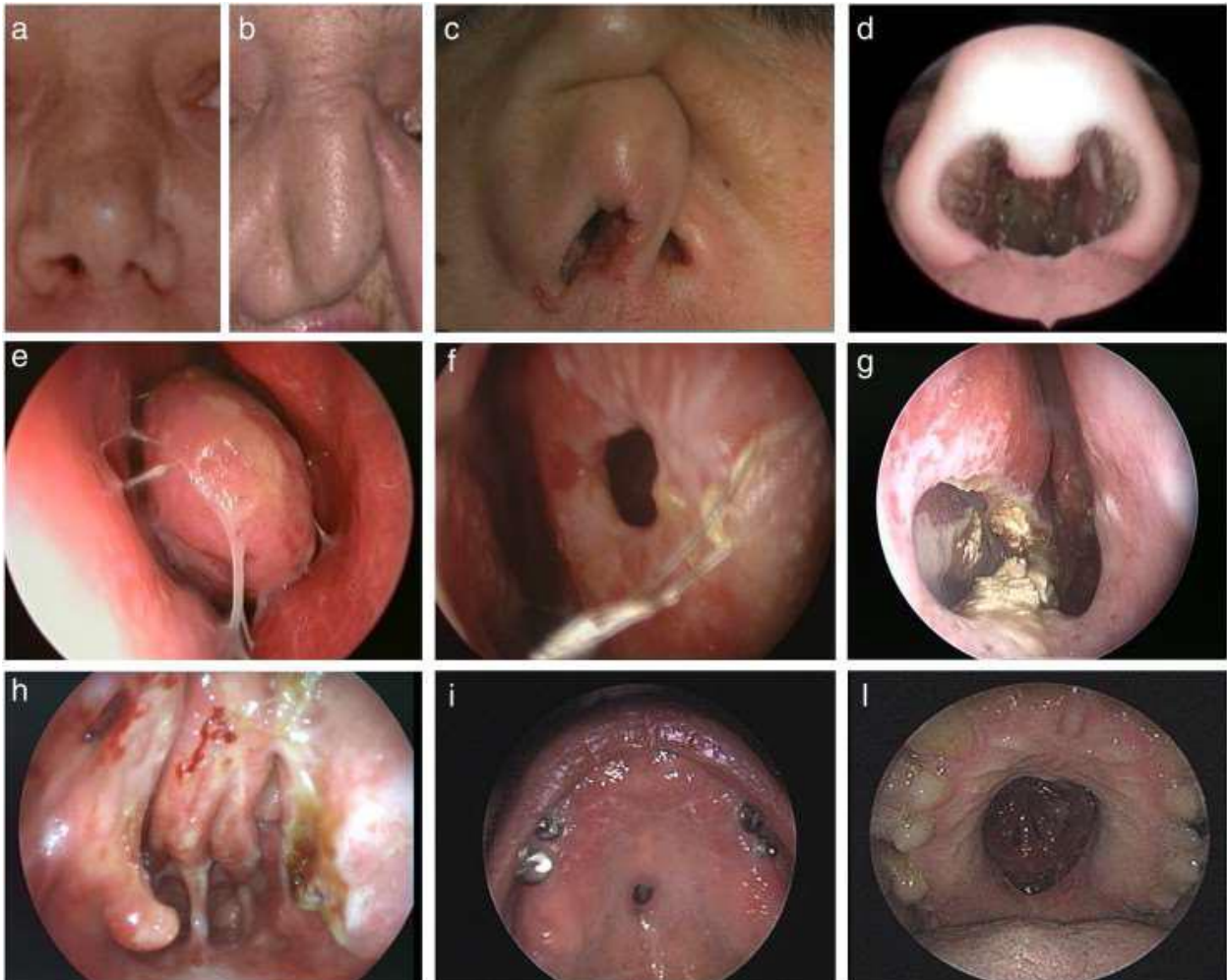


- **Biopsy**
 - **Histopathology feature:-**
 - Appearance microscopically :-
 - Capillaries arranged in lobules and separated by a loose connective tissue stroma, often infiltrated by inflammatory cells the
 - Surface may have ulcerations.
- ✓ **DDx :-**
 - The appearance can be very similar to that of a hemangioma
 - Angiofibroma
 - Hamartoma
 - Arteriovenous malformation
 - Nasal polyp
 - Kaposi's Sarcoma In the oral cavity
 - Meningocele
 - Wegener's granulomatosis
 - Metastases from highly vascularized tumors (e.g., kidney or thyroid carcinoma)
- ✓ **Rx:-**
 - Total excision of the lesion by endoscopic surgery techniques has been recommended & cautery the base
 - Visualization of the mass and surrounding anatomy, thus allowing the surgeon to remove the mass completely
 - Incomplete resection can lead to recurrence

❖ Foreign Body Reactions

➤ Cocaine-induced midline granuloma

- ✓ Nasal inhalation of cocaine can cause necrosis and destruction of nasal and paranasal structures
- ✓ Facial pain, epistaxis, nasal crusting, septal perforation, destruction of medial maxillary wall, inferior and medial turbinates,
- ✓ Can easily be confused with (limited) WG
- ✓ c-ANCA is specific for WG; p-ANCA is nonspecific, (10% WG)
- ✓ No systemic symptoms
- ✓ **Histology:**
 - Extensive necrosis, acute/chronic inflammatory changes
 - Absence of necrotizing granulomas, multinucleated giant cells, vasculitis distinguishes it from WG
- ✓ **Treatment:**
 - Stop use of cocaine
 - Topical steroids
 - Antibiotics



❖ **IMMUNODEFICIENCY DISEASES**

- **SINUSITIS IN THE IMMUNOCOMPROMISED PATIENT**
- **AIDS AND THE NASAL AIRWAY**

❖ **CUTANEOUS DISEASES**

- **PEMPHIGUS VULGARIS**
- **PEMPHIGOID**
- **SCLERODERMA**
- **BEHÇET DISEASE**

❖ **MUCOCILIARY DISEASES**

- **PRIMARY CILIARY DYSKINESIA**
- **CYSTIC FIBROSIS**

❖ **HEMATOLOGIC DISEASES**

- **HEREDITARY HEMORRHAGIC TELANGIECTASIA**

Otology:

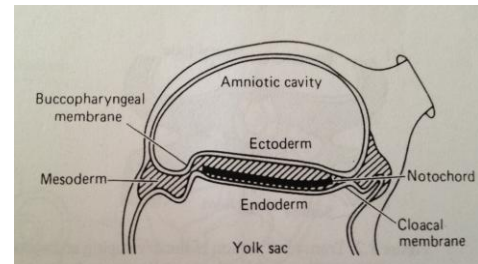
- 1- Ear Embryology**
- 2- External Ear Anatomy**
- 3- Middle Ear Anatomy**
- 4- Inner Ear Anatomy**
- 5- Eustachian Tube Anatomy, Physiology and Disorders**
- 6- Physiology of Auditory System**
- 7- Audiology and Hearing Assessment**
- 8- Approach to Hearing Loss in Adults**
- 9- Congenital Hearing Loss and Neonatal Hearing Screening**
- 10- Congenital Disorders of External Ear**
- 11- Disorders of External Ear**
- 12- Disorders of Tympanic Membrane**
- 13- Acute Otitis Media**
- 14- Otitis Media With Effusion**
- 15- OME and VT Guidelines Summary**
- 16- Chronic Suppurative Otitis Media (CSOM), Cholestatoma and Tympanomastoidectomy**
- 17- Ossiculoplasty**
- 18- Complications of Otitis Media**
- 19- Otosclerosis**
- 20- CPA and Petrous Apex Lesions**
- 21- Temporal Bone Trauma**
- 22- Facial Nerve Paralysis and Rehabilitation**
- 23- CSF Otorrhea**
- 24- Hearing Aids and Implantable Hearing Devices**
- 25- Cochlear Implantation in Chronic Ear**
- 26- Cochlear Implantation in Inner Ear Malformations**
- 27- Inner Ear Malformations (Summary)**
- 28- Labyrinthine Ossifications**
- 29- Auditory Neuropathy**
- 30- Physiology of Vestibular System**
- 31- Evaluation of a Dizzy Patient**
- 32- Disorders of Vestibular System**
- 33- Approach to Tinnitus**

Embryology of Ear

- Prenatal development divided in to separate periods:

1. First period (Pre-embryonic):

- From implantation to end of 3rd week.
- Three layers develop (ectoderm, mesoderm & endoderm) containing the notochord.



2. Second period (Embryonic):

- 35 days (until the end of 8th week)
- **Major systems and organs are formed**, embryo has external shape that recognizable as human.
- **Mesoderm** give raise to **Pharyngeal arches**.
- **Ectoderm** around the notochord give raise to the **Neural crest**

3. Fetal period:

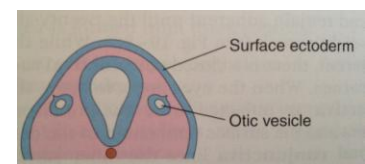
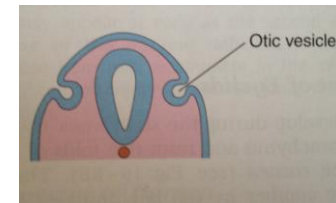
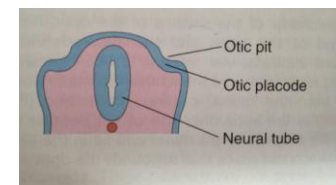
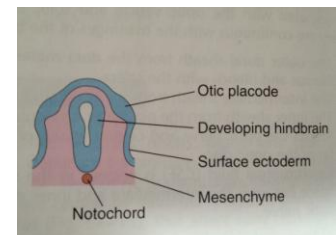
- Remaining 7 months
- **Change of position and shape of structure**

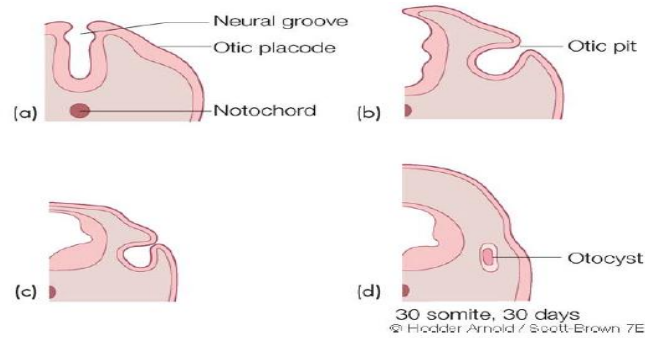
Development of Ear

- Composed of 3 anatomical parts:
 1. External ear: (auricle, EAC and tympanic membrane)
 2. Middle ear
 3. Internal ear

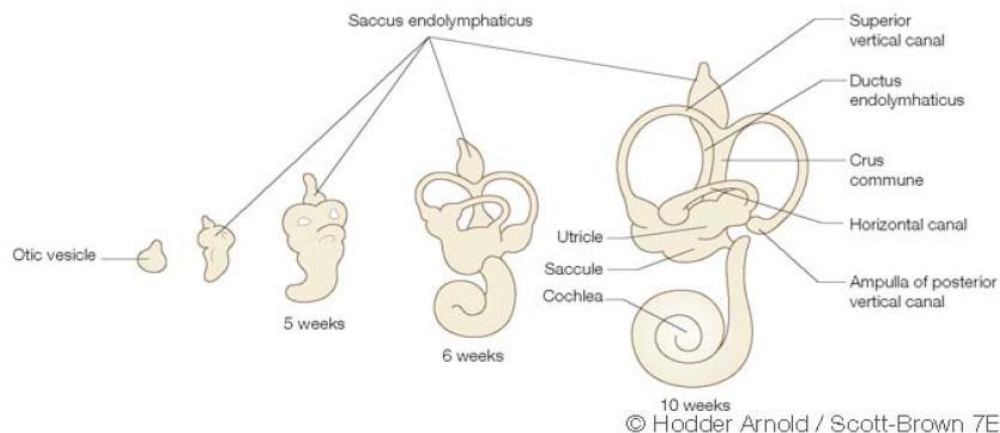
Development of INNER EAR:

- 1st to develop from the 3 parts.
- **Starts in 3rd-4th week of fetal life**
- **Complete by the 16th week.**
- **By 4th week:**
- Thickening of surface Ectoderm (**Otic Placode**) on each side of caudal part of hindbrain (myelencephalon)
- Its form is stimulated by notochord and paraxial mesoderm
- It invaginates and sinks deep into the underlying mesenchyme to form (**Otic Pit**)
- Edges of otic pit comes together to form (**Otic Vesicle or Otocyst**) which is considered as (**Primordium of membranous labyrinth**)
- Otic vesicle loses its connection with surface ectoderm.





- A diverticulum grows from the otic vesicle and elongates to form **Endolymphatic duct and sac (1st to develop)**
- Otocyst is differentiated into 2 parts:
 - 1. Dorsal Utricular part:**
 - Endolymphatic duct, utricle & semicircular ducts)
 - 2. Ventral Saccular part:**
 - Saccule & cochlear duct



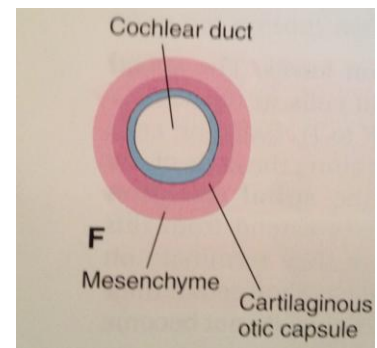
Development of Utricular part of membranous labyrinth takes place **Earlier** than Sacular part.

- From the Dorsal utricular part:
- 3 disclike diverticula grows out from the utricular part, the central parts of these diverticula fuse and disappear (**Semicircular Ducts**)
- Attached to the utricle, later enclosed in **semicircular canals** of **bony labyrinth** (from mesenchyme around otic vesicle)
- At one end of each semicircular duct a dilatation **Ampullae**, inside it differentiate the specialized receptors area (**cristae ampullares**)
- In the utricle and saccule (**maculae**)

- From the ventral saccular part:
- Tubular diverticulum grows (**Cochlear duct**) and coils to form the **membranous cochlea**
- Sacculle connected with cochlea by **Ductus reuniens (narrowest segment)**
- **Organ of corti** (Spiral organ) differentiate from cells in the wall of cochlear duct
- Ganglion cells of the 8th nerve migrate along the coils of membranous cochlea and form **spiral ganglion**
- Nerve processes extend from this ganglion to the spiral organ and terminate on the hair cells
- **Periotic Duct: within the cochlear aquaduct**, connects the scala tympani to the posterior cranial fossa

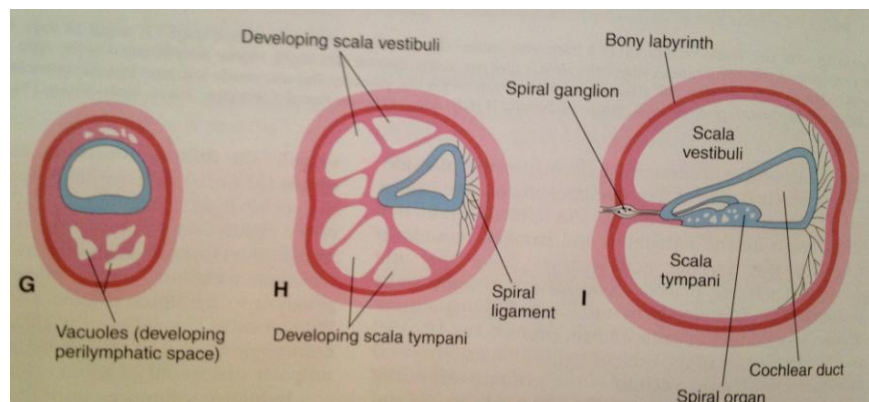
Bony labyrinth:

- Mesenchyme around the otic vesicle condenses and differentiates into Cartilaginous (Otic Capsule)
- Transforming growth factor-B1 may play role
- As the membranous labyrinth enlarges, vacuoles appear in the otic capsule which soon coalesce to form **perilymphatic space**
- Membranous labyrinth become suspended in perilymph



- The perilymphatic space related to cochlea develops two divisions:

1. **Scala Vestibuli**
2. **Scala Tympani**



- Cartilaginous otic capsule later **ossifies** to form the **bony labyrinth** (**Start ossification at 16 weeks**)

- Variable number of centers that finally fuse without leaving telltale suture lines. (From periosteal and endochondral ossification)
- The dense bony mass is the Petrous bone
- **INNER ear reach Adult SHAPE (NOT SIZE) by the middle of the fetal period (20-22 weeks)**
- **Cochlea is developed sufficiently by 20 weeks of gestation and the fetus can hear in the womb of the mother.**
- Channels within the otic capsule includes **oval window** where **part of the otic capsule becomes the stapes footplate and the annular ligament**, thereby allowing sound from the middle ear to enter the labyrinthine fluids

Table 225.4 Development of communication channels passing through labyrinth.

	Channel
Internal auditory meatus	Persisting channel in cartilage model around VII and VIII nerves
Subarcuate fossa	Persisting vascular channel
Vestibular aqueduct	Fifth and sixth ossification centres fuse around the endolymphatic duct
Cochlear aqueduct	Resorption of precartilag
Fossula ante fenestram	Resorption of precartilag
Fossula post fenestram (inconstant)	Resorption of cartilage
Oval window	Otic capsule becomes footplate of stapes and annular ligament
Round window	Persisting cartilage becomes round window niche and membrane

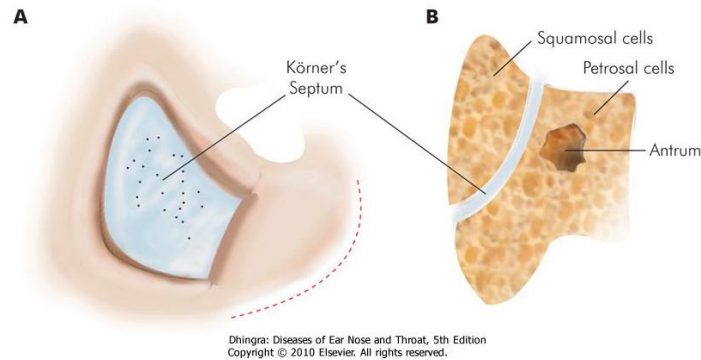
Development of External and Middle ear:

- External and middle ears are **independent** of the development of inner ear
- We can see malformed and non-functional inner ear in the presence of normal external and middle ears, and vice versa.
- External and middle ears mainly derived from **1st and 2nd Pharyngeal ARCHES.**

Pharyngeal Arch	Nerve	Muscles	Skeleton
1. Mandibular	V. Trigeminal; mandibular divisions	Mastication (temporal; masseter; medial, lateral pterygoids); mylohyoid; anterior belly of digastric; tensor palatine, tensor tympani	Premaxilla, maxilla, zygomatic bone, part of temporal bone, Meckel's cartilage, mandible, malleus, incus, anterior ligament of malleus, sphenomandibular ligament
2. Hyoid	VII. Facial	Facial expression (buccinator; auricularis; frontalis; platysma etc); posterior belly of digastric; stylohyoid; stapedius	Stapes; styloid process; stylohyoid ligament; lesser horn and upper portion of body of hyoid bone

Development of Middle ear:

- **Tubotympanic Recess** from the Endoderm of **1st Pharyngeal POUCH.**
- 1. Proximal part form **Eustachian tube**
- 2. Distal part become **Tympanic cavity** (which envelop the ossicles, their tendons, ligament and corda typani)
- Late fetal period, expansion of tympanic cavity gives rise to **mastoid antrum** (in the petromastoid part of the temporal bone)
- **Middle ear reaches Adult SHAPE at Birth.**
- **No Mastoid air cells present at Birth.**
- **Mastoid** develops from the Squamous and Petrous bones.
- Petrosquamosal suture may persist as a bony plate (**Korner's septum**) separating superficial squamosal cells from the deep petrosal cells.
- **By 2-3 years of age the mastoid cells are well developed and produce conical projections of the temporal bone (mastoid process)**



Pharyngeal Arch	Nerve	Muscles	Skeleton
1. Mandibular	V. Trigeminal: mandibular divisions	Mastication (temporal; masseter; medial, lateral pterygoids); mylohyoid; anterior belly of digastric; tensor palatine, tensor tympani	Premaxilla, maxilla, zygomatic bone, part of temporal bone, Meckel's cartilage, mandible, malleus, incus, anterior ligament of malleus, sphenomandibular ligament
2. Hyoid	VII. Facial	Facial expression (buccinator; auricularis; frontalis; platysma etc); posterior belly of digastric; stylohyoid; stapedius	Stapes; styloid process; stylohyoid ligament; lesser horn and upper portion of body of hyoid bone

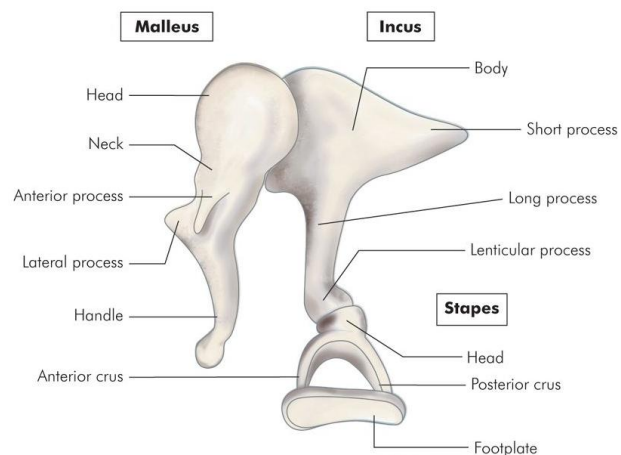
- Ossicles, muscles and nerves in the table above

1. First Branchial Arch (Meckel's cartilage):

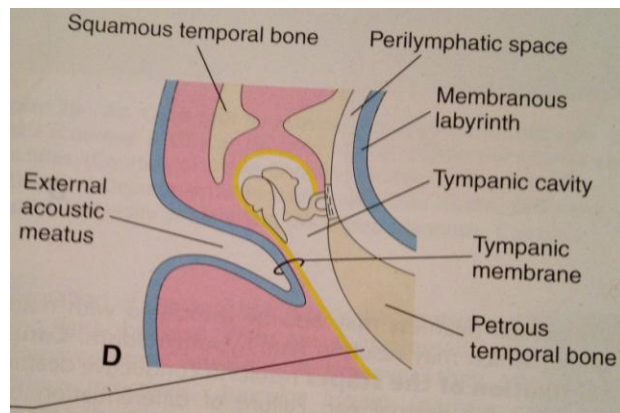
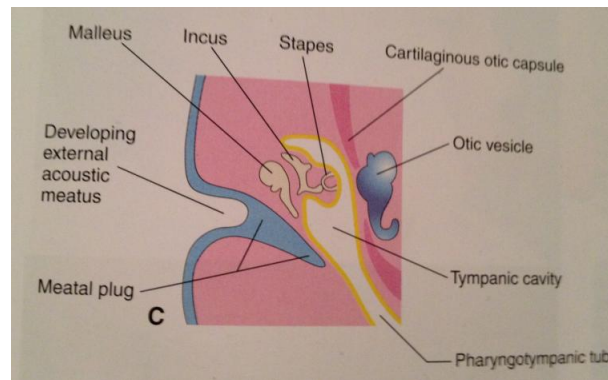
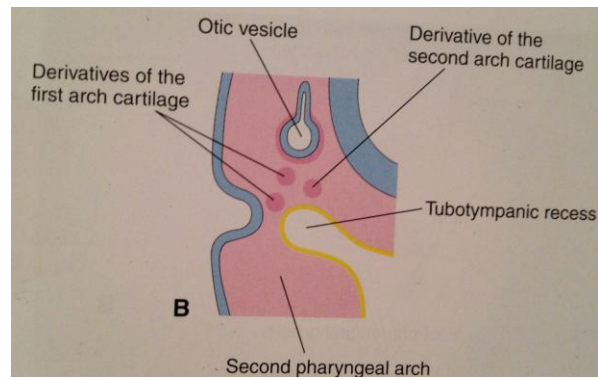
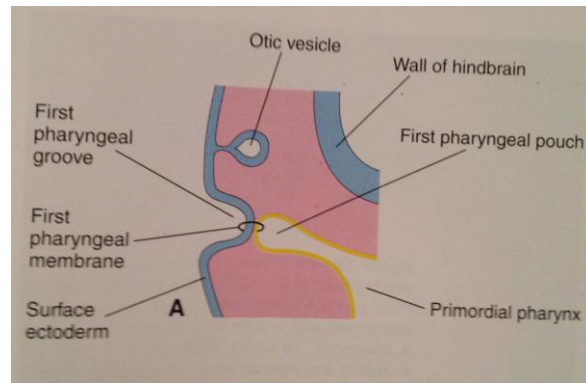
- Malleus Head and neck (**Mesoderm**)
- Anterior Malleal ligament,
- Incus Body and Short process (**Mesoderm**)

2. Second Branchial Arch (Reichert's cartilage):

- Handle of the Malleus
- Long process and Lenticular process of the incus
- Stapes (except vestibular part of footplate and Annular ligament from Otic capsule)

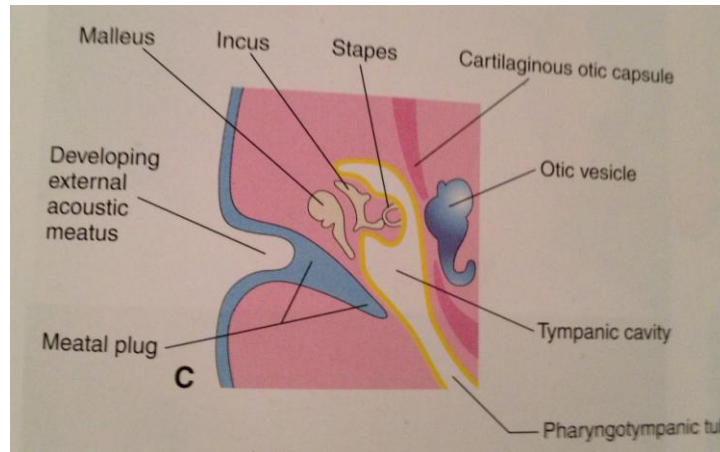


- Upper parts of ossicles derived from 1st ARCH
- Lower parts of ossicles derived from 2nd ARCH



Development of External Ear:

- **External acoustic meatus** develop from **1st pharyngeal GROOVE**
- Ectodermal cells at the bottom of it proliferate to form a solid epithelial plat (**Meatal plug**)



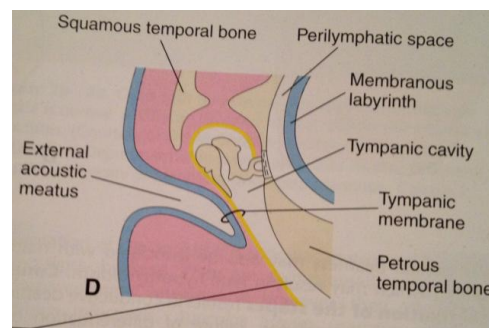
- Late in fetal period central part of the plug degenerate forming a cavity that becomes the internal part of the external acoustic meatus

Degeneration of Plug starts at **7th month** from Medial to Lateral

Failure of recannulization results in aural atresia, and may be normal boney but atresia in cartilage only - Canal cholesteatoma

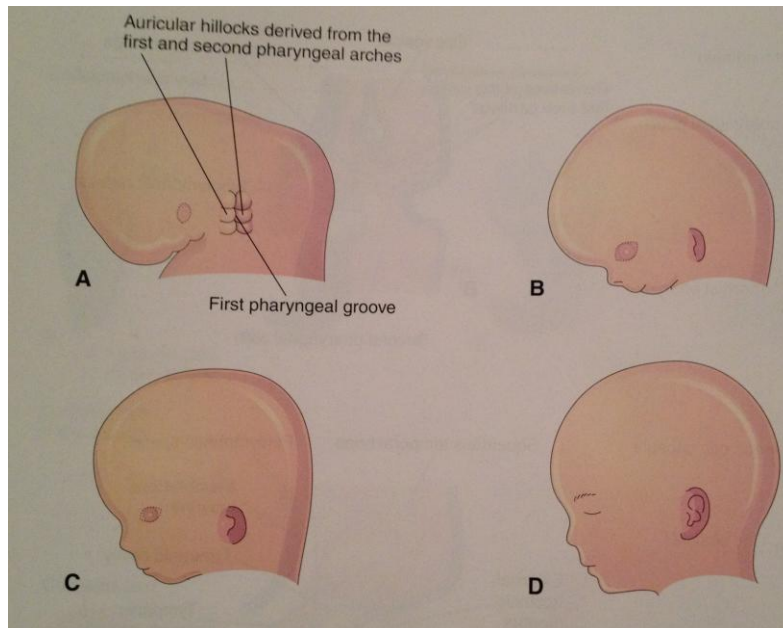
- The meatus short at birth
- **EAC reach Adult LENGTH by 9th year**
- **Tympanic membrane** from **1st pharyngeal MEMBRANE** which separate the 1st groove from 1st pouch.

1. Ectoderm of the 1st GROOVE form Squamous outer layer of TM (thin skin)
2. Mesoderm from 1st & 2nd ARCH form collagenic fibers between the 2 layers.
3. Endoderm of 1st POUCH form inner mucosal layer (ciliated columnar)



Auricle

- Develops from 6 Mesenchymal proliferations in the **1st & 2nd pharyngeal ARCHES (Auricular hillocks)**
- It surrounds the 1st pharyngeal Groove
- Initially, Auricle begin to develop low in the base of the neck
- As the mandible develop it move to its normal position (more lateral and cranial).



- **Tragus** develops from the **Tubercle of the 1st ARCH**
- **Rest of the pinna** develops from the remaining **5 Tubercles of the 2nd ARCH.**

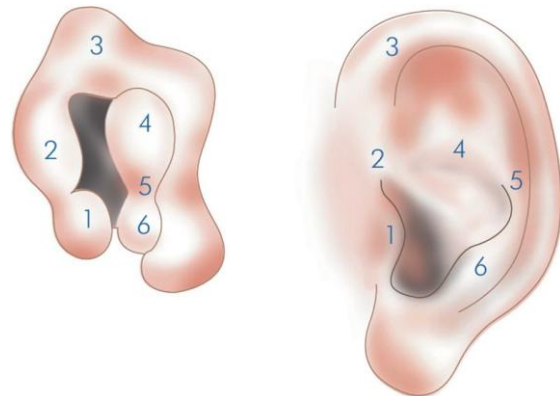
- **Pasha:**

1. First Branchial Arch:

- Hillock 1-3:
- 1. Tragus
- 2. Helical crus
- 3. Helix

2. Second Branchial Arch:

- Hillock 4-6:
- 4. Antihelix crus
- 5. Antihelix
- 6. Lobule and antitragus



- **Ear lobule is the last to develop**

Faulty fusion between 1st and 2nd ARCHs tubercles causes **Preauricular sinus or cyst** which is commonly seen between the tragus and crus of helix.

- By 20th week, Pinna achieves **Adult SHAPE**

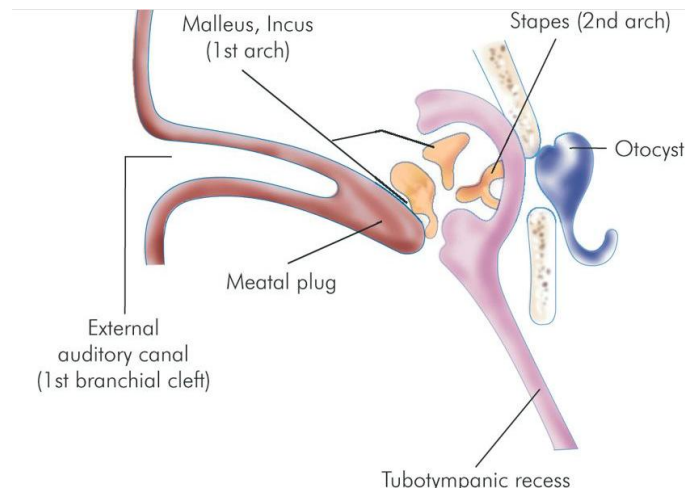


Table 1-3. Timing of development of the ear in the week of gestation*

Development	Pinna	Meatus	Middle ear	Vestibular labyrinth	Cochlea
Begins	6th	8th	3rd	3rd	3rd
Completes	20th	28th	30th	20th	20th

*Source: Gulya, A.J. Developmental Anatomy of the Ear. In Glasscock and Shambaugh ed. Surgery of the Ear. Philadelphia: W.B. Saunders Company, 1990.

Temporal bone:

- Derived from 4 separate morphological elements fuse together
1. Tympanic bone.
 2. Squamous bone.
 3. Petromastoid complex
 4. Styloid process

Some Anomalies

Inner ear:

(i) *Sheibe's dysplasia.*

- Most common inner ear anomaly.
- *Cochleosaccular dysplasia.*
- Dysplasia is seen in the cochlea and saccule.
- Bony labyrinth is NORMAL.
- Superior part of membranous labyrinth (utricle and semicircular ducts) is also NORMAL.
- Inherited as an autosomal recessive non-syndromic trait.

(ii) *Alexander's dysplasia.*

- Affects only the basal turn of membranous cochlea.
- Only high frequencies are affected.
- Residual hearing is present in low frequencies and can be exploited by amplification with hearing aids.

(iii) *Bing-Siebenmann dysplasia.*

- Complete absence of membranous labyrinth.

(iv) *Michel aplasia.*

- Complete absence of bony and membranous labyrinth.
- Petrous apex is absent.
- External and middle ears may be completely unaffected.
- No hearing aids or cochlear implantation can be used.

(v) *Mondini's dysplasia.*

- Only basal coil is present or cochlea is 1.5 turns.
- Absence of osseous spiral lamina.
- Incomplete partition between the scalae
- Unilateral or bilateral.
- May be seen in Pendred, Waardenburg, Branchio-oto-renal, Treacher-Collins and Wildervanch syndromes.

(vi) *Enlarged vestibular aqueduct.*

- Vestibular aqueduct is enlarged (>2 mm)
- Endolymphatic sac is also enlarged and can be seen on T₂ MRI.
- Early onset sensorineural hearing loss, which is progressive.
- Vertigo may be present.
- Perilymphatic fistula may occur.

(vii) *Semicircular canal malformations.*

- Both superior and lateral or only lateral semicircular canal malformations may be seen.
- They can be identified on imaging techniques

External ear:

- Auricular appendages (skin tags)
- Low-set slanted ear
- Absence of auricle
- Microtia
- Preauricular sinus
- Atresia of external acoustic meatus

Middle ear:

- Congenital fixation of stapes

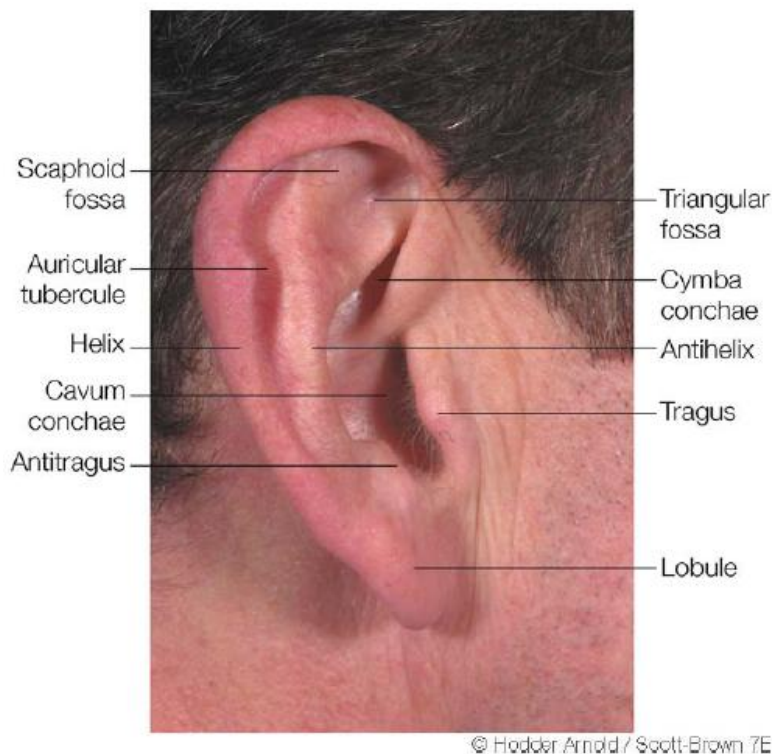
Anatomy of External Ear

- Consists of:

1. Auricle or pinna
2. External acoustic canal
3. Tympanic membrane

1. Auricle or Pinna:

- Projects at a variable angle from the side of the head
- External ear is less than 2-3 cm from the head,
- At an angle of less than 25 degrees from the side of the head.
- Function in collecting sound.
- Lateral surface of the auricle has characteristic prominences and depressions different in every individual even among identical twins.
- This unique pattern is comparable to fingerprints.

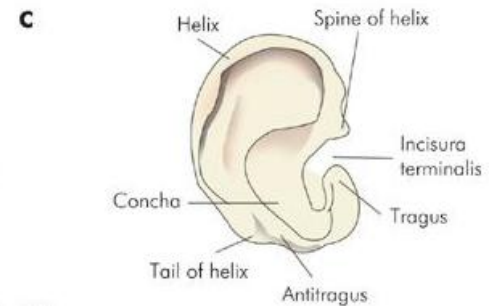


- The medial (cranial) surface of the auricle has elevations corresponding to the depressions on the lateral surface, and possesses corresponding names, for example the eminentia conchae.
- Post-auricular sulcus is the depression behind the ear next to the head

- Auricle is formed from **Elastic fibrocartilage** and is a continuous plate Except for a narrow gap between the tragus and the anterior crus of the helix, where it is replaced by a dense fibrous tissue band (**Incisura terminalis**) which is site for an endaural incision because it will not cut through cartilage.



- Lobule lies below the antitragus and is soft, also composed of fibrous and adipose tissue (no cartilage).
- Cartilage of auricle extends about 8 mm down the ear canal to form its Lateral third of EAC



- Cartilage of the auricle is covered with perichondrium from which it derives its supply of nutrients (**cartilage itself is avascular**).
- Stripping the perichondrium from the cartilage, as occurs following injuries that cause haematoma, can lead to cartilage necrosis and formation of Crumpled up 'boxer's ears' cowlflower ear.
- Skin of the pinna is Thin and closely attached to the perichondrium on the lateral surface
- On the medial (cranial) surface, there is a definite subdermal adipose layer that allows dissection during pinnaplasty surgery.
- Skin of the auricle is covered with **Fine hairs** and, most noticeably in the concha and the scaphoid fossa, there are **sebaceous glands** opening into the root canals of these hairs.
- On the tragus and intertragic notch **coarse, thick hairs** may develop in the middle-aged and older male.

- **Extrinsic Ligaments of Auricle:**

- Connects Cartilage of the auricle Temporal bone.

1. Anterior ligament:

- From the tragus and from a cartilaginous spine on the anterior rim of the crus of the helix to the root of the zygomatic arch.

2. Posterior ligament:

- From medial surface of the concha to the lateral surface of mastoid prominence.

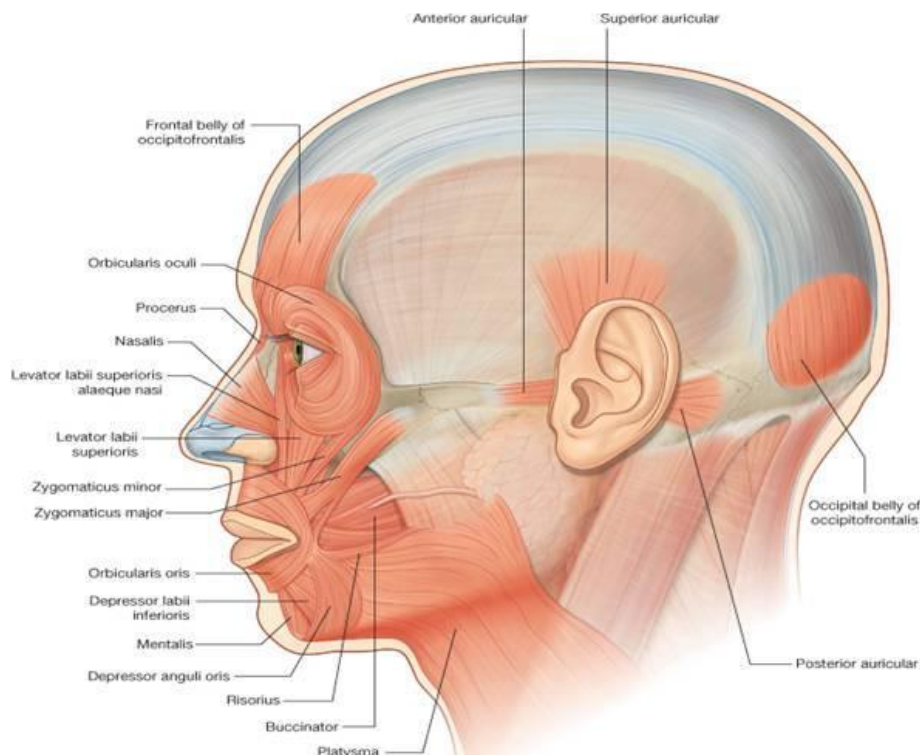
- **Intrinsic Ligaments of Auricle:**

- Connect various parts of the cartilaginous auricle.
- Connects between helix and tragus and another runs from the antihelix to the posteroinferior portion of the helix.

- **Extrinsic Muscles of Auricle:**

- 3 muscles radiate out from the auricle to insert into the epicranial aponeurosis.
- Auricularis anterior, superior and posterior.
- Supplied by Facial nerve CN-VII

Muscles of the External Ear				
Auricularis anterior	Epicranial aponeurosis	Auricle	Facial nerve	Small amount of auricular movement in some individuals
Auricularis superior	Epicranial aponeurosis	Auricle	Facial nerve	Small amount of auricular movement in some individuals
Auricularis posterior	Epicranial aponeurosis	Auricle	Facial nerve	Small amount of auricular movement in some individuals



- [Intrinsic Muscles of Auricle:](#)
- 6 in number.
- Small, inconsistent and without useful function.

[Blood supply for the pinna:](#)

- Branches of External carotid Artery.

1. Posterior auricular Artery:

- [The dominant artery.](#)
- Supply:
 - o Medial surface (except the lobule).
 - o Concha
 - o Middle and lower portions of the helix.
 - o Lower part of the antihelix.

2. Anterior Auricular Artery:

- Branch from Superficial Temporal artery.
- Supply:
 - o Upper portions of the helix
 - o Antihelix
 - o Triangular fossa
 - o Tragus
 - o Lobule.

3. Small auricular Artery:

- Branch from Occipital artery.
- Assist the posterior auricular in supplying the medial surface.

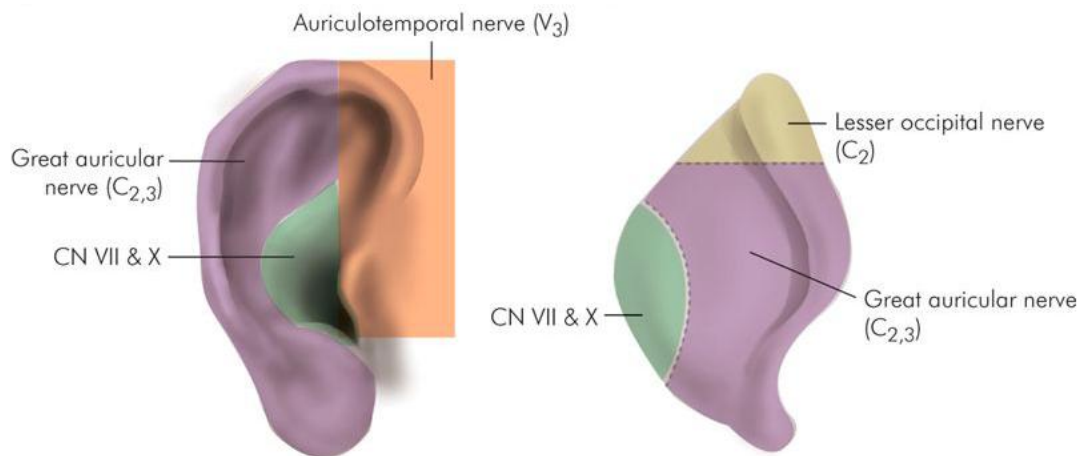
- [The lymphatic drainage:](#)

- Posterior surface --> Lymph nodes at the mastoid tip
- Tragus and from upper part of the anterior surface to --> Preauricular nodes
- Rest of auricle --> Upper deep cervical nodes.

- Nerves supply of auricle:

Table 225.1 Sensory innervation of the auricle.

Nerve	Derivation	Region supplied
Greater auricular	Cervical plexus C2, 3	Medial surface and posterior portion of lateral surface
Lesser occipital	Cervical plexus C2, 3	Superior portion of medial surface
Auricular	Vagus X	Concha and antihelix
		Some supply medial surface (eminetia concha)
Auriculotemporal	Vc mandibular	Tragus, crus of helix and adjacent helix
Facial VII		Probably supplies small region in the root of concha

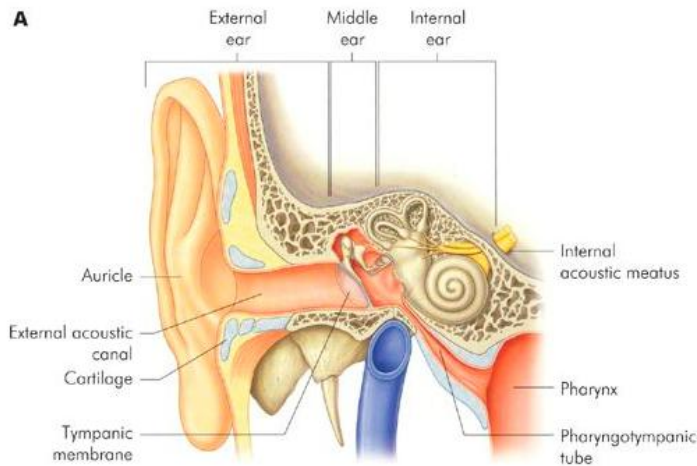
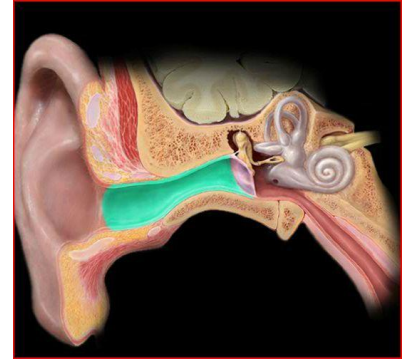


Dhingra: Diseases of Ear Nose and Throat, 5th Edition
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Figure 1.3 Nerve supply of pinna. (A) Lateral surface of pinna. (B) Medial or cranial surface of pinna.

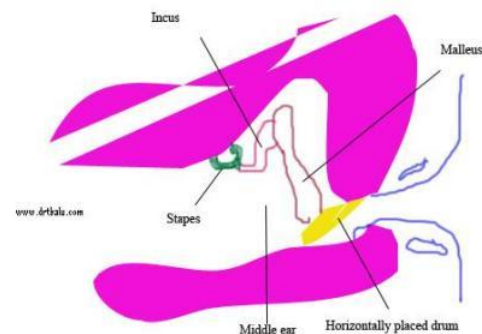
- Pinna can be source of several graft materials for the surgeon.
- Cartilage from the tragus, perichondrium from the tragus or concha, and fat from the lobule are frequently used for reconstructive surgery of the middle ear.
- Conchal cartilage has also been used to correct the depressed nasal bridge while the composite grafts of the skin and cartilage from the pinna are sometimes used for repair of defects of nasal ala.

2. External auditory canal

- Extends from Concha of auricle to Tympanic membrane.
- 2.4 cm Long.
- Cartilaginous in Lateral One-third.
- Bony in Medial two-thirds.
- In Adults, it is Not a straight tube.
- Outer part is directed:
Upwards, backwards and medially
- Inner part is directed:
Downwards, forwards and medially.
- To examine tympanic membrane, the pinna has to be Pulled upwards, backwards and laterally so as to bring the two parts in alignment.



- In Neonate,
- Tympanic portion of temporal bone is not yet developed.
- No bony external meatus
- Tympanic membrane is more horizontally placed.
- Auricle must be gently drawn downwards and backwards for the best view of the tympanic membrane.



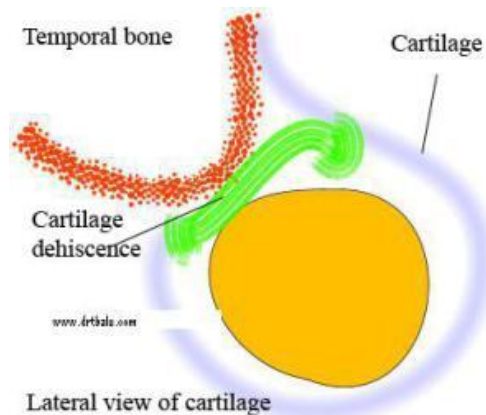
- Antero-inferior wall of EAC is slightly longer (31 mm) than Postero-superior wall (25 mm) because of Antero-inferior inclination of the ear drum.

EAC is divided into two parts:

1. Cartilaginous
2. Bony.

Cartilaginous Part:

- Outer 1/3
- 8 mm long.
- Continuation of pinna cartilage .
- Surrounds by incomplete cylinder of cartilage which is deficient in its superior portion.
- This defect is bridged by dense fibrous tissue that is attached to the Squamous portion of temporal bone.
- Cartilaginous canal is attached to rim of the bony canal by fibrous bands.
- Constriction at junction of the cartilage and bony part (1st constriction)



- Fissures of Santorini:
 - o 2 horizontal fissures located Antero-inferiorly in the cartilaginous portion.
 - o Render more flexibility to the external canal.
 - o Lymphatic channels that connect the lateral cartilaginous EAC to the parotid and glenoid fossa region
 - o Allow infections and tumor to pass between the external canal and the parotid gland.
- Skin covering the cartilaginous canal:
 - o Thick
 - o Contains ceruminous and sebaceous glands.
 - o Contains Hair which is only confined to the outer canal (most numerous at lateral end of the canal, less numerous medially and totally absent from the bony cartiagenous junction).
 - o Furuncles are seen only in outer third.

Bony Part

- Medial 2/3
- 16 mm long
- Composed of a complete cylinder of bone extending laterally from the ear drum.
- Narrower than cartilaginous portion and becomes smaller closer to tympanic membrane.
- Anterior and Inferior walls:
 - o Tympanic portion of temporal bone.
- Posterior wall:
 - o Mastoid portion of temporal bone.
- Superior wall:
 - o Squamous portion of temporal bone.
- Medial end of the bony canal is marked by a groove, tympanic sulcus, which is absent superiorly.
- **Tympanic bone makes up the greater part of the canal**, and also carries the sulcus.
- 2 suture lines in the canal wall:

1. TympanoSquamous:

- Anteriorly
- Transmits Auricular branch of Glossopharyngeal nerve (**Jacobson's nerve**)

2. TympanoMastoid:

- Posteriorly
- Transmits Auricular branch of vagus Nerve (**Arnold's nerve**).
- Evident in the posteriorinferior portion of the canal wall during surgical procedures like elevation of the tympanomeatal flap.

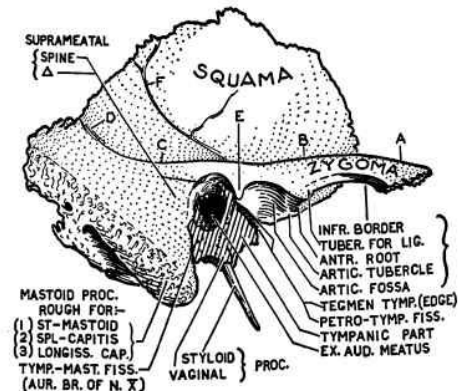
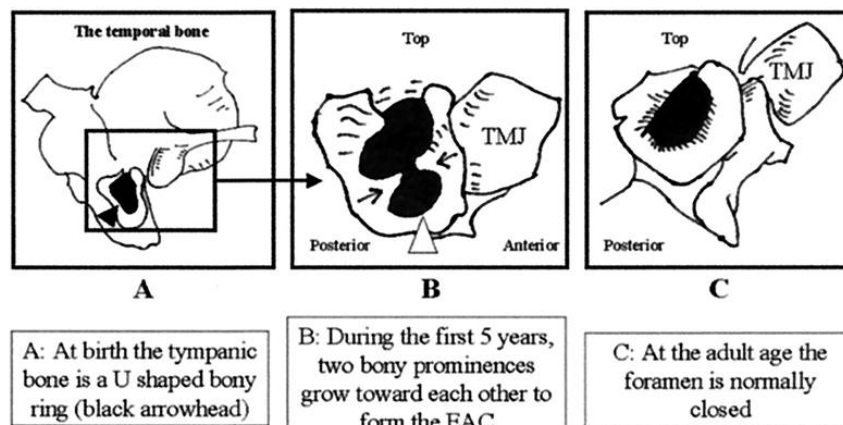


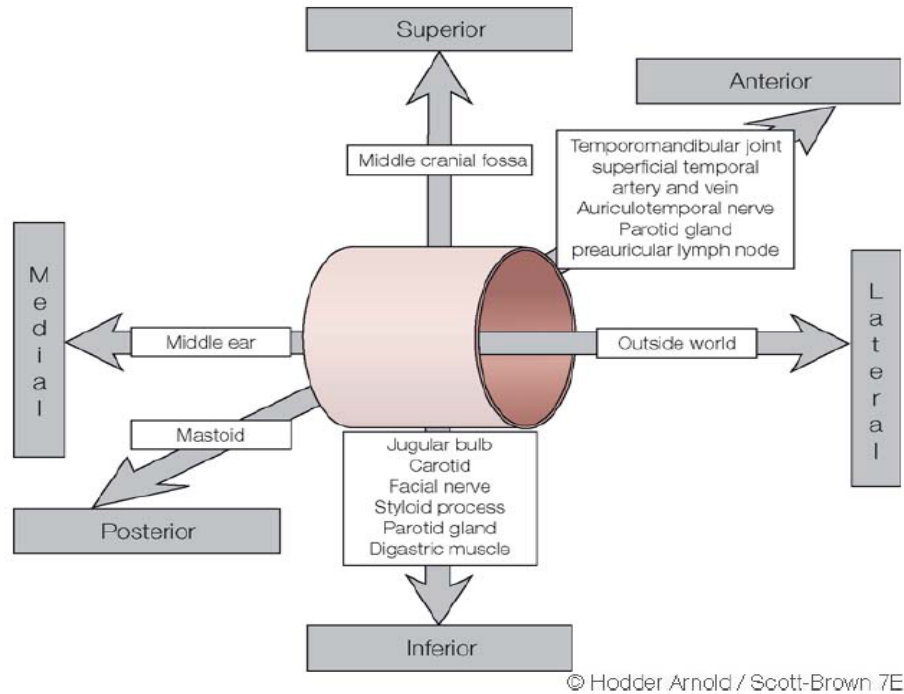
FIG. 835. Temporal Bone—lateral aspect



- Henle's spine:
 - Projection produced by temporal bone in Postero-superior aspect of external auditory canal.
 - Important landmark for mastoid surgery
- Isthmus:
 - Narrowing in the bony canal.
 - 6 mm lateral to tympanic membrane
 - *2nd constriction.*
 - **Narrowest part of EAC.**
- Anterior recess:
 - Recess located in Antero-inferior part of the deep meatus beyond the isthmus
 - Acts as a cesspool for discharge and debris in cases of external and middle ear infections.
- Condyle of the mandible and glenoid fossa produce a convexity in Anterior bony canal wall limiting the visualisation of the ear drum.
- This prominence and the Isthmus predispose foreign body entrapment in Antero-inferior portion of the medial end of the external canal and are difficult to remove.
- Foramen of Huschke:
 - Deficiency in the Antero-inferior part of the bony canal.
 - Found in children up to 4 years old or sometimes in adults.
 - Connects the bony EAC to parotid and glenoid fossa.
 - Permitting infections to and from the parotid)
- Skin covering the bony canal:
 - Thin
 - Devoid of hair and glands.
 - Continuous over the tympanic membrane.
- **Skin lining the whole external canal is the only keratinising epithelium that lacks Eccrine glands.**

Relations of External Acoustic Meatus:

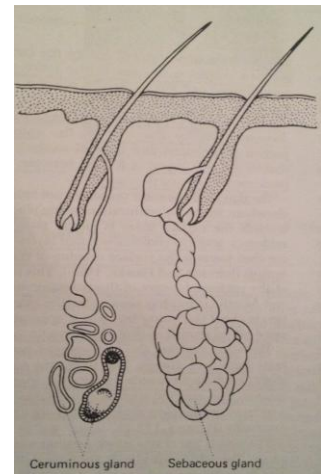
- Superiorly: Middle cranial fossa
 - Posteriorly: Mastoid air cells and the facial nerve
 - Inferiorly: Parotid gland
 - Anteriorly: Temporomandibular joint
 - Postero-superior part of deeper canal near the tympanic membrane is related to the mastoid antrum.
- ("Sagging" of this area may be noticed in acute mastoiditis)**



- **Nerve supply to External Auditory Canal:**
- Anterior wall and roof:
 1. Auriculotemporal (V_3) of Trigeminal nerve. (CN-V).
- Posterior wall and floor:
 1. Auricular branch of vagus (CN-X) (**Arnold's nerve**).
 2. Facial nerve also supply posterior wall.
 - Hitzelberger sign: Numbness at facial nerve area in patient with acoustic neuroma.
- **Blood supply of External Auditory Canal:**
- Branches from External carotid Artery:
 - 1. Auricular branch of Superficial Temporal Artery:**
 - Supply Roof and Anterior portion of the canal.
 - 2. Deep auricular Artery:**
 - Branch of 1st part of maxillary artery.
 - Supply Anterior wall of the canal and outer surface of TM
- **Vein drain in the external jugular, maxillary & pterygoid plexus**
- **Lymphatic drainage follows the auricle.**

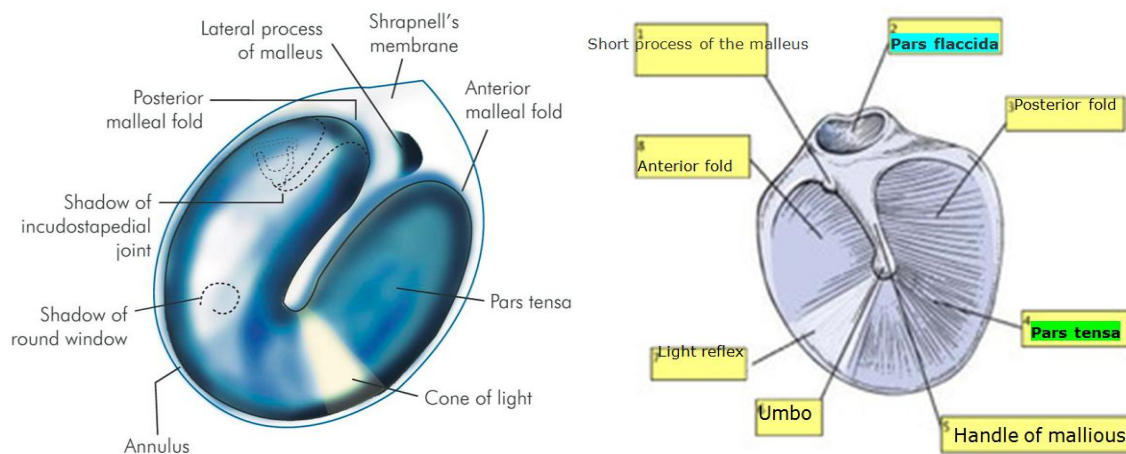
Physiology:

- Properties of External Auditory canal:
 - Lateral growth of the epidermis with the consequence that layers of keratin are shed towards the surface opening of the external meatus.
 - Rate of migration 0.05 - 0.1 mm/day.
 - Same thing for epidermal layer of TM
- Volume of the external canal is about **0.85 ml²**.
- Normal flora of the outer ear canal is predominantly **Gram-positive Bacteria:** Staphylococcus epidermidis, Staphylococcus aureus, Diphtheroids (Corynebacterium).
- **Glands of External Auditory canal:**
 1. Ceruminous glands:
 - Modified apocrine sweat glands.
 - Simple coiled tubular gland
 - Open into the root canal of the hair follicles
 - Produce a watery, white secretion that slowly darkens
 - Lay deep in the dermis
 2. Sebaceous gland
 - Simple or branched alveolar glands
 - Typical like elsewhere.
 - Secrete sebum
 - Form their secretion by passive breakdown of cells.
- **Wax:** Mixture of sebum (from sebaceous glands) + watery secretions (from ceruminous glands) + Desquamated epithelium.
 - Two distinct forms:
 1. Dry wax: Yellowish or gray
 2. Wet wax: Yellowish brown
- Functions of Wax:
 - Wax has acidic PH
 - Bacteriostatic properties.
 - Contains bactericidal enzymes, amino acids, and immuno globulins which helps to prevent infection.
 - Protecting tympanic membrane.
 - Keep moisture of ear canal.
 - has a self cleansing action
 - Traps the dirt and dust and move it outwards by migration of skin from the lateral part of tympanic membrane to outside.
 - With aging, Cerumen becomes harder, drier, less likely to be cleared due to atrophy of modified apocrine sweat glands.



3. Tympanic membrane

- At the medial end of the external auditory meatus.
- Between EAC and middle ear.
- Forms majority of the lateral wall of the tympanic cavity.
- Obliquely set
- Postero-superior part is more lateral than Antero-inferior part (EAC slightly longer Anteriorly)
- Oval in shape, being broader above than below.
- Forming an angle of about 55° with the floor of the meatus.
- Height 9-10 mm
- Width 8-9 mm
- Thickness 0.1 mm
- Surface Area: 70-80 mm²
- Vibrating surface area: 55 mm²
- Most of the circumference is thickened to form a fibrocartilaginous ring, the **tympanic annulus**, which sits in a groove in the tympanic bone, the tympanic sulcus.
- The sulcus does not extend into the notch of Rivinus at the roof of the canal, which is formed by part of the squama of the temporal bone.
- From the superior limits of the sulcus, the annulus becomes a fibrous band which runs centrally as anterior and posterior malleolar folds to the lateral process of the malleus
- Handle of malleus is clearly visible within the tympanic membrane.



- This leaves a small, triangular region of tympanic membrane above the malleolar folds within the notch of Rivinus, called the **pars flaccida** (Shrapnel's Membrane), which does not have a tympanic annulus at its margins.

- **Pars Tensa** forms the rest of the tympanic membrane and is concave towards the ear canal.
- Each segment is slightly convex between the lateral attachment of the annulus and the centre of the membrane where the tip of the malleus handle is attached at the umbo.
- Both the pars tensa and pars flaccida comprise 3 layers.
 1. Outer epithelial layer:
 - Epidermis.
 - Continuous with the skin of the external meatus.
 2. Middle fibrous layer:
 - Lamina propria.
 - Outer radiating fibrous layer.
 - Inner circular fibrous layer.
 3. Inner mucosal layer:
 - Continuous with the lining of the tympanic cavity)
- In the pars flaccida, the lamina propria is less marked (thin) and the orientation of the collagen fibers seems random.

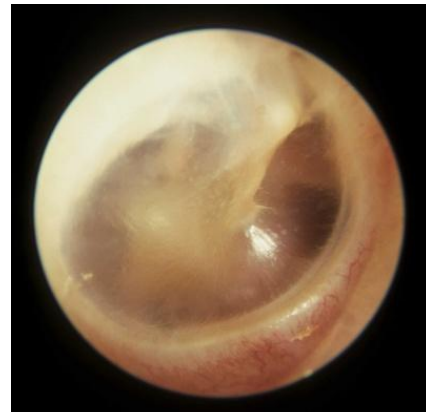
Normal TM:

1. Color:
 - Pearly grey.
 - Shiny.
 - Translucent.
 - No bulging or retraction.
2. Consistency:
 - Smooth.
3. Landmarks:
 - Cone-shaped light reflection of the otoscope light
 - Short process of malleus
 - Handle of malleus
 - Umbo
 - Anterior and posterior folds

Left TM



Right TM

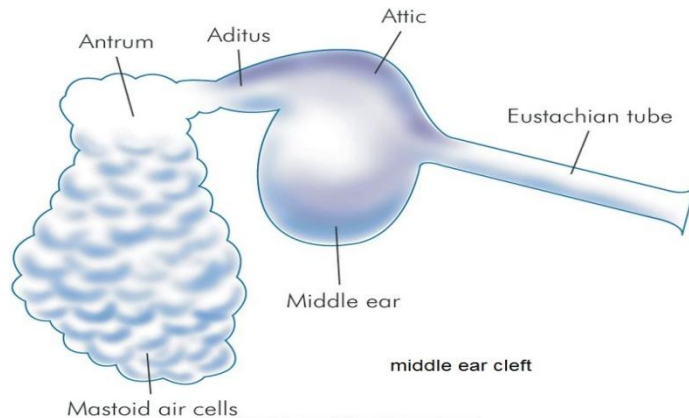


- Blood supply of the tympanic membrane:
 - From branches supplying both the external auditory meatus and the middle ear.
 - Interconnect through extensive anastomoses within the connective tissue layer of the lamina propria.
- 1. Epidermal vessels** from the **Deep auricular branch of the maxillary artery** coming from the external auditory meatus,
- 2. Mucosal vessels** arise from **Anterior tympanic branches of the maxillary artery, stylomastoid branch of the posterior auricular artery** and **probably from the middle meningeal artery.**
- Nerve supply of TM:
 1. Anterior half of lateral surface:
 - Auriculotemporal (V_3) of Trigeminal Nerve (CN-V)
 2. Posterior half of lateral surface:
 - Auricular branch of vagus (CN X) (**Arnold's nerve**).
 3. Medial surface:
 - Tympanic branch of CN IX (**Jacobson's nerve**).

Anatomy of Middle Ear

- **Middle Ear Cleft:**

1. Middle ear
2. Eustachian tube
3. Aditus
4. Antrum
5. Mastoid air cells



- Lined by mucous membrane and filled with air within the temporal bone.
- Extends much beyond the limits of tympanic membrane which forms its lateral boundary and is sometimes divided into:

1. Epitympanum (Attic):

- o Area above the Pars Tensa, Medial to Pars Flaccida and scutum and lateral to Lateral SCC prominence.

2. Mesotympanum:

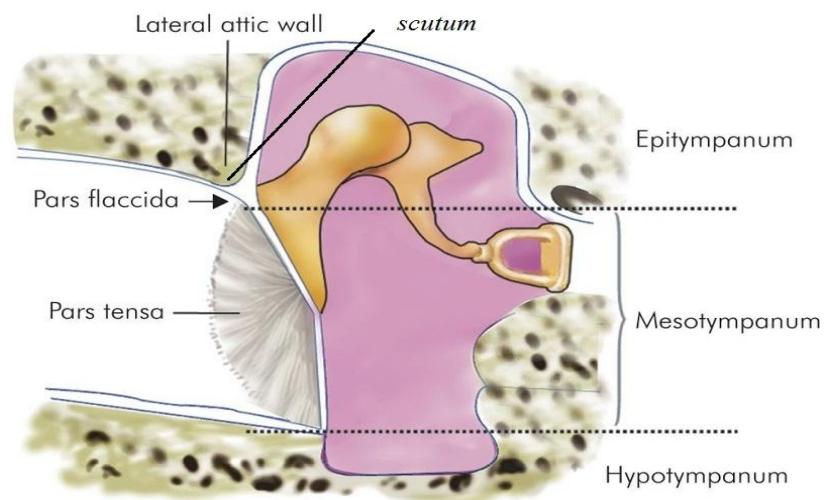
- o Area opposite to Pars Tensa.

3. Hypotympanum:

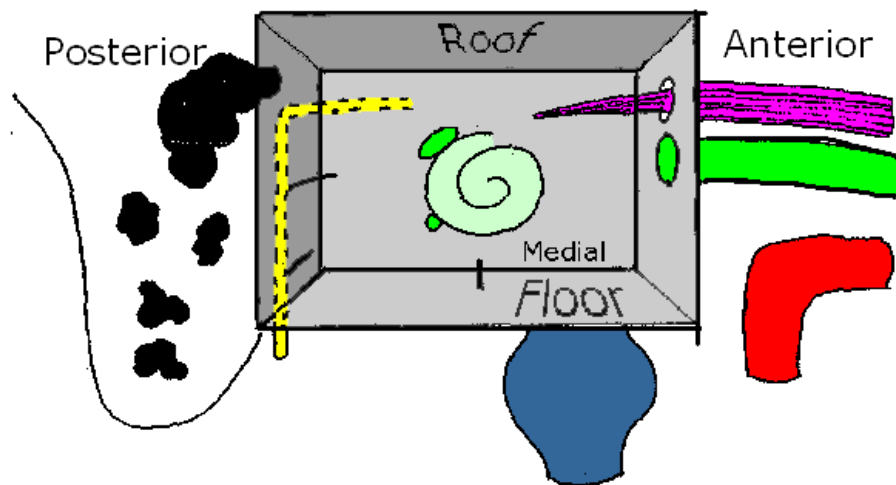
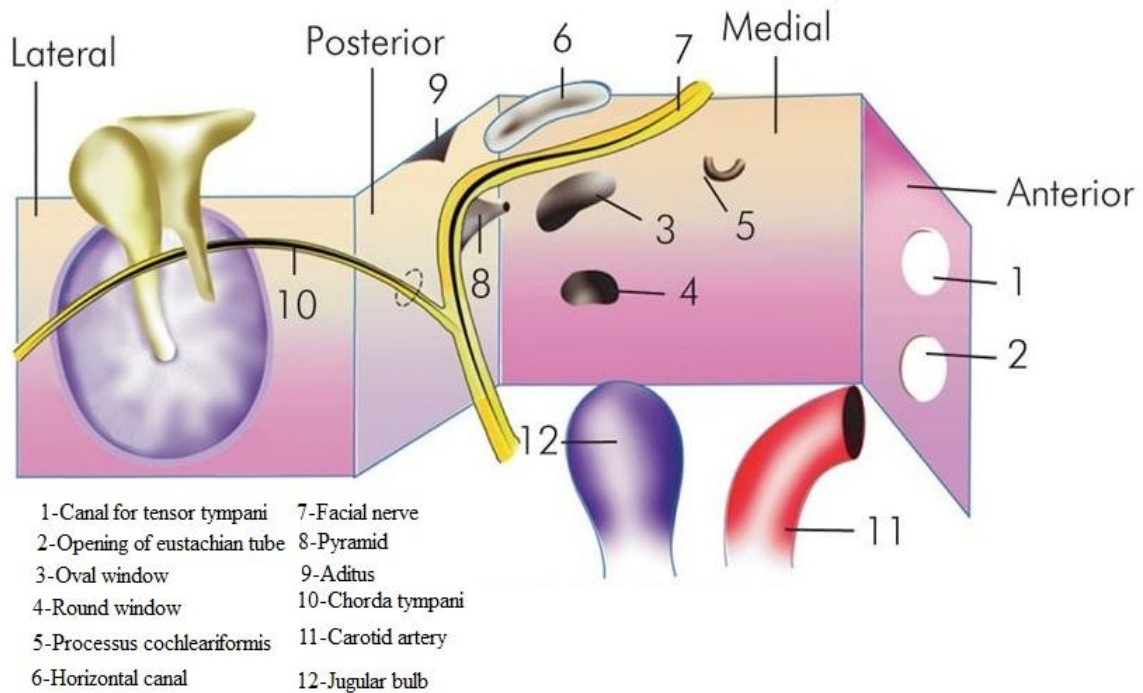
- o Area below the level of Pars Tensa.

4. Protympanum:

- o Area around the tympanic orifice of the Eustachian tube.

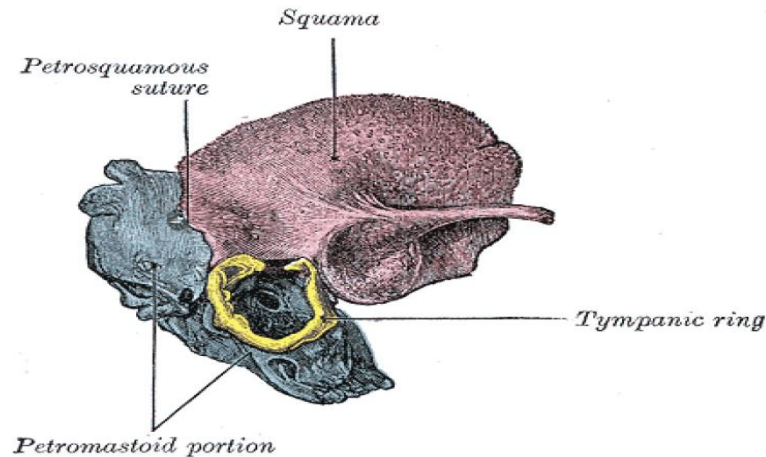


- Middle ear can be likened to a Six-sided Box with a Roof, Floor, Medial, Lateral, Anterior and Posterior walls



Roof

- Thin plate of bone called **Tegmen Tympani**.
- Separates tympanic cavity from the dura of **Middle cranial fossa**.
- Extends posteriorly to form the Roof of the **Aditus and Antrum**.
- Formed from both **Petrous and Squamous** portions of temporal bone with suture line in between known as petrosquamous suture line.
- PetroSquamous suture line:
 - o Unossified in the young and close in adult life.
 - o Provide a route of access for infection into the extradural space in children .
 - o Veins from the tympanic cavity pass through this suture line to the **Superior Petrosal Sinus**.

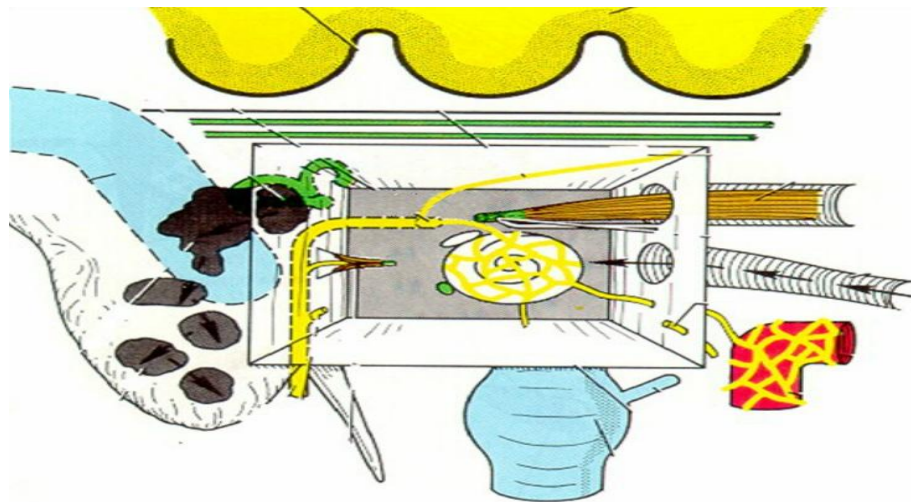


Floor

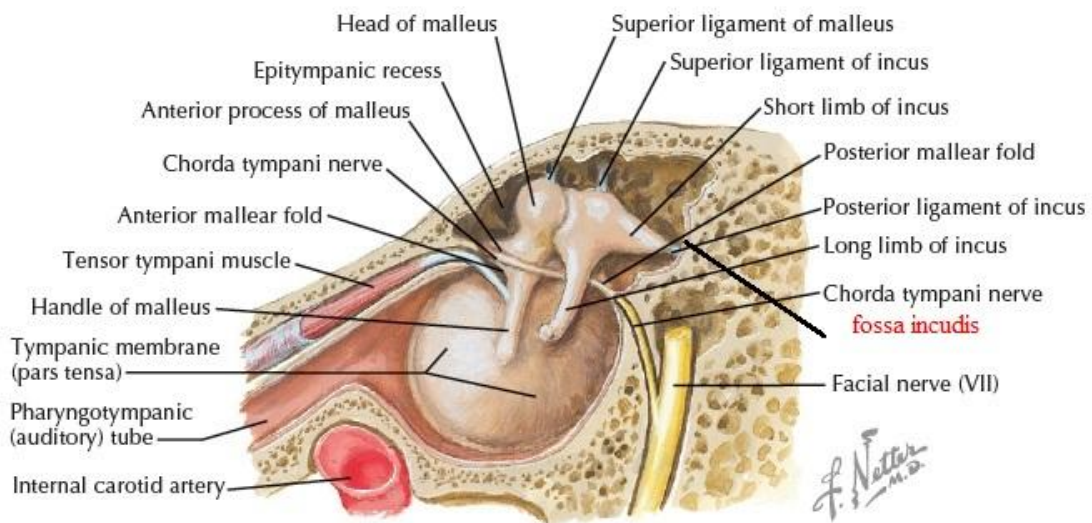
- Thin plate of bone.
- Narrower than the roof of the middle ear cavity
- Separates tympanic cavity from the **jugular bulb**.
- Its thickness can vary according to the height of the jugular fossa.
- Sometimes, it is congenitally deficient and the jugular bulb may then project into the middle ear; separated from the cavity only by **fibrous tissue & mucosa**.
- **Tympanic Canaliculus:**
 - o Small opening at junction of Floor and Medial wall of the cavity
 - o Allows the entry of **Tympanic branch of the Glossopharyngeal nerve** "**Jacobson's Nerve**" into the middle ear from its origin below the base of the skull.

Anterior wall

- o The narrowest wall because Medial and lateral walls converge anteriorly.
- o 5 openings perforates the Anterior wall.
- o **Lower 1/3 of Anterior wall:**
 - Thin plate of bone covering **Internal Carotid artery** as it enters the skull and before it turns Anteriorly.
 - Perforated by 3 openings:
 1. Superior and Inferior Caroticotympanic nerves carrying sympathetic fibers to the tympanic plexus.
 2. Tympanic branches of internal carotid artery.
- o **Middle 1/3 of Anterior wall:**
 - Perforated by 2 openings:
 1. Upper opening for the **Canal of Tensor Tympani Muscle** that subsequently runs along the medial wall of the tympanic cavity.
 2. Lower one for the **Eustachian tube** (oval and 5 x 2 mm in size)
- o **Upper 1/3 of Anterior wall:**
 - Pneumatized and may house the **Anterior Epitympanic sinus**.
 - **Epitympanic recess:**
 - o Small niche anterior to ossicular heads
 - o Can **hide residual cholesteatoma in canal wall up surgery**.



Lateral wall of tympanic cavity: medial (internal) view



Posterior Wall:

- Lies close to the mastoid air cells.

Aditus:

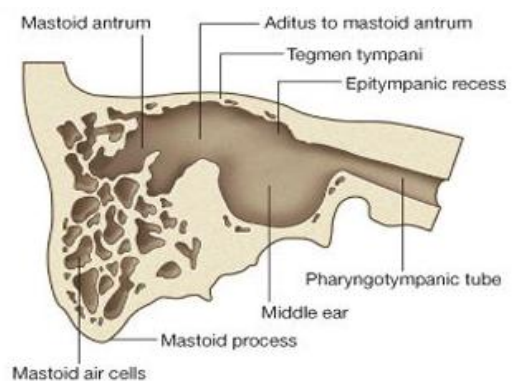
- o Opening through which Attic communicates with the antrum.
- o Lies above the pyramid, near the junction with the Roof of the middle ear.

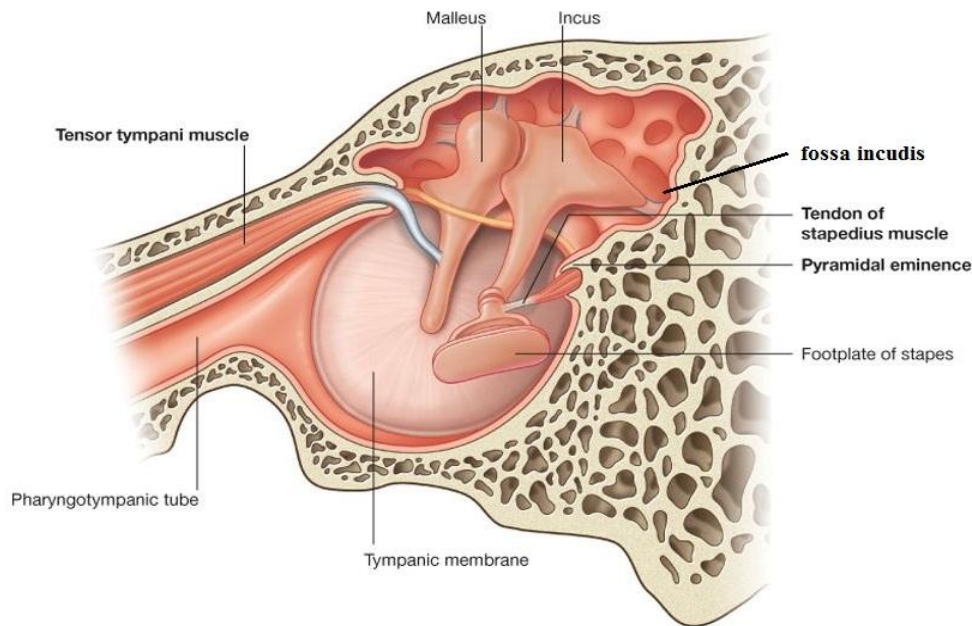
Fossa Incudis:

- o Small depression below the Aditus.
- o Houses:
 1. **Short process of Incus**
 2. **Short Incudal ligament.**

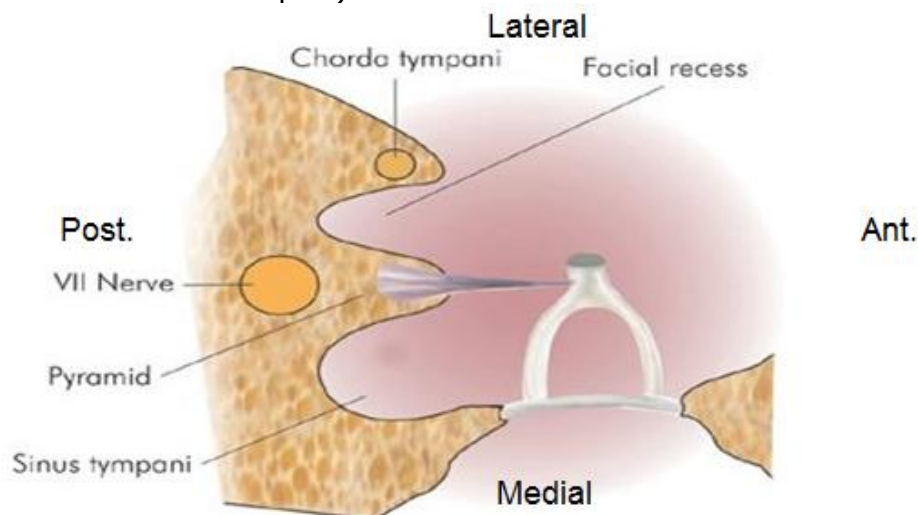
Pyramid:

- o Bony projection from the posterior wall with its apex pointing Anteriorly.
- o Below Fossa incudis.
- o Contains Tendon of Stapedius muscle to get attachment to the Neck of and Posterior crus of Stapes.



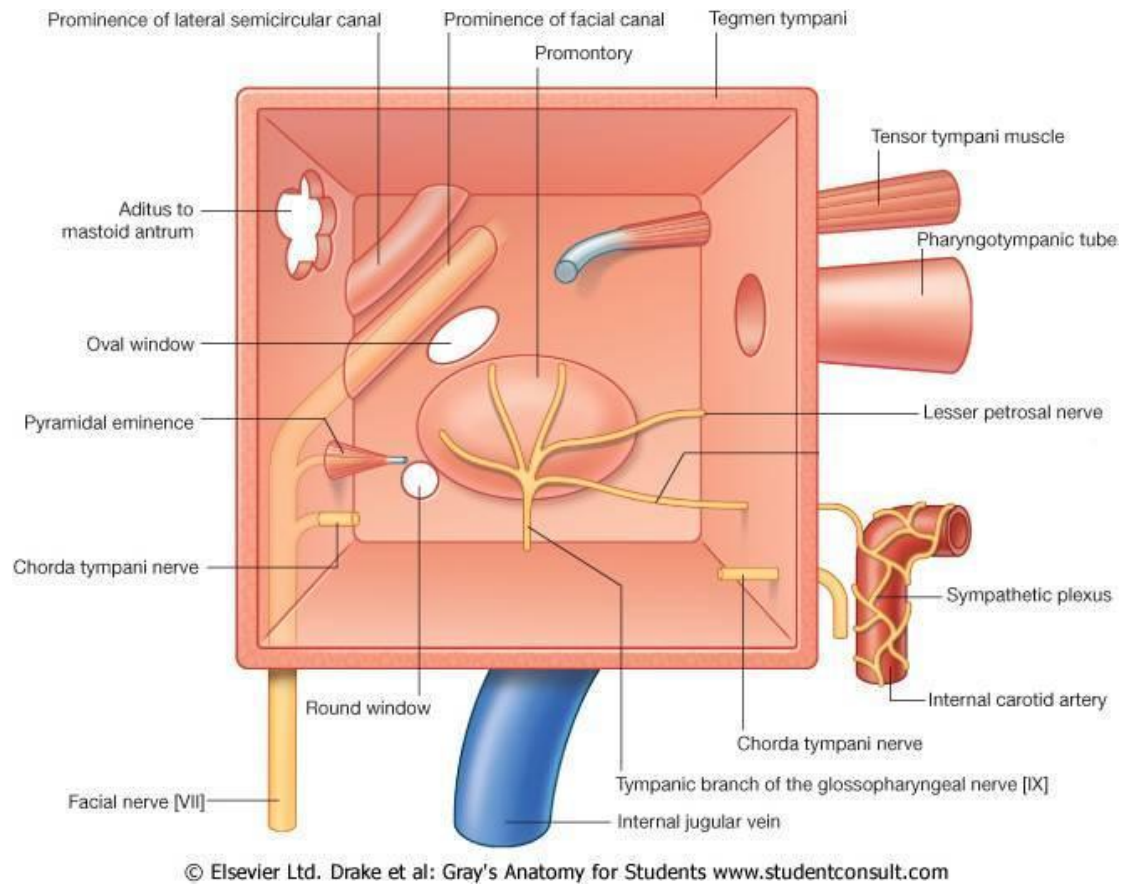


- **Facial Nerve:** Runs in the posterior wall just behind the **pyramid**.
- **Facial Recess:**
 - o Also called posterior sinus (supra pyramidal recess)
 - o Depression in the posterior wall **Lateral to Pyramid**.
 - o Bounded Medially by **Vertical part of Facial nerve (CN-VII)**
 - o Bounded Laterally by **Chorda tympani and tympanic annulus**.
 - o Bounded from Above by **Fossa incudis**.
 - o Surgically, facial recess is important, as direct access to the mesotympanum can be made through this into the middle ear without disturbing posterior canal wall (canal wall up or "intact canal wall technique").



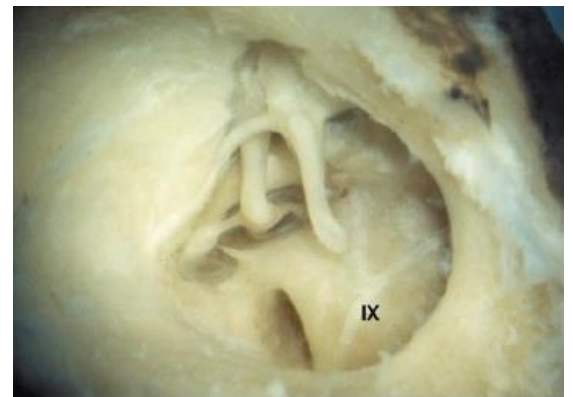
Facial recess lies lateral and sinus tympani medial to the pyramidal eminence and vertical part of the facial nerve.

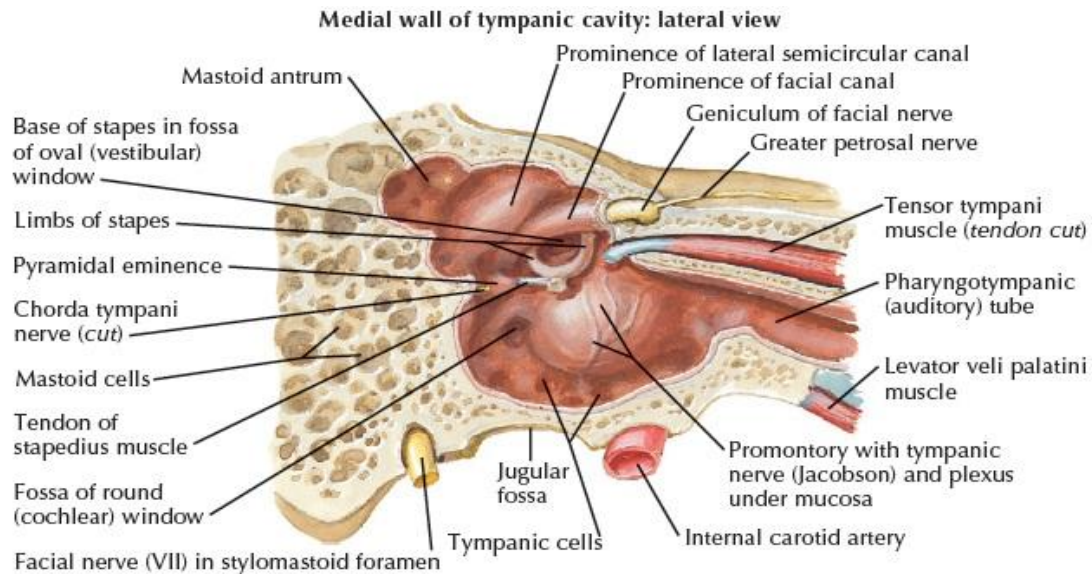
- **Facial canal:** Passes Supero-inferiorly immediately posterior to the middle ear until it terminates at the stylomastoid foramen.
- **Posterior cranial fossa and sigmoid sinus** located posterior to the posterior wall



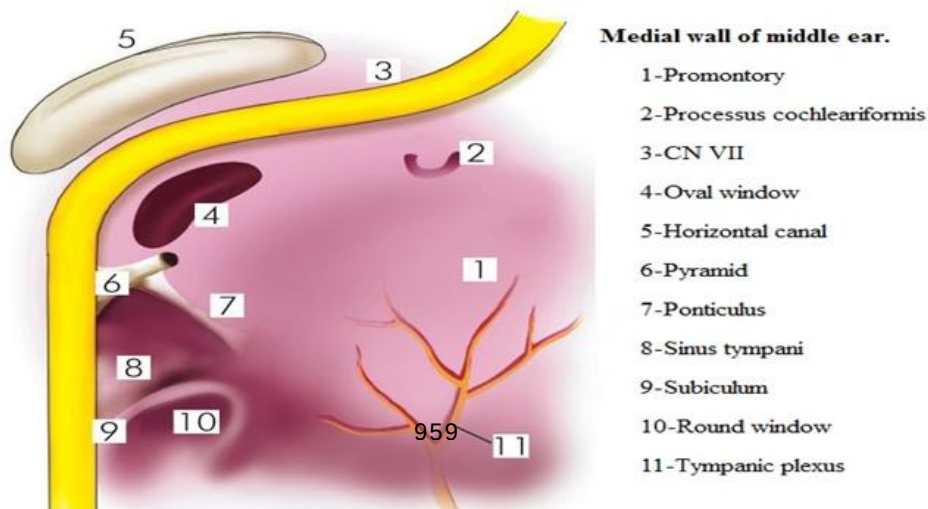
Medial wall

- Separates the tympanic cavity from the internal ear.
- **Promontory:**
 - o Most prominent portion in the medial surface due to the **basal coil of cochlea**
 - o Has small grooves on its surface containing the nerves which form **Tympanic plexus**.
 - o Sometimes the groove containing the **tympanic branch of the Glossopharyngeal nerve "Jacobson's Nerve"** may be covered by bone "small canal".
- **Oval window:**
 - o Also called Fenestra vestibuli.
 - o Located Posterior and Superior to the promontory.
 - o Connects tympanic cavity with vestibule
 - o Closed by foot plate of stapes and its surrounding annular ligament.
 - o Its size naturally varies with the size of the footplate, but on average it is **3.25 mm long and 1.75 mm wide**.
 - o The long axis of the fenestra vestibuli is **Horizontal**.
 - o Lies at the bottom of a depression or niche known as fossula that can be of varying width depending on the position of the facial nerve superiorly, and the prominence of the promontory inferiorly.



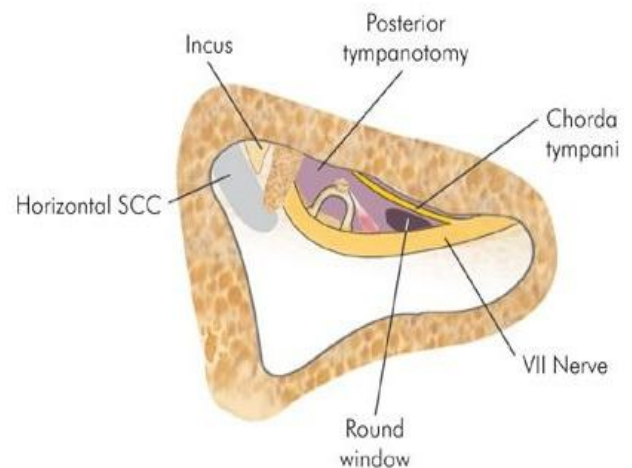


- Round window:
- Also called Fenestra cochlea.
- Lies **Inferior and posterior** to oval window niche.
- Round window niche is most commonly **Triangular in shape, with Anterior, Posterosuperior and Posteroinferior walls.**
- Posterosuperior and posteroinferior walls meet posteriorly leading on to sinus tympani.
- **Subiculum** is Posterior extension of promontory separating oval and round windows.
- Covered by the secondary tympanic membrane.
 - o Membrane is usually out of sight, obscured by the overhanging edge of the promontory forming the niche and mucosal folds within it.
 - o Roughly oval in shape, about 2.3 x 1.9 mm in dimension.
 - o Made up of Three layers: **Outer mucosal, Middle fibrous and Inner endothelial layer.**
 - o Does not lie at the end of Scala tympani but forms part of its floor.
 - o It tends to curve towards the Scala tympani of the basal coil of the cochlea, so that it is **concave** when viewed from the middle ear.
 - o Ampulla of Posterior SCC is the closest vestibular structure to this membrane.
 - o Nerve supplying Ampulla of Posterior SCC (**Singular nerve**) runs 1 mm behind and parallel to the posterior portion of the membrane
 - o It is a landmark for the position of the singular nerve during surgical procedures like singular neurectomy for treatment of intractable BPPV.



- **Facial nerve Canal:**
- Also called Fallopian canal.
- Runs Above promontory and oval window in an Antero-posterior direction.
- Behind the oval window, the facial canal starts to turn inferiorly as it begins its descent in the Posterior wall of the tympanic cavity
- Its bony covering may sometimes be congenitally dehiscent and the nerve may lie exposed making it very vulnerable to injuries or infection.
- Facial nerve canal is marked Anteriorly by **Processus Cochleariformis**
- It has a smooth rounded lateral surface that often has Microdehiscences and when the bone is thin or the nerve exposed by disease, there are **two or three straight blood vessels clearly visible along this line of nerve**. These are the only straight blood vessels in the middle ear and indicate quite clearly that the facial nerve is very close.

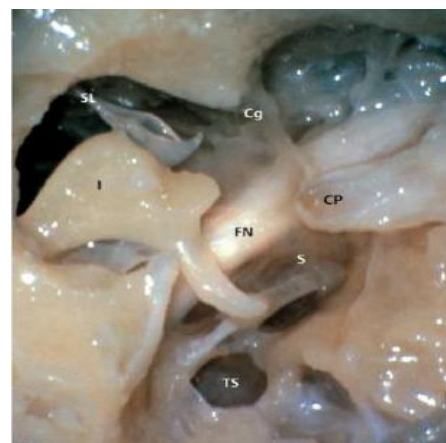
- **Prominence of Lateral SCC:**
- Above facial nerve canal.
- Forms Medial wall of the Epitympanum.
- Major feature of the Posterior portion of the Epitympanum, lying posterior and extending a little lateral to the facial canal.
- During a cortical mastoidectomy, the Triangular relationship of Lateral SCC, Short process of incus and Facial nerve is often quite helpful.



- In well aerated mastoid bones, labyrinthine bone over Superior SCC may be prominent, running at right angles to the lateral canal and joining it anteriorly at a swelling which houses the ampullae of the two canals.

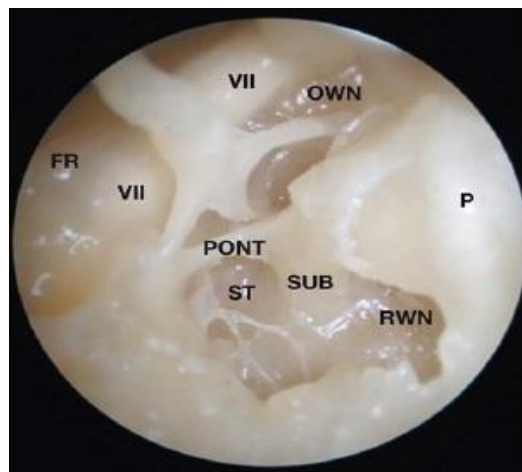
- **Processus Cochleariformis:**
- Hook-like projection just Anterior to the oval window.
- Tendon of Tensor Tympani takes a turn here to get attachment to the Neck of malleus.
- Cochleariform process also marks the level of the Geniculate ganglion of Facial nerve which is an important landmark for surgery of the facial nerve.

- **Cog Process:**
- Small bony bar Anterior and Superior to Cochleariform process which separates Anterior Epitympanum from rest of the attic.
- Facial Nerve runs **between** Cog process & cochleariform process.
- Geniculate ganglion lies deep and medial to cog.



Cg cog
Cp processus cochleariformis
I incus
S stapes
TS tympanic sinus

- **Sinus Tympani (Infra Pyramidal Recess):**
- Most constant depression present in Retrotympanic area.
- Posterior extension of mesotympanum and lies deep to both the promontory and the pyramid "facial nerve"
- Lies Medial to pyramid "facial nerve" .
- Bounded by the:-
 - o **Subiculum** Below.
 - o **Ponticulus** Above. "Arise from promontory above subiculum and runs to the pyramid on the posterior wall of the cavity"
- Occasions it can communicate with the mastoid air cells..
- The sinus can extend as far as 9 mm into the mastoid bone when measured from the tip of the pyramid.
- Its importance is that **cholesteatoma** (which has extended here from mesotympanum) can **hide** to which access is difficult.
- **Most Inaccessible site** in the middle ear and mastoid.

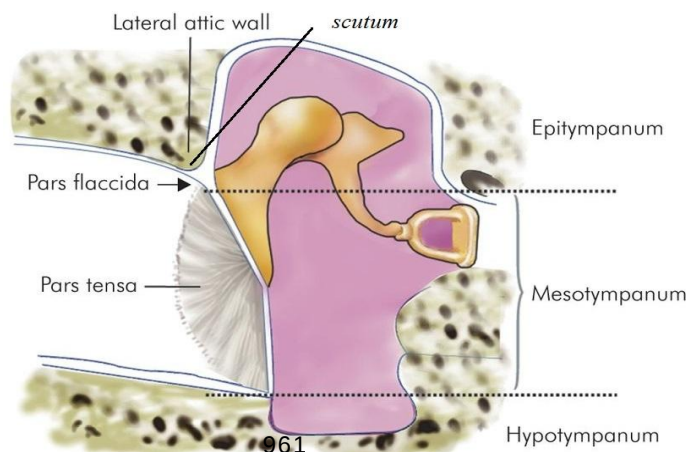


posterior medial wall of the tympanic cavity

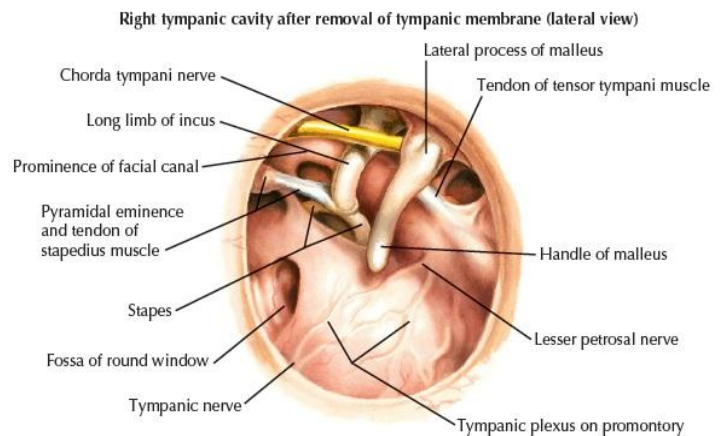
oval window niche (OVN).
round window niche (RWN).
promontory (P)
facial nerve (VII)
facial recess (FR)
The sinus tympani (ST)
ponticulus (PONT)
subiculum (SUB).

Lateral Wall:

- Separates middle ear from the external ear.
- Formed mainly by tympanic membrane, partly by the ring of bone into which this membrane is inserted.
- This ring of bone is incomplete at its upper part, forming a notch (notch of Rivinus), close to which are Three small openings.
- **Centrally:** Formed largely by **Tympanic membrane**, with the malleus attached to the membrane at the umbo
- **Superiorly:** Formed by **Scutum** "outer attic wall" bony lateral wall of Epitympanum
- **Inferiorly:** form by bony lateral wall of the **Hypotympanum**



- **Tympanic membrane:**
Semitransparent and forms a 'window' into the middle ear.
- It is possible to see some structures of the middle ear through the normal tympanic membrane (**Long process of Incus, incudostapedial joint and the round window**).



- **Chorda Tympani nerve:**
 - o Enters the middle ear through Posterior Canaliculus.
 - o Runs on the medial surface of the tympanic membrane between the handle of malleus and long process of incus, Above the attachment of tendon of tensor tympani.
 - o Lies along the tympanic membrane and malleus until exiting through the Anterior Canaliculus.
- **Scutum:**
 - o Thin bone portion and **easily eroded** by cholesteatoma, leaving a telltale sign on a high resolution Coronal CT scan.
- **Three openings present in medial surface of lateral wall of tympanic cavity:**

1. Posterior Canaliculus:

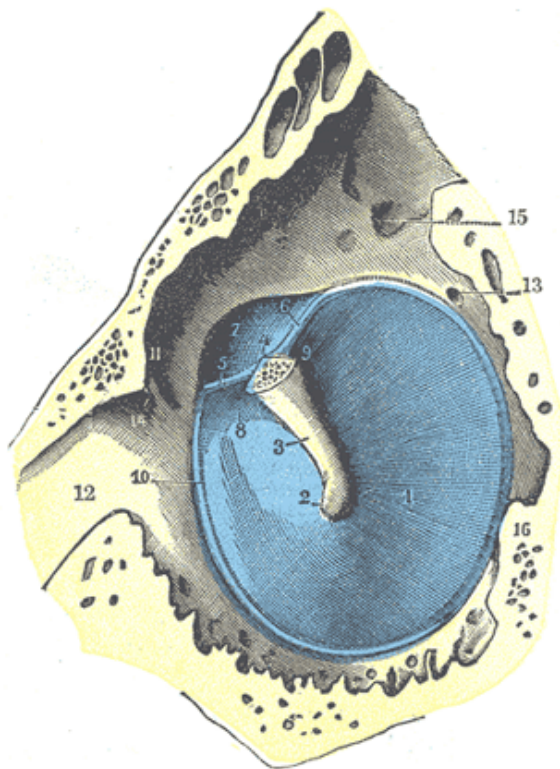
- o Situated at junction of Lateral and Posterior walls of the tympanic cavity immediately behind the tympanic membrane.
- o Present at the level of Upper end of the handle of the malleus.
- o Leads to bony canal which descends through the posterior wall of the tympanic cavity in front of the facial nerve canal.
- o The canal ends in facial nerve canal near the Stylomastoid foramen.
- o Also known as canal for chorda tympani nerve.
- 1. **Chorda tympani nerve** Enters the tympanic cavity through this opening.
- 2. Transmits **Stylomastoid artery** which usually accompanies the chorda tympani nerve.

2. PetroTympanic (Glaserian) Fissure:

- o A small slit about 2 mm long which opens Anteriorly just above the attachment of the tympanic membrane.
- 1. Houses **Anterior Process of Malleus**
- 2. Receives the **Anterior malleolar ligament**.
- 3. Transmits **Anterior tympanic branch of maxillary artery** to Tympani cavity.
- 4. If the Anterior Canaliculus is inconsistent, **Chorda Tympani Nerve** leaves through this fissure.

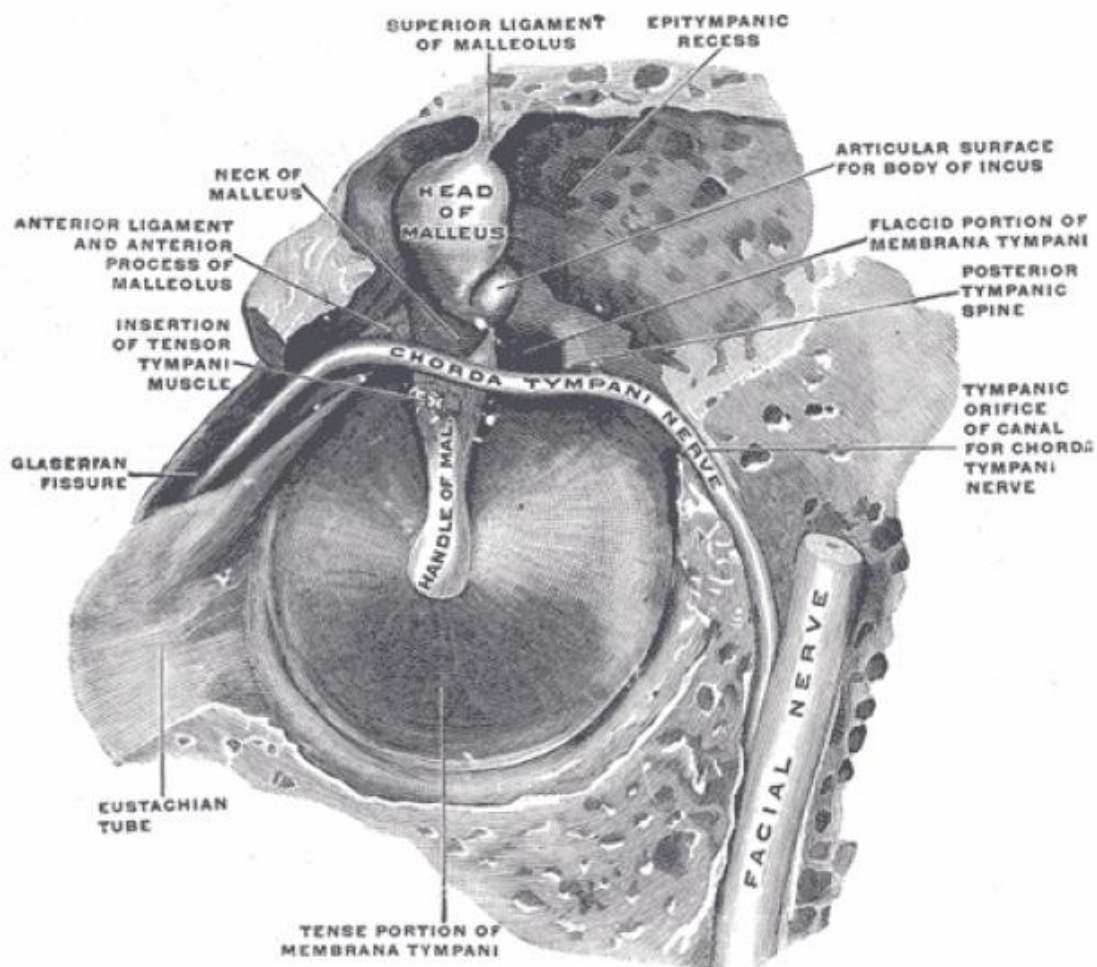
3. Canal of Hugier (Anterior canaliculus):

- o Lies medial to PetroTympanic (Glaserian) Fissure.
- o **Chorda tympani nerve** Leaves the tympanic cavity through this.

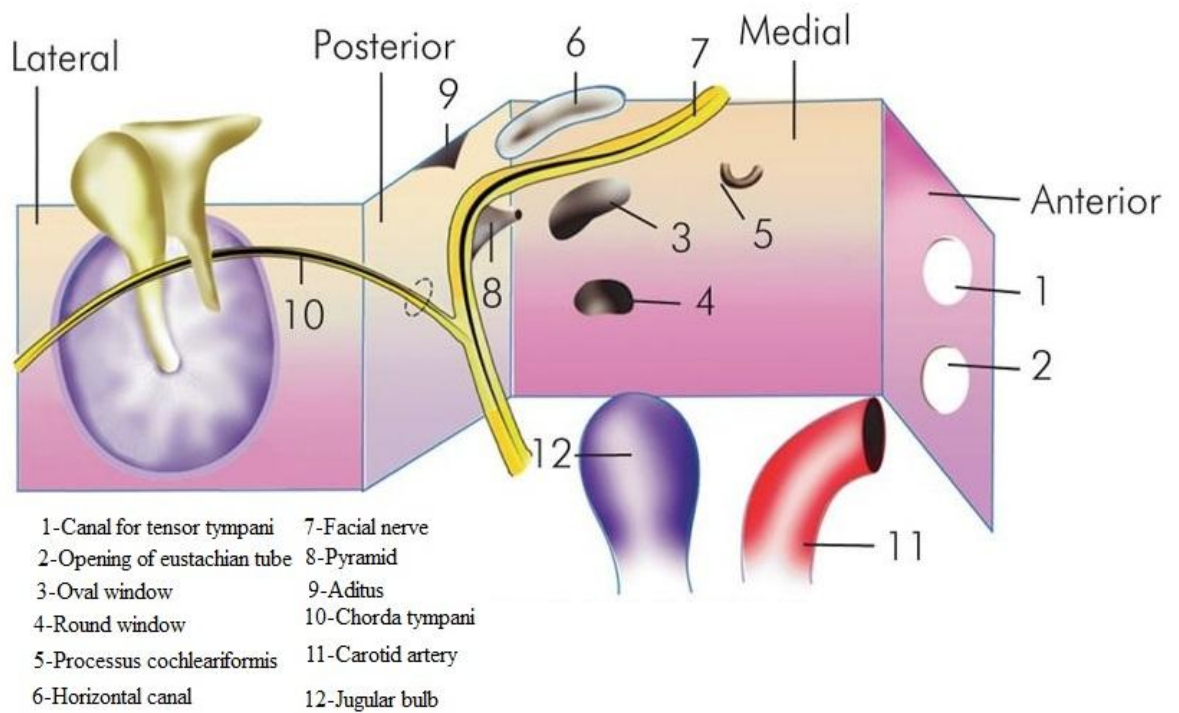
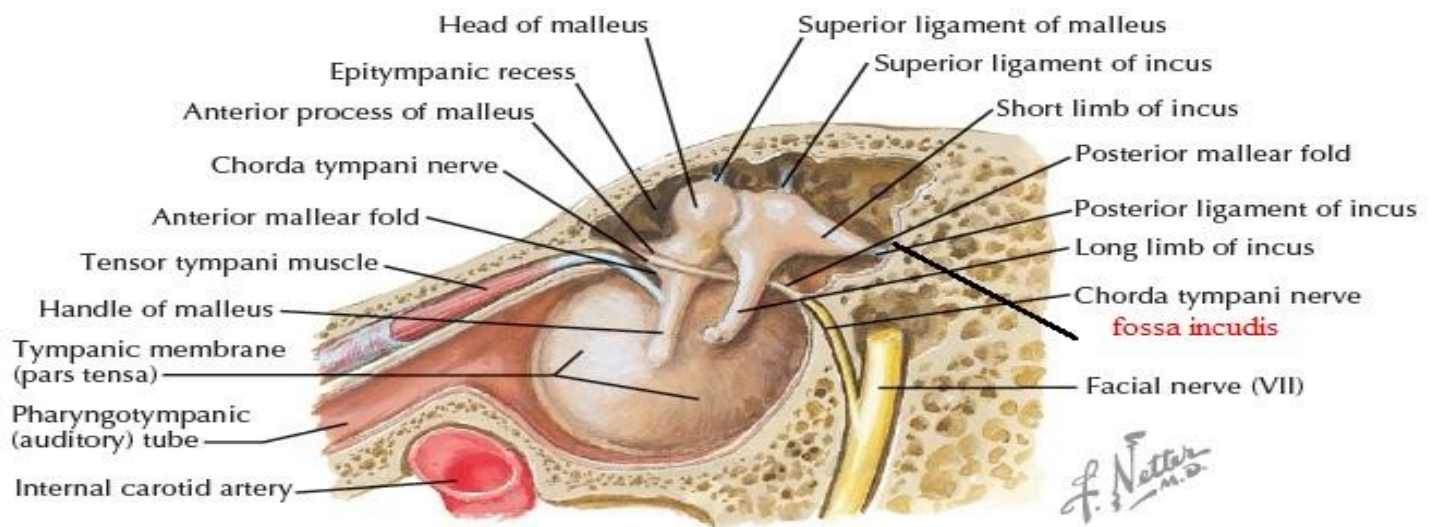


The tympanic membrane viewed from within.

1. Tympanic membrane.
2. Umbo.
3. Handle of the malleus.
4. Lateral process.
5. Anterior tympanomalleolar fold.
6. Posterior tympanomalleolar fold.
7. Pars flaccida.
8. Anterior pouch of Tröltsch.
9. Posterior pouch of Tröltsch.
10. Fibrocartilaginous ring.
11. Petrotympanic fissure.
12. Auditory tube.
13. Posterior Canaliculus.
14. Canal of Huguier.
15. Fossa incudis for short crus of the incus.
16. Styloid prominence.

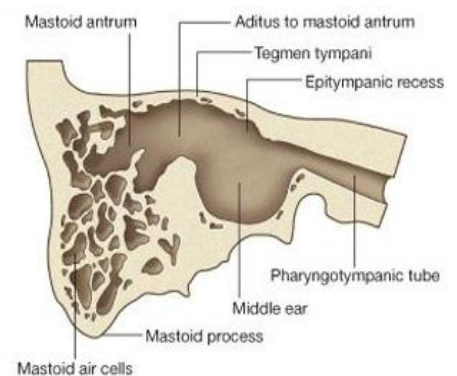
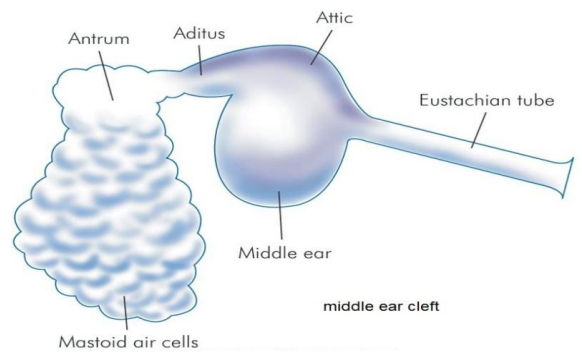


Lateral wall of tympanic cavity: medial (internal) view

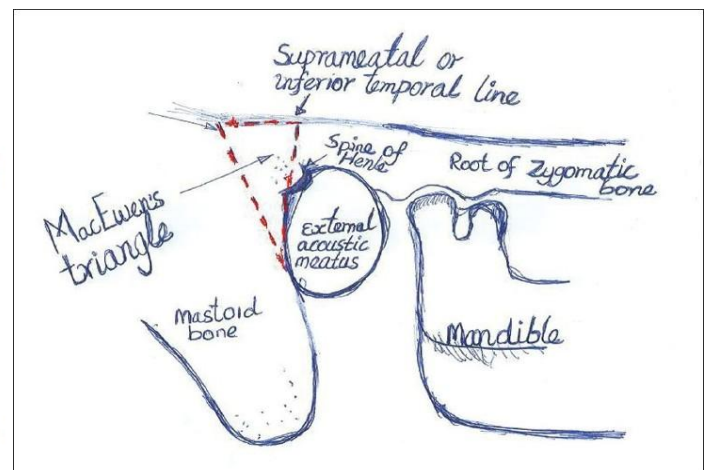
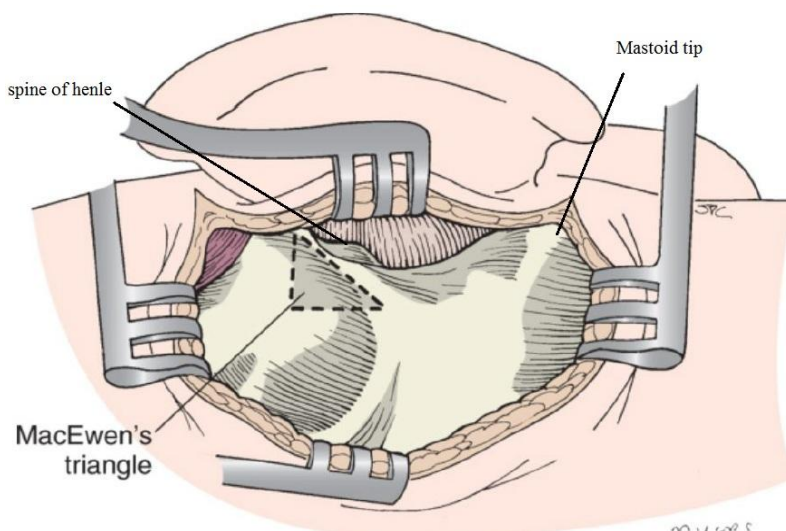


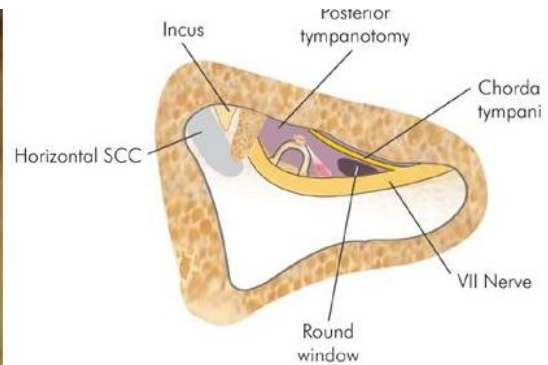
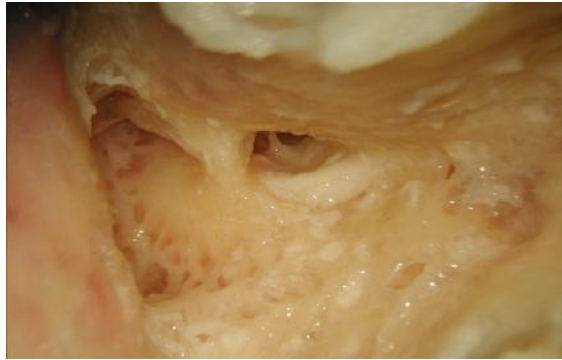
- **Mastoid Antrum**

- Large, air-containing space in the upper part of mastoid (Petrus part of the temporal bone).
- Communicates with the attic through the aditus.
- Roof is formed by the **Tegmen Antri** which is a continuation of the **Tegmen Tympani** and separates it from the **Middle cranial fossa**.
- Antrum (**NOT the air cells**) is well developed at birth.
- By adult life Antrum has a volume of 2 mL.
- Medial wall relates to **Posterior SCC**.
- Lateral wall is formed by a plate of bone which is on an average **1.5 cm** thick in the adult.
- This bone is marked externally on the surface of mastoid by **Suprameatal (MacEwen's) Triangle**:
 - Important landmark to locate the mastoid in mastoid surgery.
 - Located Postero-Superior to External auditory meatus.

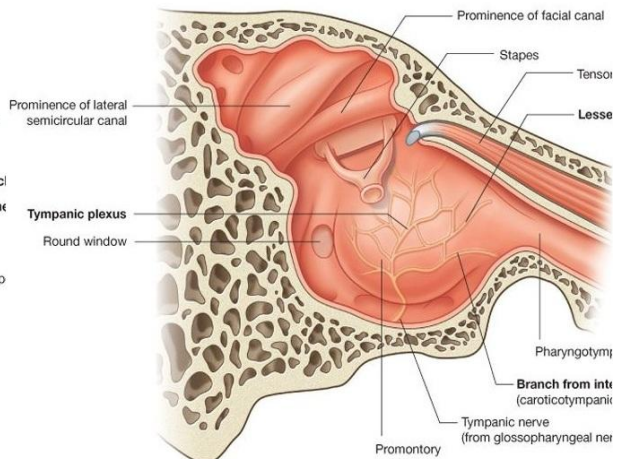
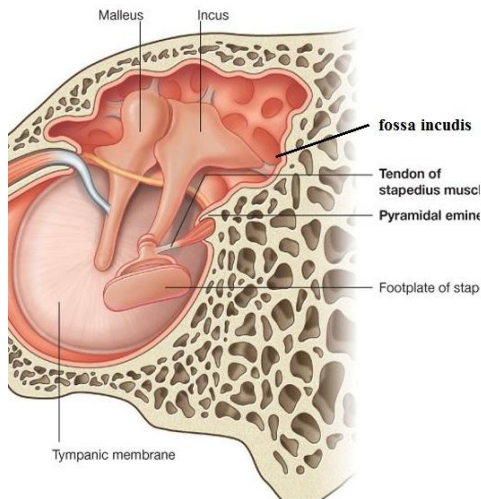


- **Suprameatal (MacEwen's) triangle boundaries:**
 1. Superiorly by: Supramastoid crest (Temporal line)
 2. Anteriorly by: Postero-Superior margin of External auditory meatus. (**spine of henle**)
 3. Posteriorly by: Vertical tangent to the posterior margin of meatus.





- **Aditus:**
- Opening through which the attic communicates with the antrum.
- Lies between:
 - o **Medially:** The bony prominence of the horizontal SCC
 - o **Laterally:** Fossa Incudis to which is attached the short process of incus.
- Facial nerve courses just below the aditus.



- **Mastoid and Its Air Cell System:**
- Mastoid consists of bone cortex with a "honeycomb" of air cells underneath.
- Acting as a reservoir of air to limit pressure changes within the middle ear.
- Facial nerve is embedded in bone in its Petrous part but exits at the stylomastoid Foramen.
- **In infants, Mastoid process is undeveloped and the Facial nerve is very superficial.**
- Mastoid process starts to develop at age of 1 year.
- Complete development at age of 2 years.
- **Lining of the mastoid** is a **Flattened, Non-ciliated epithelium without goblet cells or mucus glands.**

- Depending on development of air cell, three types of mastoid have been described :-

1. Well-Pneumatised:

- Cellular.
- Mastoid cells are well developed and intervening septa are thin.

2. Diploetic:

- Mastoid consists of marrow spaces and a few air cells.

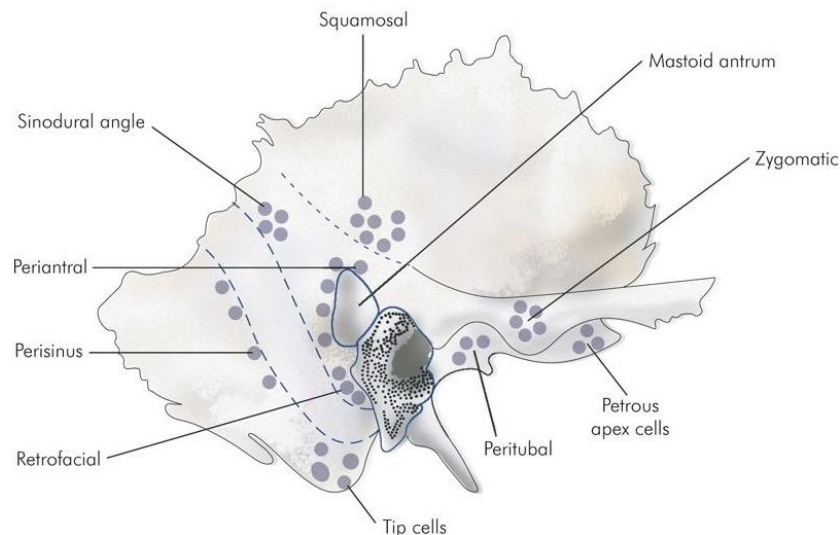
3. Sclerotic:

- Acellular
- No air cells or marrow spaces.
- Occurs in 20 % of adult temporal bones.
- Seen in individuals with Chronic ear disease.
- Mastoid antrum may be the only air-filled space in the mastoid mastoids & antrum is usually small and the sigmoid sinus is anteposed.

- ❖ **With any type of mastoid pneumatisation, Antrum is always present.**

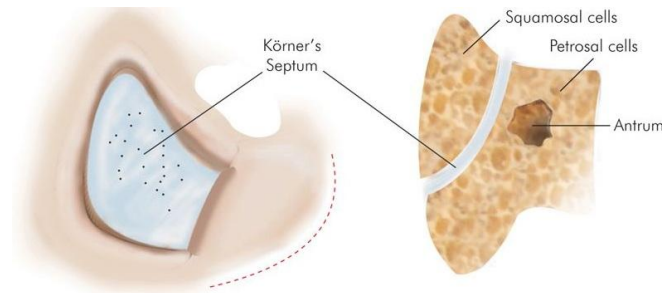
- Depending on the location, mastoid air cells are divided into:

1. **Zygomatic cells** (in the root of zygoma).
 2. **Tegmen cells** (extending into the tegmen tympani).
 3. **Perisinus cells** (overlying the sinus plate).
 4. **Retrofacial cells** (round the facial nerve).
 5. **Perilabyrinthine cells** (located above, below and behind the labyrinth, some of them pass through the arch of superior semicircular canal. These cells may communicate with the petrous apex).
 6. **Peritubal** (around the eustachian tube. Along with hypotympanic cells they also communicate with the petrous apex).
 7. **tip cells** which are quite large and lie medial and lateral to the digastric ridge in the tip of mastoid.
 8. **Marginal cells** (lying behind the sinus plate and may extend into the occipital bone).
 9. **Squamosal cells** (lying in the squamous part of temporal bones).
- Abscesses may form in relation to these air cells and may sometimes be located far from the mastoid region.

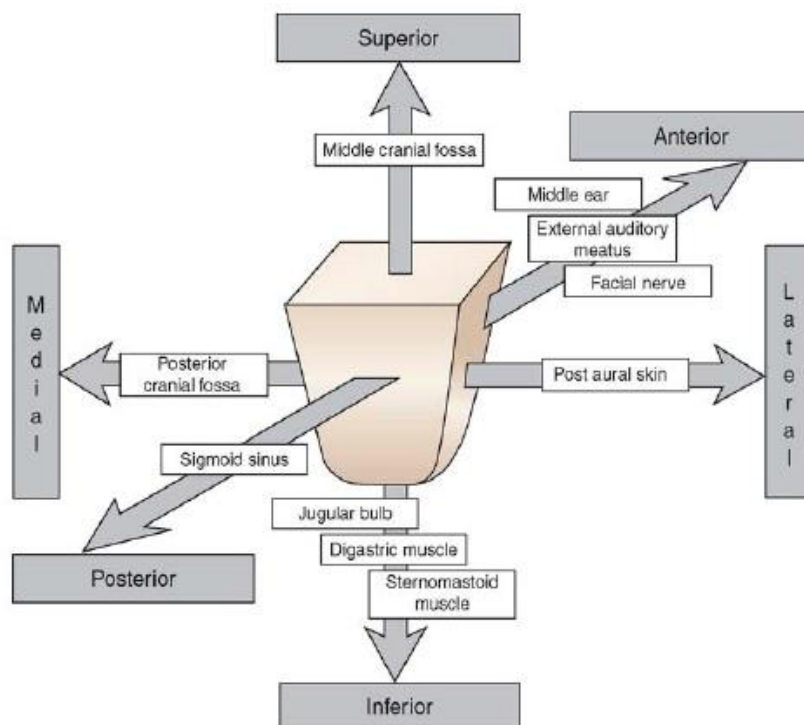


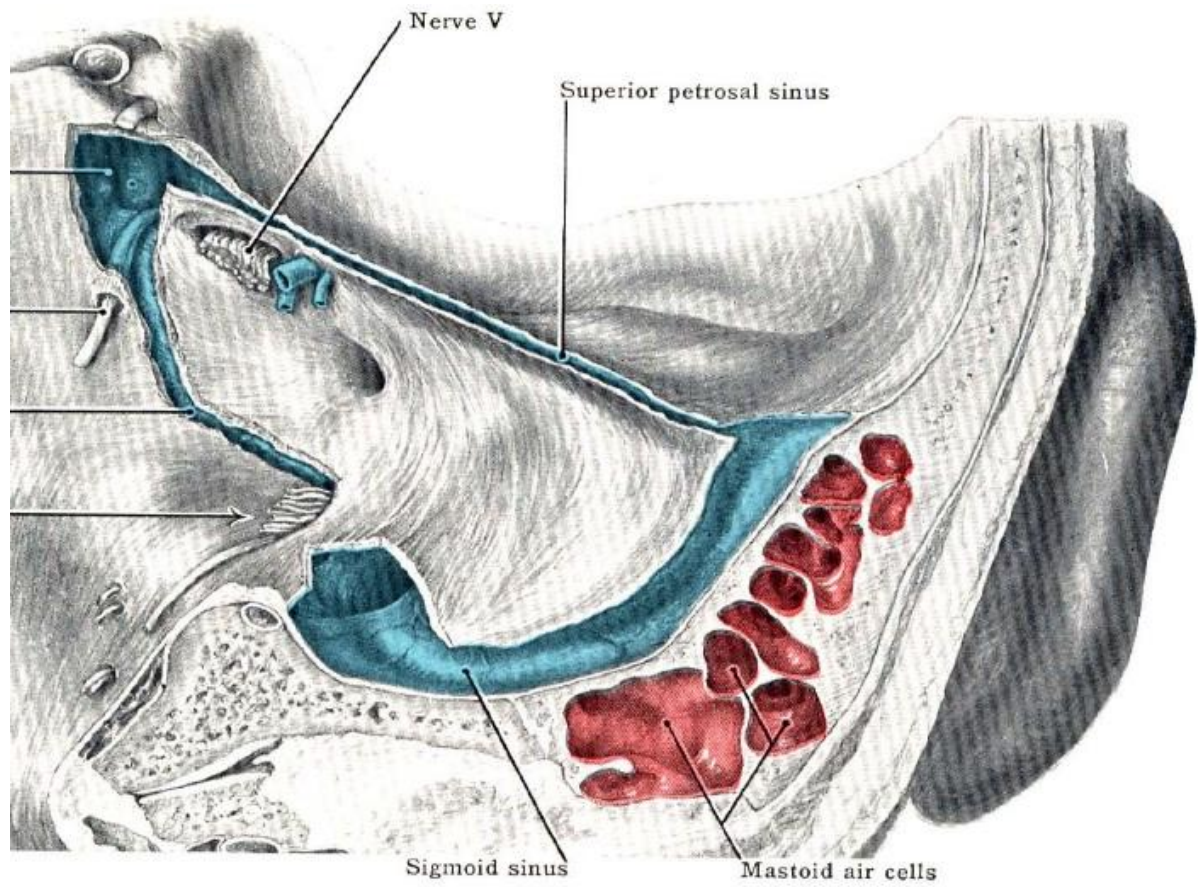
- **Development of Mastoid**

- Mastoid develops from **Squamous and Petrous bones**.
- Petrosquamosal suture may persist as a bony plate (**Körner's septum**) separating superficial squamosal cells from the deep petrosal cells.
- Körner's septum is surgically important as it may cause difficulty in locating Antrum and the deeper cells; and thus may lead to incomplete removal of disease at Mastoidectomy .
- Mastoid Antrum cannot be reached unless the Körner's septum has been removed.



- Posterior to mastoid is the **Sigmoid sinus**.
- It curves downwards only to turn sharply upwards to Pass medial to Facial nerve and then becomes the dome of the jugular bulb in the middle ear space.
- The posterior belly of the digastric muscle forms a groove in the base of the mastoid bone.

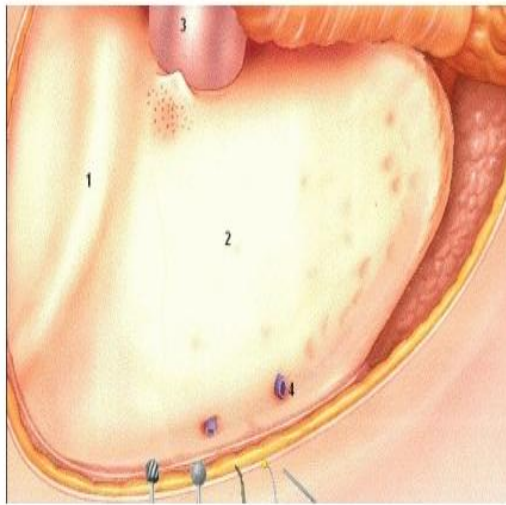




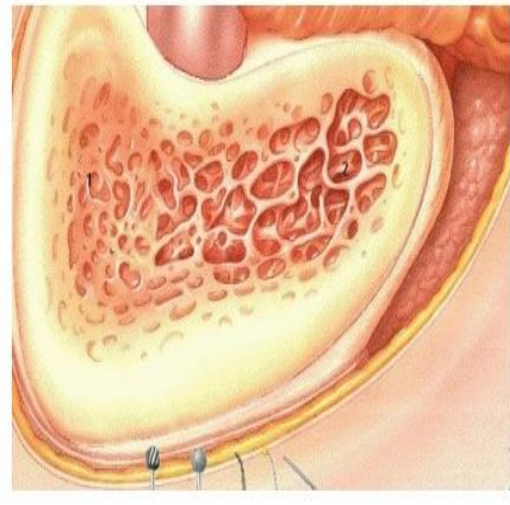
Air Cells—Dural Sinuses

Temporal Bone Dissection LATERAL APPROACH

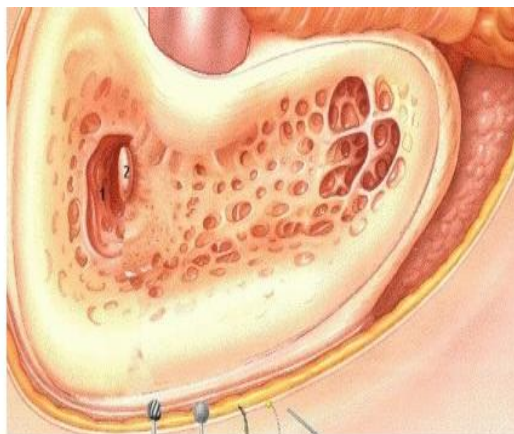
http://drfling.hyperphp.com/Notes/Temporal_Bone_Dissection_Lateral%20Approach.htm



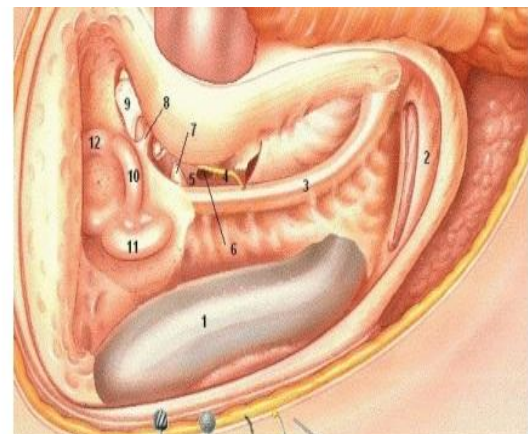
- 1. Temporal line
- 2. Mastoid cortex
- 3. External auditory canal skin
- 4. Emissary veins



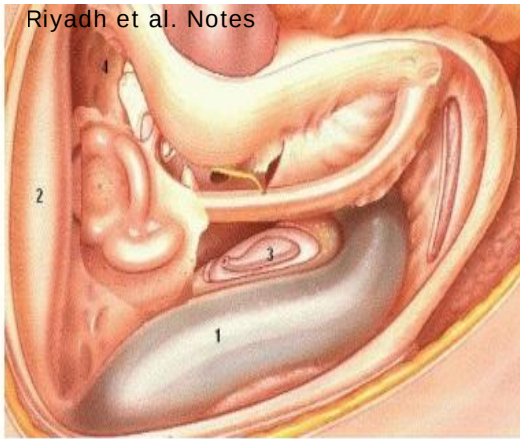
- 1. Koerner's septum
- 2. Mastoid air cells



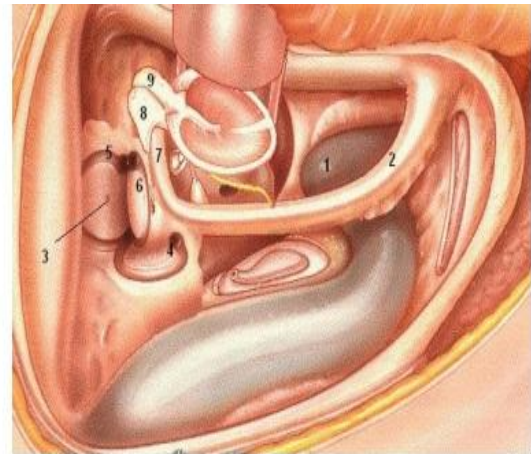
- 1. Mastoid antrum
- 2. Horizontal semicircular canal



- 1. Sigmoid sinus
- 2. Digastric ridge and posterior belly of digastric muscle
- 3. Facial nerve (skeletonized mastoid segment)
- 4. Chorda tympani
- 5. Facial recess
 - triangle bound by: chorda tympani, facial nerve and incus buttress
- 6. Round window
 - opens into cochlear scala tympani
- 7. Pyramidal eminence containing stapedius muscle
- 8. Incus buttress
- 9. Incus body
- 10. Horizontal semicircular canal
- 11. Posterior semicircular canal
- 12. Superior semicircular canal



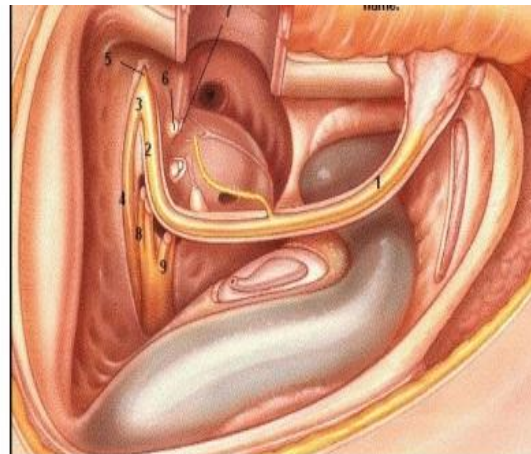
- 1. Sigmoid sinus
- 2. Tegmen tympani
- 3. Endolymphatic sac
 - in dura of posterior fossa between posterior SCC and sigmoid sinus
 - enters temporal bone at **operculum** which lies posteroinferiorly from IAC
 - lies at or below a line projected from axis of horizontal SCC (**Donaldson's line**)
- 4. Epitympanum



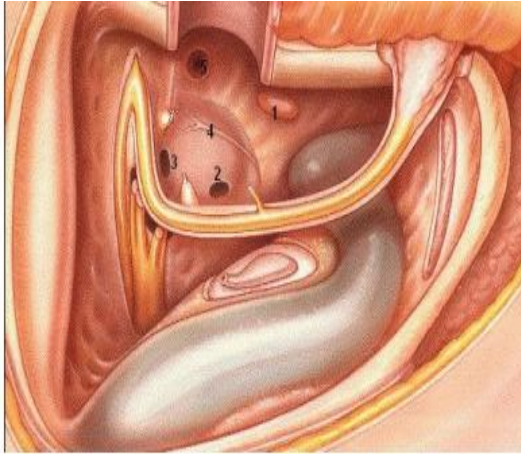
- 1. Jugular bulb
- 2. Facial nerve (mastoid segment)
- 3. Subarcuate artery
- 4. Posterior semicircular canal (ampullated end)
- 5. Superior semicircular canal (ampullated end)
- 6. Horizontal semicircular canal (ampullated end)
- 7. Facial nerve (tympanic segment)
- 8. Incus
- 9. Malleus



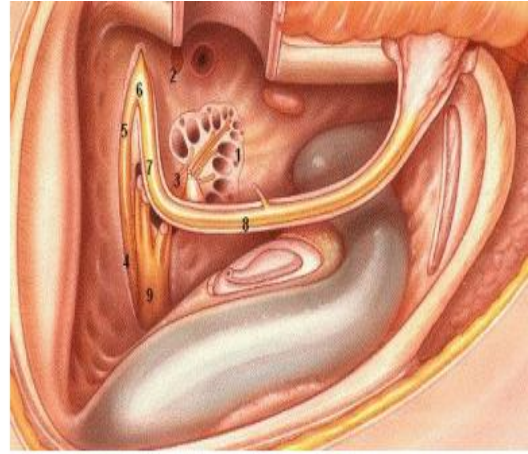
- 1. Facial nerve (tympanic segment)
- 2. Facial nerve (mastoid segment)
- 3. Chorda tympani
- 4. Eustachian tube
- 5. Jacobson's nerve (CN IX)
- 6. Promontory of cochlea
- 7. Vestibule
 - elliptical recess of utricle
 - spherical recess of saccule
- 8. Sinodural angle



- 1. Facial nerve (mastoid segment)
- 2. Facial nerve (tympanic segment)
- 3. Geniculate ganglion
- 4. Facial nerve (labyrinthine segment)
- 5. Greater superficial petrosal nerve
- 6. Cochleariform process
- 7. Tendon of tensor tympani
- 8. Superior vestibular nerve
- 9. Inferior vestibular nerve

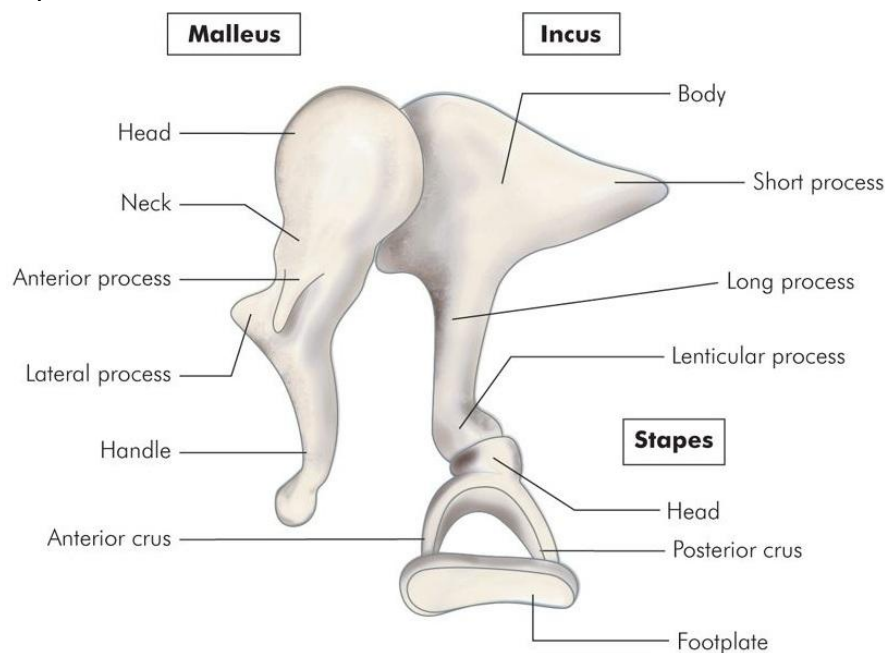


- 1. Internal carotid artery
- 2. Round window
- 3. Oval window
- 4. Jacobson's nerve and cochlear promontory



- 1. Cochlea
 - scala vestibuli ("superior" to basilar membrane in cochleostomy)
 - scala tympani ("inferior" to basilar membrane in cochleostomy)
- 2. Tensor tympani muscle (cut)
- 3. Cochlear nerve to modiolus
- 4. Facial nerve (meatal segment)
- 5. Facial nerve (labyrinthine segment)
- 6. GSPN
- 7. Facial nerve (tympanic segment)
- 8. Facial nerve (mastoid segment)
- 9. Vestibular nerve

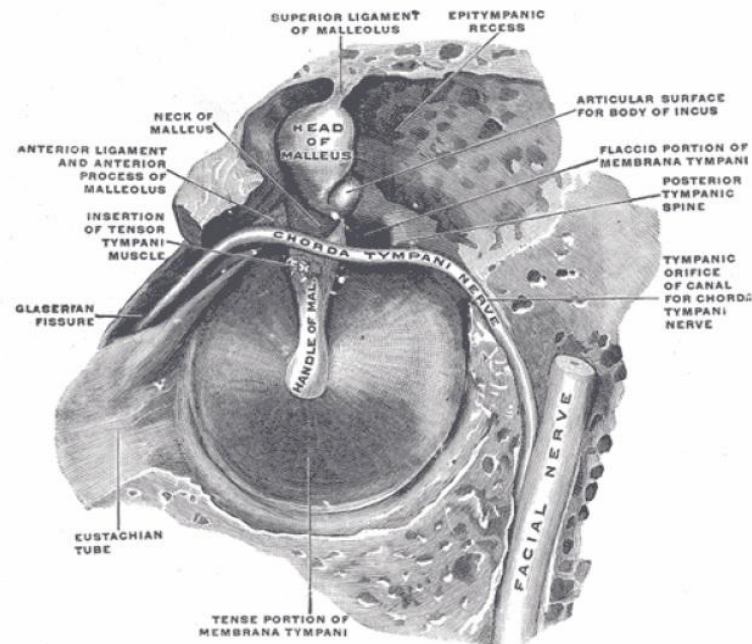
- **Contents of Tympanic cavity:**
- Most important content of the middle ear is AIR which flows into the middle ear through a patent Eustachean tube.
- 3 Ossicles: (**malleus - Incus - Stapes**)
- 2 muscles: (**Tensor Tympani - Stapedius**)
- 2 Nerves: (**Chorda tympani - Tympanic plexus**).
- **Ossicles:**
- Three ossicles: **Malleus (Hammer)- Incus (Anvil) - Stapes (Stirrup)**
- Form a semi-rigid bony chain for conducting sound.
- Held in position by Ligaments, Muscles and Interossicular joints.
- Malleus is most Latero-Inferior and attached to TM.
- Incus is most Latero-Superior.
- Stapes is most medial and attached to the oval window.



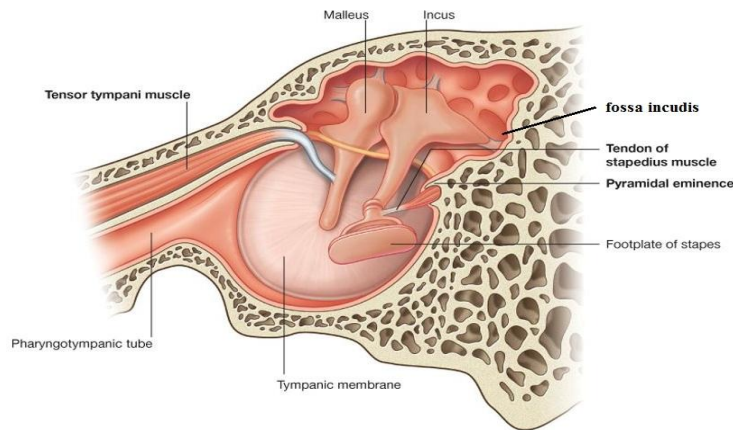
- **Malleus:**
- Largest ossicle
- Measuring up to 9 mm length
- Shape look like a **Hammer**
- Has Head, Neck and 3 processes (Handle, Lateral and Anterior Process) arising from below the neck.
- **Head of malleus:**
- Lie in the Attic "Epitympanum".
- Divides Attic into anterior and posterior portion.
- During surgical procedures for attic cholesteatoma clipping of this head will improve the exposure in the attic region.
- Supported by superior ligament, which runs upward to Tegmen tympani.
- Has a saddle-shaped facet on its Postero-Medial surface to articulate with the Body of the incus by synovial joint.



- Cog:
 - Projection in Lower part of Head of Malleus.
 - NOT eroded by cholestelema and can be used as surgical landmark in mastoid surgery if handle of malleus is eroded by cholestelema.
- **Neck of malleus:**
 - Also lie in the attic "Epitympanum".
 - Below the neck the bone broadens and gives rise to Handle, Lateral and Anterior Process.
- **Handle of malleus (Manubrium):**
 - Runs downwards, Medially and slightly backwards between the mucosal and fibrous layers of the tympanic membrane.
 - Medial surface of handle, near its upper end, is a small projection into which the tendon of the Tensor tympani muscle inserts.
 - Its most prominent lower part is Umbo.
- **Lateral process of malleus:**
 - Forms a knob-like projection on outer surface of tympanic membrane.
 - Gives attachment to Anterior and posterior malleal (malleolar) folds from the tympanic annulus.
- **Anterior process of malleus:**
 - A slender Anterior ligament arises from anterior process to insert into the petrotympanic fissure



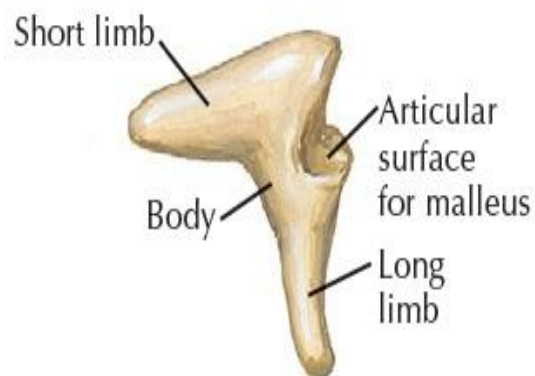
- Chorda tympani nerve crosses the **upper part** of Malleus Handle on its medial surface Above the insertion of Tendon of tensor tympani, but **below** the neck of the malleus itself.
- Neck of the malleus connects the Handle with the head and Amputation of the head by cutting through the neck leaves both chorda tympani and tensor tympani intact.



- **Incus:**
- has a body and 2 process (Short and Long process)
- Shape look like **Anvil**.
- Body and Short process lie in the Attic.

- **Body of Incus:**

- Has a cartilage-covered facet corresponding to that on the malleus.
- Body is suspended by the **Superior Incudal Ligament** that is attached to the tegmen tympani.



- **Short Process of Incus:**

- Projects backwards from the body to Lie in the Fossa incudis to which it is attached by a **Short suspensory ligament**

- **Long Process of Incus:**

- Hangs vertically and descends into the Mesotympanum behind and medial to the handle of the malleus
- At its tip, there is a small medially directed called **Lenticular process** attaches to the head of stapes.
- Long process of the incus has poor blood supply and prone for undergoing necrosis in disease conditions.
- 1st part to be eroded in cholesteatoma.

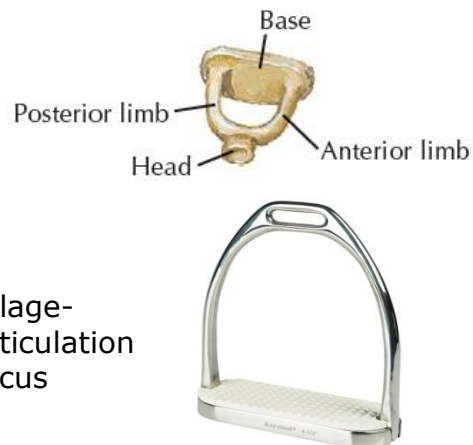


- **Lenticular process of Incus:**

- Sometimes been called the fourth ossicle because of its incomplete fusion with the tip of Long process of incus, giving the appearance of a separate bone or at least a sesamoid bone.

- **Stapes:**

- Has a head, neck, Anterior and Posterior crura (Limb) And a Footplate (Base)
- Footplate is held in oval window by Annular ligament.
- Shape look like **Stirrup**.



- **Head of Stapes:**

- Points laterally and has a small cartilage-covered depression for a synovial articulation with the Lenticular process of the Incus

- **Neck of Stapes:**

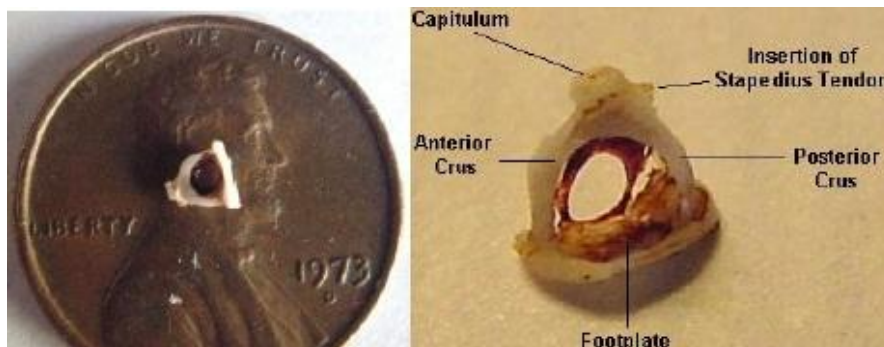
- Stapedius tendon inserts into Posterior part of Neck and Upper portion of Posterior crus.

- **Crura of Stapes:**

- Neck of the stapes gives rise to two crura.
- There is great variation in the shape of the two crura.
- Posterior crus is Longer than Anterior crus.
- Anterior crus is thinner and less curved than Posterior crus.
- The two crura join the footplate.

- **Footplate of Stapes:**

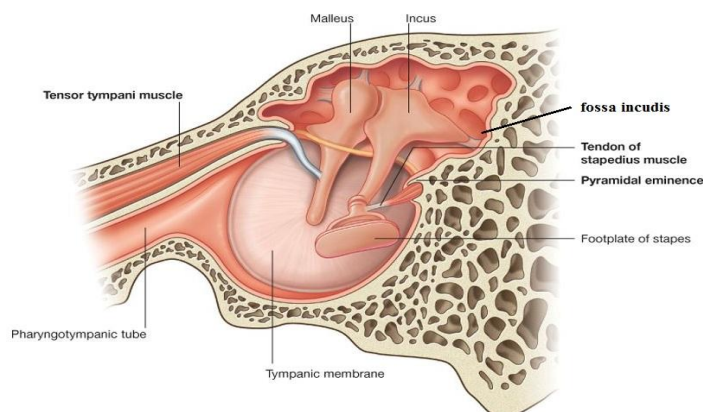
- Has a convex superior margin, and almost straight inferior margin and curved anterior and posterior ends.
- Average dimensions of the footplate 3 mm long and 1.4 mm wide
- It lies in the oval window where it is attached to the bony margins by the Annular ligament.
- Long axis of the footplate is almost horizontal, with the posterior end being slightly lower than the anterior.



- **Ossicular joints are synovial type.**

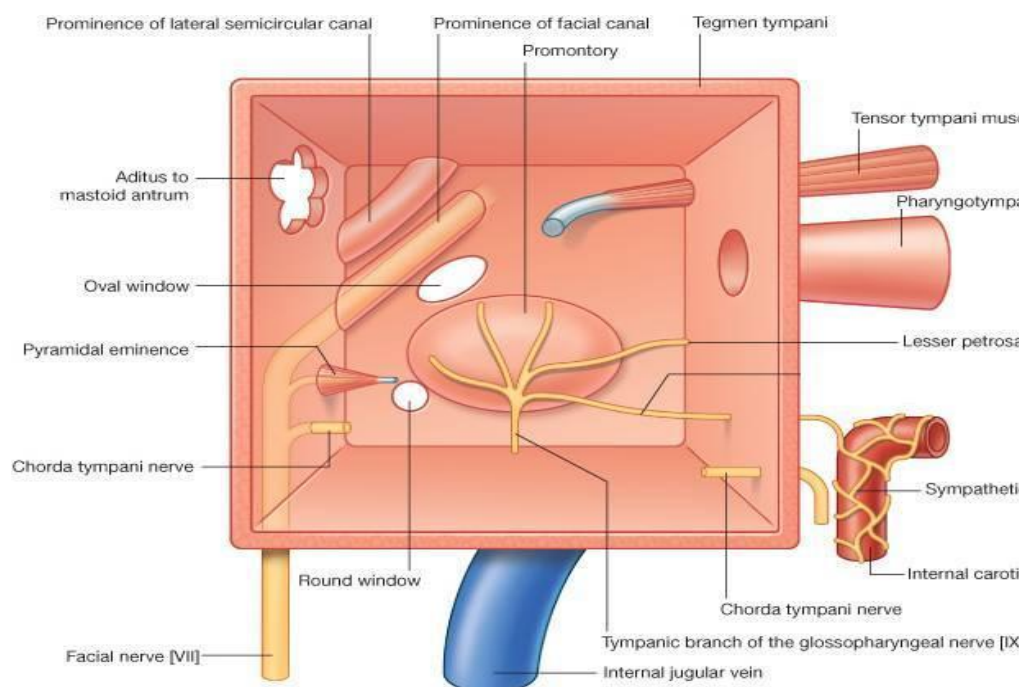
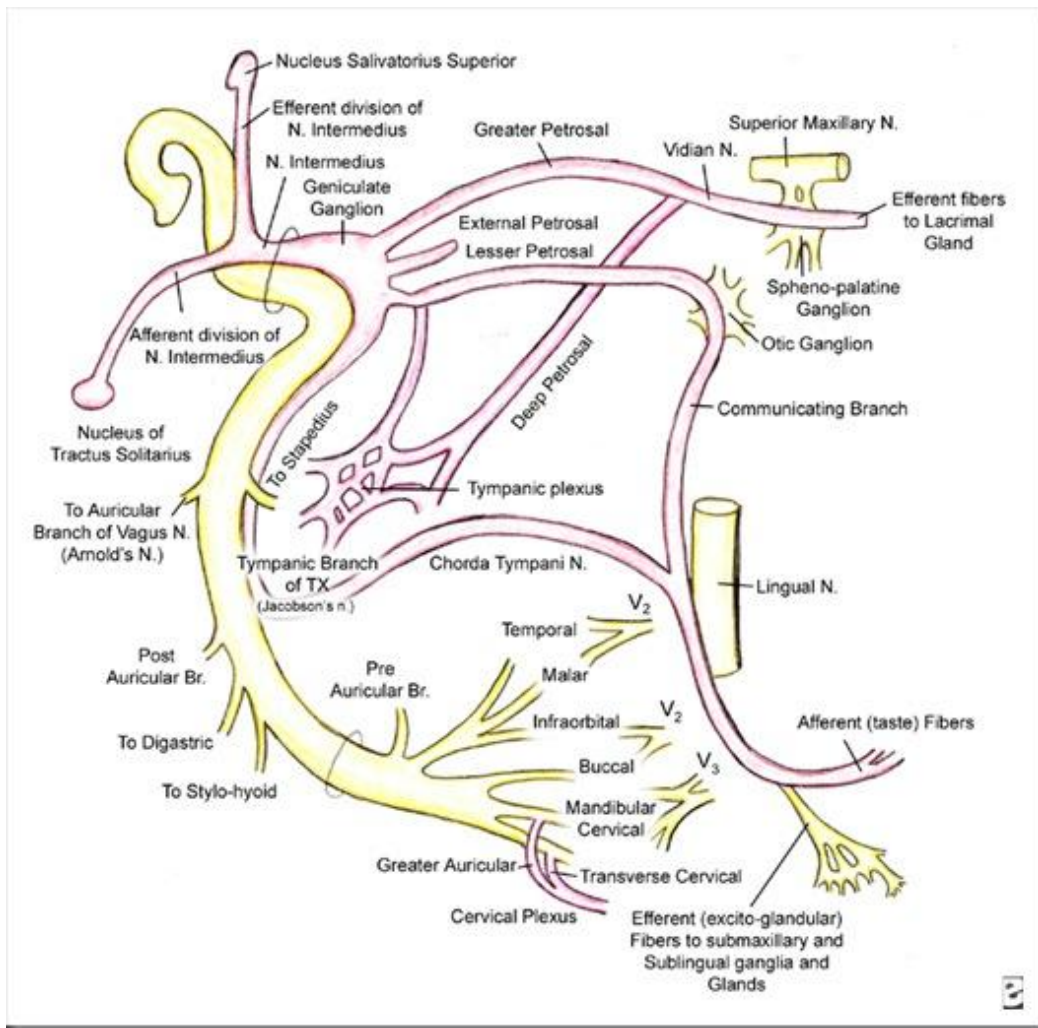
- Malleo-incudal joint: Saddle joint
- Incudo-stapedial joint: Ball and socket joint.

- **Intratympanic Muscles:**
- 2 muscles: (**Tensor tympani and Stapedius**)
- **Tensor Tympani Muscle:**
- 1st Arch Muscle.
- Supplied by Medial Pterygoid Nerve which is branch of Mandibular nerve (V₃) (Trigeminal Nerve)
- It is a long slender muscle arising from the walls of the bony canal lying above the Eustachian tube.
- Parts of the muscle also arise from the Cartilaginous portion of the Eustachian tube and Greater wing of the sphenoid.
- Then it passes backwards into the tympanic cavity lying on the medial wall of the middle ear just below the level of the facial nerve.
- The bony covering of the canal is often deficient in its tympanic segment where the muscle is replace by its tendon.
- This tendon enters the **Processus cochleariformis** on the medial wall, where it is held down by a transverse tendon as it turns through a right angle to pass laterally and insert into the **Medial Aspect of Upper end of the Malleus Handle.**
- It tenses the tympanic membrane by holding the handle of the malleus thus helping the middle ear in better sound perception.

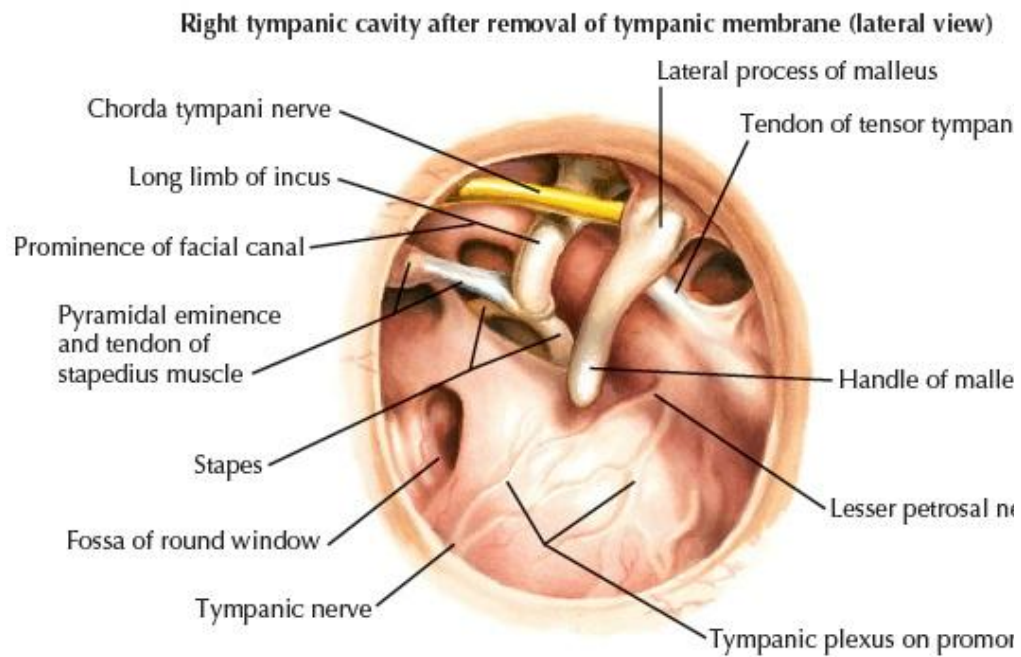


- **Stapedius Muscle:**
- 2nd arch muscle.
- Supplied by a branch of Facial nerve (CN-VII).
- Smallest muscle in the body.
- Arises from walls of the conical cavity within the pyramid.
- A slender tendon emerges from the apex of the pyramid and inserts into the **Neck and Posterior crus of stapes.**
- On contraction, this muscle rocks the stapes backwards holding it firm against the annular ligament preventing excessive transmission of sound into the inner ear.
- Protective role by dampen very loud sounds.
- Preventing noise trauma to the inner ear.
- Patients with facial nerve palsy have hyperacusis because of lack of action of this muscle.

- **Tympanic Plexus:**
- Lies on the promontory.
- Formed by:
 1. Tympanic branch of Glossopharyngeal nerve . (Jacobson's nerve)
 2. Caroticotympanic nerves (sympathetic fibers from the plexus around the internal carotid artery).
- Tympanic plexus gives innervations to:
 1. Medial surface of TM.
 2. Mucous membrane lining the tympanic cavity, Bony Eustachean tube, Mastoid Antrum and its air cells.
 3. **Secretomotor fibers** to Parotid gland.
- Tympanic plexus provide the following branches:
 1. Branches to the mucous membrane lining the tympanic cavity, eustachean tube, mastoid antrum and its air cells
 2. A Deep branch joining the Greater superficial petrosal nerve.
 3. Lesser superficial petrosal nerve, which contain all the parasympathetic fibers of Glossopharyngeal nerve (CN-IX) to supply the parotid gland.
 - **Lesser superficial petrosal nerve** leaves the middle ear through a small canal below the tensor tympani muscle where it receives parasympathetic fibers from Facial nerve (CN-VII) nerve by way of a branch from the geniculate ganglion.
 - The full nerve passes through the temporal bone to emerge lateral to the greater superficial petrosal nerve on the floor of the middle cranial fossa, outside the dura.
 - It then passes through the foramen ovale with the Mandibular nerve and Accessory meningeal artery to the otic ganglion.
 - Post ganglionic fibers from the otic ganglion supply secretomotor fibers to the parotid gland by way of the auriculotemporal nerve.
- Course of secretomotor fibers to the parotid:
- Inferior salivary nucleus → CN IX → Tympanic branch → Tympanic plexus → Lesser petrosal nerve → Otic ganglion → Auriculotemporal nerve → Parotid gland.
- Section of Tympanic branch of glossopharyngeal nerve (**Jacobson's nerve**) can be carried out in the middle ear in cases of **Frey's syndrome**.
- Frey's syndrome are redness and sweating on the cheek area adjacent to the ear appear when the affected person eats as a side effect of parotid gland surgery or due to injury to Auriculotemporal nerve.
- Auriculotemporal branch of the Trigeminal nerve carries sympathetic fibers to the sweat glands of the scalp and parasympathetic fibers to the parotid gland.
- As a result of severance and inappropriate regeneration, the parasympathetic nerve fibers may switch course, resulting in sweating in the anticipation of eating, instead of the normal salivatory response.



- **Chorda Tympani Nerve:**
- Branch of Facial nerve (CN-VII)
- Enters the middle ear through **Posterior canaliculus**, at the junction of **Lateral and posterior walls**.
- Runs on the medial surface of the tympanic membrane between the Handle of malleus and Long process of Incus, above the attachment of tendon of tensor tympani.
- Continue forwards and leave by way of the **Canal of Hugier (Anterior canaliculus)**, which subsequently joins the **Petrotympanic fissure**.
- Carries **Taste from Anterior 2/3 of tongue**.
- Supplies **Secretomotor fibers to Submaxillary and Sublingual Salivary glands**.



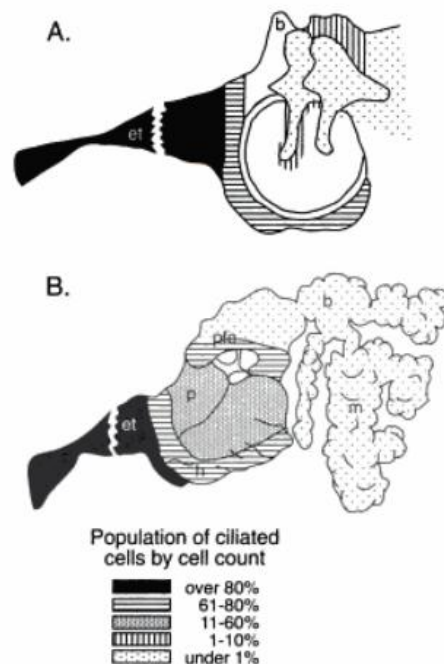
- **Lining of Middle Ear Cleft:**
- Mucous membrane of the Nasopharynx is continuous with that of Middle ear, Aditus, Antrum and Mastoid air cells.
- Histologically, Epithelium of middle ear cavity is varies according to the location.
- **Eustachian Tube Lining:**
- **Pseudostratified Ciliated Columnar Epithelium**
- With several mucous glands in the submucosa.
- At its nasopharyngeal end, the mucosa is truly respiratory; but in passing along the tube towards the middle ear, the number of goblet cells and glands decreases, and the ciliary carpet becomes less profuse.

- **Tympanic cavity Lining:**
- Antero-Inferior Compartment (Hypotympanum):
 - o **Pseudostratified Ciliated Columnar Epithelium.**
 - o The function of this compartment is devoted primarily to mucociliary clearance.
- Middle Compartment:
 - o **Ciliated Cuboidal Epithelium.**
- Postero-Superior compartment (Epitympanum and Mastoid):
 - o **Flat Non-ciliated Epithelium.**
 - o A gas exchange occurs in this compartment.

- Mucous membrane wraps the middle ear structures (ossicles, muscles, ligaments, and nerves) like **peritoneum** wraps various viscera in the abdomen, Raising several folds and dividing the middle ear into **various compartments**.

- As a result, **Only route for ventilation of Epitympanic space from Mesotympanum** is via 2 small openings between the various mucosal folds:

1. **Anterior isthmus tympani.**
2. **Posterior isthmus tympani.**

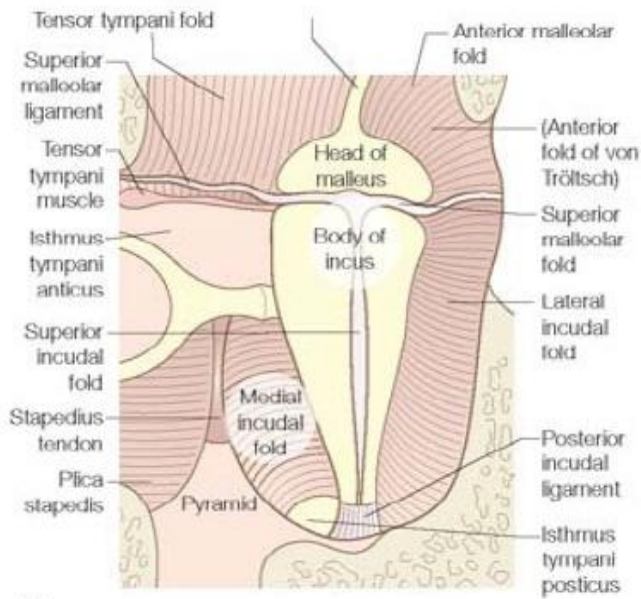


- **Prussak's Space:**
- Bordered by:
 - **Laterally:** Pars Flaccida, Laterally
 - **Medially:** Neck of the Malleus.
 - **Inferiorly:** Lateral process of Malleus
 - **Superiorly:** Lateral malleolar fold.

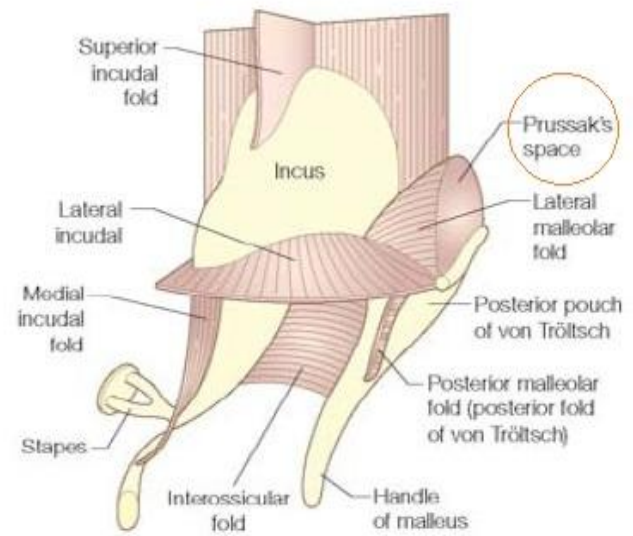
- This space can play an important role in the retention of keratin and subsequent development of cholesteatoma. (**1st site of origin of cholesteatoma.**)



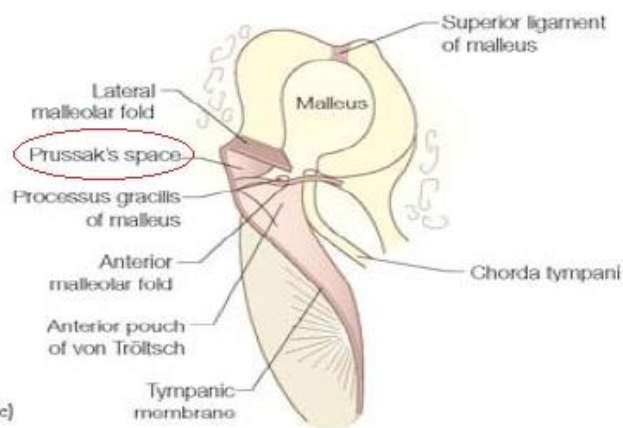
- **The mucosal folds have been described in detail by Figure below**



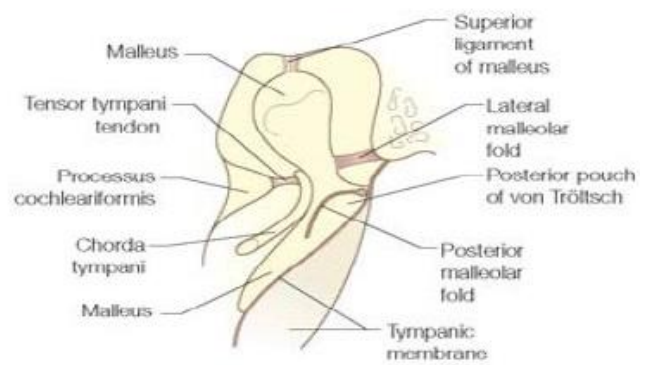
(a)



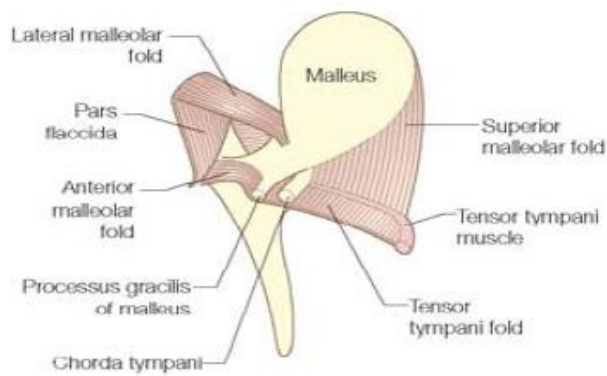
(b)



(c)



(d)



- **Blood Supply of Middle Ear**
- Middle ear is supplied by 6 Arteries.
- From both Internal and External carotid system.
- **2 Main Arteries:**
 1. **Anterior tympanic branch of Maxillary Artery:**
 - Supplies Tympanic membrane.
 2. **Posterior tympanic branch of Stylomastoid Artery:**
 - Branch of Posterior Auricular Artery.
 - Supplies Middle ear and mastoid air cells.
- **4 Minor Arteries:**
 1. **Petrosal branch of Middle Meningeal Artery:**
 - Runs along Greater Petrosal Nerve.
 2. **Superior tympanic branch of Middle Meningeal Artery:**
 - Traversing along the canal for tensor tympani muscle.
 3. **Branch of Artery of Pterygoid Canal:**
 - Runs along Eustachian tube.
 4. **Tympanic branch of internal carotid.**
- **Veins drain into:**
 1. Pterygoid venous plexus
 2. Superior Petrosal sinus

Branch	Parent artery	Region supplied
Anterior tympanic	Maxillary artery	Tympanic membrane; malleus and incus; anterior part of tympanic cavity
Stylomastoid	Posterior auricular	Posterior part of tympanic cavity; stapedius muscle
Mastoid	Stylomastoid	Mastoid air cells
Petrosal	Middle meningeal	Roof of mastoid; roof of epitympanum
Superior tympanic	Middle meningeal	Malleus and incus; tensor tympani
Inferior tympanic	Ascending pharyngeal	Mesotympanum
Branch from artery	Artery of pterygoid canal	Meso- and hypotympanum
Tympanic arches	Internal carotid	Meso- and hypotympanum

- **Lymphatic Drainage of Ear**
- Lymphatics from the middle ear drain into retropharyngeal and parotid nodes.
- Lymphatics of the Eustachian tube drain into Retropharyngeal group

Area	Nodes
Concha, tragus, fossa triangularis and external cartilaginous canal	Preauricular and parotid nodes
Lobule and antitragus	Infra-auricular nodes
Helix and antihelix	Post-auricular nodes, deep jugular and spinal accessory nodes
Middle ear and eustachian tube	Retropharyngeal nodes → upper jugular chain
Inner ear	No lymphatics

Inner Ear:

- Also called the internal ear or the labyrinth.
- Important organ of hearing and balance.
- Consists of:
 1. Bony labyrinth.
 2. Membranous labyrinth.

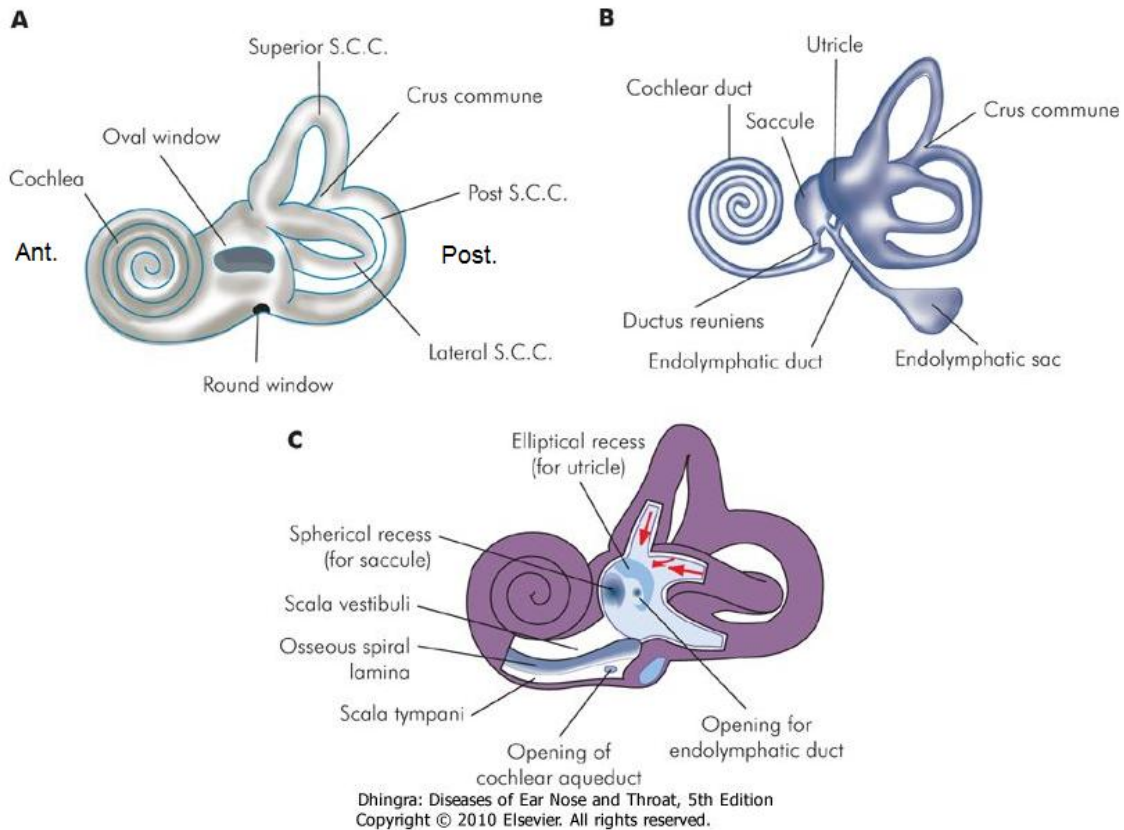


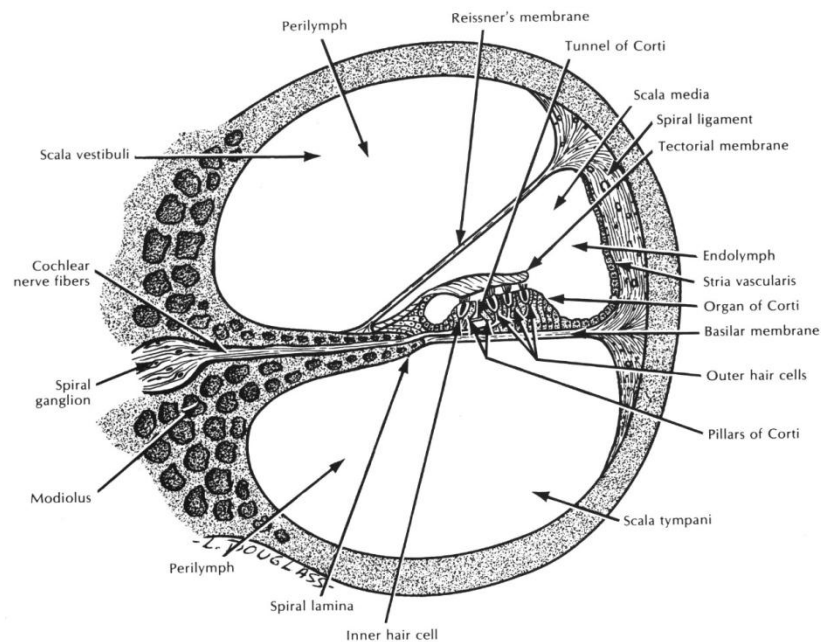
Figure 1.13 (A) Left bony labyrinth. (B) Left membranous labyrinth. (C) Cut section of bony labyrinth.

Bony Labyrinth:

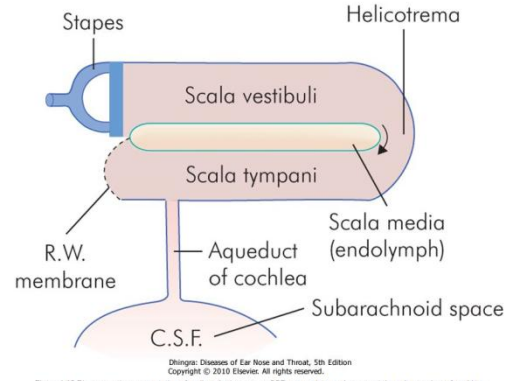
- Located in the petrous portion of the temporal bone.
- Surrounds the membranous labyrinth and contains **Perilymph**.
- Consists of:
 1. Cochlea
 2. Vestibule.
 3. Semicircular canals.

1. Cochlea:

- Anterior portion of the bony labyrinth
- Contains the cochlear duct of the membranous labyrinth.
- Coiled tube making **2.5 - 2.75 turns** round a central pyramid of bone called the **modiolus**.
- Base of modiolus is directed towards internal acoustic meatus and carries branches of the cochlear nerve to the cochlear duct.
- Around the modiolus and winding spirally like the thread of a screw, is a thin plate of bone called **osseous spiral lamina**.
- It divides the bony cochlea incompletely, and gives attachment to the basilar membrane.
- **Spiral ganglion**: contains cell bodies of the cochlear nerve, located within the central modiolous in lateral end of cochlear nerve.
- The promontory (bony bulge in the medial wall of middle ear) is due to the basal coil of the cochlea.

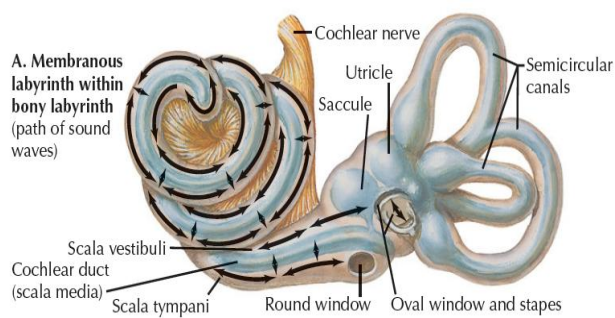
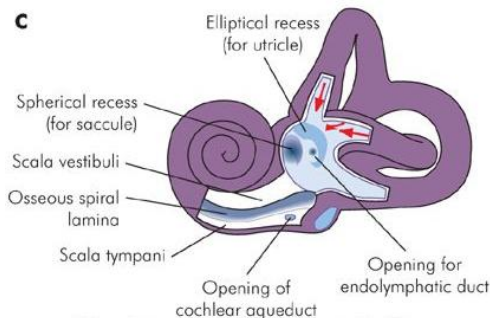


- The bony cochlea contains three compartments:
 1. Scala **V**estibuli.
 2. Scala **M**edia.
 3. Scala **T**ympani.

- **Scala Vestibuli**
 - Filled with **Perilymph**.
 - Begins in vestibule.
 - Closed by the footplate of stapes over the oval window which separates it from the air-filled middle ear.
 - Communicate with Scala tympani at the apex of cochlea through an opening called **helicotrema**.
 - **Scala Media**
 - Also called Cochlear duct, Membranous cochlea
 - Filled with **Endolymph**.
 - Begins at round window
 - **Scala Tympani**
 - Filled with **Perilymph**.
 - Closed by secondary tympanic membrane.
 - Connected with the subarachnoid space through the aqueduct of cochlea.
- 
- **Cochlear aqueduct** contains the **periotic duct**, allows perilymph to drain into CSF (Subarachnoid space), ends in posterior cranial fossa.

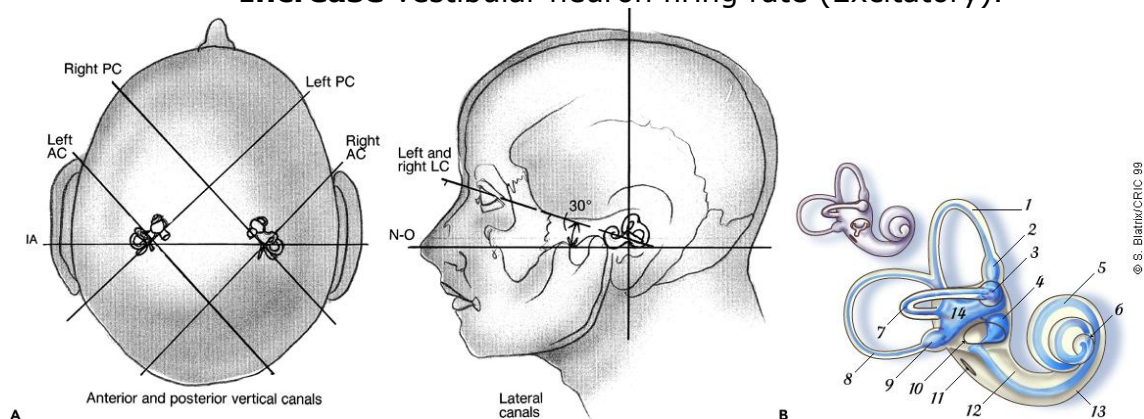
2. Vestibule:

- Middle portion of the bony labyrinth.
- Oval window lies in its lateral wall.
- Inside of its medial wall presents two recesses:
 1. Spherical recess: lodges the **Sacculle** (Anterior)
 2. Elliptical recess: lodges the **Utricle** (Posterior)
- In the posterosuperior part of vestibule are the five openings of semicircular canals.
- **Vestibular Aqueduct** contains the **endolymphatic duct** and runs from the vestibule to the posterior surface of the petrous pyramid in the posterior cranial fossa (opening is the **operculum**)



3. Semicircular Canals (SCC):

- Posterior portion of the bony labyrinth.
- Three in number, **Lateral**, **Posterior** and **Superior**.
- Lie in planes at right angles to one another.
- Each canal has an **Ampullated end** which opens independently into the Utricle and a **Nonampullated end**.
- Cupula: gelatinous layer located within each ampulla, extends to the roof of the ampulla sealing the SCC.
- Cilia are embedded in the cupula; deflections of the cupula bends stereocilia.
- Membranous labyrinth turns with head, endolymph stays but due to inertial mass; causes pressure across cupula
- The three canals open into the vestibule by five openings.
- **Lateral (Horizontal) SCC:**
 - o Inclined 30 degrees from horizontal.
 - o AmpulloPetal flow of endolymph (**TOWARD vestibule**) **Increase** vestibular neuron firing rate (Excitatory).
- **Superior (Anterior) and Posterior SCC:**
 - o Vertical canals.
 - o Share one nonampullated ends **Crus commune**.
 - o AmpulloFugal flow of endolymph (**AWAY from vestibule**) **Increase** vestibular neuron firing rate (Excitatory).



Membranous Labyrinth:

- Located within the bony labyrinth and contains **Endolymph**
- consists of:
 1. Cochlear duct
 2. Saccule
 3. Utricle
 4. Three semicircular ducts
 5. Endolymphatic duct and sac.

1. Cochlear duct

- Also called membranous cochlea or the scala media.
- Blind coiled tube located within the bony cochlea.

- Triangular on cross-section and its three walls are formed by:

1. Basilar membrane:

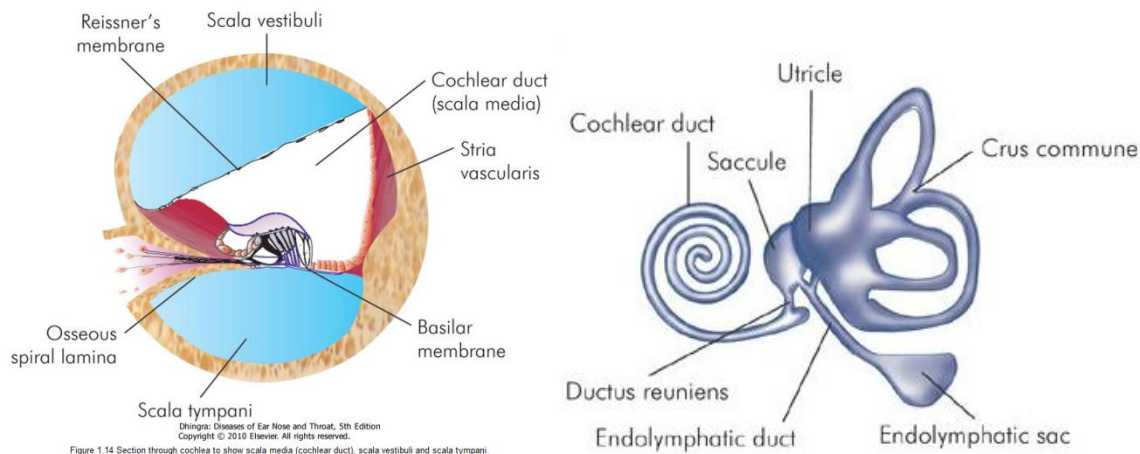
- Floor of scala media.
- Between scala media and scala tympani.
- Supports the organ of corti.
- **Base:** Stiffer and Thinner.
- **High frequencies** of sound are heard at the Basal coil.
- **Apex:** Flexible and Thicker.
- **Lower frequencies** are heard at the Apical coil.

2. Reissner's membrane:

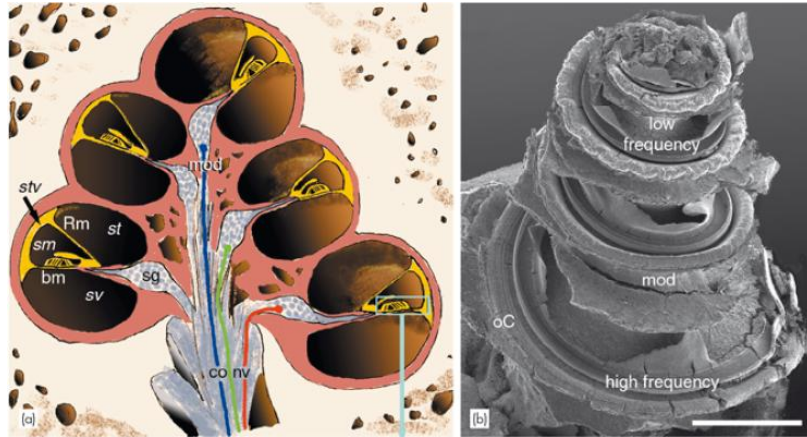
- Also called vestibular membrane.
- Roof of scala media.
- Separates scala media from the scala vestibuli.
- Two-cell layer membrane separated by a basement membrane:
- Endothelial cell layer facing the scala media.
- Mesothelial cell layer facing the scala vestibuli.

3. Stria vascularis:

- Lateral wall of scala media.
- Contains vascular epithelium.
- Support cochlear function.
- Na-K ATPase keeps membrane potential at **+80 mV**.
- Secretes of **Endolymph**.



- **Ductus reuniens:** narrowest segment connects cochlear duct to the saccule.
- **Periotic duct:** located within the **cochlear aqueduct**, connects the scala tympani to the posterior cranial fossa.



- The central bony axis of the spiral, the modiolus (mod) contains the spiral ganglion (sg) comprised of bipolar neurones that peripherally innervate the hair cells and centrally form the cochlear nerve (co nv).
- Afferent fibres representative of low (blue), middle (green) and high (red) frequency illustrate the tonotopic arrangement within the nerve.

2. **Saccul**

- Lies in the Anterior part bony vestibule.
- Anterior and perpendicular to the utricle and opposite the stapes footplate.
- Communicates with Cochlear duct (via Ductus reuniens).
- Communicates with Endolymphatic duct (via Saccular duct).
- Does NOT communicate directly with utricle
- Its sensory epithelium is called the macula.
- Saccular macula lies mostly in vertical plane.
- Detects Vertical linear acceleration and change in Gravity
- Can be surgically decompressed by perforating the stapes footplate in Meniere's disease.

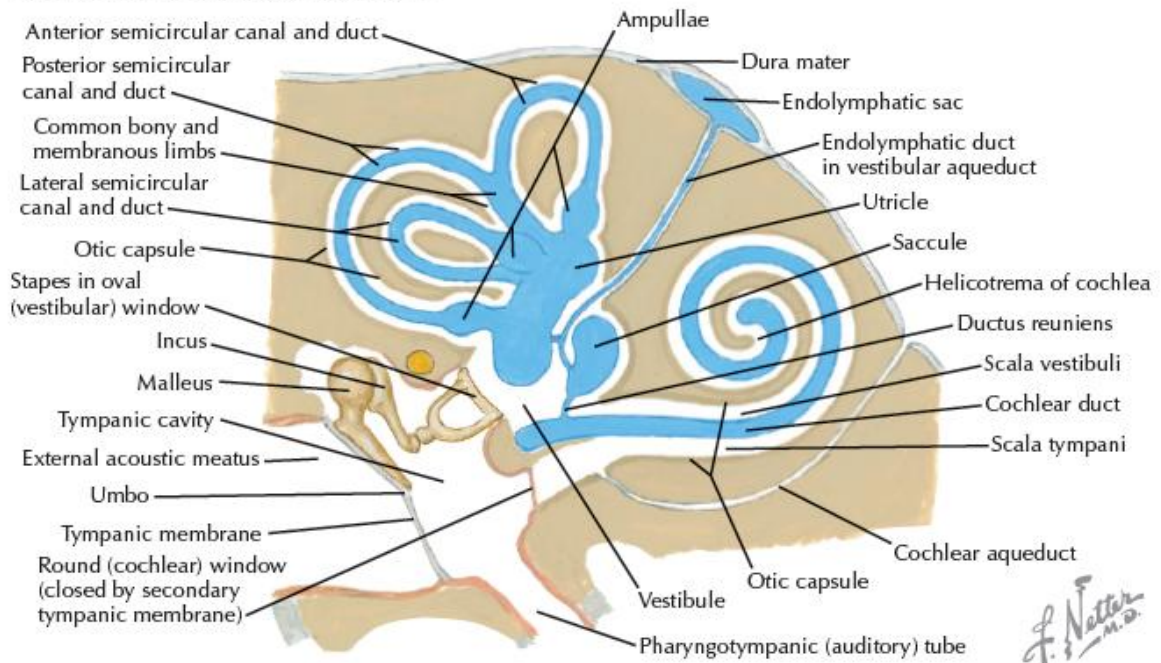
3. **Utricle**

- Utricle lies in the Posterior part of bony vestibule.
- Parallel to earth and aligned with Lateral SCC.
- Superior to Saccul.
- Receives the five openings of the three semicircular ducts.
- Communicates with Endolymphatic duct (via Utricular duct).
- The sensory epithelium of the utricle is also called the macula.
- Macula utriculi lies mostly in horizontal plane.
- Detects Horizontal linear acceleration.

4. **Semicircular ducts**

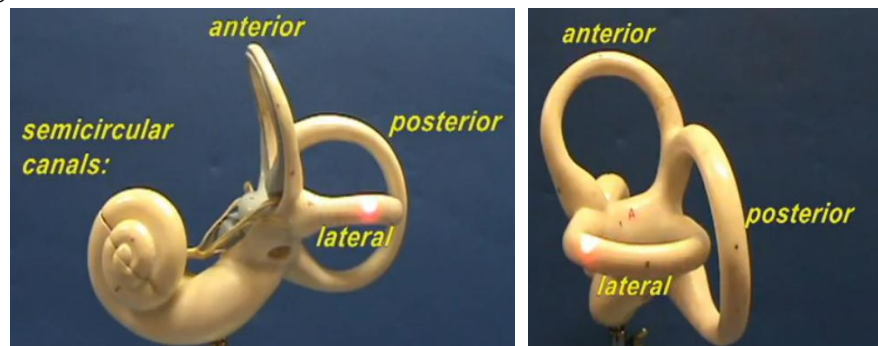
- Three in number and correspond exactly to the three bony canals.
- Open in the utricle.
- Sensory receptors are located in the ampullated end and known as Crista Ampullaris, which contains hair cells.
- Concerned with for Angular acceleration.

Bony and membranous labyrinths: schema



5. Endolymphatic Duct and Sac

- Endolymphatic duct:
 - Formed by the union of two ducts, one each from the saccule and the utricle.
 - Contained within the **vestibular aqueduct**.
 - Its terminal part is dilated to form endolymphatic sac which lies between two layers of dura on the posterior surface of petrous bone.
- Endolymphatic sac:
 - **Site of endolymph absorption.**
 - The 1st to appear and the last to stop growth.
 - Surgically important, it is exposed for drainage or shunt operation in Meniere's disease.
 -



Inner Ear Fluids and their Circulation

1. Perilymph :

- Within the bony labyrinth.
- Resembles Extracellular fluid (ECF) and CSF.
- Rich in **Na** ions. ($\text{Na} > \text{K}$)
- Contributes to electrical potential of **0 mV** in scala vestibuli and scala tympani.
- Formed from the filtrate of blood and diffusion of CSF.
- Communicates with CSF through the **aqueduct of cochlea** which opens into the scala tympani near the round window.
- In fact this duct is not a direct communication but contains connective tissue resembling arachnoid through which perilymph percolates.
- Changes in blood composition are reflected much more rapidly in perilymph than in CSF.
- Perilymph leaves the ear by drainage through venules and through the middle ear mucosa.

2. Endolymph

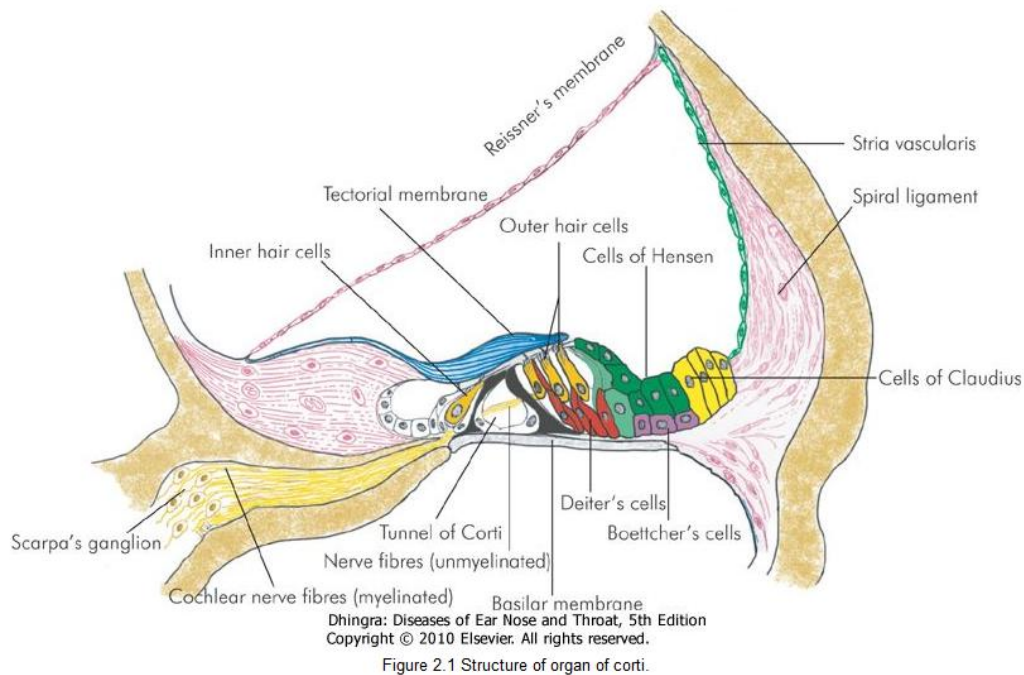
- Within the membranous labyrinth.
- Resembles Intracellular fluid (ICF).
- Rich in **K** ions. ($\text{K} > \text{Na}$)
- Contributes to positive DC resting potential of **+80 mV** in scala media
- Secreted by the secretory cells of the stria vascularis of the cochlea and by the dark cells (present in the utricle and also near the ampullated ends of semicircular ducts).
- Gets absorbed through endolymphatic sac which lies in the subdural space.

Table 1-2. Composition of inner ear fluids

	Endolymph	Perilymph	CSF
Na ⁺ (mEq/L)	5	140	152
K ⁺ (mEq/L)	144	10	4
Protein (mg/dL)	126	200-400	20-50
Glucose (mg/dL)	10-40	85	70

Organ of Corti

- Sense organ of hearing.
- Situated on the basilar membrane of Scala media.



Important components of the organ of corti:

1. Tunnel of Corti

- Formed by the inner and outer rods.
- Contains a fluid called **cortilymph** (similar to Perilymph).
- Exact function of the rods and cortilymph is not known.

2. Hair cells

- Receptor cells of hearing.
- Transduce mechanical sound energy into electrical energy.
- **Inner hair cells:**
- Principal transducer of motion from the basilar membrane to nerve impulse.
- Single row.
- Fewer in number.
- Rounded, Flask-like shape with nucleus in the center.
- Low intracellular glycogen.
- Few stereocilia in curvilinear shape.
- Loose connection to tectorial membrane.
- Completely surrounding by supporting cells.
- Afferent innervation: **Type I** (Radial, bipolar, myelinated), form 95% of fibers of the cochlear nerve.
- Each inner hair cell is innervated by 10-20 neurons (Low hair to ganglion ratio) --> cochlear nucleus.
- Efferent innervation: Sparse

➤ **Outer hair cells:**

- Cochlear amplifier which amplify motion from the basilar membrane.
- **Source of otoacoustic emissions.**
- 3 rows.
- More numerous.
- Cylindrical shaped with nucleus at base.
- High intracellular glycogen.
- Many stereocilia in "w" or "v" shape.
- Tight connection to tectorial membrane.
- Supported only at base.
- **Afferent innervation: Type II** (Spiral, pseudomonopolar, unmyelinated), form 5% of fibers of the cochlear nerve.
- Each 10 outer hair cells are innervated by one neuron (High hair to ganglion ratio) --> cochlear nucleus.
- **Efferent innervation:** from the auditory cortex down to the cochlear nuclei, additional contributions from the superior olive join and terminate predominantly on the outer hair cells.

3. Supporting cell

- Provide nutrients and structural support.
- Deiters' cells are situated between the outer hair cells and provide support to the latter.
- Hensens' cells lie outside the Deiters' cells.
- Claudius' cells

4. Tectorial membrane

- Fibrogelatinous structure
- Arises from the bony spiral lamina.
- Tips of stereocilia of the **outer hair cells** are partially embedded in the tectorial membrane.
- Vibration of the basilar membrane causes shearing forces at the tectorial membrane which then results in stimulation of the hair cells.

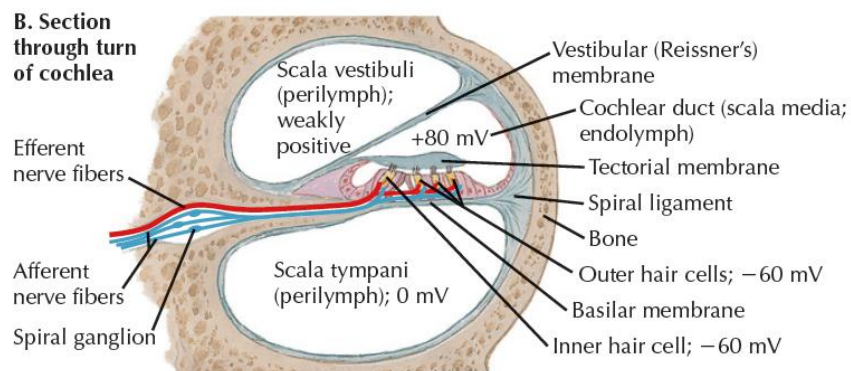


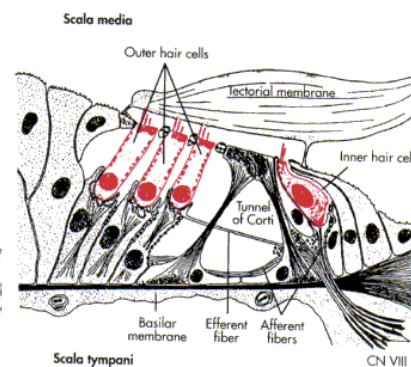
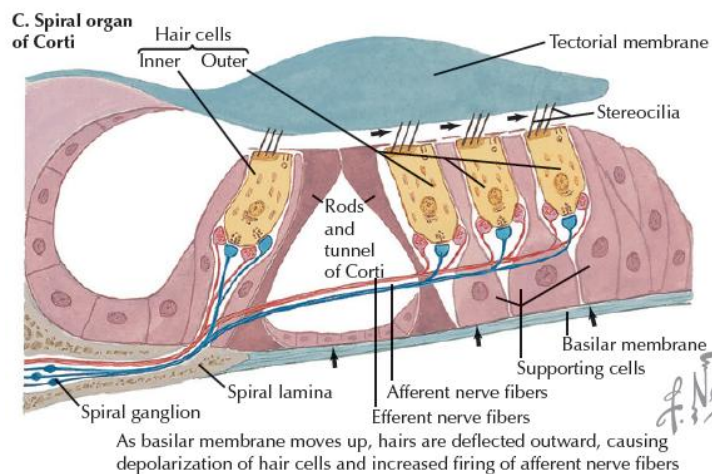
TABLE 129.1. STRUCTURE AND INNERVATION OF INNER AND OUTER HAIR CELLS

Characteristic	Inner Hair Cells	Outer Hair Cells
Number	3,500	12,000
Shape	Flask	Cylindrical
Stereocilia		
No. of hair cells	Few	Many
Arrangement	3 or 4 rows; rows slightly curved	6 or 7 rows; rows arranged in V or W shape
Attachment to tectorial membrane	None or loosely connected	Longest stereocilia firmly embedded
Ultrastructure		
Position of nucleus cell body	Center	Base
Cytoplasmic organelles	Scattered	Adjacent to cell membrane
Presynaptic specializations	Large	Small or absent (synaptic bars and vesicles)
Glycogen content	Low	High
Relation to supporting cells	Completely surrounded	Supported only at surface and base
Afferent innervation		
Ganglion cells	Type I	Type II
Number of ganglion cells	27,000	2,100
Hair cell to ganglion cell ratio	1.8:1	5.7:1
Efferent innervation		
Source	Lateral superior olivary complex	Medial superior olivary complex
Postsynaptic target	Afferent dendrites	Base of hair cell

From Neely JG, Dennis JM, Lippe WR. Anatomy of the auditory end organ and neural pathways. In: Cummings CW, ed. *Otolaryngology—head and neck surgery*. St. Louis: Mosby, 1986:2571, with permission.

Table 2-1. Differences between inner and outer hair cells

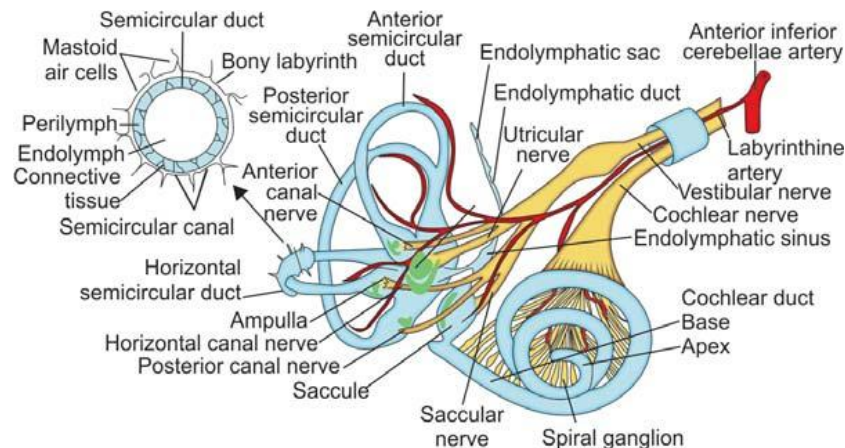
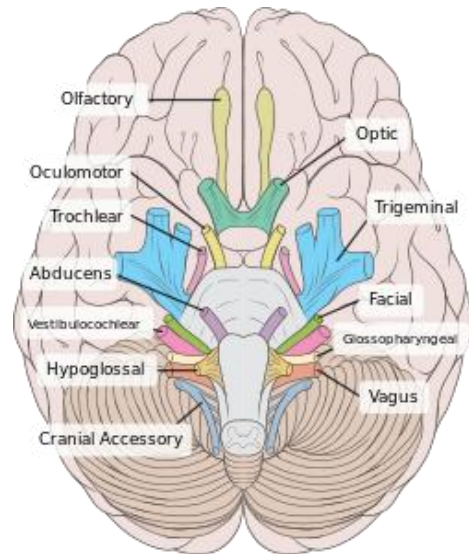
	Inner hair cells	Outer hair cells
Total no.	3500	12,000
Rows	One row	Three or four rows
Shape	Flask-shaped	Cylindrical
Nerve supply	Primarily afferent fibres and very few efferent	Mainly efferent fibres and very few afferent
Development	Develop earlier	Develop late
Function	Transmit auditory stimuli	Modulate function of inner hair cells
Vulnerability	More resistant	Easily damaged by ototoxic drugs and high intensity noise



- **Sensory Innervation of Inner Ear:**

- **Vestibulocochlear Nerve (CN VIII)**

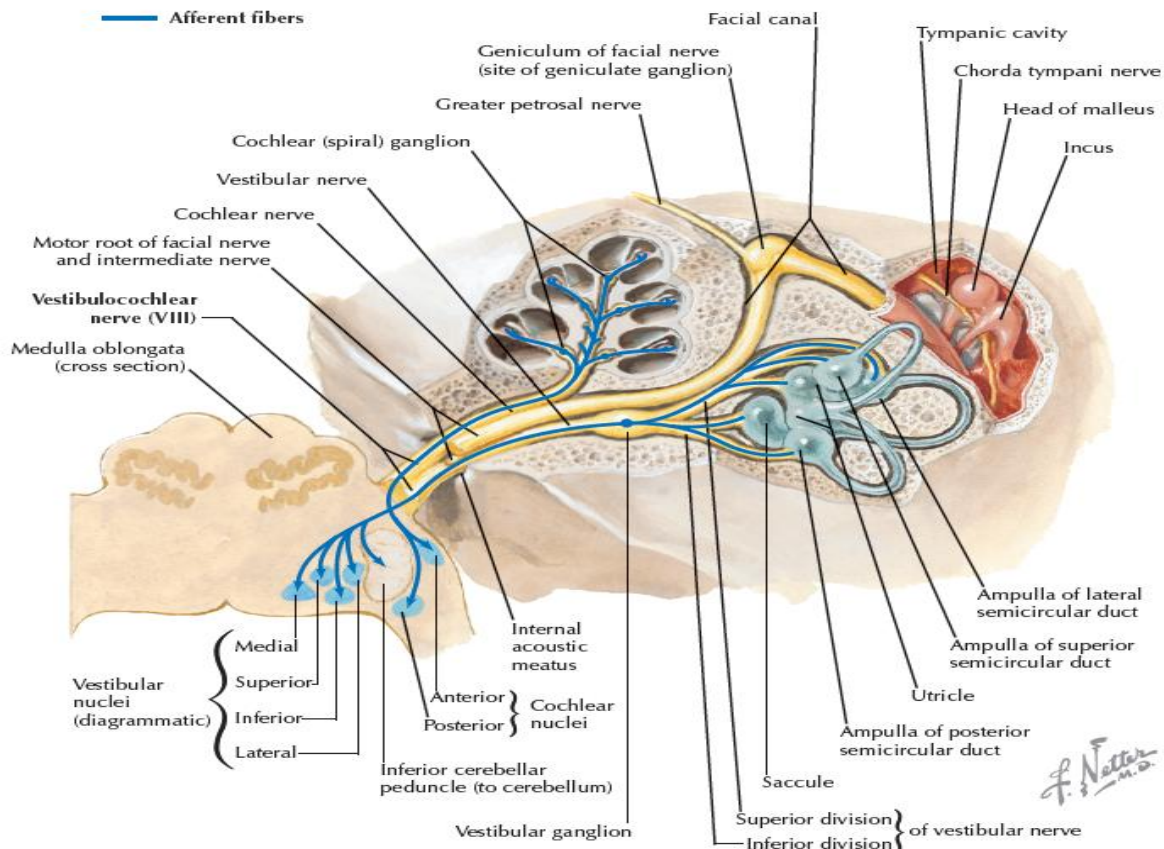
- Emerges between the pons and medulla oblongata.
- Enters the internal acoustic meatus with the Facial nerve
- Divides into vestibular branches and the cochlear branch.
- In the internal auditory meatus, the vestibular and cochlear nerves merge.
- During their course to the brainstem, the Facial nerve becomes located further up the brain.
- A small arterial branch from the **Anterior Inferior Cerebellar Artery (AICA)** runs between the CN-VII and CN-VIII on the brainstem.
- It can be seen during vestibular schwannoma surgery and be used as a landmark.



- **Vestibular Nerve:**

- Nerve cell bodies are located in the vestibular ganglion (Scarpa's ganglion).
- Divides into superior and inferior branches.
 - **Superior vestibular Nerve:**
 1. Ampulla of the **Superior** SSC.
 2. Ampulla of the **Lateral** SSC.
 3. Macula of the **Utricle**.
 4. The Antero-superior portion of macula of the saccule.
 - **Inferior vestibular Nerve:**
 1. Ampulla of the **Posterior** SSC.
 2. Main portion of macula of the **Saccule**.

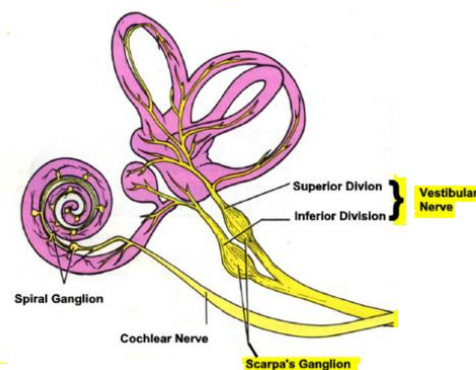
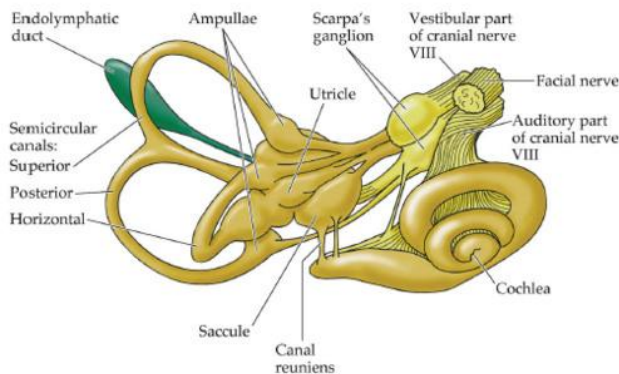
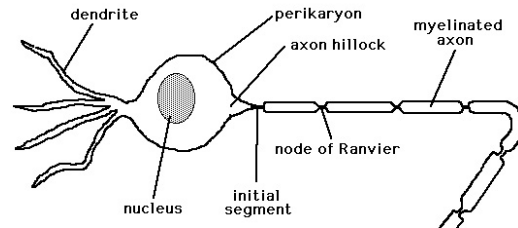
- **Auditory Sensory System:**
- **Cochlear Nerve:**
- Acoustic information from the hair cells is transferred by the Cochlear nerve to the ipsilateral cochlear nuclear complex in the brain stem.
- Cochlear nerve is composed of Afferent fibers from spiral ganglion neurones just central to the osseus spiral lamina.
- **Spiral ganglion:**
- Cell body of Cochlear nerve.
- Follows the course of the organ of Corti inside the modiolus.
- Sends two types of Afferent fibers type I that innervate the inner hair cells and type II that innervate the outer hair cells.
- Majority of spiral ganglion neurones (95%) are type I and innervate the inner hair cells.
- 50,000 neurons innervate cochlea
- **95% synapse directly on Inner hair cells (Type I neurons):**
 - Predominantly Afferent.
 - 10-20 of these neurons innervate each inner hair cells
- **5% synapse directly on Outer hair cells (Type II neurons):**
 - Predominantly Efferent.
 - Each type II neuron branches to innervate ~ 10 outer hair cells
- Efferent fibres to the hair cells come from the olivocochlear bundle. Their cell bodies are situated in superior olivary complex.
- Each cochlea sends innervation to both sides of the brain.



- **Vestibular Sensory System:**

- **Scarpa's ganglion:**

- Also called Vestibular ganglion.
- Cell body of vestibular nerve.
- Consists of bipolar neurons located in the lateral part of the internal auditory canal.
- Consists of superior and inferior group of cells associated with the superior and inferior vestibular nerve.
- Numbers of both vestibular hair cells and nerve cells in Scarpa's ganglion are found to be reduced in the ears of older people.
- Like the bodies of the human spiral ganglion, the perikarya of the vestibular ganglion cells are unmyelinated and surrounded by a thin sheath of Schwann cell.



- **Three distinct types of Vestibular Afferents**

1. Calyx:

- Terminate exclusively on **type I hair cells**
- May terminate on one or several hair cells
- Thicker axons
- Terminate in central zone of crista ampullaris and near striola
- Irregularly firing; sensitive to galvanic stim, not angular mov't

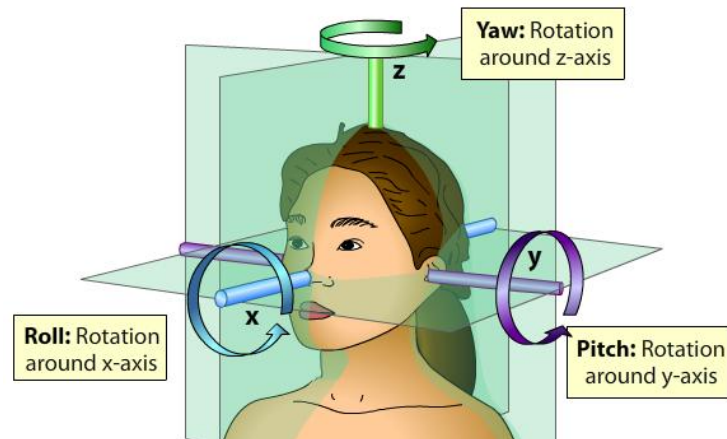
2. Dimorphic:

- Endings on **both** type I and type II hair cells
- Most prevalent (probably)
- Can be thin or thick
- Terminate in central, intermediate and peripheral zones
- Combo of both wrt sensitivity/firing; depends on location

3. Bouton:

- Terminate only on **type II hair cells**
- Thinner axons
- Terminate in peripheral zone of crista ampullaris
- Lower galvanic and natural stim thresholds; regular firing with slower conduction velocities.

- All vestibular epithelium has efferent fibres; function unknown.
- The vestibular system plays a Lesser role for the control of human posture and balance.
- The main peripheral component of the vestibular system is the labyrinth.
- Labyrinth has sensory cells known as Hair cells that transduce physical motion into neural impulses.
- Motions in the labyrinth are due to:
 - o Head movements
 - o Inertial effects due to gravity
 - o Ground-borne variations
- Labyrinth consists of:
 - o 2 otolith organs (Utricle and Saccule)
 - o 3 SCC.
- Utricle and saccule are specialized primarily to respond to:
 - o Linear accelerations of the head.
 - o Static head position relative to the gravitational axis
- SCC are specialized for responding to:
 - o Rotational accelerations of the head in 3 planes.



- **Neuroepithelium of Vestibular system:**
- Composed of sensory hair cells in contact with a gelatinous membrane.
- Hair cell bundles in each vestibular organ have specific orientations.
- The organ as a whole is responsive to displacements in ALL directions.
- **Glutamine** is the neurotransmitter.
- There are now data suggesting that the sensory vestibular epithelia may regenerate in mammals including man, which may explain the return of symptoms after gentamicin treatment in patients with Meniere's disease.

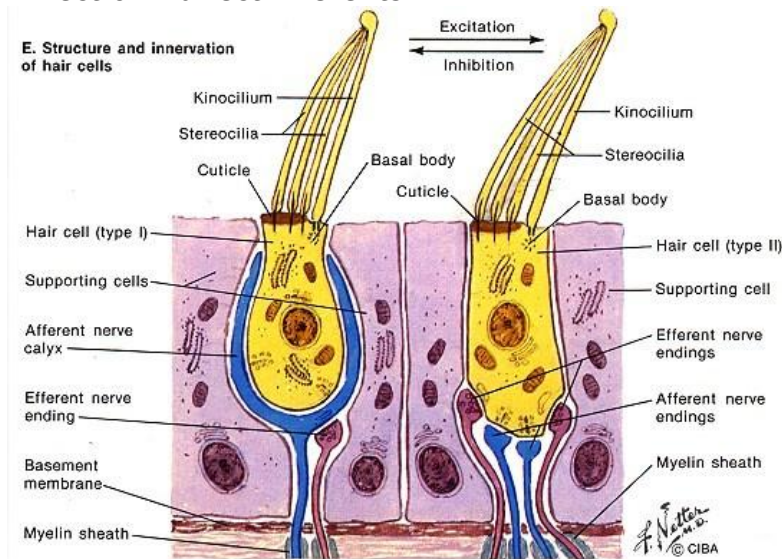
- Two types of sensory hair cells:

1. Type I:

- Flask-shaped.
- Surrounded by **one** nerve **Chalice** (cup or goblet) formed by the terminal end of the **Afferent nerve** fiber of the vestibular nerve.
- Correspond to the inner hair cells of the organ of Corti.
- Stereocilia & kinocilium arrangement.

2. Type II:

- Cylindrical in shape.
- Same arrangement of stereocilia & kinocilia as the type I cells.
- One or **more Afferents**
- Direct or indirect Efferents



- Hair cell arrangement:

- The upper surface of the hair cell contains approximately 70 stereocilia and one kinocilium arranged with the longest stereocilia positioned adjacent to the kinocilium.
- The upper surface of the cell contains a thicker region, called the '**Cuticular plate**', into which the stereocilia extend their rootlets.

1. Kinocilium:

- Tallest.
- Near edge of top hair cell
- A true cilium demonstrating the 9 + 2 arrangement of microtubules.

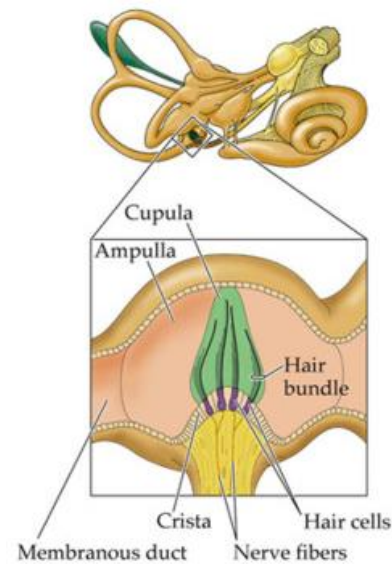
2. Stereocilia:

- Arranged in rows.
- Sterocilia closer to kinocilia are longer.

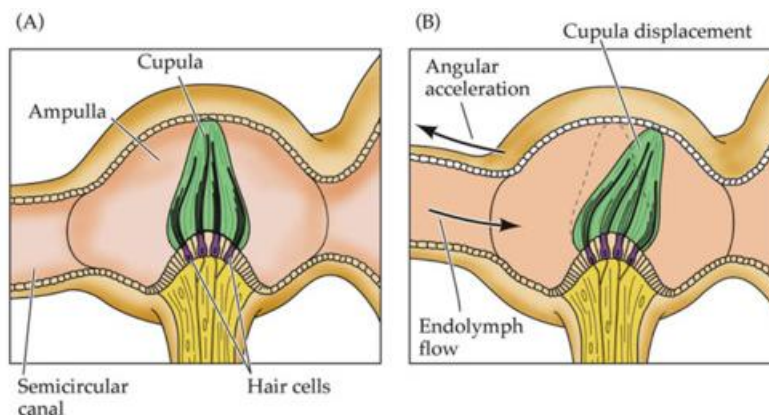
- Deflection of stereocilia **TOWARD** the kinocilium results in **Increased** vestibular neuronal firing rate.
- Deflection of stereocilia **AWAY** from the kinocilium results in **Decreased** vestibular neuronal firing rate.
- Hair cells in utricle and saccule act similarly to SCC in regard to the kinocilia and stereocilia, however, the utricle and saccule's hair cells are arranged in a specific pattern.
- Sensory Hair cells are found in:
 1. **Ampullae of SCCs.**
 2. **Macula of Otolith organs (utricle and saccule).**

Ampullae of SCC:

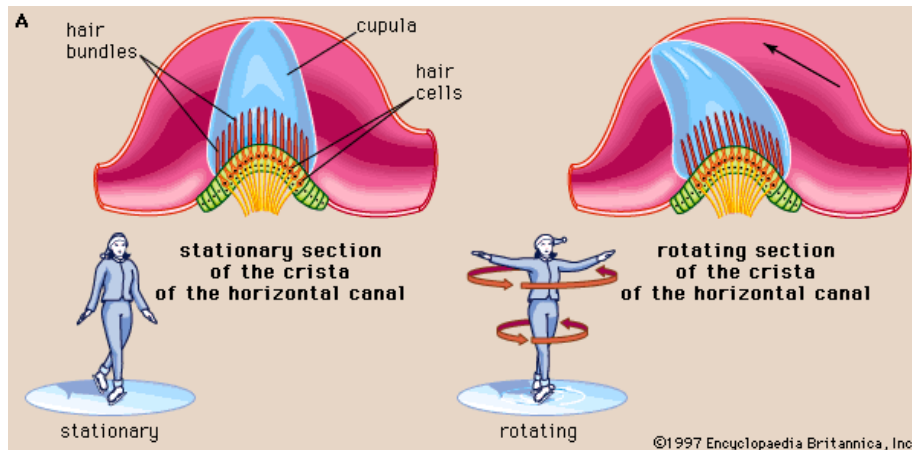
- Ampullae are bulbous expansion located at the base of SCC next to the utricle.
- Houses the sensory epithelium known as **Crista** that contains the hair cells
- Hair bundles extend out of the crista into a gelatinous mass known as the **Cupula**.
- Cupula bridges the width of the ampulla, forming a viscous barrier through which endolymph cannot circulate.
- The relatively compliant cupula is distorted by movements of the endolymph.
- When the head turns in the plane of one of the semicircular canals, the inertia of the endolymph produces a force against the cupula, distending it away from the direction of the head movement.



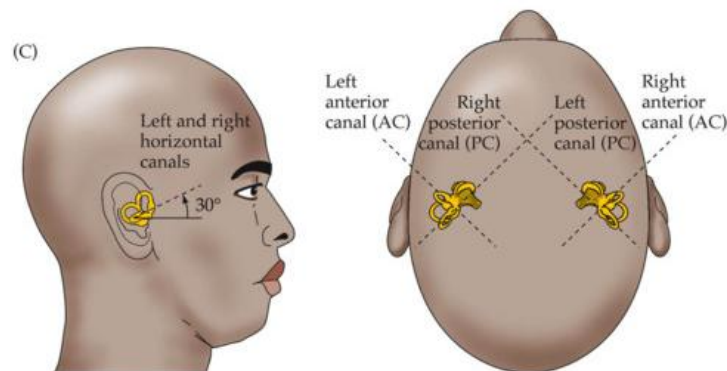
- Distention of the cupula away from the direction of head movement causes a displacement of the hair bundles within the crista.
- Linear accelerations of the head produce EQUAL forces on the 2 sides of the cupula, so the hair bundles are NOT displaced



- SCC sense Head rotation either arising from self induced movements or from angular accelerations of the head imparted by external forces.
- Unlike the saccular and utricular maculae, ALL hair cells in the crista within each SCC are organized with their kinocilia pointing in the SAME direction.
- When the cupula moves in the **Appropriate direction**, the entire population of hair cells is **Depolarized** and the activity in ALL of the innervating axons is **Increased**
- When the cupula moves in the **Opposite direction**, the population is **Hyperpolarized** and neuronal activity **Decreases**.

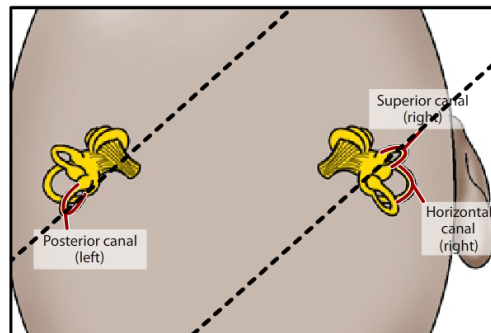


- The specific gravity of the cupula is approximately **1.0**, which is same as that of the Endolymph.
- This matching of the specific gravity is necessary to prevent the cupula from floating upward in certain head positions and causing an enduring nystagmus.
- Disruption of this match in specific gravity is likely the cause of **Postalccoholic Nystagmus**.

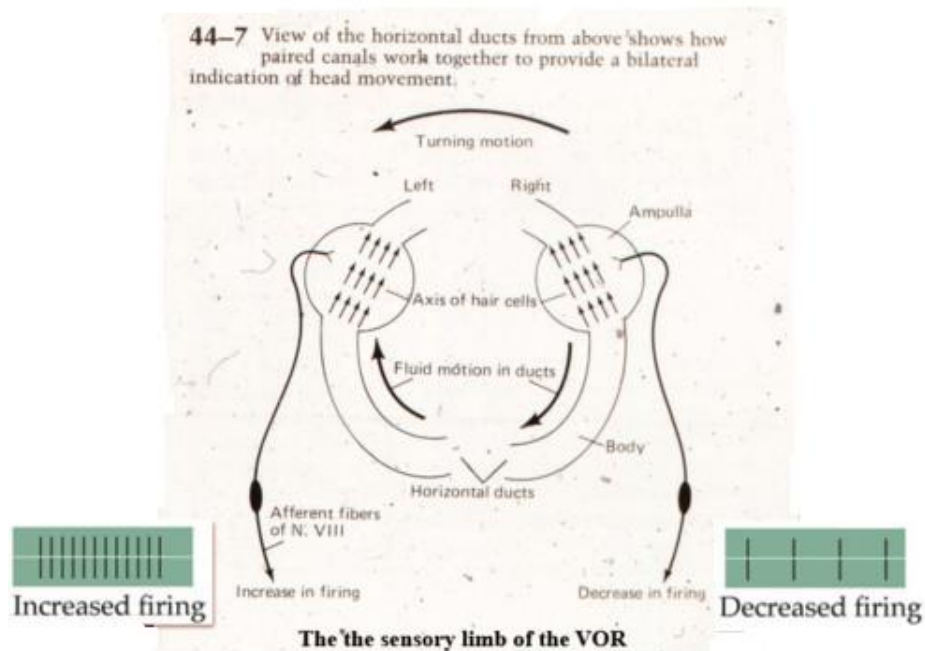


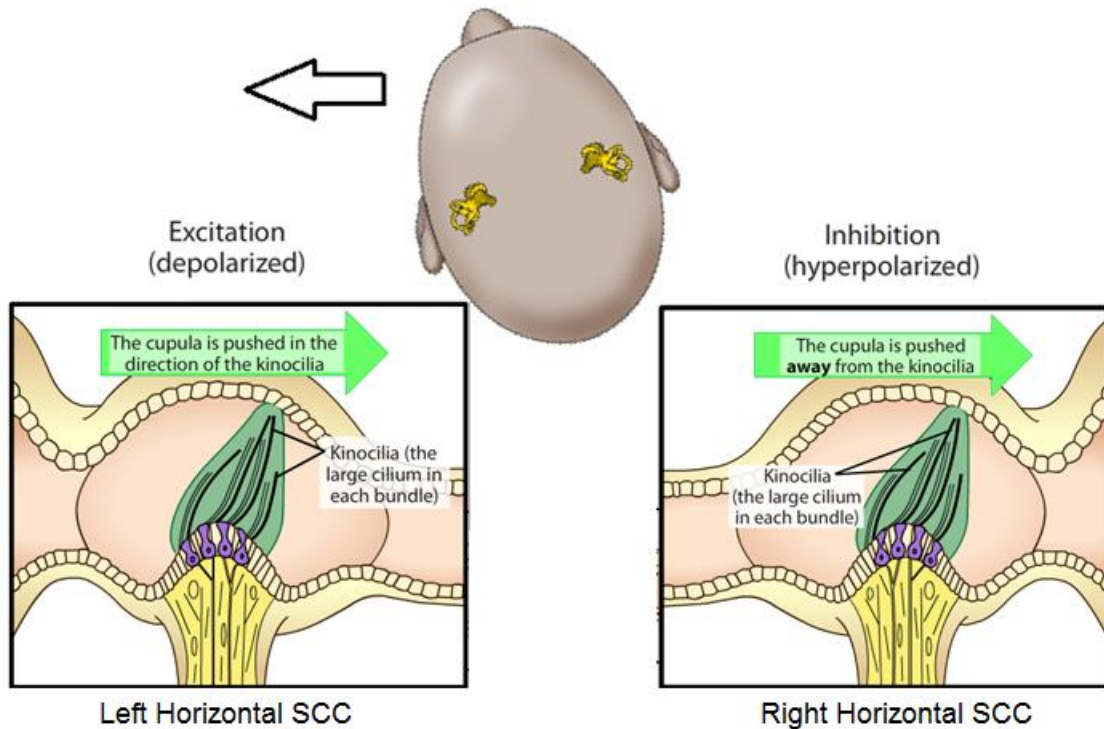
- Each semicircular canal works in concert with the partner located on the other side of the head that has its hair cells aligned oppositely.

- Three pairs:
 - 2 pairs of horizontal canals
 - Superior canal on each side working with the Posterior canal on the other side, both are in same plane.



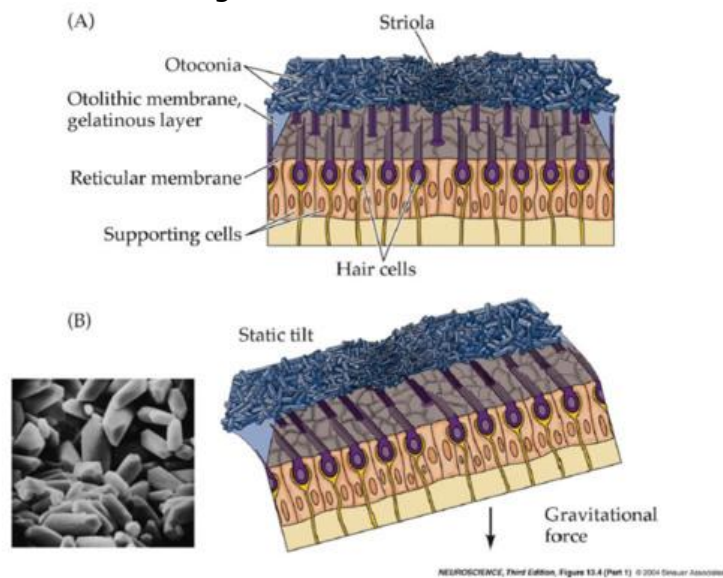
- Head rotation deforms the cupula in opposing directions for the 2 partners, resulting in opposite changes in their firing rates.
- Orientation of the horizontal canals makes them selectively sensitive to rotation in the horizontal plane.
- Hair cells in the canal Toward which the head is turning are Depolarized, while those on the other side are Hyperpolarized.



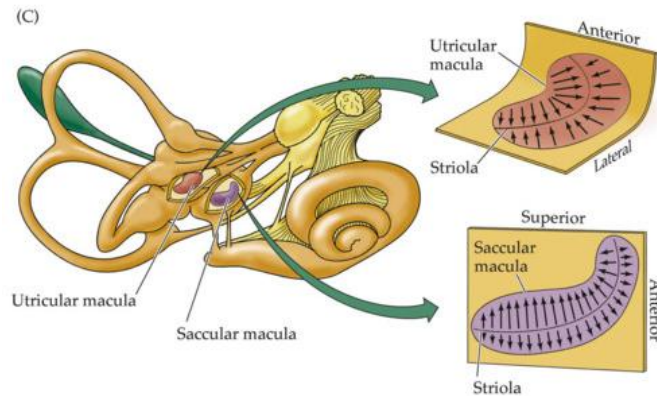


- **In Horizontal SCC:**
 - Kinocilia are located on the Utricular side.
 - Afferents are stimulated when endolymph flows against the utricle.
 - Displacement of stereocilia **TOWARD** the Utricle (**AmpulloPetal**) causes **Increased** vestibular neuronal firing rate (depolarization).
 - Displacement of stereocilia **AWAY** from the Utricle (**AmpulloFugal**) causes **Decreased** vestibular neuronal firing rate (Hyperpolarization).
- **In Vertical SCC (Posterior and Superior):**
 - Kinocilia are located on the Semicircular canal side.
 - Afferents are stimulated when endolymph flows against the Ampulla.
 - Displacement of stereocilia **TOWARD** the Utricle (**AmpulloPetal**) causes **Decreased** vestibular neuronal firing rate (Hyperpolarization).
 - Displacement of stereocilia **AWAY** from the Utricle (**AmpulloFugal**) causes **Increased** vestibular neuronal firing rate. (depolarization).

- **Macula of Otolith organs**
- Both the Saccule and Utricle contain a thickened sensory epithelium called Macula.
- Macula consists of :
 - Hair cells
 - Supporting cells
- **Gelatinous layer:**
 - Overlying the hair cells and their hair bundles.
- **Otolithic membrane:**
 - Fibrous structure above the gelatinous layer.
- **Otoconia:**
 - Also called Statoconia.
 - Calcium crystals containing material consisting of a multitude of small cylindrical and hexagonally shaped bodies with pointed ends
 - Anchored and partially embedded in the otolithic membrane.
 - Make the otolithic membrane heavier than the structures and fluids surrounding it.

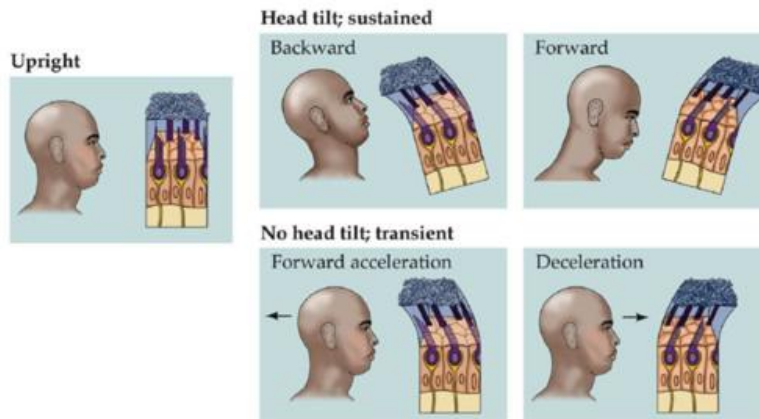
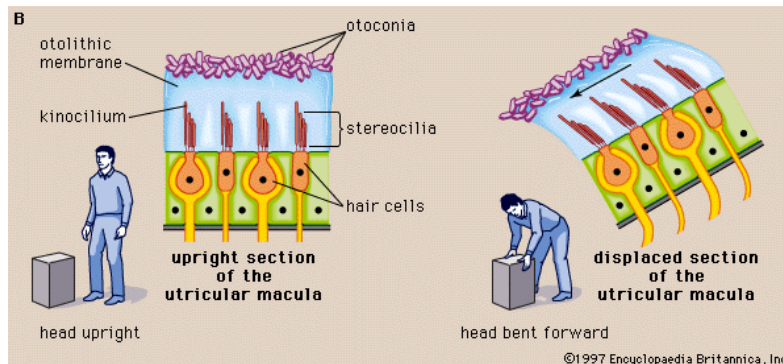


- Because of the heaviness by the otoconia, when the head tilts, gravity causes the membrane to shift relative to the sensory epithelium.
- The resulting shearing motion between the otolithic membrane and the macula displaces the hair bundles, which are embedded in the lower gelatinous surface of the membrane.

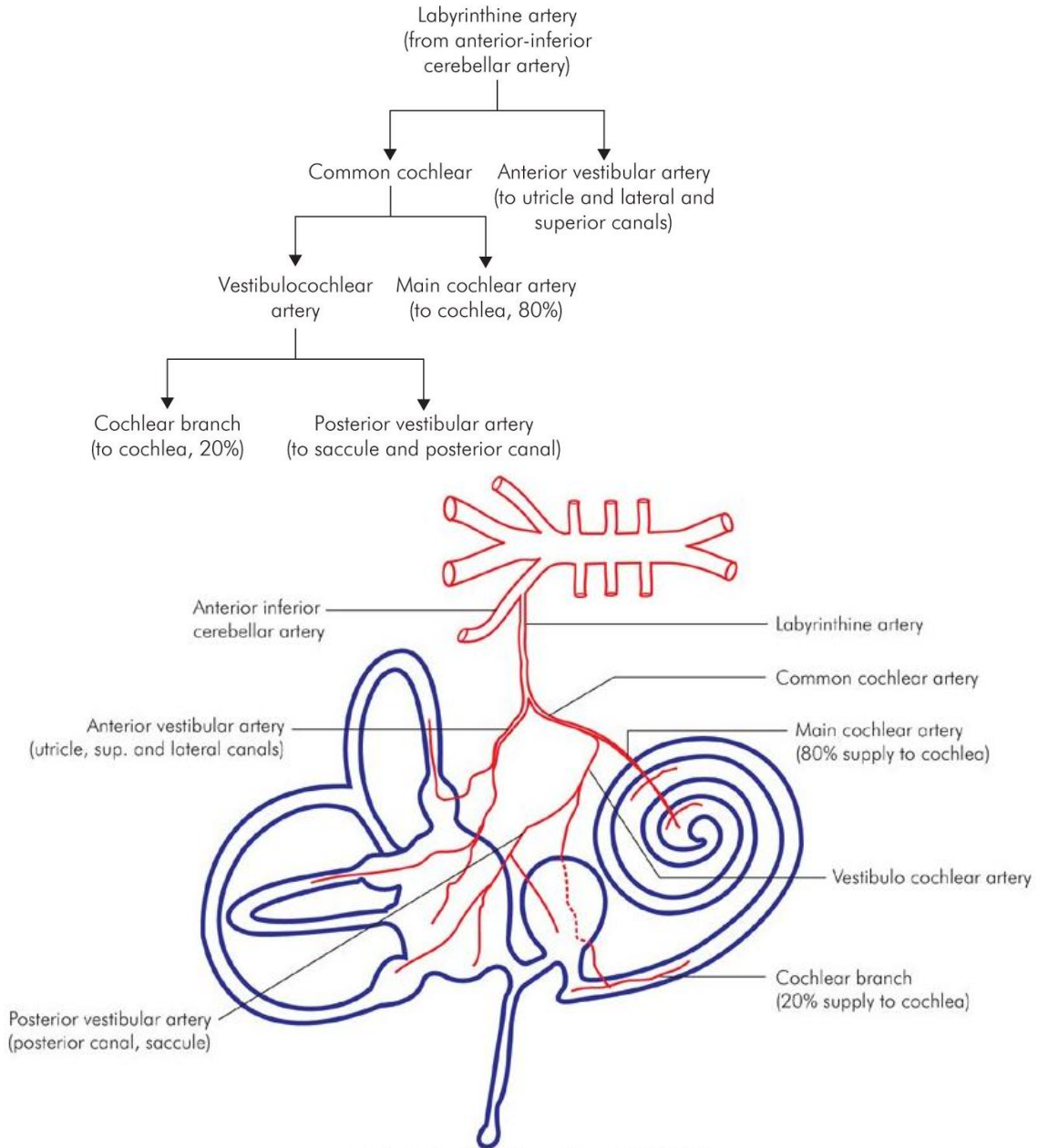


- **Striola:**
 - Specialized area in the utricle and saccule.
 - Divides the hair cells into 2 populations with **OPPOSING** polarities.
 - Contain more Type I cells.
 - Forms an axis of mirror symmetry such that hair cells on opposite sides of the striola have opposing morphological polarizations.
 - A tilt along the axis of the striola will excite the hair cells on one side while inhibiting the hair cells on the other.
 - Polarization of hair cells occurs point **TOWARD** striola in the **Utricular macula** and **AWAY** from the striola in the **saccular macula**.
- **Saccular macula:**
 - Oriented in the **VERTICAL** plane.
 - Detects Vertical linear acceleration and change in Gravity.
 - Ex: Up/down and forward backward movements in the sagittal plane.
 - Polarization of hair cells occurs point **AWAY** the striola.
- **Utricular macula:**
 - Oriented in the **HORIZONTAL** plane.
 - Detects Horizontal linear acceleration.
 - Ex: Sideways head tilts and rapid lateral displacements would be detected by the utricle.
 - Polarization of hair cells occurs point **TOWARD** the striola.
- Structure of the otolith organs enables them to sense both **Static** and **Transient** displacements.
- Tilting the head relative to the gravitational axis would be a **Static** displacement sensed by the otolith organs
- Translational movements of the head would be a **Transient** displacement sensed by the otolith organs.

- Note that the maculae respond only to changes in acceleration or velocity of head movement, they do NOT report on unchanging head positions.



- Other cell types of Macula:
 - 1. Supporting cells:**
 - Secrete Extracellular macromolecules of cupula and otolith membrane.
 - 2. Dark cells:**
 - Found in all sensory epithelia of membranous labyrinth, except the saccule which is believed to lack production of endolymph.
 - Produce ionic composition of **Endolymph**.
 - Involved in the degradation and resorption of dislodged otoconia.
- **Blood Supply of Inner Ear:**
- **Labyrinthine artery**
 - Supplies the entire labyrinth.
 - Branch of the Anterior inferior cerebellar artery (AICA) (**85%-100% cases**) or Basilar artery (**<15% cases**).
 - Accompanies the vestibulocochlear nerve through the internal acoustic meatus.
 - Divides into cochlear and vestibular branches that supply the cochlear and vestibular structures



- **Anterior Vestibular Artery:**
 - Superior portion of Utricle and Sacule
 - Superior and Horizontal SCC.
- **Posterior Vestibular Artery:**
 - Inferior portion of Utricle and Sacule
 - Posterior SCC.
- **Main Cochlear Artery:**
 - ALL cochlea except One-Third of Basal turn.
- **Cochlear Branch:**
 - One-Third of Basal turn.

- Venous drainage is through three veins, namely **internal auditory vein**, **vein of cochlear aqueduct** and **vein of vestibular aqueduct** which ultimately drain into inferior petrosal sinus and lateral venous sinus.
- Blood supply to the inner ear is independent of blood supply to middle ear and bony otic capsule, and there is no cross circulation between the two.
- Blood supply to cochlea and vestibular labyrinth is segmental, therefore, independent ischemic damage can occur to these organs causing either cochlear or vestibular symptoms.
- NO LYMPHATIC DRAINAGE OF INNER EAR.

Eustachian Tube Anatomy:

- Pharyngotympanic Tube.
- A dynamic channel that links Middle ear with Nasopharynx.
- Derived from **1st Pharyngeal Pouch**.
- In Adults:
 - o 36 mm in length
 - o Reach adult size at age 7.
 - o Runs downwards, forwards and medially from its tympanic end.
 - o Forming an angle of 45° with the horizontal.

- Divided into two parts:

1. Bony Part:

- o Lateral 1/3.
- o 12 mm in length.
- o Runs through Squamous and Petrous part of Temporal bone.
- o Thin plate of bone forms the Roof, separating ET from Tensor Tympani muscle above.
- o Carotid canal lies Medially and can impinge on Bony ET.
- o In cross section, ET is Triangular or rectangular with the horizontal diameter being the greater.
- o Tympanic End of ET:
 - At the Lateral end of the Bony part.
 - Oval in shape
 - Measures 5×2 mm.
 - Situated in Anterior wall of Middle Ear, above level of floor.
- o Isthmus:
 - Narrowest part of ET.
 - 0.5 mm in Diameter.
 - At Junction of Bony part and Fibrocartilaginous part.

2. Fibrocartilaginous Part:

- o Medial 2/3.
- o 24 mm in length.
- o Made of a single piece of cartilage folded upon itself.
 - Forms the Medial lamina, Roof and a part of Lateral lamina.
 - Rest of Lateral lamina is made of Fibrous membrane.
- o Nasopharyngeal End of ET:
 - At the Medial end of the Fibrocartilaginous part.
 - Slit-like, vertically.
 - The cartilage at this end raises an elevation called **Torus Tubarius**:
 - Situated in Lateral wall of Nasopharynx.
 - 1-1.25 cm behind Posterior end of Inferior Turbinate.

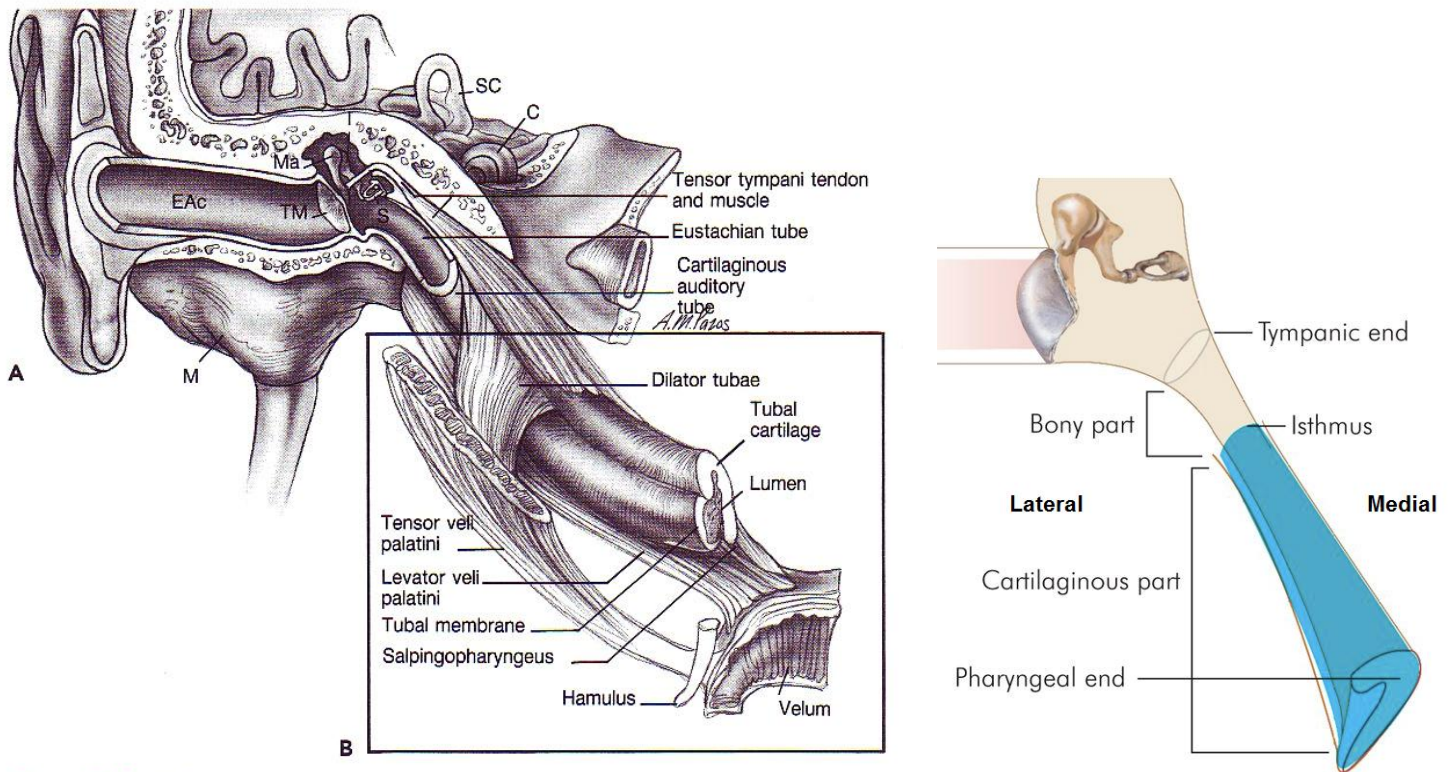
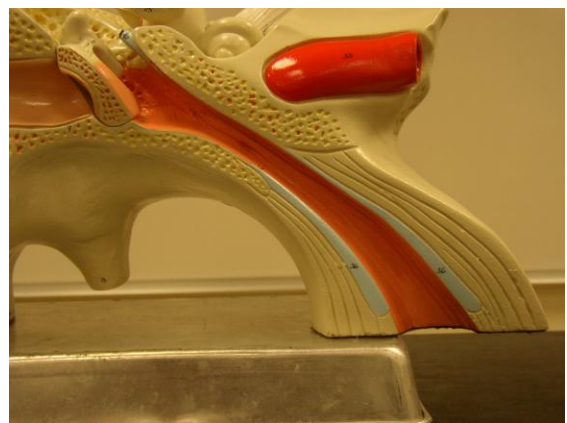
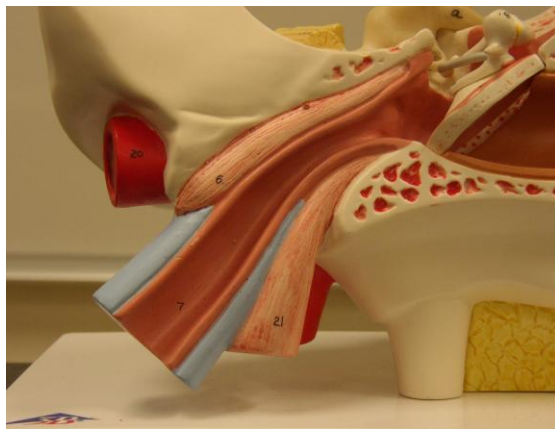


Figure 90.2 A: Complete dissection of the eustachian tube and middle ear. Especially evident are the relations between the eustachian tube, paratubal muscles, and cranial base, as well as the position of the juncture between the osseous portion of the eustachian tube and the middle ear. B: Relation between the superficial muscle bundle (tensor veli palatini) and the deep bundle (dilator tubae) to the lateral wall of the eustachian tube. C, cochlea; EAC, external auditory canal; I, incus; M, mastoid; Ma, malleus; S, stapes; SC, semicircular canals; TM, tympanic membrane.

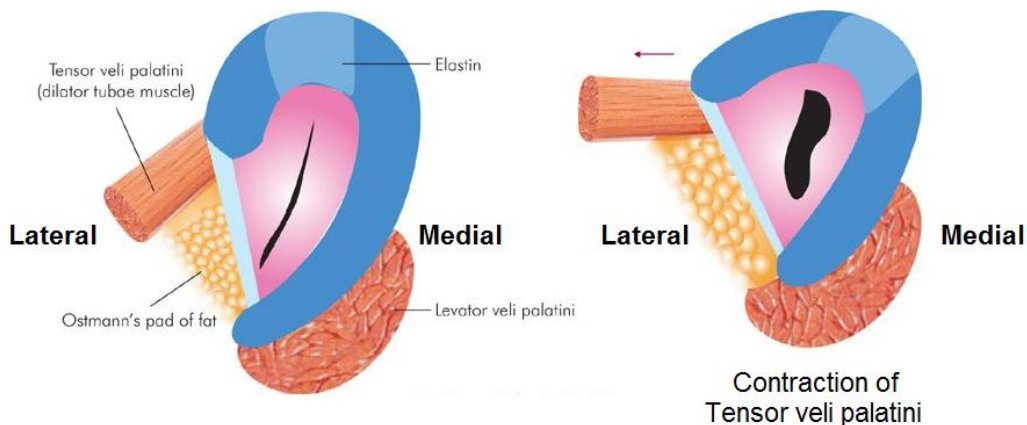


- **Lining of Eustachian Tube:**
- Pseudostratified ciliated columnar epithelium with mucous secreting goblet cells (**Respiratory Mucosa**).
- At Nasopharyngeal end of ET, the mucosa is truly respiratory; but in passing along the tube towards Middle ear, Number of goblet cells and glands decreases, and the ciliary carpet becomes less profuse.
- Cilia beat in the direction of Nasopharynx and helps to drain secretions and fluid from Middle ear into Nasopharynx.
- Submucosa of Cartilaginous part of ET is rich in seromucinous glands.

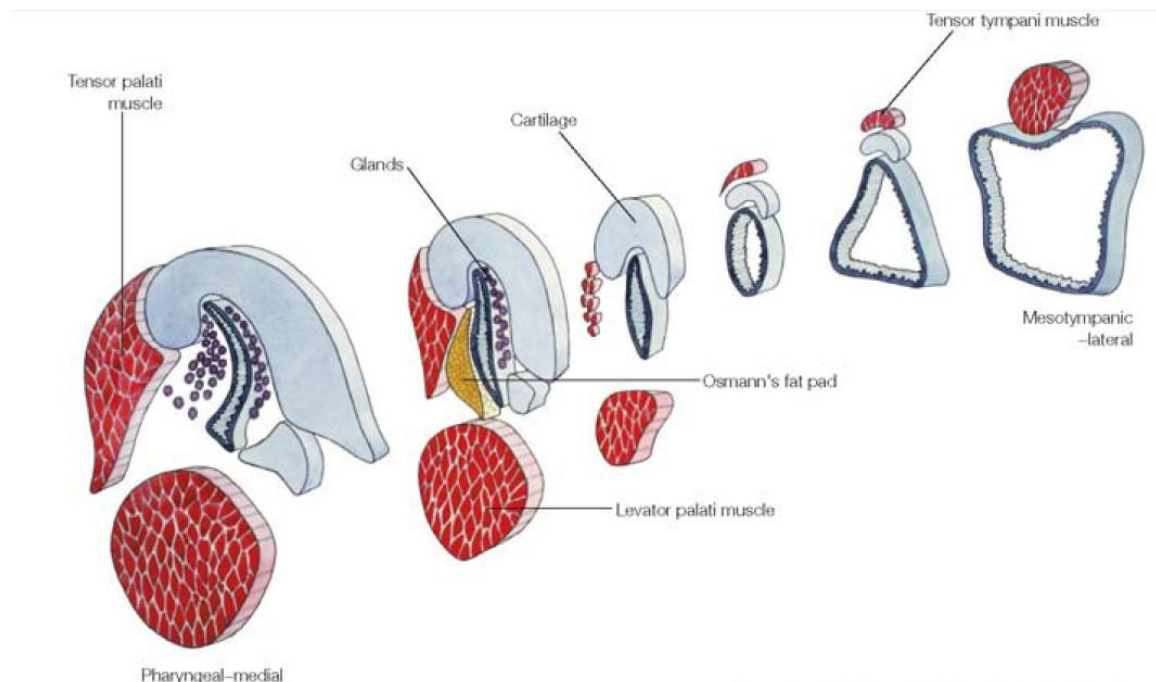
- **Muscles Related to Eustachian Tube:**

1. Tensor veli palatini.
2. Levator veli palatini.
3. Tensor Tympani.
4. Salpingopharyngeus.

MUSCLE	ORIGIN	INSERTION	NERVE SUPPLY	ACTION
Tensor Veli Palatini (Dilator Muscle)	Spine of Sphenoid. Eustachian Tube.	Forms the Palatin Aponeurosis with Tensor Veli Palatini from other side.	Nerve to Medial Pterygoid from Mandibular Branch (V3) of Trigeminal Nerve CN-V	<u>Medial fibers</u> are attached to Lateral lamina of ET. Contracts to open ET lumen. Tenses soft palate
Levator Veli Palatini	Petrous part of Temporal bone. Eustachian Tube.	Palatin Aponeurosis	Pharyngeal Plexus (Vagus Nerve CN-X)	Assist in opening ET. Raises the Palate.
SalpingoPharyngeous	Eustachian Tube.	Blends with PalatoPharyng eous.	Pharyngeal Plexus (Vagus Nerve CN-X)	Assist in opening ET. Elevates Pharynx.
Tensor Tympani	Wall of ET and wall of Its own canal.	Neck of Malleus.	Mandibular Branch (V3) of Trigeminal Nerve CN-V	Dampens down Vibrations of TM.



- **Elastin Hinge:**
- Elastin fibers found in junction of Medial and Lateral lamina at the Roof of Cartilaginous part of ET.
- Keeps ET closed by its recoil, when no longer acted upon by Tensor Veli Palatini (**Dilator Muscle**).
- **Ostmann's Fat Pad:**
- Mass of fatty tissues related **Laterally** to Membranous part of Cartilaginous tube.
- Keeps ET closed and protect it from reflux of Nasopharyngeal secretions.



- **Arterial blood supply of ET:**
 1. Ascending Pharyngeal Artery.
 2. Middle Meningeal Artery.
- **Venous blood drainage of ET:**
- Pharyngeal plexus
- **Lymphatics drainage of ET:**
- Retropharyngeal nodes.
- **Nerve Supply of ET:**
- Tympanic branch of **CN-IX:**
 - o Sensory and Parasympathetic Secretomotor Fibers to ET mucosa.

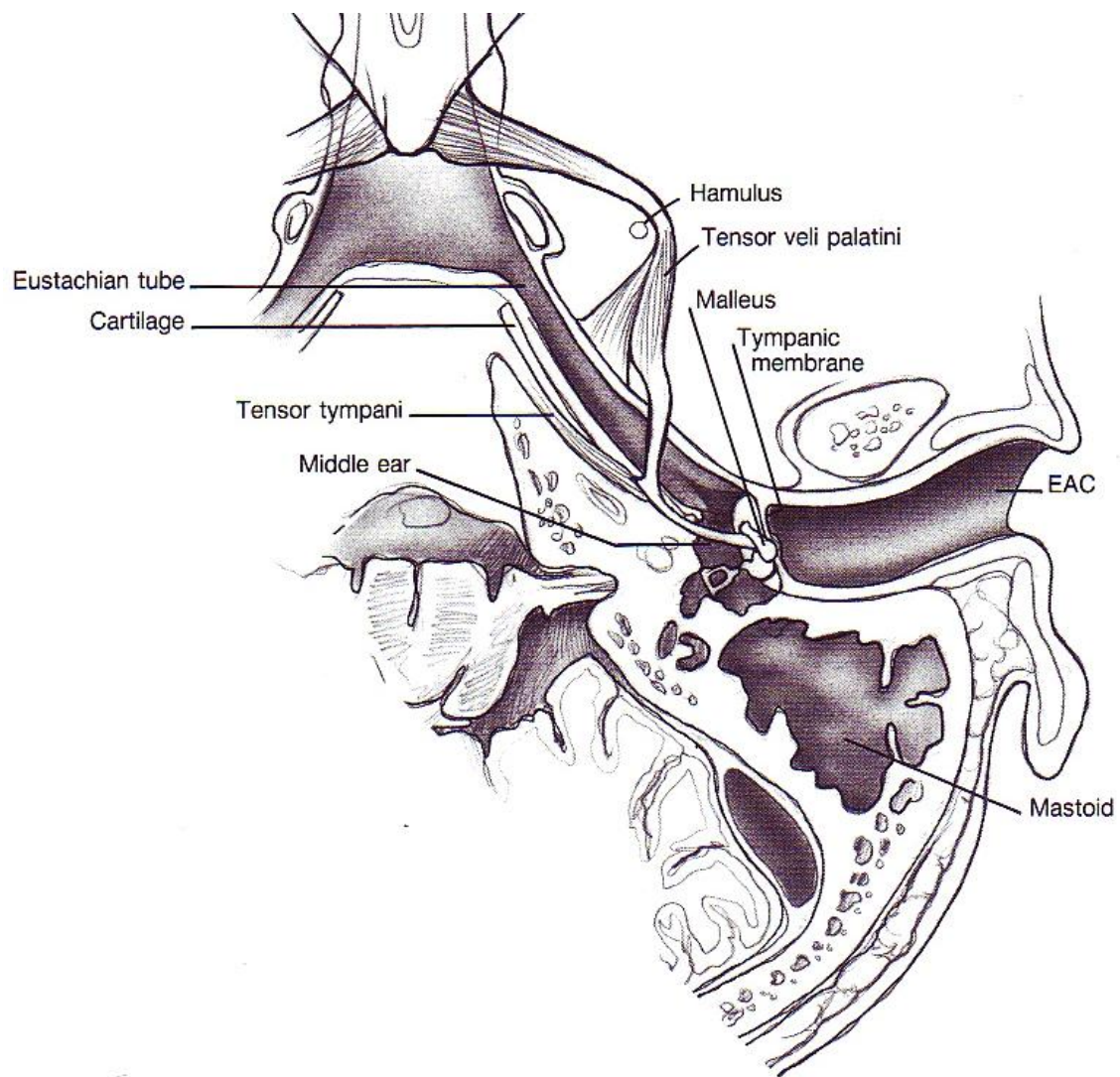


Figure 90.3 Drawing shows the tensor veli palatini muscle attachment along the lateral wall of the eustachian tube, including its course around the hamulus of the pterygoid bone and its attachment into the posterior margin of the hard palate. EAC, external auditory canal.

- **Eustachian Tube in Infants:**
 - Shorter.
 - Wider.
 - More Horizontal.
 - More susceptible to regurgitation from Nasopharynx into Middle Ear with feeding.

Table 9-1. Differences between infant and adult eustachian tube

	Infant	Adult
Length	13-18 mm at birth (about half as long as in adult)	36 mm (31-38 mm)
Direction	More horizontal. At birth it forms an angle of 10° with the horizontal At age 7 and later it is 45°	Forms an angle of 45° with the horizontal
Angulation at isthmus	No angulation	Angulation present
Bony versus cartilaginous part	Bony part is slightly longer than 1/3 of the total length of the tube and is relatively wider	Bony part 1/3; cartilaginous part 2/3
Tubal cartilage	Flaccid. Retrograde reflux of nasopharyngeal secretions can occur	Comparatively rigid. Remains closed and protects the middle ear from the reflux
Density of elastin at the hinge	Less dense; tube does not efficiently close by recoil	Density of elastin more and helps to keep the tube closed by recoil of cartilage
Ostmann's pad of fat	Less in volume	Large and helps to keep the tube closed

- **Movement of ET:**
 1. Passive closure by:
 - Elastic Recoil.
 - Passive pressure from surrounding tissue.
 2. Active opening by:
 - Tensor veli platini muscle.
- **Main Functions of Eustachian Tube:**
 1. Ventilation and Regulation of Middle Ear Pressure.
 2. Protection against:
 - Nasopharyngeal sound pressure.
 - Reflux of Nasopharyngeal secretions.
 3. Middle Ear clearance and drainage of secretions.
- **Ventilation and Regulation of Middle Ear Pressure:**
- For Normal hearing:
 - Pressure on two sides of TM should be Equal.
 - Negative or Positive Pressure in Middle ear affects hearing.
- ET should open periodically to equilibrate Air pressure in Middle ear with Ambient pressure.
- Normally, ET remains closed and opens intermittently during:
 - Swallowing
 - Yawning
 - Sneezing
- **ET opening function is Poor in:**
 - Recumbent position and during sleep due to Venous Engorgement.
 - Infants and Young children.

- **Protective Functions:**
- High sound pressures from Nasopharynx can be transmitted to Middle ear if Tube is open and interfering with normal hearing (**Patulous Eustachian Tube**)
- ET remains closed normally and protects Middle ear against these sounds.
- ET also protects from reflux of Nasopharyngeal secretions into the Middle ear.
- This reflux occurs more readily if:
 - ET is wide in diameter (patulous tube).
 - Short in length, (infants).
 - Perforated TM (cause for persistence of middle ear infections).
 - High pressures in Nasopharynx (Forceful nose blowing, closed-nose swallowing as in adenoid hypertrophy or bilateral nasal obstruction)
- **Clearance of Middle ear secretions:**
- Mucous membrane of ET and Anterior part of Middle ear is lined by Ciliated columnar cells.
- Cilia beat in direction of Nasopharynx to clear secretions and debris in Middle ear.
- Clearance function is further augmented by active opening and closing of the tube.
- **Eustachian Tube Function Tests**
- 1. Valsalva Test.
- 2. Toynbees's Test
- 3. Tympanometry
- **Valsalva Test:**
- Principle:
 - To build positive pressure in Nasopharynx so that Air enters ET into Middle ear.
- Method:
 - Exhalation against a closed airway.
 - Closing the Mouth and pinching the Nose.
- Results:
 - Normally:
 - Air enters Middle ear through ET.
 - TM move outwards (by Otoscope).
 - TM perforation:
 - Hissing sound is produced.
 - Failure of this test does not prove blockage of ET because only 65% of persons can successfully perform this test.
- Contraindications:
 - Presence of atrophic scar in TM which can rupture.
 - Presence of infection of Nose and Nasopharynx (Risk of Reflux).



- **Toynbee's Test:**
- **Principle:**
 - To build Negative pressure in Nasopharynx so that Air enters ET From Middle ear.
- **Method:**
 - Swallowing with pinched Nose.
- **Results:**
 - Normally:
 - Air enters Nasopharynx from Middle ear through ET.
 - TM move inwards (by Otoscope).
- **Tympanometry (Inflation-deflation Test):**
- **Principle:**
 - Normal ET is able to equilibrate Middle ear and Ambient pressure.
- **Method:**
 - Positive and Negative pressures (-200 or +200 mm of H2O) are created in EAC.
 - Patient swallows repeatedly (5 times in 20 seconds).
- **Results:**
 - Normally:
 - Equilibration of positive and negative pressures to Ambient pressure.
 - Done both in patients with perforated or intact TM.
 - Can be used to find the patency of VT.
- **Disorders of Eustachian Tube (ET)**
- **ET Obstruction:**
- Normally, ET is closed.
- Opens intermittently during swallowing, yawning and sneezing due to contraction of Tensor veli palatini muscle.
- Air (O₂, Co₂, Nitrogen, Water vapour) normally fills Middle ear and Mastoid.
 - Lower O₂ & CO₂ and higher N₂ compared to Atmosphere.
 - Similar to venous gas.
- **If ET is Blocked:**
 - Initially, Oxygen is Absorbed then other gases are Absorbed.
 - → Negative pressure in Middle ear.
 - → Retraction of TM.
 - → If Negative pressure is increased
 - → ET will be locked.
 - → Collection of Transudate and later Exudate.
 - → Atelectatic ear / Perforation.
 - → Retraction pocket / Cholesteatoma.
 - → Erosion of Incudo-Stapedial joint.

- **Causes of ET Ostruction**

1. URTI.

2. Allergy.

3. Sinusitis.

4. DNS.

5. Hypertrophic Adenoid:

- Mechanical obstruction of ET.
- Reservoir for Pathogenic organisms.
- Adenoidectomy helps with OME and Recurrent AMO.

6. Nasopharyngeal Tumor/mass.

7. Cleft Palate:

- Abnormalities of Torus Tubarius (High elastin density making tube difficult to open).
- Tensor veli palatini muscle does not insert into Torus tubarius in 40% cases of cleft palate.
- OME is common even after cleft palate repair.
- Requires insertion of VT.

8. Down's syndrome.

- Poor tone of Tensor Veli Palatini muscle.
- Abnormal shape of Nasopharynx.
- OME is common.
- Requires insertion of VT.

- **Signs and Symptoms of ET Obstruction:**

- Otalgia, CHL, Popping sensation, Tinnitus and Vertigo.
- Retracted TM, Congested TM, Transudate behind TM imparting it an Amber colour and a Fluid level.

- **Retraction Pockets and ET:**

- Air passes from ET to Mesotympanum, Attic, Aditus, Antrum and Mastoid air cell system.

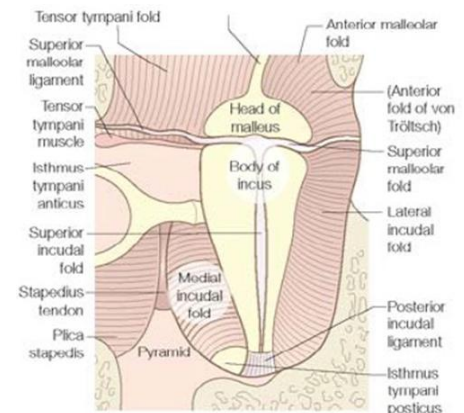
- **Mesotympanum communicates with Attic via:**

1. Anterior Isthmus:

- Between Tensor Tympani tendon and Stapes.

2. Posterior Isthmus:

- Between Stapedius tendon and Pyramid, and Short Process of Incus.



- Any obstruction in the pathways of ventilation can cause Retraction pockets or Atelectasis of TM:
 - **Obstruction of ET** → Total Atelectasis of TM.
 - **Obstruction in Middle ear** → Retraction pocket in posterior part of Middle ear while Anterior part is ventilated.
 - **Obstruction of Isthmi** → Attic Retraction pocket.
 - **Obstruction at Aditus** → Cholesterol granuloma and collection of mucoid discharge in mastoid air cells, while Middle ear and Attic appear Normal.
- Principles of Management of Retraction pockets and Atelectasis of middle ear would entail Correction/repair of the irreversible pathologic processes and establishment of the ventilation.
- **Patulous Eustachian Tube:**
- ET is abnormally patent.
- Mainly idiopathic.
- Acquired:
 - Scarring post Adenoidectomy.
 - Rapid weight loss.
 - Pregnancy (Third trimester).
 - Multiple Sclerosis.
- **Signs and Symptoms:**
- Distorted Autophony (Abnormal perception of one's own breath and voice sounds – **most common**)
 - Echoing occasionally severe enough to interfere with speech production
- Fluctuating Aural fullness.
- Roaring Tinnitus synchronous with nasal respiration.
- Audible respiratory sounds.
- Synchronous TM movement with Respiration can be seen on otoscopy.
- Symptoms improve when Supine.
- Self-limiting and does not require treatment.
- Weight gain, oral administration of potassium iodide is helpful but some long-standing cases may require cauterisation of the tubes or insertion of a grommet.

Physiology of Auditory System:

External Ear

- ✓ Head and external ear play passive role in hearing
- ✓ pinna helps to differentiate sound in front of or behind listener
- ✓ EAC: open at one end, so behaves like a quarter-wave resonator;
- ✓ resonant frequency (f_0) depends on length, not curvature (for 2.5 cm, it **equals 3.5 kHz (8 kHz for infants)**)
- ✓ Head: attenuates sounds if width of head is greater than sound's wavelength (> 2 kHz); called **head shadow effect**;
- ✓ interaural intensity difference of **5-15 dB** helps with localization;
- interaural time difference of **0.6 ms** helps too
- ✓ Interaural intensity difference.
- ✓ Interaural time difference.
- ✓ Pinna effect.
- ✓ The auditory brainstem processes interaural timing and amplitude differences between ears to determine the location of the sound source.

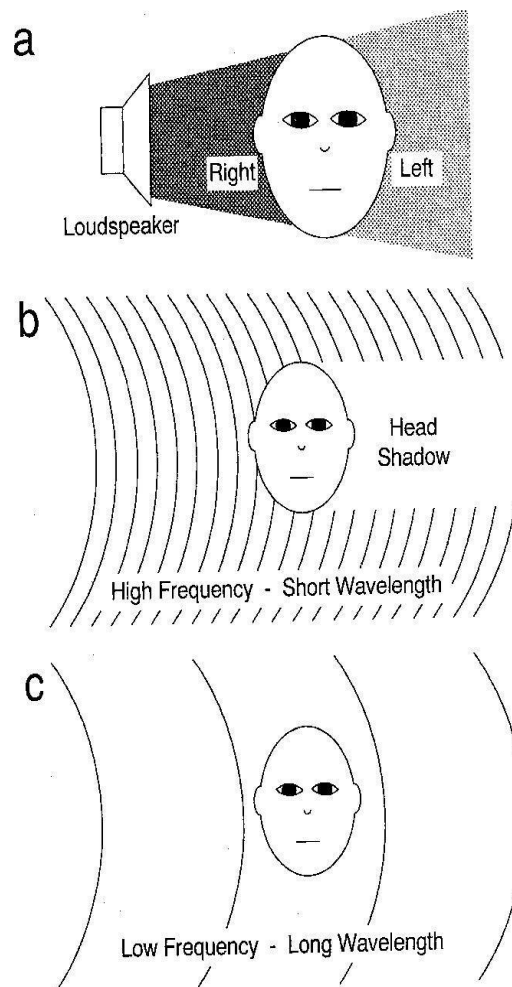


FIGURE 2-17 Sound coming from a loudspeaker off to the right side (a) arrives differently at the two ears. An acoustical shadow (the head shadow effect) occurs for high-frequency sounds because their wavelengths are small compared to the size of the head (b). Because of their large wavelengths, low-frequency sounds are not subjected to a head shadow because they are able to bend around the head (c).

Middle Ear

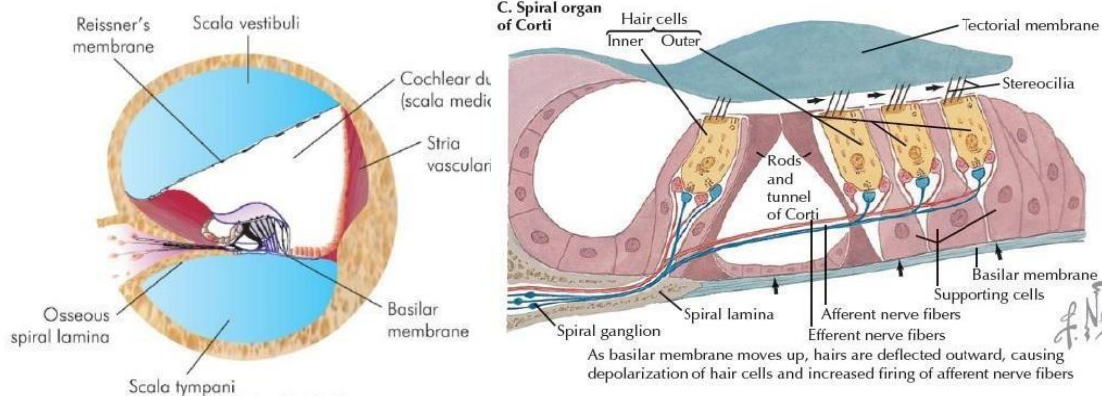
- Effects of conductive system defects and hearing loss:
 - ✦ Isolated TM perforation: 30-45 dB CHL
 - ✦ TM perforation and ossicular chain discontinuity: 40-50 dB CHL
 - ✦ **Intact TM and ossicular chain discontinuity: 55-60 dB CHL**
- Roles of TM in addition to above: protects middle ear from EAC contents; provides air cushion to prevent insufflation of foreign material from the NP via the ET
- Less than half of the power entering the middle ear gets to cochlea; much is absorbed by ligaments and the rest of the middle ear
(remember that a 50% loss of power is a loss of only 3 dB intensity)

Middle ear muscles: tensor tympani (CN V) and stapedius (CN VII)

- ✦ Stapedius contracts in response to loud sounds (> 80 dB SPL); stiffens ossicular chain; limits low frequency (< 2 kHz) sound transmission to cochlea; **10 ms latency**; no good for bursts

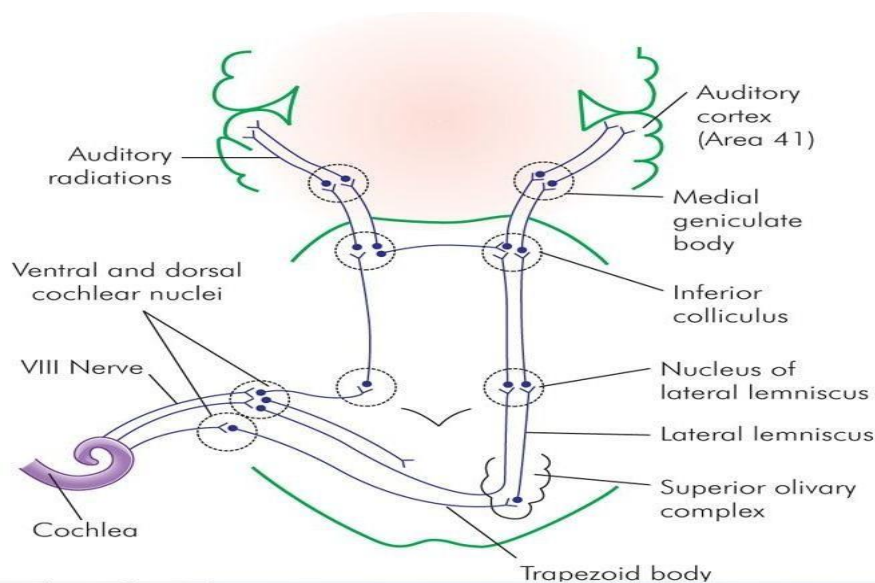
Auditory Neural Pathways and their Nuclei:

- ✓ Hair cells are innervated by dendrites of bipolar cells of spiral ganglion which is situated in **Rosenthal's canal** (canal running along the **osseous spiral lamina**).



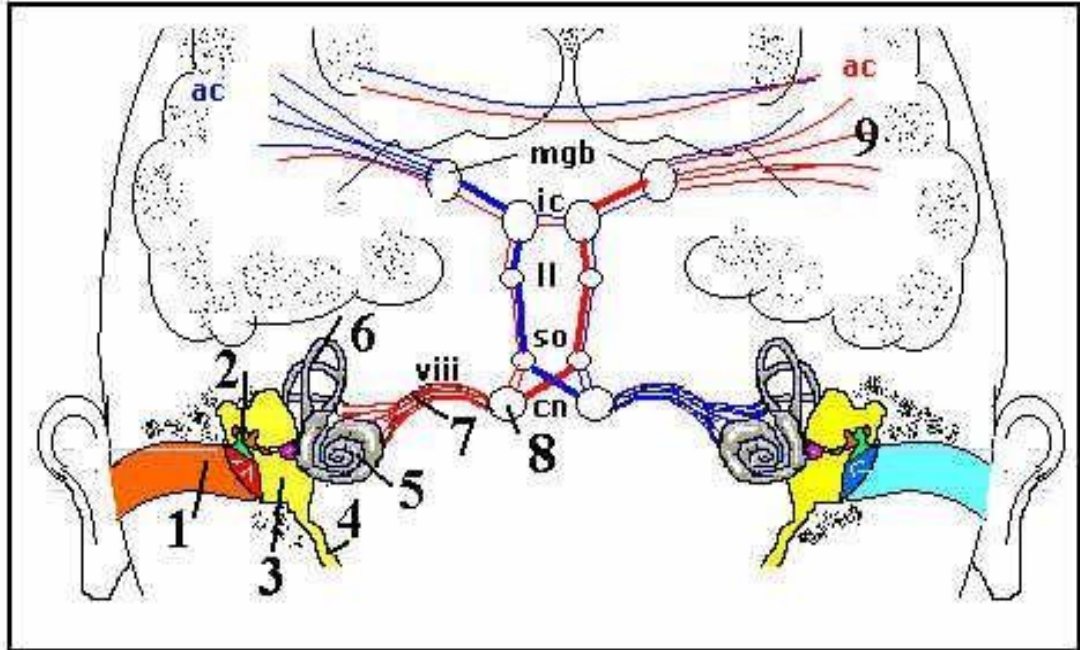
- ✓ Axons of these bipolar cells **form** the **cochlear division of CN VIII** and end in the **cochlear nuclei**, the **dorsal and ventral**, on **each side of the medulla**.
- ✓ the ascending auditory pathways, sequentially, from below upwards, are:
Remember the mnemonic E.COLI-MA

- Eighth nerve
- Cochlear nuclei
- Superior olivary complex
- Nucleus of lateral lemniscus
- Inferior colliculus
- Medial geniculate body (thalamus)
- Auditory cortex.



- Auditory pathways from the right cochlea.
- Note bilateral route through brainstem and bilateral cortical representation.

- ✓ Most fibres from the cochlear nucleus cross to contra sup olivary complex; some stay ipsi
- ✓ The auditory fibres travel via the ipsilateral and contralateral routes and have multiple decussation points. Thus each ear is **represented in both cerebral hemispheres**.
- ✓ The area of cortex, concerned with hearing is situated in the **"superior temporal gyrus"** within sylvian fissure of temporal lobe" (**Brodmann's area 41**).

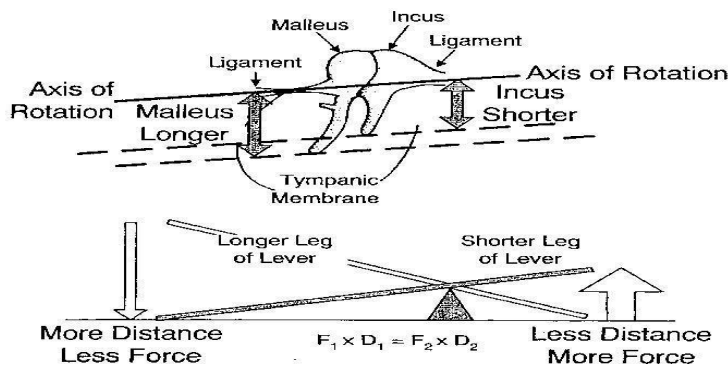


Mechanism of Hearing

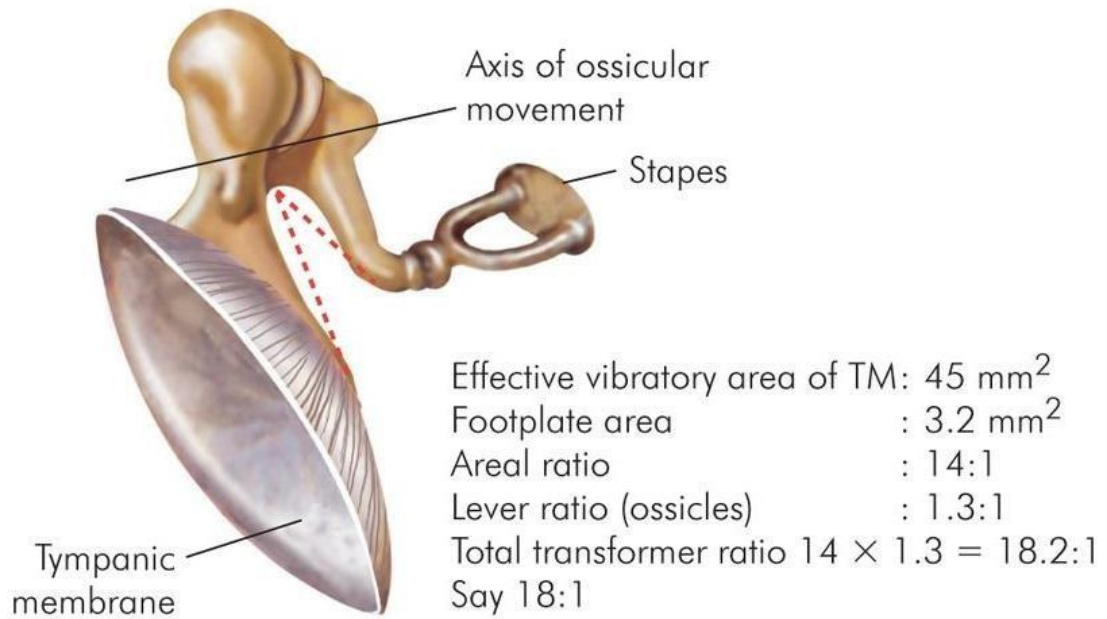
- ✓ sound signal in the environment is collected by the **pinna**, passes through external auditory canal and **strikes** the tympanic membrane.
- ✓ Vibrations of the tympanic membrane are transmitted to stapes footplate through a chain of ossicles coupled to the tympanic membrane.
- ✓ Movements of stapes footplate cause pressure changes in the labyrinthine fluids which move the **basilar membrane**.
- ✓ move the **basilar membrane** stimulates the hair cells of the **organ of Corti**.
- ✓ **hair cells** act as transducers and **convert the mechanical energy into electrical impulses** which travel along the auditory nerve.
- ✓ Thus, the mechanism of hearing can be broadly divided into:
 - **1.** Mechanical conduction of sound (**conductive apparatus**).
 - **2.** Transduction of mechanical energy to electrical impulses (**sensory system of cochlea**).
 - **3.** Conduction of electrical impulses to the brain (**neural pathways**).

1. Conduction of Sound

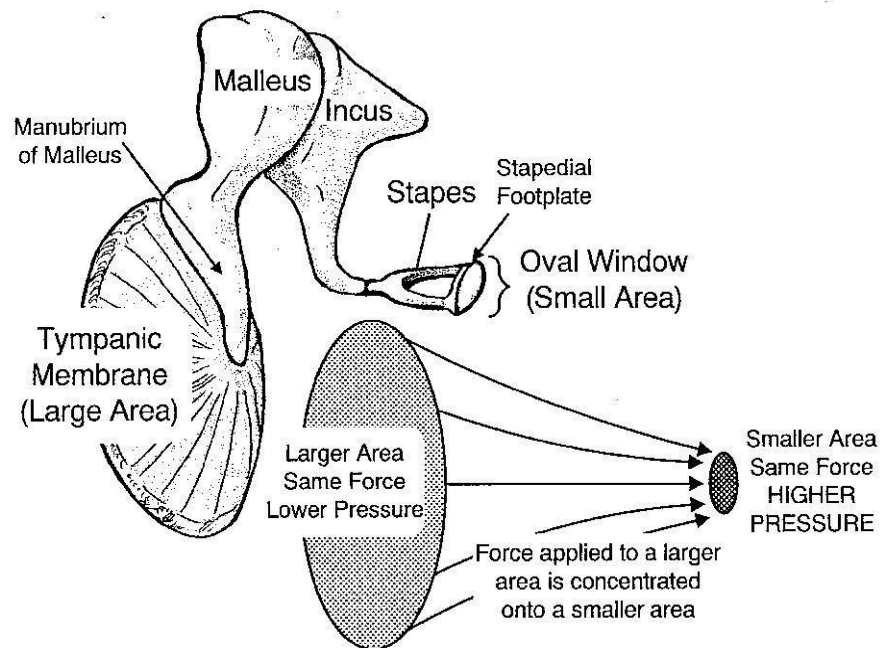
- ✓ person under water cannot hear any sound made in the air because 99.9% of the sound energy is reflected away from the surface of water because of the impedance offered by it.
- ✓ similar situation exists in the ear when air-conducted sound has to travel to cochlear fluids. Nature has compensated for this loss of sound energy by interposing the middle ear which converts sound of greater amplitude, but lesser force, to that of lesser amplitude but greater force ((ME act as mechanical transformer))
- ✓ This function of the middle ear is called impedance matching mechanism or the transformer action. to compensate the sound energy lost during its transmission from air to liquid medium. it is accomplished by:



- (a) **Lever action of the ossicles.** Handle of malleus is 1.3 times longer than long process of the incus, providing a mechanical advantage of 1.3.
- (b) **Hydraulic action of tympanic membrane**
 - ✓ The area of tympanic membrane is much larger than the area of stapes footplate, the average ratio between the two being 21:1.
 - ✓ As the effective vibratory area of tympanic membrane is only two-thirds, the effective areal ratio is reduced to 14:1, and this is the mechanical advantage provided by the tympanic membrane. The product of areal ratio and lever action of ossicles is 18:1.
 - According to some workers (Wever & Lawrence) out of a total of 90 mm² area of human tympanic membrane, only 55 mm² is functional and given the area of stapes footplate (3.2 mm²), the areal ratio is 17:1 and total transformer ratio (17 × 1.3) is 22.1 or approximately 25–30 dB gain [increase in sound pressure](#)

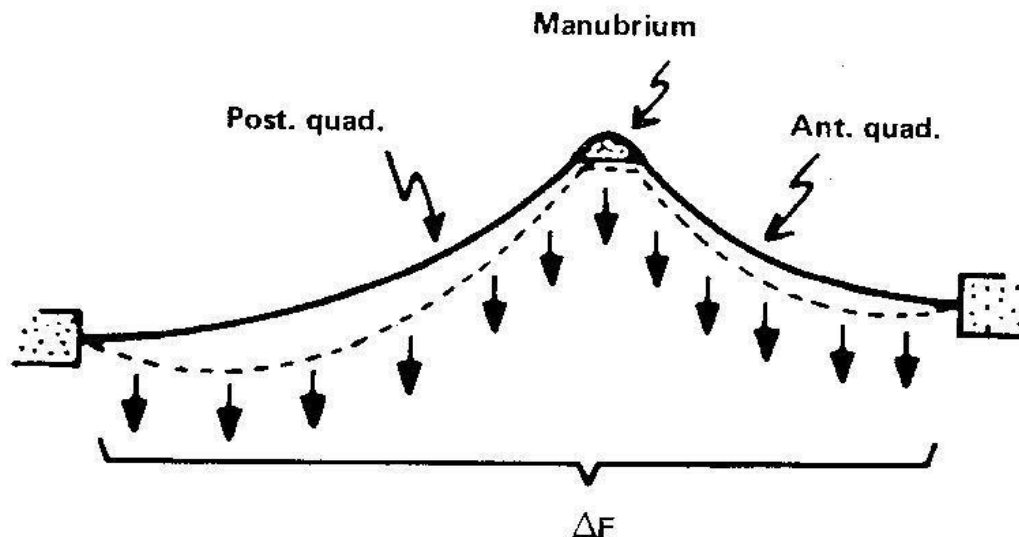


□



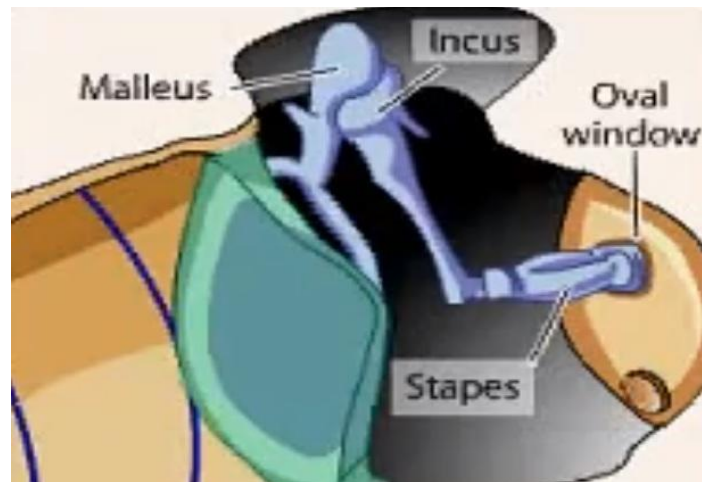
- **(c) Curved membrane effect.**

- ✓ Movements of tympanic membrane are more at the periphery than at the center where malleus handle is attached. This too provides some Force.
- ✓ The curved membrane buckling advantage is 2 to 1



Phase differential between oval and round windows

- ✓ Sound waves striking the tympanic membrane do not reach the oval and round windows simultaneously.
- ✓ There is a preferential pathway to the oval window because of the ossicular chain. when oval window is receiving wave of compression, the round window is at the phase of **rarefaction**.
- ✓ If the sound waves were to strike both the windows simultaneously, they would cancel each other's effect with no movement of the perilymph and no hearing.
- ✓ This acoustic separation of windows is achieved by the presence of intact tympanic membrane protects the round window .
- ✓ Phase differential between the windows contributes 4 dB when tympanic membrane is intact.

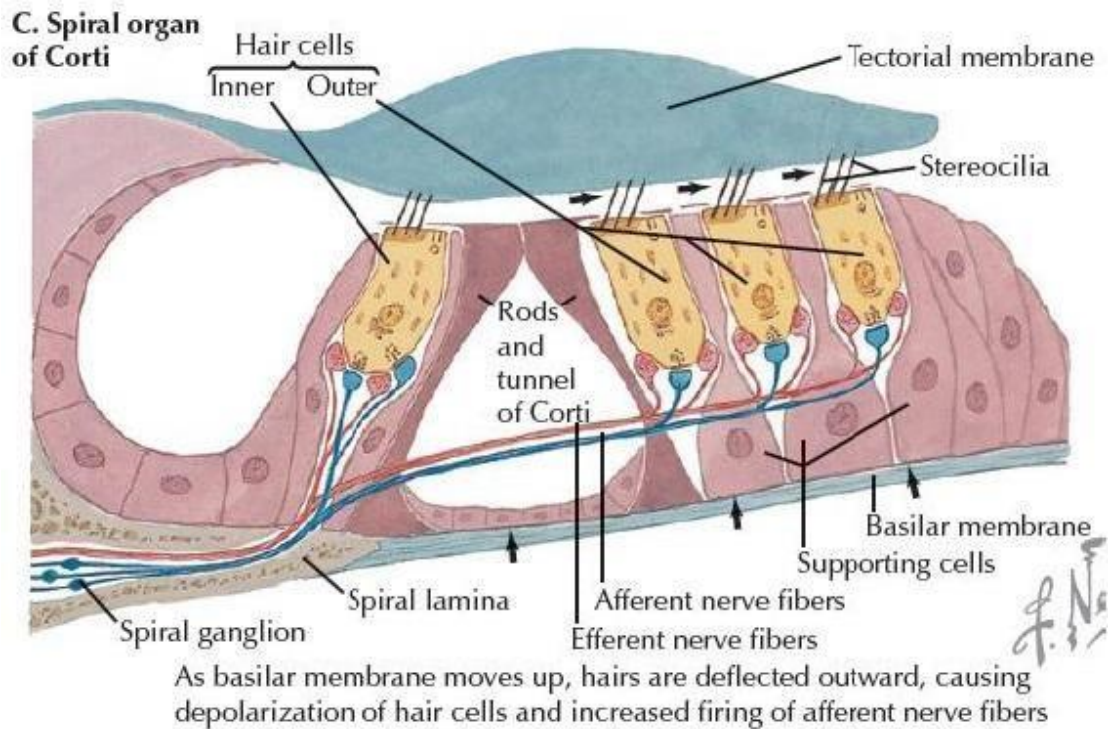


Natural resonance of external and middle ear

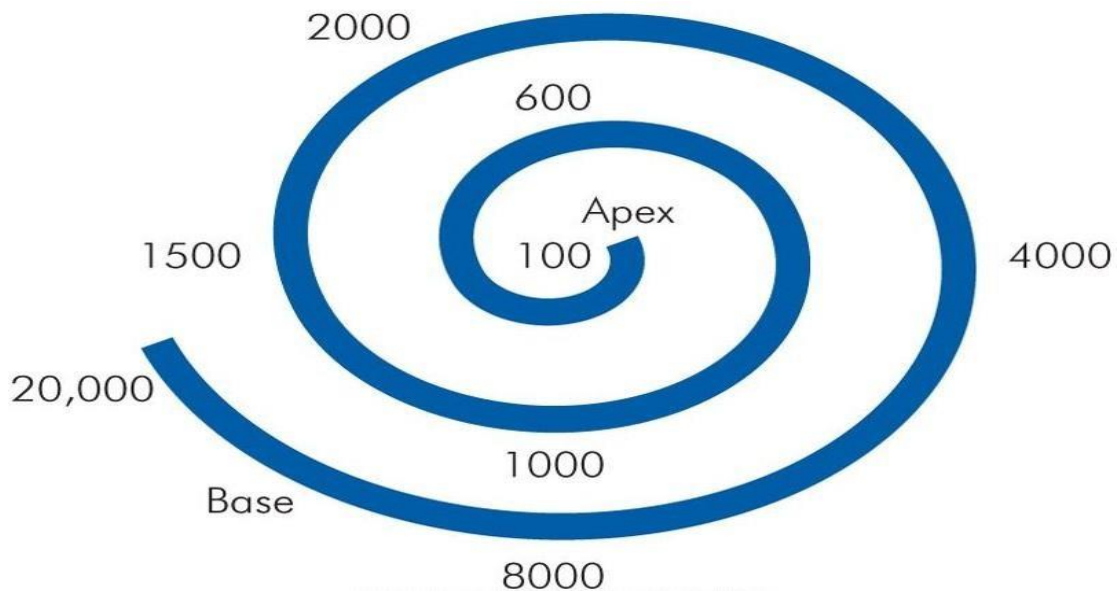
- ✓ **Resonance Frequencies:** natural frequencies that vibrate a mass with the least amount of force
- ✓ Inherent anatomic and physiologic properties of the external and middle ear allow **certain frequencies** of sound to pass more easily to the inner ear due to their natural resonances.
 - **Concha:** 4000–6000 Hz
 - **EAC:** 2000–3000 Hz (primary frequencies of speech)
 - **TM:** 800–1600 Hz
 - **Ossicles:** 500–2000 Hz (most efficiently transmitted by ossicular chain)
 - **Middle Ear:** 800 Hz
- ✓ Thus greatest sensitivity of the sound transmission is between 500 and 3000 Hz and these are the frequencies most important to man in day to day conversation
- ✓ EAC and pinna give gain of 15 dB at 3 kHz range (10 dB from 2 to 5 kHz); this is one of the reasons for 4 kHz notch with noise-induced hearing loss (boilermaker notch).

2. Transduction of Mechanical Energy to Electrical Impulses

- ✓ Movements of the stapes footplate, transmitted to the cochlear fluids, move the basilar membrane, setting up shearing force between the tectorial membrane and the hair cells.
- ✓ The distortion of hair cells gives rise to cochlear microphonics which trigger the nerve impulse



- ✓ **Place Theory (Tonotopic Organization of the Cochlea)** A sound wave, depending on its frequency, reaches maximum amplitude on a **particular place on the basilar membrane** and stimulates that segment (travelling wave theory of von Békésy).
- ✓ Higher frequencies are represented in the basal turn of the cochlea and the progressively lower ones towards the apex



3. Neural Pathways

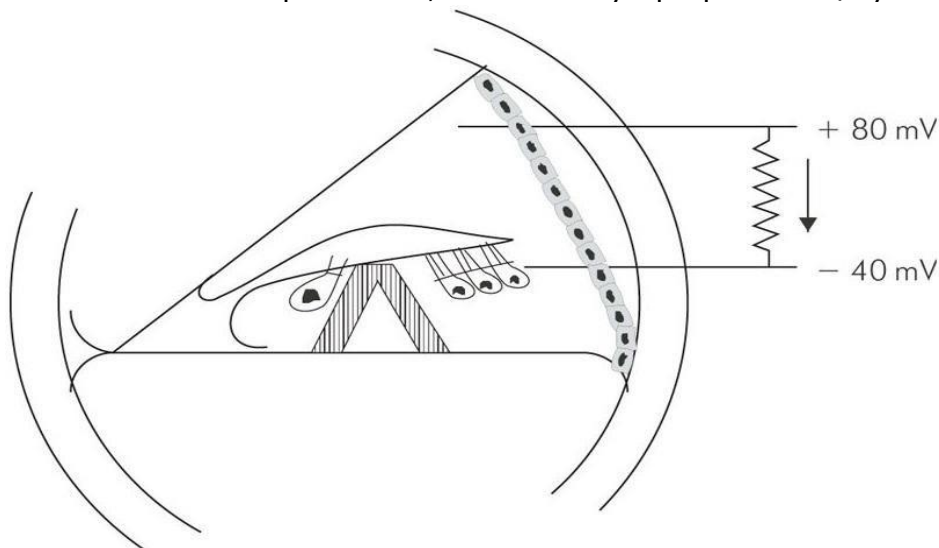
- ✓ Hair cells get innervation from the bipolar cells of spiral ganglion.
- ✓ Central axons of these cells collect to form cochlear nerve which goes to ventral and dorsal cochlear nuclei. From there, both crossed and uncrossed fibres travel to the superior olivary nucleus, lateral lemniscus, inferior colliculus, medial geniculate body and finally reach the auditory cortex of the temporal lobe.

Electrical Potentials of Cochlea and CN VIII

Four types of potentials have been recorded; **three** from the **cochlea** and **one** from **CN VIII fibres**. They are:

1. Endocochlear (Endolymphatic) potential

- ✓ It is a direct current (DC) potential recorded from **scala media**.
- ✓ It is **+80 mV** and is generated from the stria vascularis by Na^+/K^+ -ATPase pump and provides **source of energy** for cochlear transduction.
- ✓ It is **present at rest** and does **not require sound stimulus**.
- ✓ This potential provides a sort of "battery" of the cochlea, to drive the current through hair cells when they move in response to a sound stimulus.
- ✓ **Stimulation of hair cells produces intracellular potential of -40 mV.** This provides flow of current of 120 mV through the top of hair cells.
 - o **Metabolic presbycusis:** HL associated with altered endo-cochlear potentials; from endolymph problems/hydrops



2. Cochlear microphonic (CM)

- ✓ It is an alternating current (AC) potential.
- ✓ voltage recorded within cochlea near round window
- ✓ When basilar membrane moves in response to sound stimulus, **electrical resistance at the tips of hair cells changes allowing flow of K^+ through hair cells and produces voltage fluctuations called cochlear microphonic.**
- ✓ wave form mirrors motion of basilar membrane

3. Summating potential (SP)

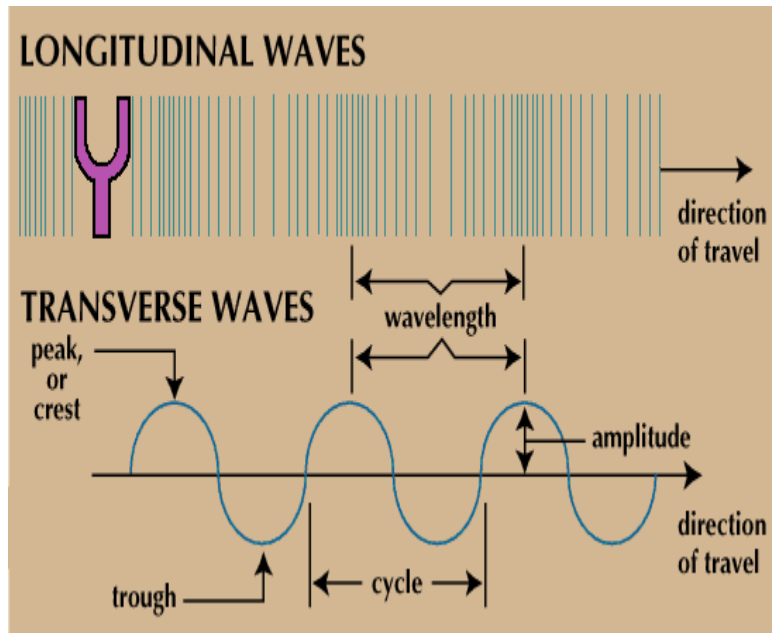
- ✓ It is a DC potential and follows "envelope" of stimulating sound.
- ✓ It is produced by hair cells.
- ✓ It may be negative or positive.
- ✓ SP has been used in diagnosis of **Mènière's disease**.
- ✓ It is superimposed on VIII nerve action potential.
- ✓ They differ from action potentials in that:
 - ✦ They are graded rather than all or none phenomenon,
 - ✦ have no latency
 - ✦ not propagated
 - ✦ no post-response refractory period.

4. Compound (**Whole nerve**) action potential

- ✓ It is an all or none response of **auditory nerve fibres**.
- ✓ recorded with gross electrode near the round window or auditory nerve, with high-frequency signals
 - ✦ Amplitude increases with intensity from 40 to 50 dB
 - ✦ Latency decreases as stimulus intensity increases

Audiology and Hearing Assessment:

- **Sound:**
 - Form of energy produced by a vibrating object.
- **Sound wave:**
 - Consists of compression and rarefaction of molecules of the medium (air, liquid or solid) in which it travels.



- **Sound velocity:**
 - Different in different media.
 - In the air, at 20°C, at sea level, sound travels 344 m/sec.
 - Travels faster in liquid and more faster in a solid medium.

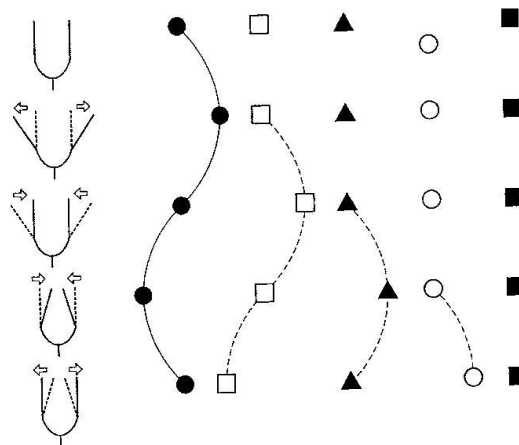
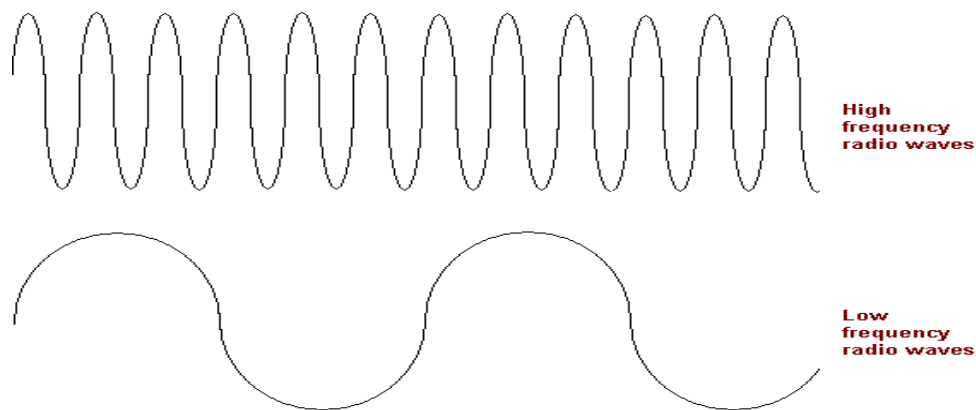


Figure 1-4. Sound is initiated by transmitting the vibratory pattern of the sound source to nearby air particles, and then the vibratory pattern is passed from particle to particle as a wave. Notice how it is the pattern of vibration that is being transmitted, whereas each particle oscillates around its own average location.

- **Sound Pitch (Hz):**

- Subjective sensation produced by Frequency of sound.
- Higher the frequency, greater is the pitch.
- Sound Frequency is the number of cycles/second.
- Measured by Hertz (Hz).
- Sound of 1000 Hz means 1000 cycles/second.
- Normal person can hear frequencies of 20-20,000 Hz.
- Routine audiometry tests only 125-8000 Hz.
- Frequencies of 500, 1000 and 2000 Hz are called speech frequencies as most of human voice falls within this range.
- Pure tone average is the average threshold of hearing in these three speech frequencies which corresponds to the speech reception threshold.



- **Pure tone:**

- Single frequency sound.
- Sound of 250, 500 or 1000 Hz.
- In pure-tone audiometry, we measure the threshold of hearing in decibels for various pure tones from 125 to 8000 Hz.

- **Complex sound:**

- Sound with more than one frequency.
- Human voice is a complex sound.

- **Loudness (dB):**

- Subjective sensation produced by Intensity of sound.
- More the intensity of sound, greater the loudness.
- Measured in decibels.

Whisper	=	30 dB
Normal conversation	=	60 dB
Shout	=	90 dB
Discomfort of the ear	=	120 dB
Pain in the ear	=	130 dB

- **Hearing Threshold:**
 - Minimal intensity which a normal healthy person can hear.
 - Vary from person to person.
 - Zero level on the audiometer (0 dB):
 - The least intensity for average normal ear to perceive a specific frequency 50% of the time
- **Hearing level (HL):**
 - Sound pressure level (SPL) produced by an audiometer at a specific frequency.
 - It is measured in decibels with reference to audiometric zero.
 - If an audiometer delivers a sound at 70 dB, it is represented as 70 dB HL.
- **Sensation level (SL):**
 - Level of sound above the threshold of hearing for an individual.
 - If someone is tested at 40 dB SL, it means he was tested at 40 dB above his threshold.
 - For a normal person, this would be a sound of $0 + 40$, i.e. 40 dB HL, but for one with a hearing loss of say 30 dB, it would be $30 + 40$, i.e. 70 dB HL.
 - In other words, sensation level refers to the sound which will produce the same sensation, as in normally hearing person.
 - In speech audiometry, discrimination scores are tested at 30 to 40 dB SL.
 - Stapedial reflex is elicited with a sound of 70-100 dB SL
- **Noise:**
 - Aperiodic complex sound.
 - Used for masking by keeping one ear busy by noise while the other is being tested.
 - Types of noise:
 - **Narrow-band noise:**
 - Contains certain frequencies with smaller frequency range.
 - Used to mask the test frequency in PTA.
 - **Broad-band noise:**
 - Contains all frequencies in audible spectrum.
 - Used for masking in speech audiometry.

Assessment of Hearing:

- Hearing loss can be of three types:

1. Conductive Hearing Loss (CHL):

- Any disease process interfering with the conduction of sound from External ear to Stapediovestibular joint.
- Cause may lie in:
 1. External Ear (Obstructions)
 2. Tympanic Membrane (Perforation)
 3. Middle Ear (Fluid)
 4. Ossicles (Fixation or Disruption)
 5. Eustachian Tube (Obstruction)
 6. Inner ear (SCDS/Third window effect)

2. Sensorineural Hearing Loss (SNHL):

- Cochlear HL:
 - Hearing loss due to lesions of Cochlea.
 - Examples:
 - Noise-induced SNHL
 - Ototoxicosis
 - Presbycusis
 - Meniere's Disease
 - Labrynthitis
 - Autoimmune
 - Inner ear malformation
- Retro-cochlear HL:
 - Hearing loss due to lesions of CN-VIII or central auditory system.
 - Examples:
 - CPA tumors

Table 15-1. Results of various tests to differentiate a cochlear from a retrocochlear lesion

	Normal	Cochlear lesion	Retrocochlear lesion
• Pure tone audiogram	Normal	Sensorineural hearing loss	Sensorineural hearing loss
• Speech discrimination score	90-100%	Below 90%	Very poor
• Roll over phenomenon	Absent	Absent	Present
• Recruitment	Absent	Present	Absent
• SISI score	0-15%	Over 70%	0-20%
• Threshold tone decay test	0-15 dB	Less than 25 dB	Above 25 dB
• Stapedial reflex	Present	Present	Absent
• Stapedial reflex decay (page 109)	Normal	Normal	Abnormal
• E.R.A	Normal interval between wave I & V	Normal interval between wave I & V	Wave V delayed or absent

3. Mixed Hearing Loss:

- Both CHL and SNHL are present in the same ear.
 - Air-Bone gap indicates CHL
 - Impairment of bone conduction indicates SNHL.
- Examples:
 - Advance Otosclerosis.
 - Advance CSOM.
 - Mixed pathology.

- **While assessing Auditory function it is important to find out:**

1. Type of Hearing Loss:

- CHL
- SNHL
- Mixed HL

2. Degree of Hearing Loss:

- Mild
- Moderate
- Moderately Severe
- Severe
- Profound
- Total

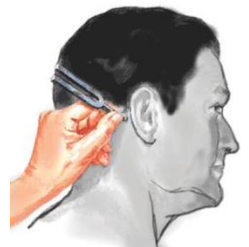
3. Site of Lesion:

- If CHL:
 - External Ear
 - TM
 - Middle Ear
 - Ossicles
 - Eustachian tube
 - Inner ear defects
- If SNHL:
 - Cochlear
 - Retrocochlear

4. Cause of Hearing Loss:

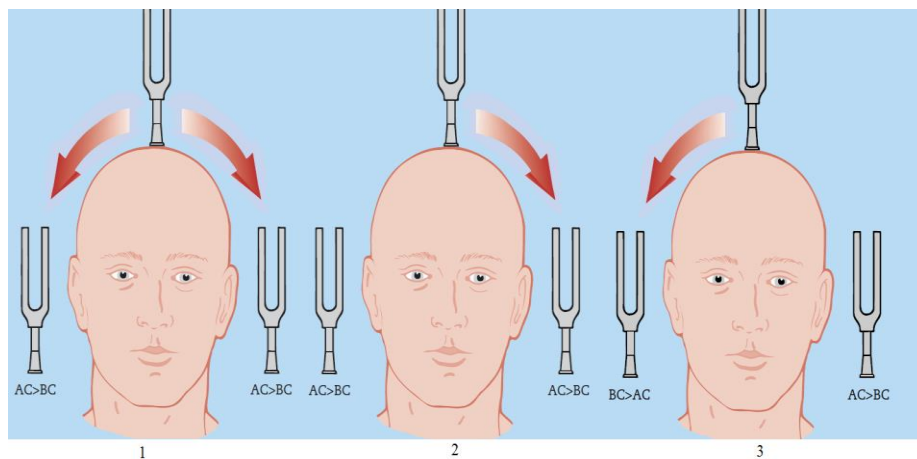
- Congenital
- Traumatic
- Infective
- Neoplastic
- Degenerative
- Metabolic
- Ototoxic
- Vascular
- Autoimmune

- **Clinical Tests of Hearing:**
- Finger Friction Test:
- Rough but quick method of screening.
- Rubbing or snapping the thumb and a finger close to patient's ear.
- Tuning Fork Test:
- Performed with tuning forks of different frequencies (128, 256, 512, 1024 Hz).
- For routine clinical practice, tuning fork of 512 Hz is **ideal**.
 - o Forks of Lower Frequencies (128 and 256 Hz):
 - Produce sense of bone vibration.
 - o Forks of Higher Frequencies (1024 Hz):
 - Have a shorter decay time.
- **Air Conduction (AC):**
 - o Vibrating fork is placed vertically about 1 cm away from the opening of external auditory meatus.
 - o Sound waves are transmitted through TM, Middle ear and Ossicles to Inner ear.
 - o Testing both Conducting mechanism and Cochlea
 - o Normally, hearing through air conduction is louder and heard twice as long as through Bone conduction route.
- **Bone Conduction (BC):**
 - o Footplate of vibrating tuning fork is placed firmly on Mastoid bone.
 - o Cochlea is stimulated directly by vibrations conducted through the skull bones.
 - o BC is a measure of Cochlear function only.
- The clinically useful tuning fork tests include:
- **1. Rinne Test:**
- Air conduction (AC) is compared with bone conduction (BC).
- Vibrating tuning fork placed vertically within 1 cm of Opening of External auditory meatus (AC) then immediately placed on the mastoid bone (BC).
 - o **(+ve) Rinne Test: (AC > BC)**
 1. Normal Hearing
 2. SNHL
 - o **(-ve) Rinne Test: (BC > AC)**
 1. CHL (>15-30 dB HL)
 2. Severe unilateral SNHL with Crossover (False negative):
 - Responds to bone conduction testing which comes from the opposite normal ear because of transcranial transmission of sound.
 - Correct diagnosis can be made by masking the Non-test ear while testing for bone conduction.



2. Weber Test:

- Vibrating tuning fork is placed in Forehead, Vertex or Maxillary teeth and the sound travels directly to the cochlea via bone.
- More sensitive than Rinne test (Lateralizes with 5 dB of conductive hearing loss).
- **Normally:**
 - o Heard equally in both ears.
- **CHL:**
 - o Lateralized to Diseased ear.
- **SNHL:**
 - o Lateralized to Better ear.



1. Tuning fork tests showing:
 - o Positive Rinne bilaterally
 - o Weber test referred equally to each ear.
 - o Dx: Normal Hearing.
2. Tuning fork tests showing:
 - o Positive Rinne bilaterally.
 - o Weber test referred to Left ear (Better ear).
 - o Dx: SNHL in Right ear.
3. Tuning fork tests showing:
 - o Negative Rinne on Right ear.
 - o Positive Rinne on Left ear.
 - o Weber test referred to Right ear (Diseased ear).
 - o Dx: CHL in Right ear.

Table 4-1. Tuning fork tests and their interpretation

Test	Normal	Conductive deafness	SN deafness
Rinne	AC > BC (Rinne positive)	BC > AC (Rinne negative)	AC > BC
Weber	Not lateralised	Lateralised to poorer ear	Lateralised to better ear

Prediction of Air-Bone gap can be made if tuning forks of 256, 512 and 1024 Hz are used:

- **Air-bone gap of > 5 dB:**
 - o Webber lateralized to the affected side
 - o (+ve) Rinne test for 256, 512 and 1024 Hz.
- **Air-bone gap of > 15 dB:**
 - o Webber lateralized to the affected side
 - o **(-ve)** Rinne test for **256** Hz.
 - o (+ve) Rinne test for 512 and 1024 Hz.
- **Air-bone gap of > 30 dB:**
 - o Webber lateralized to the affected side
 - o **(-ve)** Rinne test for 256 and **512** Hz
 - o (+ve) Rinne test for 1024 Hz.
- **Air-bone gap of > 45 dB:**
 - o Webber lateralized to the affected side
 - o **(-ve)** Rinne test for 256, 512 and **1024** Hz.
- **Behavioral (Subjective) Audiometric Tests of Hearing:**
 1. Behavioral Observation Audiometry (BOA): **<5 months old.**
 2. Visual Response Audiometry (VRA): **5 months to 3 years old.**
 3. Conventional Play Audiometry (CPA): **3 to 5 years old.**
 4. Pure Tone Audiometry (PTA): **>5 years old.**
 5. Speech Audiometry: **>5 years old.**



Behavioral Observation Audiometry (BOA):

- Limited for infants **<5 months old**.
- BOA is NOT accurate in estimating hearing threshold due to:
 - o Infant responses are variable and occur at levels above hearing threshold.
- Physiologic measures (ABR) are necessary for estimation of hearing threshold in this age group.
- BOA can rule out severe and profound hearing loss only.
- Method:
 - o Auditory stimuli are introduced via speaker.
 - o Infant are observed for a response (eye widening, startle, head turn, cessation of sucking).

Visual Response Audiometry (VRA):

- Limited for children between **5 months to 3 years old**.
- VRA is accurate in estimating hearing threshold in cooperative child using standard audiometric techniques.
- Method:
 - o Child is trained to turn his head toward auditory stimulus with visual stimulus (toy animation, flashing light).
 - o Auditory stimulus is delivered by insert earphones (for ear-specific response) or by sound field (speakers).



Figure 100.1 VRA. An 10-month-old infant is conditioned to turn his head to a video display when an auditory stimulus is detected.

Conditioned Play Audiometry (CPA):

- Limited for children between **3 to 5 years old**.
- CPA is accurate in estimating hearing threshold in cooperative child using standard audiometric techniques.
- Method:
 - Child is conditioned to respond to pure tones by playing a simple game using toys such as placing a marble in a box.
 - Each correct performance of the act is reinforced with encouragement or reward.



Pure Tone Audiometry (PTA):

- Most common measurement of hearing sensitivity (threshold).
- Accurate method in estimating hearing threshold for **>5 years old**.
- Method:
 1. Single-frequency sound (pure tone) is produced at selected sound intensities (db).
 2. Presented at different frequencies:
 - Octave frequencies:
 - 250, 500, 1000, 2000, 4000 and 8000 Hz.
 - Standard frequencies tested.
 - High-frequency audiometry for frequencies greater than 8,000 Hz and up to 20,000 Hz is indicated in patients at risk for ototoxicity.
 - Inter-octave frequencies:
 - 750, 1500, 3000, and 6000 Hz.
 - **Tested only if hearing thresholds between two adjacent octave frequencies are >20 dB.**
 - Inter-octave hearing loss is a characteristic of noise-induced cochlear dysfunction.
 3. Starting with AC followed by BC:
 - Air-Conduction (AC):
 - Represents conduction from auricle to cochlea.
 - Sound introduced via either:
 - Insert earphones:
 - Transducer of choice.
 - More comfortable.
 - Greater interaural attenuation.
 - Supra-aural headphones:
 - Less comfortable to children
 - Less interaural attenuation
 - Measures of the function of whole hearing system (external, middle and inner ear).
 - Bone-Conduction (BC):
 - Represents conduction from skull bones to the inner ear (bypassing external and middle ear).
 - Sound introduced via bone oscillator placed over the mastoid.
 - Measure of Cochlear function.
 - Not measured at 8000 Hz.
 - Should always be better than air conduction.
 - Indications of BC testing:
 1. Pediatric patients
 2. Abnormal AC threshold.
 3. Suspecting SCDS.
 4. Detect hearing threshold for each frequency:
 - Lowest signal intensity (dB) at which the patient perceives 50% of pure tones at each frequencies.
 5. Then charted in Audiogram.

Modality	Ear	
	Right	Left
AC unmasked	○	×
AC masked	△	□
BC unmasked	<	>
BC masked	⌈	⌋
No response	↻	↷

- Interpretation:

○ **Type of Hearing Loss:**

- Determined by evaluating bone and air conduction thresholds and the difference between bone and air conduction (Air-bone gap/ABG).
- ABG measures of the degree of conductive deafness.
- ABG should be at least 10 db.
- Conductive Hearing Loss (CHL):
 - Normal bone conduction thresholds.
 - Abnormal air conduction thresholds.
 - Presence of ABG.
 - Maximum CHL is 60 dB which is found in:
 - Aural atresia.
 - Ossicular chain discontinuity with intact TM.
- Sensorineural Hearing Loss (CHL):
 - Abnormal bone and air conduction thresholds.
 - No ABG.
- Mixed Hearing Loss (MHL):
 - Abnormal bone and air conduction thresholds.
 - Presence of ABG.

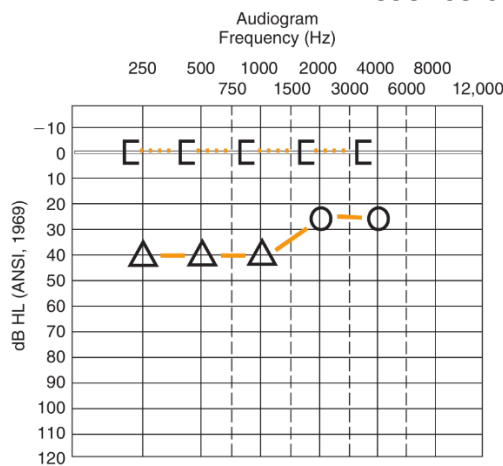


FIGURE 133-2. Audiogram with a normal bone-conduction threshold and an elevated air-conduction threshold—an air-bone gap. ANSI, American National Standards Institute; dB HL, decibel hearing level.

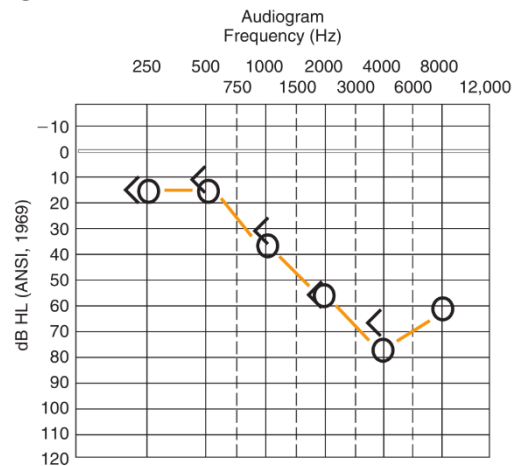


FIGURE 133-3. Audiogram showing sensorineural hearing loss. The air-conduction and bone-conduction thresholds are identically elevated. ANSI, American National Standards Institute; dB HL, decibel hearing level.

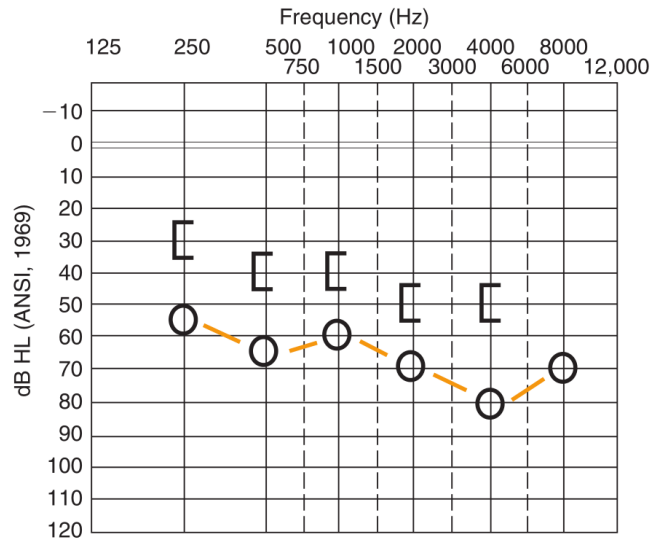
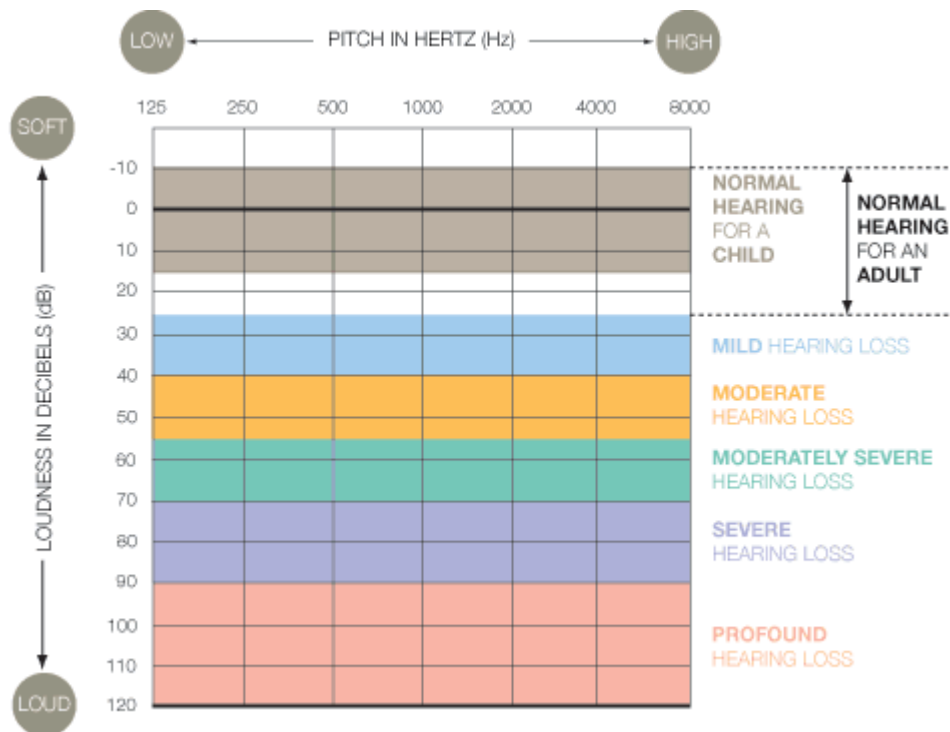


FIGURE 133-4. Audiogram showing mixed hearing loss. Both the air-conduction and bone-conduction thresholds are elevated, and an air-bone gap is evident. ANSI, American National Standards Institute; dB HL, decibel hearing level.

○ **Degree of Hearing Loss:**

- Amount of intensity (dB) that has to be raised above the normal level (0 dB) at each frequency.



Degree of hearing loss	Hearing loss range (dB HL)
Normal	-10 to 15
Slight	16 to 25
Mild	26 to 40
Moderate	41 to 55
Moderately severe	56 to 70
Severe	71 to 90
Profound	91+

Source: Clark, J. G. (1981). Uses and abuses of hearing loss classification. *Asha*, 23, 493-500.

- Pure Tone Average:
 - Measures the hearing sensitivity of speech frequencies in decibels.
 - Average AC thresholds of (500, 1000, 2000 Hz).
 - Should be within 10 dB of Speech Reception Threshold (SRT).

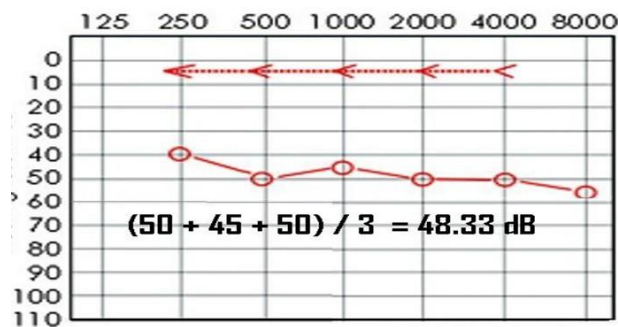


Table 5-5. Hearing loss and difficulty in hearing speech

Hearing threshold in better ear (average of 500, 1000, 2000 Hz)	Degree of impairment (WHO classification)	Ability to understand speech
0-25	Not significant	No significant difficulty with faint speech
26-40	Mild	Difficulty with faint speech.
41-55	Moderate	Frequent difficulty with normal speech.
56-70	Moderately severe	Frequent difficulty even with loud speech.
71-91	Severe	Can understand only shouted or amplified speech.
Above 91	Profound	Usually cannot understand even amplified speech.

○ **Masking:**

- Audiometric results are valid only when the patient's responses are caused by stimulation of the test ear.
- Masking is a noise that introduced into non-tested ear to prevent **crossover** from the tested ear, whenever sound stimulation exceeds inter-aural attenuation.
 - Narrow band noise is used for Pure-tone signals.
 - Wide band noise is used for speech signals.
- Inter-aural attenuation:
 - The amount of reduction in sound intensity that occurs as the signal crosses over the head from one ear to the other (The amount of sound intensity needed before crossover occurs).
 - No inter-aural attenuation for BC signals and very faint sound presented to the mastoid of one ear by BC vibrator can be transmitted through the skull to either or both inner ears.
 - Inter-aural attenuation for AC is higher with insert ear-phones (70 dB) compared to supra-aural headphones (40 dB).
- Crossover:
 - When the sound that is presented to Tested ear crosses the head via bone conduction and perceived by non-tested ear.
 - Crossover occurs at:
 - **0 dB** for Bone Conduction (BC).
 - **40 dB** for AC with supra-aural headphones.
 - **70 dB** for AC with insert earphones.
- Indications of Masking:
 1. All BC conduction tests.
 2. Difference between AC threshold of tested ear and BC threshold of non-tested ear is:
 - **> 40 dB** with supra-aural headphones.
 - **> 70 dB** with insert earphones.
- Masking Dilemma:
 - With effective masking, any signal crossing over to the ear not being tested is masked by the noise.
 - Excess levels of masking noise must be avoided because the noise can cross back over to the ear being tested (over-masking).
 - Masking dilemma occurs when effective masking to the non-tested ear can only be done at high level that causes the noise to cross over to the tested ear and interferes with accurate estimation of hearing threshold.
 - Occurs in **Bilateral severe (50-60 dB) CHL.**

- **Special PTA Tests:**

- **Short Increment Sensitivity Index (SISI Test):**

- Test for recruitment phenomenon (abnormal loudness growth).
- Patients with cochlear SNHL distinguish smaller changes in intensity of pure tone better than normal persons, CHL or retrocochlear SNHL.
- SISI test is used to differentiate cochlear from a retrocochlear SNHL.
- Method:
 - Continuous tone is presented 20 dB above hearing threshold.
 - Every 5 seconds, the tone is increased by 1 dB and 20 such blips are presented.
 - The patient should count how many times the tone changes in intensity.
 - The score is calculated by multiplying the number of correct 1 dB increases by 5, which will provide a percentage.
- Interpretation:
 - SISI score is >75% in cochlear SNHL.
 - SISI score is <20% in retrocochlear SNHL.

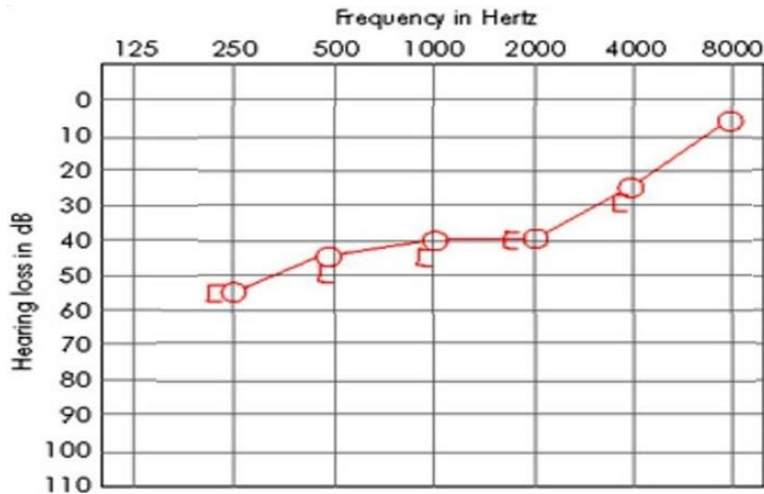
- **Tone Decay Test:**

- It is a measure of nerve fatigue.
- Used to detect retrocochlear lesions.
- Normally, a person can hear a tone continuously for 60 seconds.
- In nerve fatigue, he stops hearing earlier.
- Method:
 - A tone of 4000 Hz is presented at 5 dB SL continuously for 60 seconds.
 - If patient stops hearing earlier, intensity is increased by another 5 dB.
 - The procedure is continued till patient can hear the tone continuously for 60 seconds, or no level exists above the threshold where tone is audible for full 60 seconds.
 - The result is expressed as number of dB of decay.
- Interpretation:
 - Tone decay > 25 dB is diagnostic of a retrocochlear lesion.

- **Common Audiogram Patterns:**

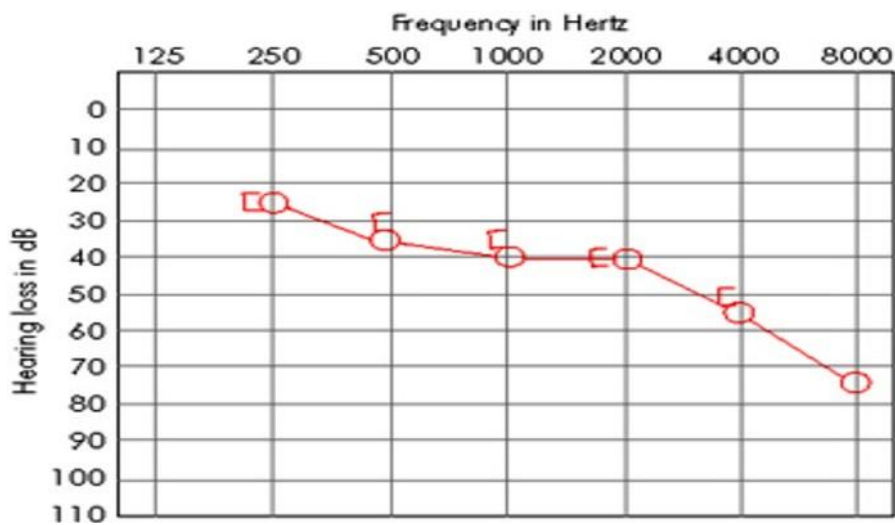
- 1. Low Frequency SNHL:**

- Meniere's disease.



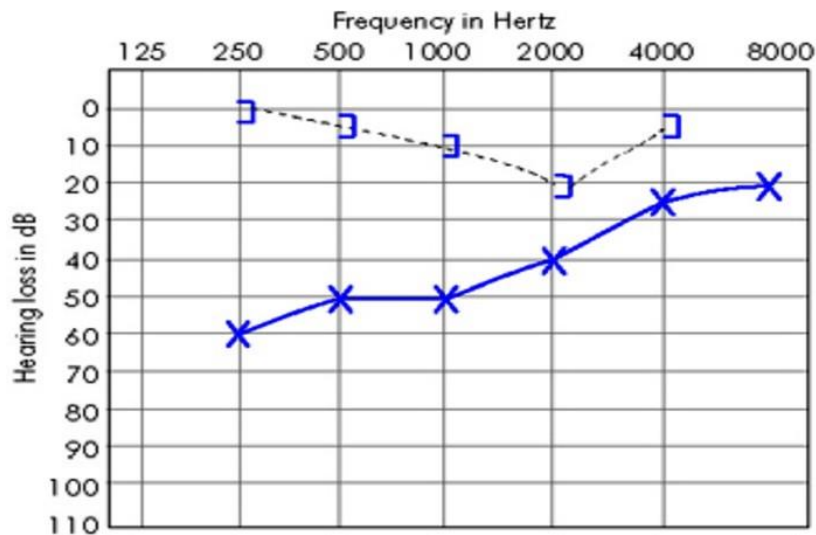
- 2. High Frequency SNHL:**

- Presbycusis
- Ototoxicity



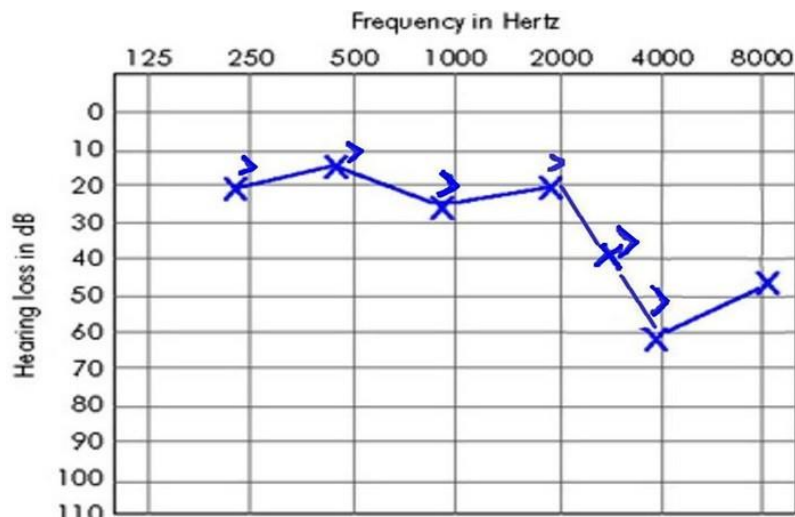
3. Carhart's Notch:

- Depression of BC occurs maximum at **2000 Hz**.
- Mechanical artifact secondary to stapes fixation and the change in the normal ossicular resonance, which is around 2000 Hz in human.
- Occurs in any condition which reduces the inertial vibration of the stapes footplate:
 - Otosclerosis
 - Tympanosclerosis
 - Ossicular fixation



4. Acoustic Dip:

- High frequency SNHL at **4000 Hz**.
- Noise-induced hearing loss.
- Because natural resonance frequency of EAC is at 3000 Hz but routine PTA tests only 4000 Hz.



5. Cookie Bite (U-Shape):

- Hereditary hearing loss.

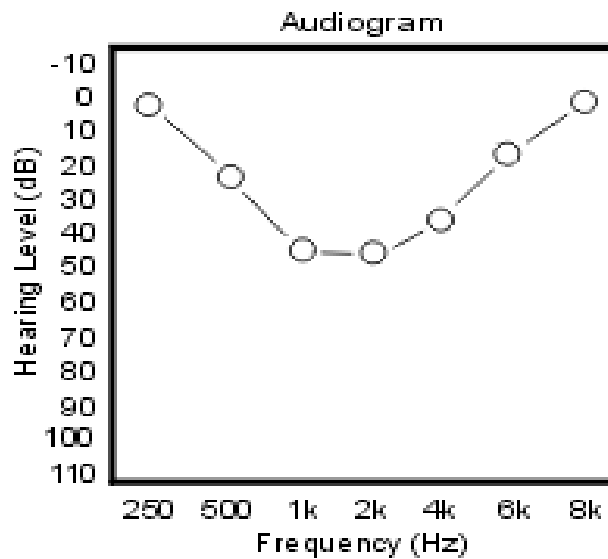


Fig. 9. Cookie-bite loss

- Uses of PTA:
 1. Measure of threshold of hearing.
 2. Predict speech reception threshold.
 3. Documentation for future reference.
 4. Prescription of hearing aid.
 5. Medicolegal purposes.

Test-retest variability in PTA threshold should be ± 5 dB

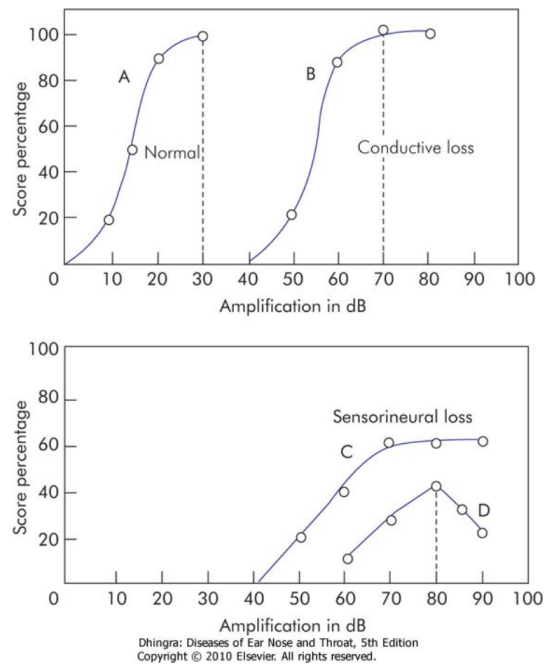
Speech Audiometry:

- Utilizes spoken voice as a sound stimulus at selected intensities.
- Measures patient's ability to hear and understand speech.
- No need to be done in patient with normal PTA.
- Two parameters are tested:
 - o Speech Reception Threshold (**SRT**).
 - o Speech Discrimination Score (**SDS**).
- **Speech Reception Threshold (SRT):**
 - o Measures hearing threshold for speech.
 - o Minimum intensity (dB) at which the patient can repeat correctly 50% of **Spondees**.
 - o Spondees are 2-syllable words with balanced accents (e.g. baseball, sunlight, day-dream).
 - o **Normally, SRT should be within 10 dB of PTA average threshold.**
 - o Indications of masking in SRT:
 - Difference between SRT of the tested ear and SRT of non-tested ear is ≥ 45 dB.
 - o **Recruitment:**
 - Increasing sound intensity (dB) produces an out-of-proportion perception of loudness.
 - Loud sound which is tolerable in normal ear may grow to abnormal levels of loudness in the recruiting ear and becomes intolerable.
 - Recruitment causes discomfort within small range of SRT.
 - **Seen in cochlear SNHL (Meniere's disease, presbycusis).**
- **Speech Discrimination Score (SDS):**
 - o Measures patient's ability to understand speech.
 - o Percentage of **Phonemes** repeated correctly after being presented at 40 dB SL above the SRT.
 - o Phoneme is a phonetically balanced (PB) single syllable words (e.g. pin, sin, day, bus, etc).
 - o SDS is best in CHL followed by cochlear SNHL followed by retrocochlear SNHL.
 - o **Roll over:**
 - Paradoxical decrease in SDS with increasing speech intensity (dB).
 - **Seen in Retrocochlear hearing loss (Vestibular schwannoma).**

Table 4-2. Ability to understand speech and its relation to speech discrimination (SD) score

A list of 50 PB words is presented and the number correctly heard is multiplied by 2.

SD score	Ability to understand speech
90-100%	Normal
76-88%	Slight difficulty
60-74%	Moderate difficulty
40-58%	Poor
<40%	Very poor

**Figure 4.3 Speech audiogram.**

- A-PB score in a normal person 100% at 30 dB.
- B-PB score in conductive hearing loss 100% at 70 dB. This curve runs parallel to that of a normal person.
 - C-Cochlear SNHL. PB max is at 70 dB and then attains a plateau.
- D-Roll over curve: PB max at 80 dB. PB scores decline as intensity increases further.

- Uses of Speech Audiometry:

1. Measures SRT:

- Correlate SRT with PTA average to diagnose non-organic (functional) hearing loss.

2. Measures SDS:

- Important for HA fitting.
- Differentiate cochlear from a retrocochlear SNHL.

Impedance Audiometry:

- Battery of Objective tests.
- Providing information about external ear, middle ear, inner ear, acoustic nerve and brain-stem function.
- Consists of:

1. Tympanometry:

- Compliance of Middle ear system.
- Middle Ear Air Pressure.
- EAC volume.

2. Acoustic (Stapedial) Reflex.

3. Otoacoustic Emissions.



- Immittance Meter:

- Consists of a probe which fits into EAC and has the following channels:

- **Oscillator:**

- Generates a tone on a frequency of **226 Hz** (1000 Hz in neonates < 6 months).
- Delivered to a probe that is sealed in Ear canal.

- **Microphone:**

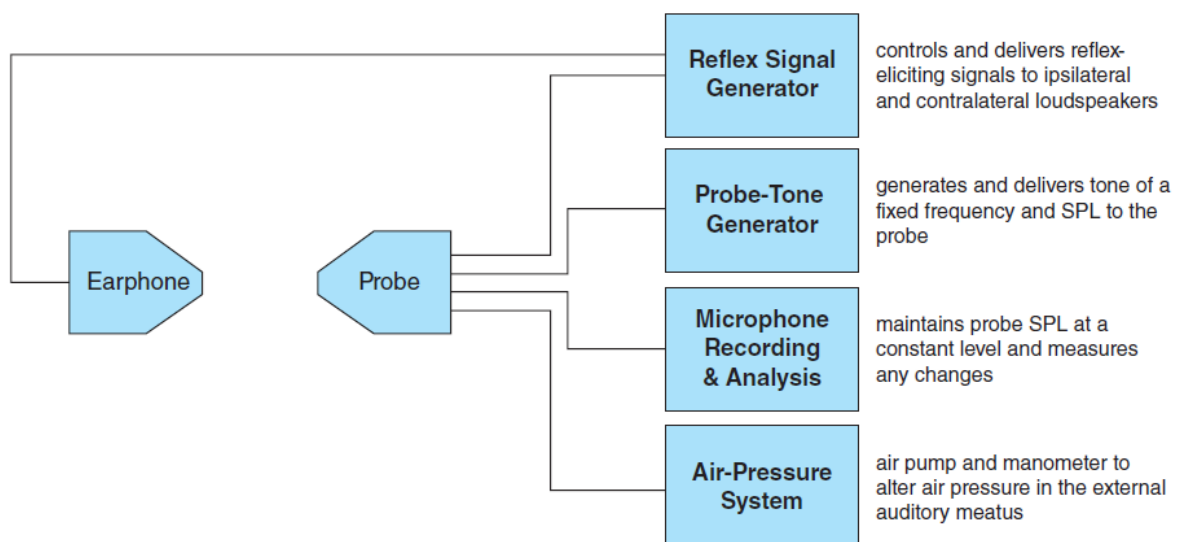
- Pick up and measure sound pressure level reflected from the tympanic membrane

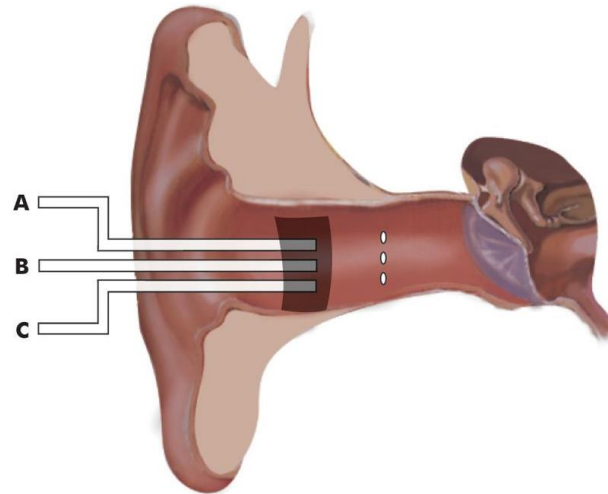
- **Air Pump:**

- Measures and changes Air pressure in the ear canal.

- **Reflex Signal Generator and Transducers (for Stapedial Reflex):**

- Delivers high-intensity signals to the ear for eliciting Acoustic Reflexes.
- Transducer is either an earphone on the ear opposite to the probe ear or a speaker within the probe itself.





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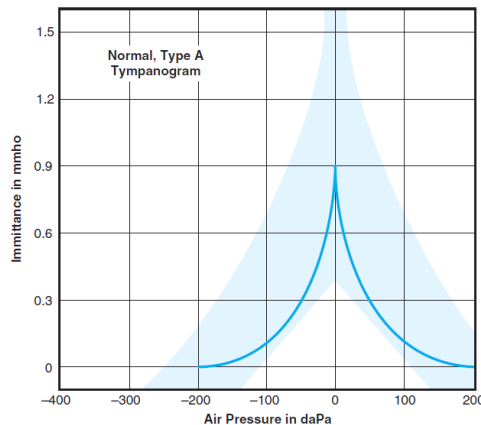
Figure 4.5 Principle of impedance audiometry. (A) Oscillator to produce a tone of 220 Hz. (B) Air pump to increase or decrease air pressure in the air canal. (C) Microphone to pick up and measure sound pressure level reflected from the tympanic membrane.

- **Tympanometry:**
- Indirect test of Middle ear function by transmission/reflection of sound energy.
- Provides an estimation of:
 - Intra-tympanic pressure
 - Eustachian tube function
 - Tympanic membrane integrity and mobility
 - Continuity of the ossicular chain.
- Transmission of sound through Middle-ear mechanism is MAXIMAL when air pressure is EQUAL on both sides of TM.
- When a sound strikes tympanic membrane, some of sound energy is absorbed while the rest is reflected.
 - Stiff tympanic membrane would reflect more of sound energy than a compliant one.
- By changing the pressures in a sealed EAC and then measuring the reflected sound energy, it is possible to find the compliance or stiffness of the tympano-ossicular system and thus find the healthy or diseased status of the middle ear.
- **Tympanogram:**
 - Plots compliance changes of TM versus Air pressure in EAC.
 - Peak:
 - Represents the point of maximum compliance.
 - EAC air pressure = Middle ear air pressure
 - Normally at 0 daPa.
- **Ear Canal Volume:**
 - Normal value in Children < **1 ml.**
 - Normal value in Adults < **2 ml.**

Types of Tympanogram:

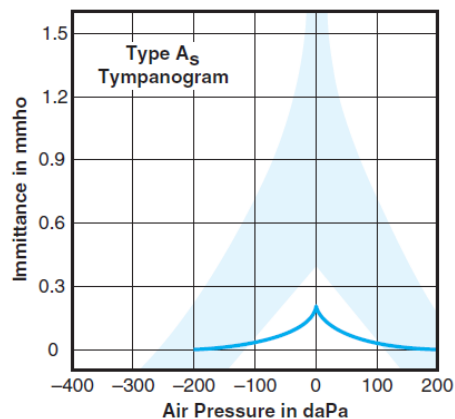
- **Type A:**

- Normal middle-ear pressure.
- Normal compliance of TM.
- Features:
 - Sharp peak at 0 daPa of Air pressure.
 - Normal range between **-100 to +50 daPa**.
 - Height of the peak (compliance) between **0.3-1.5 ml**.
- Example:
 - **Normal patient (Negative predictive value of 95%).**



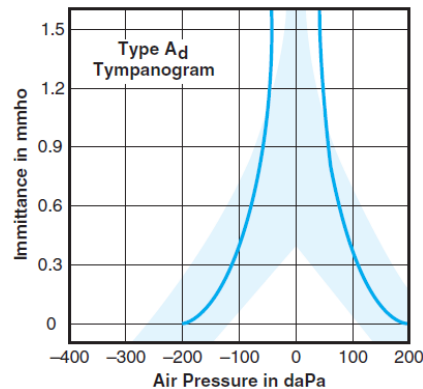
- **Type A_s:**

- Normal shape tympanogram.
- Normal middle-ear pressure.
- Reduced compliance of TM.
- Features:
 - Sharp peak at 0 daPa of Air pressure.
 - Normal range between **-100 to +50 daPa**.
 - **Shallow peak < 0.3 ml**.
- Examples:
 - Ossicular fixation (Otosclerosis, Tympanosclerosis).



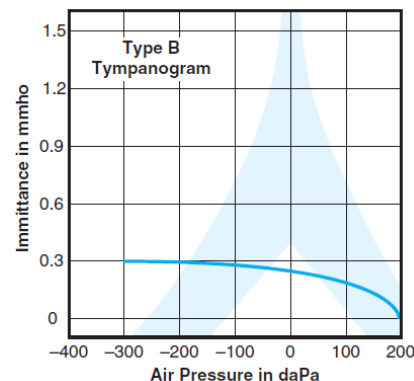
- **Type Ad:**

- Normal shape tympanogram.
- Normal middle-ear pressure.
- Increased compliance of TM.
- Features:
 - Sharp peak at 0 daPa of Air pressure.
 - Normal range between **-100 to +50 daPa**.
 - **Deep peak > 1.5 ml.**
- Examples:
 - Ossicular Discontinuity.
 - Thin and lax TM.



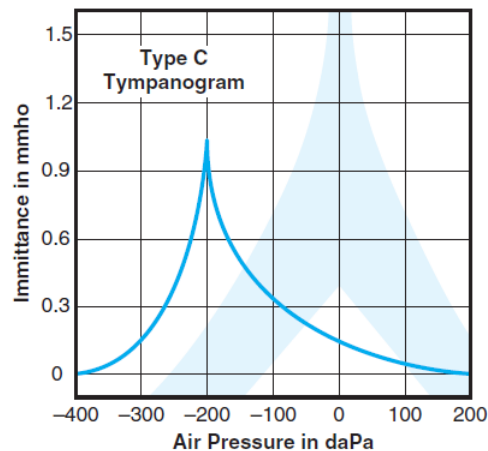
- **Type B:**

- Reduced compliance of TM.
- Features:
 - Flat or rounded shape tympanogram.
 - No change in compliance with Pressure changes.
- Examples:
 - **High canal volume:**
 - TM perforation or patent VT.
 - **Normal canal volume:**
 - **OME (positive predictive value of 90%).**
 - Thick TM.
 - **Low canal volume:**
 - EAC mass (wax, tumor).



- **Type C:**

- Normal shape tympanogram.
- Negative middle-ear pressure.
- Features:
 - Sharp peak at **< -100** daPa of Air pressure.
 - **Type C1:** Peak between **-100 to -150** daPa.
 - **Type C2:** Peak between **< -150** daPa.
 - Regardless the compliance of TM.
- Examples:
 - Eustachian Tube Dysfunction.
 - Early stages of OME.



- **Type D:**

- Normal shape tympanogram.
- Normal middle-ear pressure.
- Increased compliance of TM.
- Features:
 - Sharp peak at 0 daPa of Air pressure.
 - Normal range between **-100 to +50** daPa.
 - **Deep peak > 1.5 ml.**
 - **Notched peak.**
- Examples:
 - Scarred TM
 - Hypermobility TM

Tympanometry for testing Eustachian Tube function:

- Negative or positive pressure (-200 or +200 mm of H₂O) is created in middle ear and person is asked to swallow 5 times in 20 seconds.
- Ability to equilibrate pressure indicates **NORMAL** ET function.

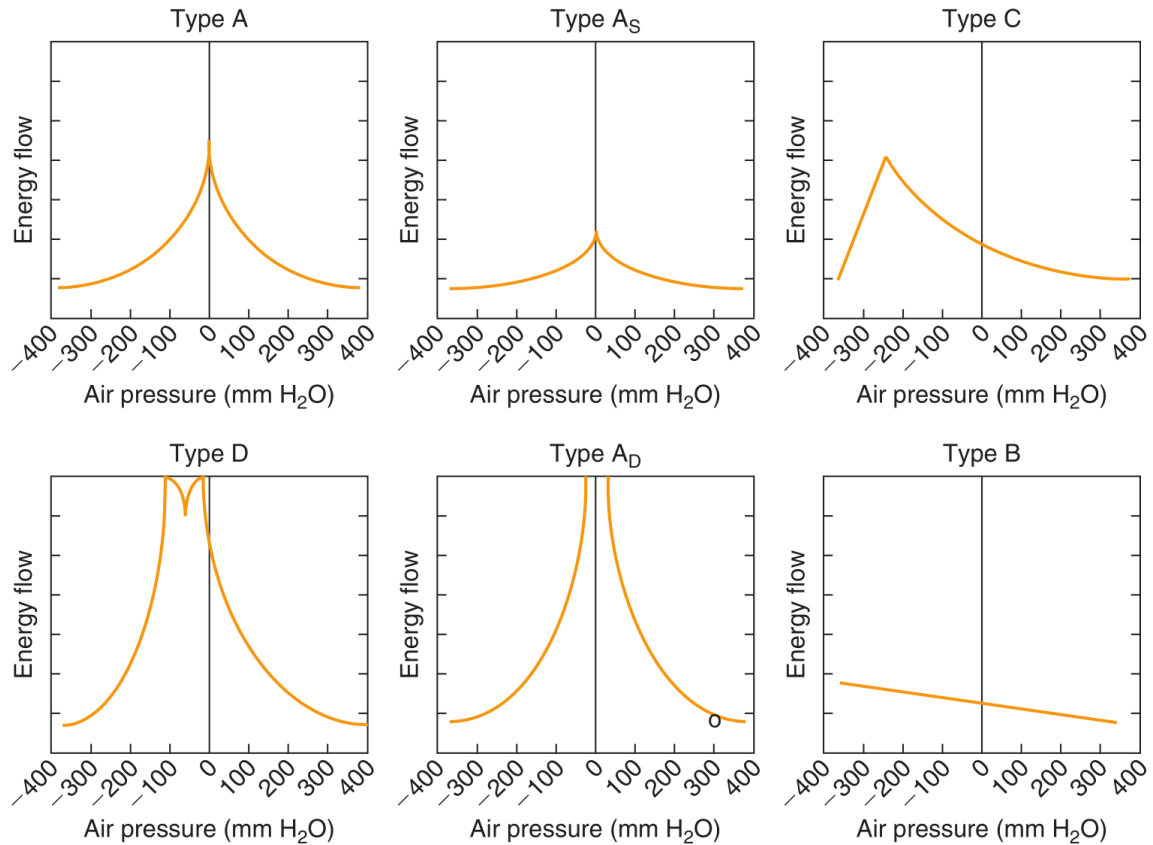


FIGURE 133-5. Classification of tympanograms: type A, type A_s, type C, type D, type A_D, and type B.

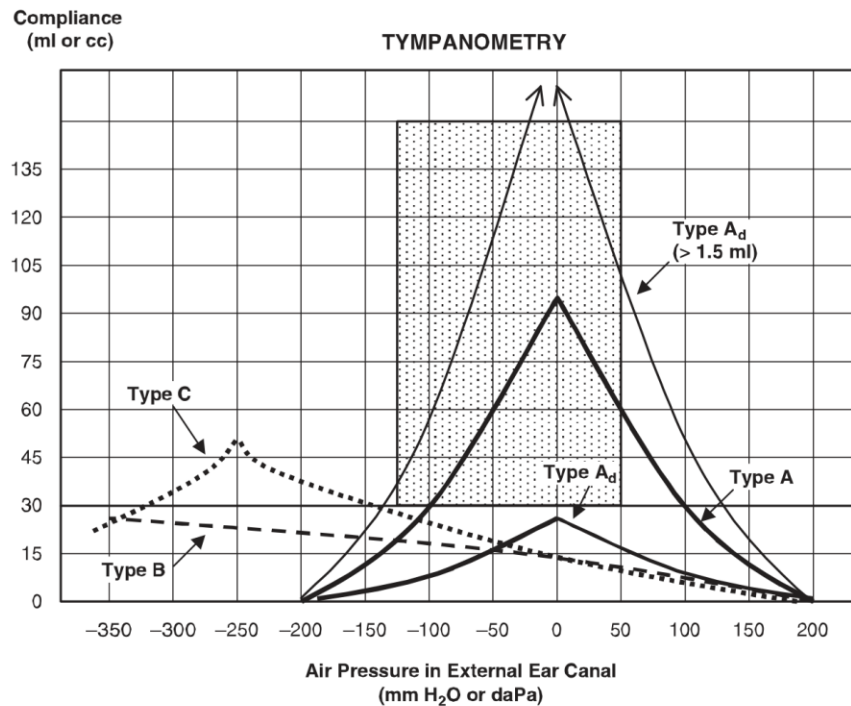


Figure 142.2 Classification system for tympanograms.

Stapedial (Acoustic) Reflex:

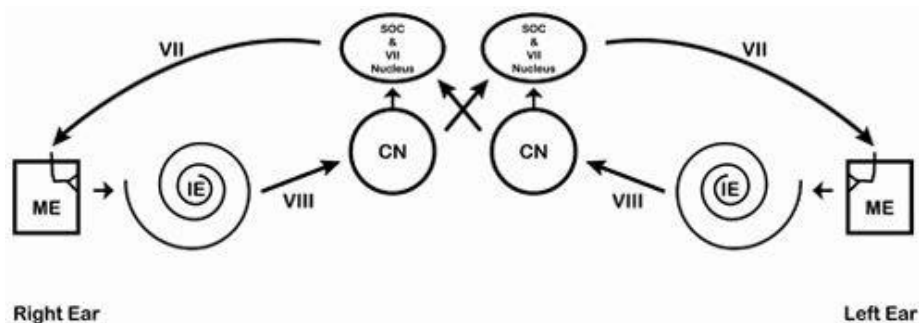
- Measure changes in TM compliance caused by contraction of stapedius muscle.
- Stapedius muscle tendon is attached to head of stapes.
 - o High intensity loud sound causes BILATERAL contraction of stapedius muscles (ipsilateral contraction stronger than contralateral contraction).
 - o When it contracts, stapedial tendon exerts tension on the stapes, stiffens the tympano-ossicular chain and reduces energy transmission through the middle ear.

Method:

- o High intensity sound (**70-100 dB HL**) above hearing threshold is introduced in each ear at 500, 1000 and 2000 Hz.
 - Sound stimuli should NOT exceed 110 dB HL
 - Duration of stimuli should NOT exceed 1 second.
- o Ipsilateral (uncrossed) and contralateral (crossed) reflexes are recorded with sound presented to each ear.
- o Normally, Stapedial reflex produces a change TM compliance in both ears which can be detected on by tympanometry.

Reflex Arc:

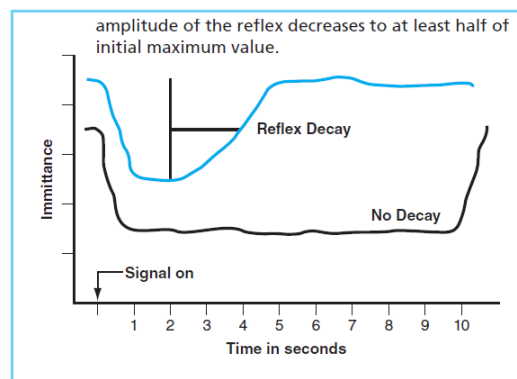
- o Ipsilateral high intensity sound (70-100 dB HL)
- o → Ipsilateral Cochlea
- o → Ipsilateral CN-VIII
- o → Ipsilateral Cochlear Nucleus
- o → Bilateral Superior Olivary Nuclei
- o → Bilateral CN-VII Nuclei
- o → Bilateral Stapedius muscles



Causes of ABSENT Stapedial reflex:

1. Any degree of CHL (Most common cause)
2. Cochlear SNHL > 60 dB
3. Any degree of Retro-cochlear SNHL
4. Brain-stem lesions
5. CN-VII dysfunction proximal to stapedial nerve

- Two parameters are tested:
 - Stapedial Reflex Threshold.
 - Stapedial Reflex Decay.
- **Stapedial Reflex Threshold:**
 - Lowest sound intensity level (dB) at which middle ear immittance change can be detected.
 - Normally elicited bilaterally at **70–100 dB SL**.
 - Allows estimation of degree of cochlear hearing loss.
 - Positive stapedial reflex at lower intensities (40-60 dB) than the usual 70 indicates Recruitment in cochlear SNHL.
- **Stapedial Reflex Decay:**
 - Measures the ability of stapedius muscle to maintain sustained contraction.
 - Method:
 - Sound signal is presented **10 dB** above stapedial reflex threshold for **10 seconds**.
 - Interpretation:
 - Normally, the patient will be able to sustained stapedial reflex.
 - Abnormal result is considered if the amplitude of the reflex decreases $\geq 50\%$ of its initial value within 5 seconds.
 - Indicate **Retro-cochlear** pathology.



Middle-Ear Condition	Tympanogram	Acoustic Reflex
Normal	A	normal
Increased mass	B	absent
Increased stiffness	A _s	absent
Excessive compliance	A _d	absent
Negative pressure	C	abnormal
TM perforation	B	absent

- Uses of Stapedial Reflex:

1. Neonatal Hearing test:

- Not frequently used since ABR and OAEs provide more information.

2. Diagnose Malingers:

- No response in PTA.
- Positive Stapedial Reflex.

3. Diagnose cochlear SNHL:

- Positive stapedial reflex at < 70 dB HL
- Indicates Recruitment.

4. Diagnose Retro-cochlear SNHL:

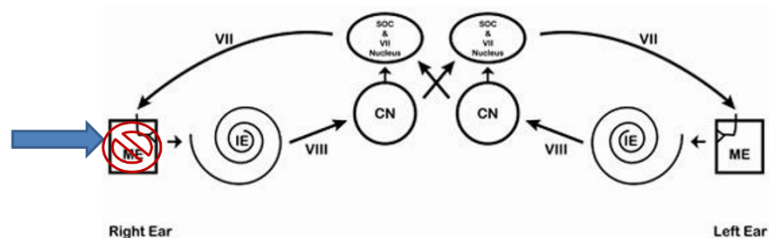
- Abnormal Stapedial Reflex decay.

5. Diagnose Facial nerve lesion proximal to stapedial nerve:

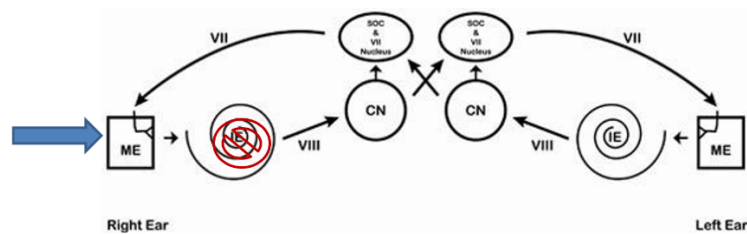
- Absence stapedial reflex without Hearing Loss.

6. Diagnose Brain-stem lesions

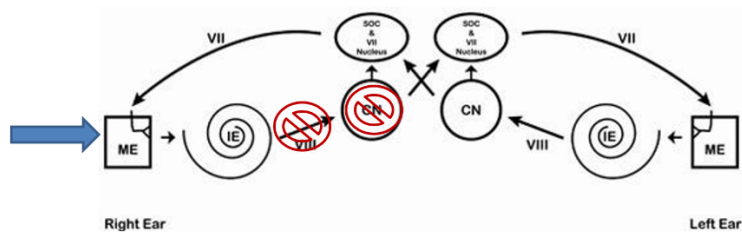
Clinical Examples:



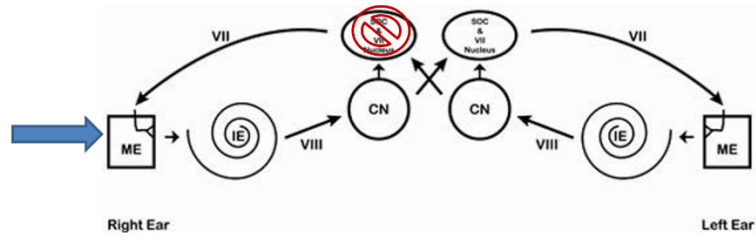
uncross	Cross	Example
Absent	Absent	Any CHL



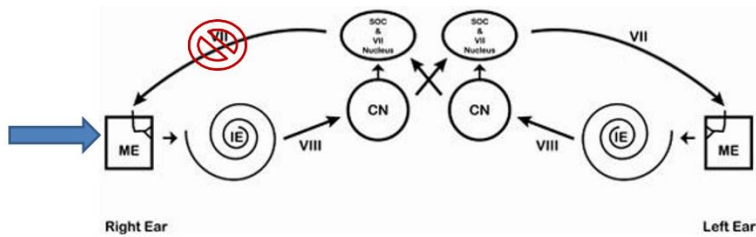
uncross	Cross	Example
Absent	Absent	Cochlear SNHL >60 db



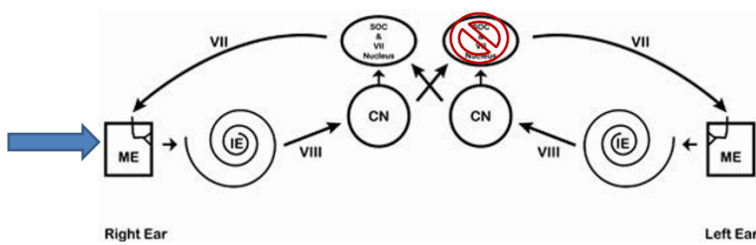
uncross	Cross	Example
Absent	Absent	Any Retro-cochlear SNHL



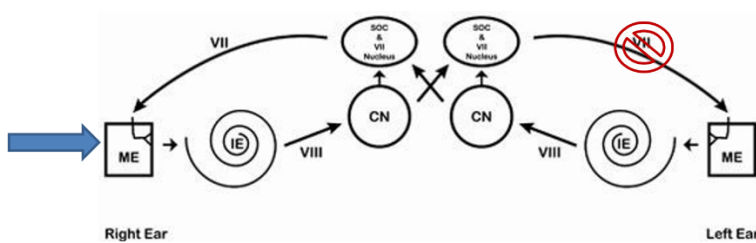
uncross	Cross	Example
Absent	Positive	Right Brainstem lesion



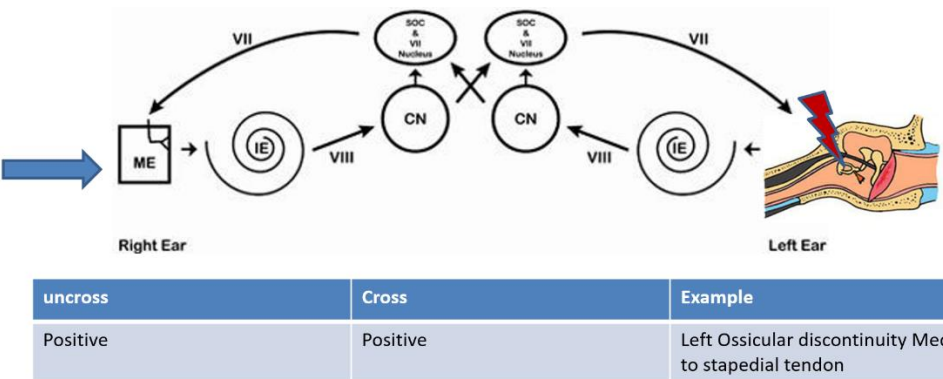
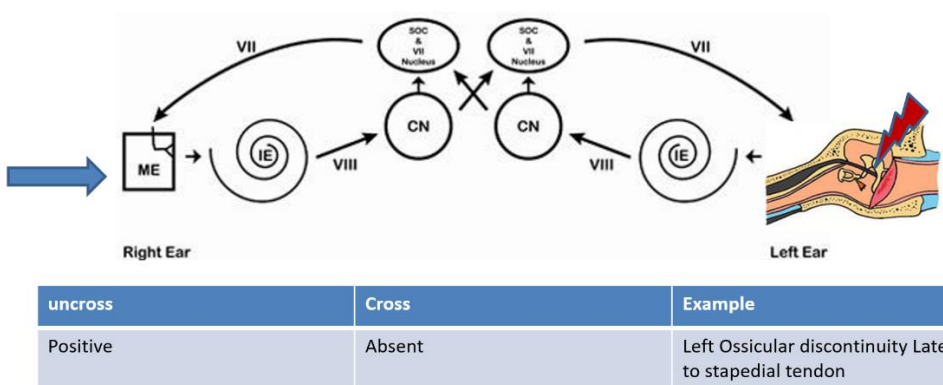
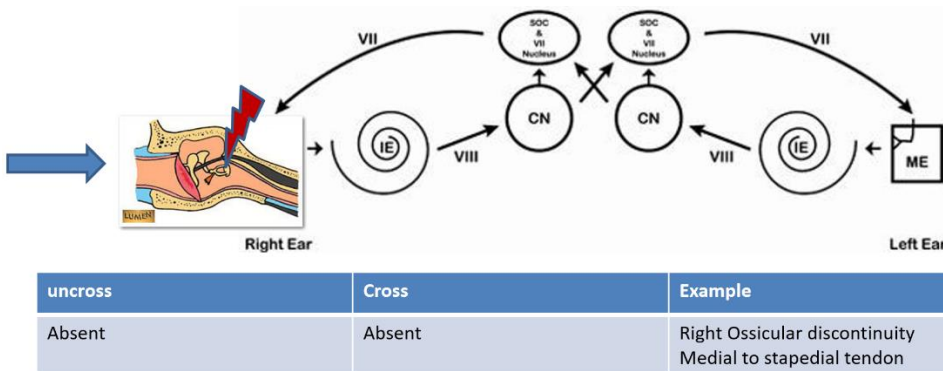
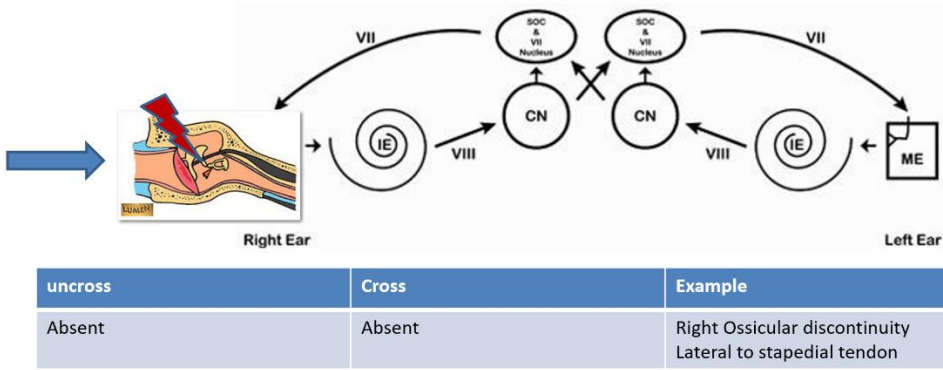
uncross	Cross	Example
Absent	Positive	Right Facial Nerve Palsy



uncross	Cross	Example
Positive	Absent	Left Brainstem lesion



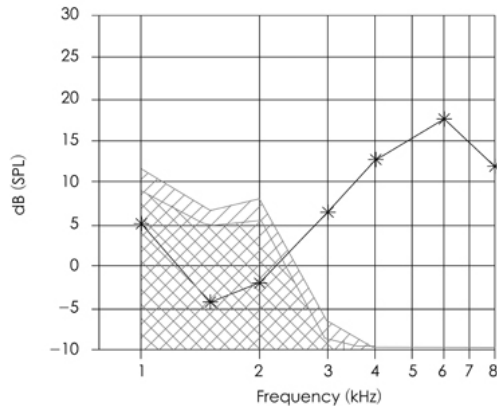
uncross	Cross	Example
Positive	Absent	Left Facial Nerve Palsy



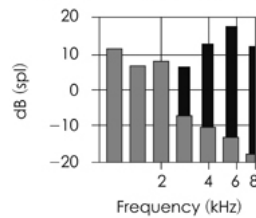
Otoacoustic Emissions (OAEs):

- Low intensity sounds produced by healthy outer hair cells (OHC) of normal cochlea.
- Evaluate the function of outer hair cells (Cochlea).
- OAEs travels in a reverse direction:
 - OHC
 - → Basilar membrane
 - → Perilymph
 - → Oval window
 - → Ossicles
 - → TM
 - → EAC
- Normal OAE indicates:
 - Normal cochlea (OHC)
 - Normal middle ear system
 - Normal external ear system
- Types of OAEs:
 - **Spontaneous OAEs:**
 - Presents only in 60% of normal people.
 - Doesn't required external stimulus.
 - **Not used for screening (not sensitive).**
 - **Evoked OAEs:**
 - 1. Transiently Evoked OAEs (TEOAEs):**
 - Presents in all normal ears.
 - Evoked by clicks or tone bursts.
 - Absent if hearing loss ≥ 30 db.
 - **Used for Neonatal hearing screening.**
 - 2. Distortion Product OAEs (DPOAEs):**
 - Presents in all normal ears.
 - Evoked by simultaneous 2 pure tone frequencies (F1 and F2).
 - Evaluate higher frequencies than TEOAE.
 - Absent if hearing loss ≥ 50 db.
 - **Used for hearing screening due to Ototoxicity and Noise induced HL.**
 - **Detect Ototoxic effects before conventional PTA but NOT before HF-PTA.**
 - 3. Stimulus-Frequency OAEs:**
 - Presents in all normal ears.
 - Evoked by a continuous pure tone stimulus.

Ear: Left
Date/Time: 2010-11-17 오후 2:55:10
Test type: DP
Stimulus: 75/65 dB 2 pts/oct
F2/F1: 1.22
Points/Oct: 2
Mode: Gen Diag
Tester ID: ABC
Data file: 7PBKBH38.DPG
Notes:



Half octave band OAE power

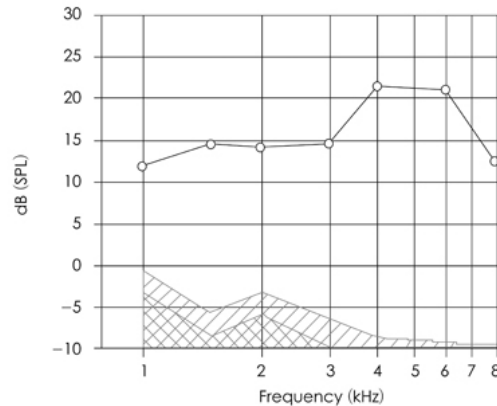


Freq (kHz)	Signal (dB SPL)	Noise (dB SPL)	SNR (dB)
1.0	5.2	11.5	-6.3
1.4	-4.4	6.6	-11.0
2.0	-2.1	7.9	-10.0
2.8	6.5	-6.8	13.3
4.0	12.8	-9.7	22.5
8.0	17.4	-12.7	30.1
9.0	11.9	-17.5	29.4

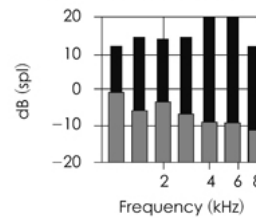
Test summary

Sum all 1/2 octave=18.9 dB SPL Ave DP 1/2 oct (1-6)=11.2 dB SPL

Ear: Right
Date/Time: 2010-11-17 오후 2:55:50
Test type: DP
Stimulus: 75/65 dB 2 pts/oct
F2/F1: 1.22
Points/Oct: 2
Mode: Gen Diag
Tester ID: ABC
Data file: 7PBKBH39.DPG
Notes:



Half octave band OAE power



Freq (kHz)	Signal (dB SPL)	Noise (dB SPL)	SNR (dB)
1.0	11.9	-0.6	12.6
1.4	14.5	-5.5	20.0
2.0	14.1	-3.1	17.2
2.8	14.4	-6.3	20.7
4.0	21.3	-8.5	29.8
8.0	20.9	-9.3	30.2
9.0	12.4	-11.2	23.6

Test summary

Sum all 1/2 octave=25.5 dB SPL Ave DP 1/2 oct (1-5)=17.7 dB SPL

- Evoked OAEs are Present in:
 1. Normal hearing level.
 2. Any Hearing Loss < 30 dB in TEOAEs.
 3. Any Hearing Loss < 50 dB in DPOAEs.
 4. Retro-cochlear SNHL.
- Evoked OAEs are Absent in:
 1. EAC pathology (wax impaction or lesions).
 2. Middle ear pathology (OME, Otosclerosis).
 3. Cochlear SNHL > 30 dB in TEOAEs.
 4. Cochlear SNHL > 50 dB in DPOAEs.

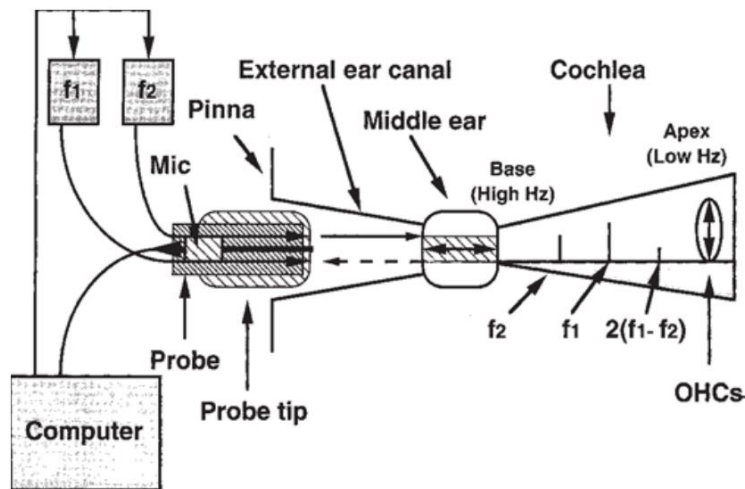


Figure 142.6 Equipment and procedure for measurement of distortion product otoacoustic emissions (DPOAEs). The two stimulus frequencies (f_1 , f_2) are presented to the ear with a soft probe (inward-pointing arrows). The DPOAEs (at the frequency $2f_1-f_2$) produced by the outer hair cells are propagated outward through the middle ear to the external ear canal. Amplitude of DPOAE in dB SPL is plotted as a function of the frequency of the stimuli (the f_2 stimulus).

- Uses of Evoked OAEs:
 1. **Neonate hearing screening (TEOAE).**
 2. **Hearing screening for Ototoxicity and Noise-induced HL (DPOAE).**
 3. **Diagnosing of Retro-cochlear SNHL:**
 - SNHL in PTA
 - Normal OAE.
 4. **Diagnosing of Malingerers:**
 - Hearing loss in PTA
 - Normal OAE.

Auditory Brainstem Response (ABR):

- Also known as:
 - Brainstem Auditory Evoked Response (BAER).
 - Brainstem Auditory Evoked Potential (BAEP).
- Non-invasive and objective test to record the activity of peripheral and central auditory systems (from cochlea to cortex) in response to an auditory signal (clicks or tone bursts).
- Auditory response is not affected by patient conscious level and can be recorded during sleeping, sedation or coma.
- Method:
 - Auditory stimuli (clicks, or tone bursts) are introduced rapidly to the ear (20-30 times/second).
 - These stimuli are detected by surface electrodes placed on the forehead and earlobe.
 - Electrical potentials (waves) are generated in response to these auditory stimuli.
 - Normally, 7 waves are produced within 10 ms after stimuli and each wave correlates to a specific anatomical location (**E-COLI**):
 - **Wave I:**
 - Originates from **Distal CN-VIII**.
 - Present at birth.
 - Stable wave.
 - Hallmark for peripheral auditory function.
 - **Wave II:**
 - Originates from **Proximal CN-VIII**.
 - **Wave III:**
 - Originates from **Cochlear Nucleus**.
 - Present at birth.
 - Stable wave.
 - **Wave IV:**
 - Originates from **Superior Olivary Complex**.
 - **Wave V:**
 - Originates from **Lateral Lemniscus**.
 - Largest wave.
 - Present at birth.
 - Stable wave.
 - Used to estimate Hearing threshold.
 - **Wave VI-VII:**
 - Originates from **Inferior Colliculus**.
 - Absolute (From stimuli to the wave), inter-wave and inter-aural latencies of each wave are calculated and compared.
 - Normal inter-wave latencies:
 - I-III = **2.0 ms**
 - III-V = **2.0 ms**
 - I-V = **4.0 ms**

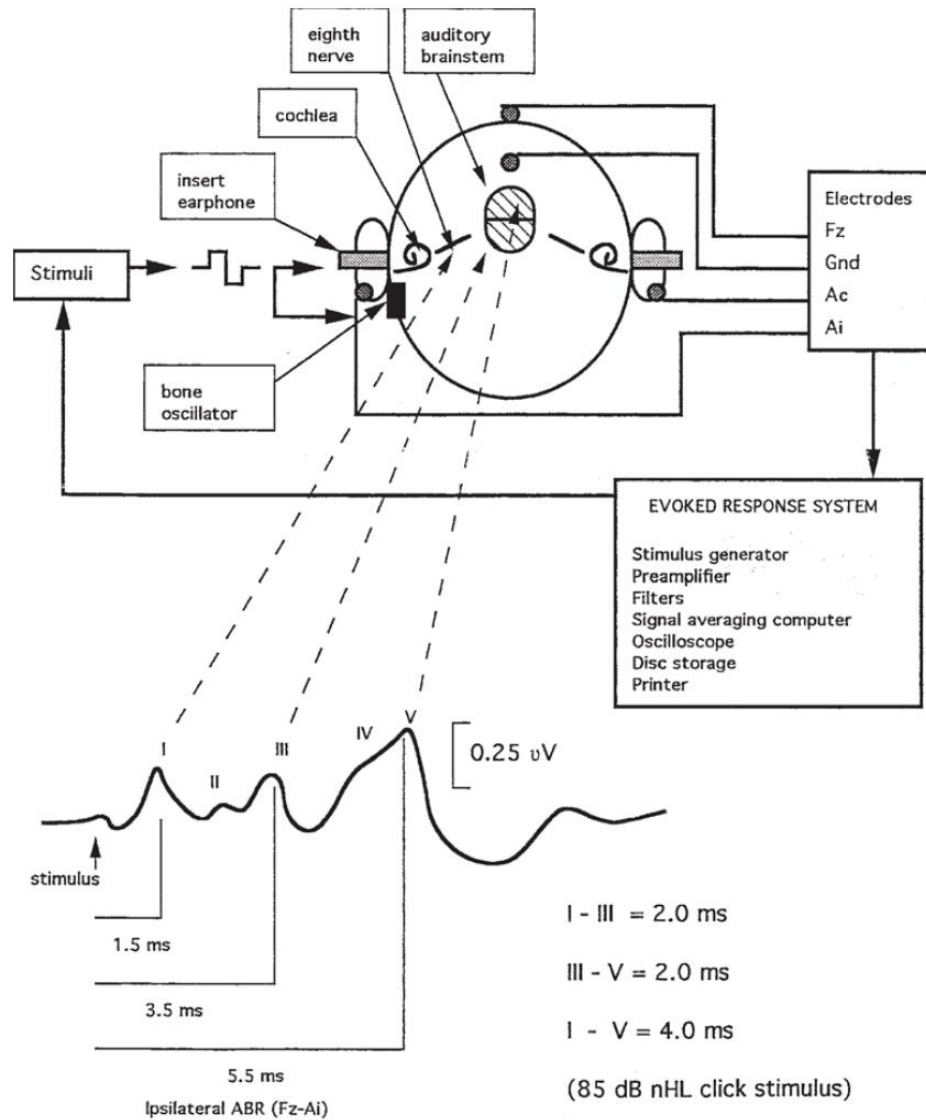


Figure 142.3 Schematic shows the instrumentation for recording the ABR and important relations between auditory anatomic features and waveform components. A simple strategy for analysis of ABR waveform in neurodiagnosis also is shown.



- Common ABR waveform patterns

- **Normal:**
 - Wave I:
 - Normal shape.
 - Normal latency.
 - Normal inter-wave latencies.
 - Good morphology.
- **CHL:**
 - Wave I:
 - **Normal shape.**
 - **Delayed latency.**
 - Normal inter-wave latencies.
 - Good morphology.
- **Cochlear SNHL:**
 - Wave I:
 - **Small or absent.**
 - **Delayed latency.**
 - Normal inter-wave latencies.
 - **Poor morphology.**
- **Retro-cochlear SNHL:**
 - Wave I:
 - Normal shape.
 - Normal latency.
 - **Delayed inter-aural wave V latency (>0.2 ms).**
 - **Delayed wave I-III latency (>2.0 ms).**
 - **Delayed inter-wave latencies.**
 - **Poor morphology.**

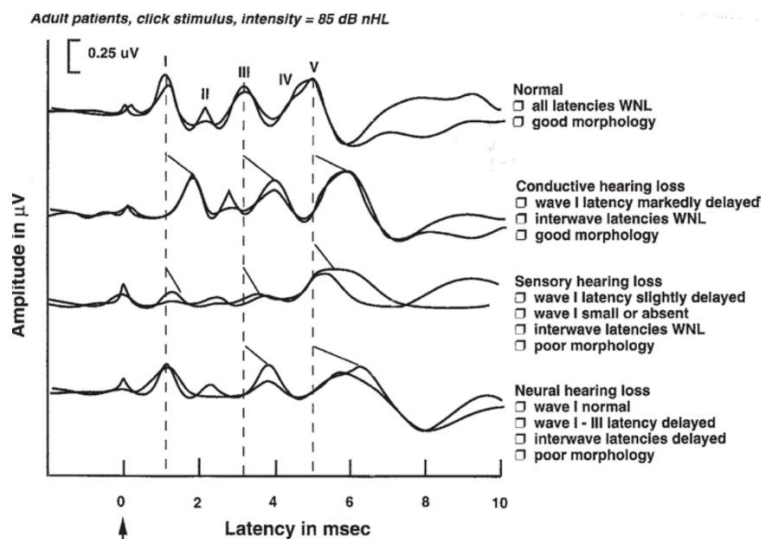


Figure 142.4 ABR patterns associated with various types of auditory dysfunction. Information about dysfunction can be inferred from the latency of specific waves and overall structure of the waveforms.

- Uses of ABR:

1. Objective estimation of hearing threshold:

- Method:
 - Wave V is observed within 10 dB of PTA.
- Indications:
 - Neonates (Hearing screening).
 - Suspected malingering.
- Limitation:
 - Maximum intensity level to elicit ABR is **80 dB**.
 - Cannot be used to estimate hearing sensitivity in severe-to-profound HL.
- Indications:
 - High Risk neonates.
 - Failed OAE.

2. Diagnosing Retro-cochlear lesions:

- Sensitivity of ABR depends on the size of VS:
 - 60-80% sensitive for small VS < 1cm.
 - 90% sensitive for medium and large VS.
- Less sensitive than MRI for small tumors resulting in false-negative results in small intra-canicular tumors.
- Not considered initial diagnostic tool for VS except in patients with contraindication for MRI (pacemaker).

3. Intra-operative monitoring:

- Used for electrophysiological monitoring of CN-VIII and auditory brainstem function during hearing preservation lateral skull base surgeries (vestibular nerve section and posterior fossa tumor resection).

Electrocochleography (ECoG):

- Objective test recording the electrical potentials arising in the peripheral auditory system (cochlea and CN VIII) in response to an auditory signal (clicks or tone bursts).
- Done under LA or GA.
- Method:
 - Surface electrode is placed on the vertex or pinna.
 - Probe is placed either in:
 - EAC adjacent to TM (non-invasive).
 - Promontory via trans-tympanic needle electrode (invasive).
 - Auditory signals (clicks) and the electrical potentials are recorded.
 - Types of recorded electrical potentials:
 - **Endocochlear Potential:**
 - DC response generated from Stria vascularis.
 - Maintained at + 80 mV by Na-K ATPase.
 - **Cochlear Microphonic Potential (CM):**
 - AC response generated from OHC.
 - **Summation Potential (SP):**
 - DC response generated from OHC.
 - **CN-VIII Action Potential (AP):**
 - All or none response generated from distal CN-VIII.
 - Equivalent to ABR wave I.
- Uses of ECoG:
 - 1. Diagnosing Ménière's disease:**
 - Tested with electrode placed in the EAC on the TM.
 - Normally SP/AP is < 30%.
 - **SP/AP > 50%** indicates Ménière's disease.
 - Sensitivity of 60%.
 - 2. Diagnosing Auditory Neuropathy:**
 - Absence of wave V on ABR with positive ECoG (CM).
 - Useful in determining whether cochlear implantation is appropriate treatment option or not.
 - 3. Intra-operative monitoring:**
 - Direct measurement of cochlear and CN-VIII function.

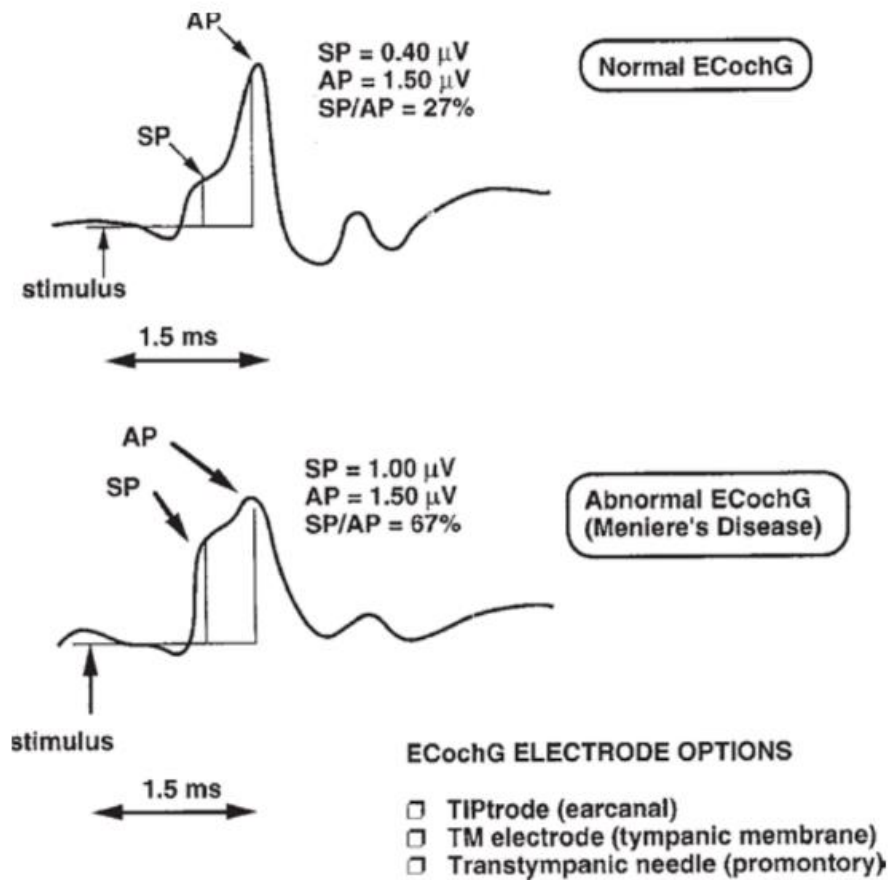


Figure 142.5 ECoG waveforms show normal relation of SP and AP and abnormally enlarged SP/AP relation for a patient with Ménière disease. Absolute and relative amplitude values for the SP and AP components and the criteria for definition of a normal response vary greatly for different electrode sites (ear canal vs. tympanic membrane vs. promontory).

Approach to Hearing Loss in Adults:

History

- Character of Hearing Loss:
 -
 - Constant vs. fluctuating.
 - Progression
 - Unilateral vs. bilateral
 - High or low tone loss.
 - Decreased speech intelligibility
- Contributing Factors:
 - Recent infection (fevers, URTI).
 - Loud noise exposure.
 - Recent trauma (barotrauma, straining, weight lifting, head injury).
 - Exacerbating factors of tinnitus (sleep, exercise, caffeine, alcohol, stress).
 - Previous otologic history (surgery, infections).
 - Toxin exposure and medications.
 - History of autoimmune disease, hypertension, diabetes, vascular disorders, TMJ disease, neurologic disease (stroke) and depression.
 - Family history of deafness.
- Associated Symptoms:
 - Aural fullness, fevers, vertigo, tinnitus, otalgia, otorrhea, weight loss, other neurologic complaints

TABLE 6–2. Common Differential Diagnosis of Hearing Loss: KITTENS Method

(K) Congenital	Infectious and Idiopathic	Toxins and Trauma	Tumor (Neoplasms)	Endocrine	Neurologic	Systemic/ PSychological
Hereditary progressive SNHL	Labyrinthitis	Ototoxic medication	Temporal Bone neoplasms	Hypothyroiditis	Multiple sclerosis	Ménière's disease
	Otitis media			Hyperlipidemia	Meningitis	Paget's disease
Congenital auricular malformations	Luetic hearing loss	Noise induced hearing loss	Cholesteatomas		Stroke	Otosclerosis
	Presbycusis	Head trauma			Central auditory processing disorder	Presbycusis
Congenital hearing loss		Lead and mercury toxicity				Cerumen impaction
		Barotrauma				Pseudohypacusis
						Autoimmune disorders

Physical Exam and Hearing Tests

- Otoscopy/Microscopy:
 - o Inspection of EAC (cerumen impaction, lesions, masses).
 - o Inspection of TM (color, thickness, presence of fluid, myringosclerosis, perforations, lesions).
 - o Valsalva test (test patency of eustachian tube by having subject perform Valsalva maneuver and inspect TM for mobility)
- Pneumatic Otoscopy:
 - o Test mobility of TM with positive and negative pressure.
 - o Fistula test (Hennebert's sign; positive pressure causes nystagmus which reverses with negative pressure, may be seen in perilymph fistulas and syphilitic labyrinthitis).
- Inspection and Palpation:
 - o Inspect outer ear for lesions, malformations, auricular pits, scars, edema, swelling, mastoid tenderness, tragal tenderness.
- Complete Head and Neck Exam:
 - o Cervical lymphadenopathy.
 - o Neurologic and vestibular exam, bruits.
- **Tuning Fork Tests**
- Rinne Test:
 - o Typically uses a 512 Hz (C1 fork) and 1024 Hz tuning forks to compare air conduction (AC) and bone conduction (BC).
 - o Strike tuning fork then place within 1 cm of the entrance to the ear canal (AC) and then immediately place on the mastoid (BC).
 - o When the tuning fork was better heard by AC then the test is referred to as **Positive** (normal ears or most SNHL).
 - o If BC is perceived louder than AC the test is referred to as **Negative** (CHL >15–30 dB HL or severe to profound SNHL with cross-over)
- Weber Test:
 - o Typically utilizes a 512 Hz (C1 fork) and 1024 Hz tuning forks.
 - o Strike tuning fork and place in center of forehead, vertex, or upper teeth.
 - o Perceived sound should **Normally** be heard **Centrally** (or in "both ears").
 - o **Unilateral SNHL** should **Lateralize to better hearing ear**.
 - o **Unilateral CHL** should **Lateralize to diseased ear**.
- **Audiometric Tests**
- Essential to identify auditory function, CHL vs. SNHL, cochlear vs. neural dysfunction, central auditory dysfunction, and pseudohypacusis.
 - o Pure Tone Audiometry (PTA)
 - o Tympanometry
 - o Otoacoustic Emissions (OAE)
 - o Auditory brainstem response (ABR)

Ancillary Tests

- **Imaging:**
- Radiographs:
 - o Special views of the temporal bone (Schuller's, Stenvere's, Towne's views) have largely been replaced by CT and MRI.
- CT Temporal Bone:
 - o Indicated to evaluate the complications of suppurative ear disease, tumors, cholesteatoma, mastoiditis, temporal bone fracture, or a congenital disorder
- MRI of Brain and Brainstem with Gadolinium:
 - o Indicated if suspect cerebellopontine angle tumors (vestibular schwannoma, meningioma), demyelinating lesions (multiple sclerosis), or petrous apex lesions (cholesterol granuloma)
- **Laboratory Studies:**
- May be considered for specific circumstances
- CBC:
 - o May suggest active inflammation or leukemic process
- Lipid Profile:
 - o High risk of atherosclerotic disease (associated with HL)
- Glucose:
 - o Screen for diabetes (associated with HL).
- Coagulation Profile:
 - o Coagulopathies are associated with HL.
- Treponemal Studies:
 - o Lyme titers and VDRL/FTA-ABS
- Immunological Profiles

- **Hearing Loss:**
- Types of hearing problems:
 1. **Hearing impairment:** Both complete and partial loss of the ability to hear.
 2. **Deafness:** Complete loss of hearing in one or both ears.
- **Degree of Hearing Loss:**
- WHO recommended the following classification on the basis of pure tone audiogram taking the average of the thresholds of hearing for frequencies of 500, 1000 and 2000 Hz.

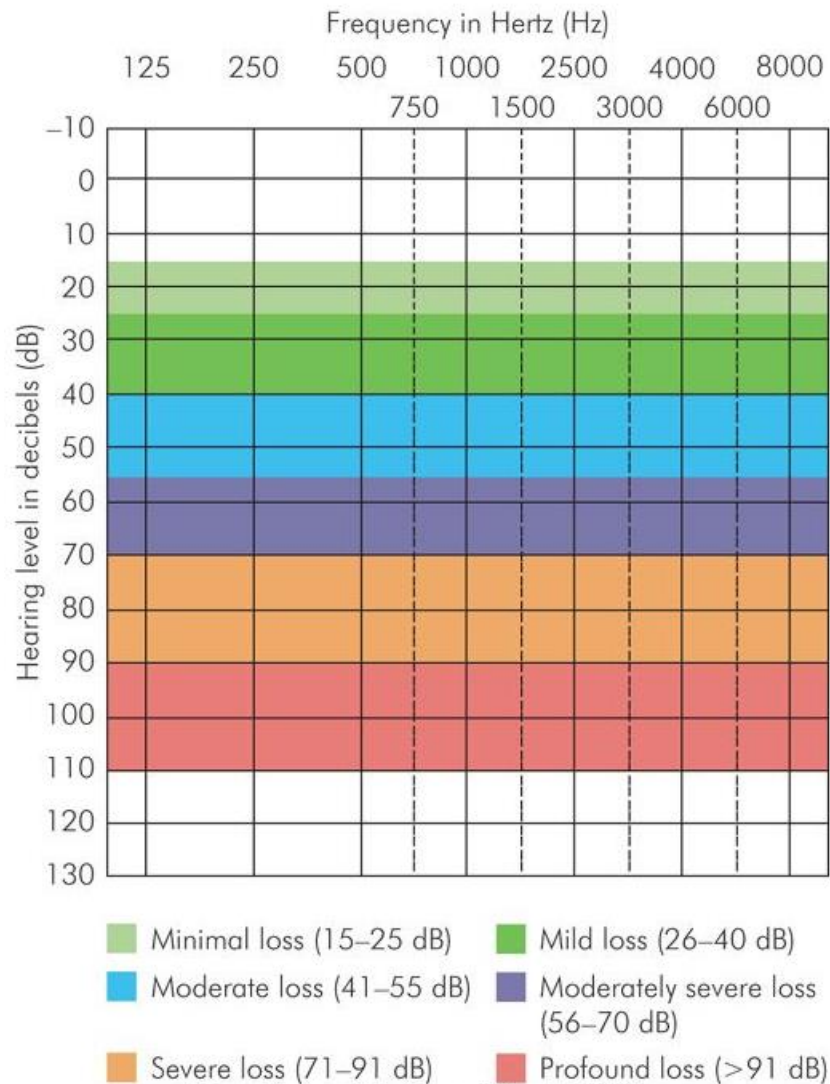
Degree of hearing loss	Hearing loss range (dB HL)
Normal	-10 to 15
Slight	16 to 25
Mild	26 to 40
Moderate	41 to 55
Moderately severe	56 to 70
Severe	71 to 90
Profound	91+

Source: Clark, J. G. (1981). Uses and abuses of hearing loss classification. *Asha*, 23, 493-500.

- There is no apparent impairment of hearing from 0 to 25 dB.
- **Disease --> Impairment --> Disability --> Handicap**
- When a disease process strikes an organ or a system it causes an **Impairment** either in structure or function, but this impairment may or may not become clinically manifested.
- When impairment affects the ability to perform certain functions in the range considered normal for that individual it is called **Disability**.
- The disability further restricts the duties and roles expected from an individual by society and is called a **Handicap**.
- To exemplify, injury (disease) to the ear may result in hearing impairment which, depending on its severity, will affect the individual's ability to hear and perform certain activities (disability) and will be termed as handicap by the society.

Table 5-5. Hearing loss and difficulty in hearing speech

Hearing threshold in better ear (average of 500, 1000, 2000 Hz)	Degree of impairment (WHO classification)	Ability to understand speech
0-25	Not significant	No significant difficulty with faint speech
26-40	Mild	Difficulty with faint speech.
41-55	Moderate	Frequent difficulty with normal speech.
56-70	Moderately severe	Frequent difficulty even with loud speech.
71-91	Severe	Can understand only shouted or amplified speech.
Above 91	Profound	Usually cannot understand even amplified speech.



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- **Calculating hearing handicap:**

1. Take an audiogram and calculate the average of thresholds of hearing for frequencies of 500, 1000, 2000 and 3000 Hz (A).
2. Deduct from it 25 dB (as there is no impairment up to 25 dB) (A-25).
3. Multiply it by 1.5 ((A-25) × 1.5)
4. This is the percentage of hearing impairment for that ear.
5. Similarly calculate the percentage of hearing impairment for the other ear.

1. Hearing loss does not begin to be handicapping until the **PTA** at 0.5, 1, 2, and 3 kHz exceeds 25 dB HL
2. Handicap grows at rate of 1.5% per decibel of HL beyond 25 dB
3. Because unilateral deafness is only a mild handicap, the two ears should not be equally weighted (5:1 favoring better ear)

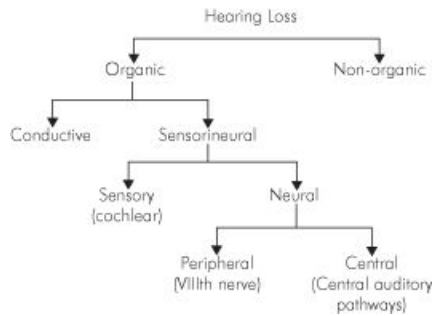
$$\textbf{Monaural Impairment (MI) = 1.5 (PTA - 25)}$$

$$\textbf{Binaural Hearing Impairment (BHI) = [5 (MI_{better}) + (MI_{worse})] / 6}$$

- **Unilateral Hearing Loss**

- Unilateral loss of hearing does not produce a serious handicap or affect speech.
- It impairs localisation of the sound source, difficulty in discrimination of speech in the presence of background noise.
- Patient has to take all precautions for the safety of the only hearing ear.
- Surgeon should be careful when he is called upon to operate on this only hearing ear.

Classification of HL:



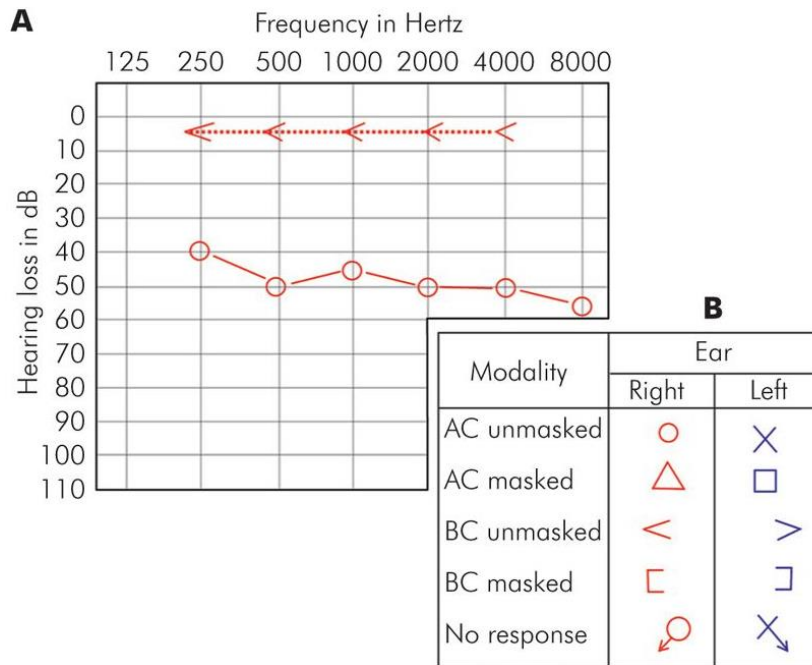
- **Conductive Hearing Loss (CHL):**
- Any disease process which interferes with the conduction of sound to reach cochlea causes conductive hearing loss.
- Lesion may lie in the external ear and tympanic membrane, middle ear or ossicles up to stapediovestibular joint.
- Characteristics of CHL:
 1. Negative Rinne test, BC > AC
 2. Weber Lateralised to Poorer ear.
 3. Normal absolute bone conduction.
 4. Low Frequencies affected more.
 5. Audiometry shows bone conduction better than air conduction with air-bone gap. Greater the air-bone gap, more is the conductive loss.
 6. Loss is not more than 60 dB.
 7. Good Speech discrimination.
- **Aetiology of CHL:**
- Congenital or acquired causes.

Table 5-1. Congenital causes of conductive hearing loss

Meatal atresia
Fixation of stapes footplate
Fixation of malleus head
Ossicular discontinuity
Congenital cholesteatoma

Table 5-2. Acquired causes of conductive hearing loss

External ear	Any obstruction in the ear canal, e.g. wax, foreign body, furuncle, acute inflammatory swelling, benign or malignant tumour or atresia of canal.
Middle ear	• (a) Perforation of tympanic membrane, traumatic or infective • (b) Fluid in the middle ear, e.g. acute otitis media, serous otitis media or haemotympanum • (c) Mass in middle ear, e.g. benign or malignant tumour • (d) Disruption of ossicles, e.g. trauma to ossicular chain, chronic suppurative otitis media, cholesteatoma • (e) Fixation of ossicles, e.g. otosclerosis, tympanosclerosis, adhesive otitis media • (f) Eustachian tube blockage, e.g. retracted tympanic membrane, serous otitis media.



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Figure 5.1 (A) Audiogram of right ear showing conductive hearing loss with A-B gap. (B) Symbols used in audiogram charting.

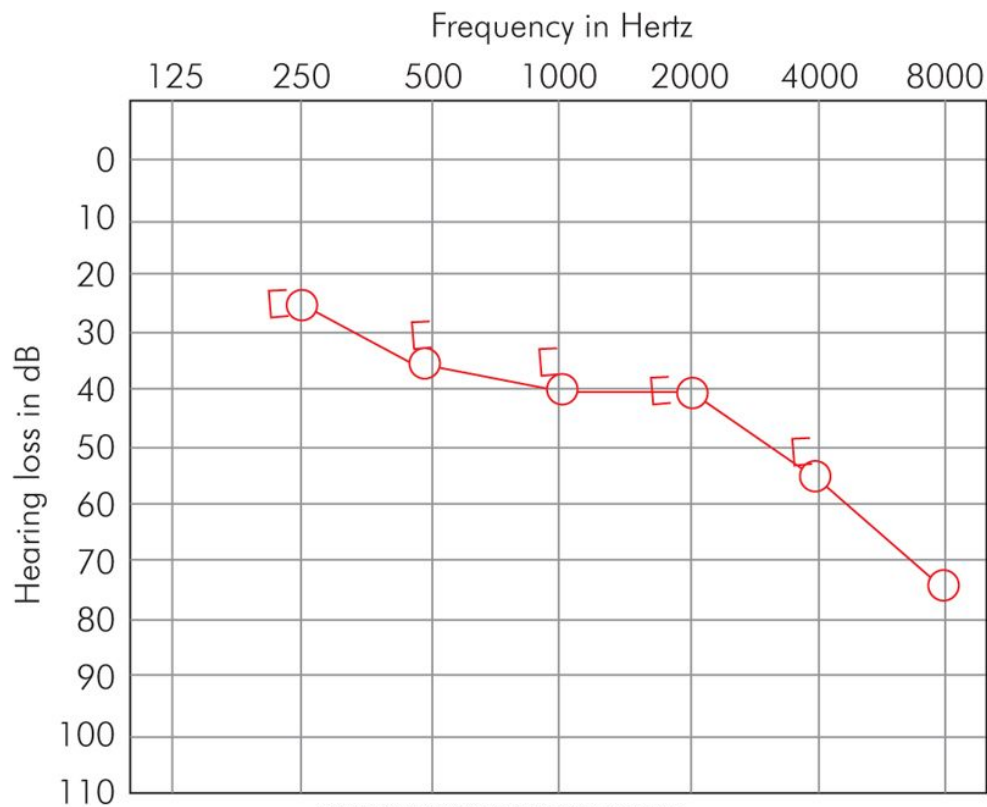
Average Hearing Loss Seen in Different Lesions of Conductive Apparatus

1. Complete obstruction of ear canal:	30 dB
2. Perforation of tympanic membrane (It varies and is directly proportional to the size of perforation):	10-40 dB
3. Ossicular interruption with intact drum:	54 dB
4. Ossicular interruption with perforation:	38 dB
5. Malleus fixation:	10-25 dB
6. Closure of oval window:	60 dB

Note here that ossicular interruption with intact drum causes more loss than ossicular interruption with perforated drum.

- **Management of CHL:**
- Most CHL can be managed by medical or surgical means.
- Consists of:
 1. **Removal of canal obstructions:**
 - e.g. impacted wax, foreign body, osteoma or exostosis, keratotic mass, benign or malignant tumours, meatal atresia.
 2. **Removal of Middle ear fluid:**
 - Myringotomy with or without grommet insertion.
 3. **Removal of Middle ear mass:**
 - Tympanotomy and removal of small middle ear tumours or cholesteatoma behind intact tympanic membrane.
 4. **Stapedectomy:**
 - In otosclerotic fixation of stapes footplate.
 5. **Tympanoplasty:**
 - Repair of perforation, ossicular chain or both.
 6. **Hearing aid:**
 - In cases, where surgery is not possible, refused or has failed.

- **Sensorineural Hearing Loss (SNHL):**
- Results from lesions of the cochlea, VIIIth nerve or central auditory pathways.
- May be present at birth (congenital) or start later in life (acquired).
- Characteristics of SNHL:
 - o Positive Rinne test, air AC > BC.
 - o Weber Lateralised to Better ear.
 - o Reduced absolute bone conduction.
 - o More often involving High frequencies.
 - o No gap between air and bone conduction curve on audiometry.
 - o Loss may exceed 60 dB.
 - o Poor Speech discrimination.
 - o Difficulty in hearing in the presence of noise.



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Figure 5.6 Audiogram of right ear showing sensorineural loss with no A-B gap.

- **Aetiology of SNHL**
- Congenital
 - Present at birth and is the result of anomalies of the inner ear or damage to the hearing apparatus by prenatal or perinatal factors.
- Acquired
 - Appears later in life.
 - Genetic or non-genetic causes
 - Genetic causes may manifest late (delayed onset) and affect only the hearing, or be a part of a larger syndrome affecting other systems of the body as well.
- Common causes of acquired SNHL include:
 1. Labyrinthitis
 2. Trauma to labyrinth or VIIIth nerve, e.g. fractures of temporal bone or concussion of labyrinth or ear surgery,
 3. Noise-induced hearing loss,
 4. Ototoxic drugs,
 5. Presbycusis,
 6. Meniere's disease,
 7. Acoustic neuroma,
 8. Sudden hearing loss,
 9. Familial progressive SNHL,
 10. Systemic disorders, e.g. diabetes, hypothyroidism, kidney disease, autoimmune disorders, multiple sclerosis, blood dyscrasias.

Table 15-1. Results of various tests to differentiate a cochlear from a retrocochlear lesion

	Normal	Cochlear lesion	Retrocochlear lesion
• Pure tone audiogram	Normal	Sensorineural hearing loss	Sensorineural hearing loss
• Speech discrimination score	90-100%	Below 90%	Very poor
• Roll over phenomenon	Absent	Absent	Present
• Recruitment	Absent	Present	Absent
• SISI score	0-15%	Over 70%	0-20%
• Threshold tone decay test	0-15 dB	Less than 25 dB	Above 25 dB
• Stapedial reflex	Present	Present	Absent
• Stapedial reflex decay (page 109)	Normal	Normal	Abnormal
• E.R.A	Normal interval between wave I & V	Normal interval between wave I & V	Wave V delayed or absent

- **Diagnosis of SNHL:**
- History:
 - Congenital or acquired
 - Stationary or progressive
 - Associated with other syndromes or not
 - Involvement of other members of the family and possible aetiologic factors.

- Severity of Deafness:
 - o Mild, moderate, severe, profound or total.
 - o This can be found out on audiometry.
- Type of audiogram:
 - o Whether loss is high frequency, low frequency, mid-frequency or flat type.
- Site of lesion:
 - o i.e. cochlear, retrocochlear or central.
- Radiology:
 - o CT temporal bone for evidence of bone destruction (congenital cholesteatoma, glomus tumour, middle ear malignancy or acoustic neuroma).
- Laboratory tests
 - o Depend on the Aetiology suspected,
 - o Blood counts (leukaemia)
 - o Blood sugar (diabetes),
 - o Thyroid functions (hypothyroidism),
 - o Serology for syphilis.
 - o Kidney function tests, etc.
- Management of SNHL:
- Early detection of SNHL is important as measures can be taken to stop its progress, reverse it or to start an early rehabilitation program, so essential for communication.
- Syphilis of the inner ear:
 - o Treatable with high doses of penicillin and steroids with improvement in hearing.
- Hearing loss of hypothyroidism:
 - o Reversed with replacement therapy.
- Serous labyrinthitis:
 - o Reversed by attention to middle ear infection.
- Meniere's disease:
 - o Early management prevents further episodes of vertigo and hearing loss.
- SNHL due to perilymph fistula:
 - o Corrected surgically by sealing the fistula in the oval or round window with fat.
- Ototoxic drugs:
 - o Should be used with care and discontinued if causing hearing loss.
 - o It may be possible to regain hearing, total or partial, if the drug is stopped
- Noise induced hearing loss:
 - o Prevented from further deterioration if the person is removed from the noisy surroundings.

- **Non-organic Hearing Loss (NOHL)**
- Pseudohypacusis.
- Subjective loss of hearing by a patient with absence of organic pathology.
- 1. Malingering:**
 - There is a motive to claim some compensation for being exposed to industrial noises, head injury or ototoxic medication.
- 2. Psychogenic.**
- Patient may present with any of the three clinical situations:
 - Total hearing loss in both ears
 - Total loss in only one ear
 - Exaggerated loss in one or both ears.
- Responsibility of the physician is to find out:
 - Is the patient malingering?
 - If so, what is his actual threshold of hearing?
- **Diagnosis:**
- 1. High index of suspicion:**
 - Patient hesitates or expresses confusion.
 - History is inconsistent with hearing loss recorded on tests.
 - Makes exaggerated efforts to hear.
 - Frequently making requests to repeat the question.
 - Placing a cupped hand to the ear.
- 2. Tuning forks:**
 - Stenger test:
 - Principle is that, if a tone of two intensities, one greater than the other, is delivered to two ears simultaneously, only the ear which receives tone of greater intensity will hear it.
 - Take two tuning forks of equal frequency, strike and keep them say 25 cm from each ear.
 - Patient will claim to hear it in the normal ear only.
 - Now bring the tuning fork on the side of feigned deafness to within 8 cm, keeping the tuning fork on the normal side at the same distance.
 - Patient will deny hearing anything even though tuning fork on normal side is where it could be heard earlier.
 - A Normal person or patient with true deafness should continue to hear on the normal side.
 - Patient should be blind-folded during this test.
 - Lombard Test:
 - Noisy background is gradually introduced below the subject's recorded response threshold.
 - Subject is asked to read aloud.
 - Pseudohypacusis is suspected if the volume of the reader's voice increases as the masking noise increased.

3. PTA:

- Inconsistent results on repeated PTA:
 - Normally, the result of repeat tests are within **±5 dB**.
 - **A variation greater than 15 dB is diagnostic of NOHL.**
- Absence of shadow curve:
 - Normally, a shadow curve can be obtained while testing bone conduction, if the healthy ear is not masked.
 - This is due to transcranial transmission of sound to the healthy ear.
 - Crossover should be normally occur at 0 dB for BC and 40 db for AC.
 - **Absence of this curve in a patient complaining of unilateral deafness is diagnostic of NOHL.**
- Stenger test:
 - As described previously.

4. Speech Audiometry:

- Inconsistent results on repeated SRT:
 - Normally, the result of repeat tests are within **±5 dB**.
 - **A variation greater than 15 dB is diagnostic of NOHL.**
- Inconsistency in PTA and SRT:
 - Normally, pure tone average (PTA) of frequencies (500, 1000 and 2000 Hz) is within 10 dB of SRT.
 - **SRT better than PTA by more than 10 dB points to NOHL.**

5. Acoustic reflex threshold:

- Normally, stapedial reflex is elicited at 70-100 dB SL and it should be absent if tests suggests significant hearing loss.
- **If patient claims total deafness but the reflex can be elicited, it indicates NOHL.**

6. Auditory Brainstem Response (ABR):

- **It is very useful in NOHL and can establish hearing acuity of the person to within 5-10 dB of actual thresholds.**

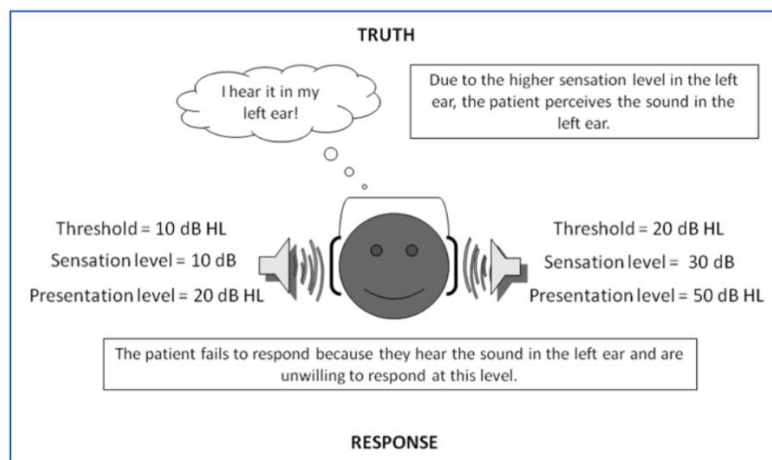


FIGURE 17-7. Positive Stenger test. The patient perceives the tone in the ear with the higher sensation level, the left ear. However, the patient has previously provided invalid responses, which would suggest that hearing in the left ear is actually poorer than it is. The patient cannot then admit that the tone was heard in the left ear. When the Stenger test tone is presented, the patient chooses not to respond to the tone.

- **Specific forms of HL:**

1. Labyrinthitis:

- Viral labyrinthitis:
 - Reach the inner ear by Blood stream.
 - Affecting stria vascularis, endolymph and organ of Corti.
 - Measles, Mumps and CMV are well documented to cause labyrinthitis.
 - Several other viruses, e.g. rubella, herpes zoster, herpes simplex, influenza and EBV are clinically known to cause deafness but direct proof of their invasion of labyrinth is lacking.
- Bacterial labyrinthitis:
 - Reach labyrinth through the middle ear (tympagogenic) or through CSF (meningogenic).
 - Sensorineural deafness following meningitis is a well-known clinical entity.
 - Bacteria can invade the labyrinth along nerves, vessels, cochlear aqueduct or the endolymphatic sac.
 - Membranous labyrinth is totally destroyed.
- Syphilitic labyrinthitis:
 - SNHL is caused both by congenital and acquired syphilis.
 - Congenital syphilis is of two types:
 1. Early form: manifesting at the age of 2 years.
 2. Late form: manifesting at the age of 8-20 years.
 - Syphilitic involvement of the inner ear can cause:
 1. Sudden SNHL which may be unilateral or bilateral. The latter is usually symmetrical in high frequencies or is a flat type.
 2. Meniere's syndrome with episodic vertigo, fluctuating hearing loss, tinnitus and aural fullness-a picture simulating Meniere's disease.
 3. Hennebert's sign. A positive fistula sign in the absence of a fistula due to fibrous adhesions between the stapes footplate and the membranous labyrinth.
 4. Tullio phenomenon in which loud sounds produce vertigo.
 - Diagnosis of otosyphilis:
 - Clinically: (interstitial keratitis, Hutchinson's teeth, saddle nose, nasal septal perforation and frontal bossing)
 - Laboratory tests:
 - FTA-ABS and VDRL or RPR tests from CSF.
 - Treatment of otosyphilis:
 - IV penicillin and steroids.

2. Familial Progressive SNHL:

- Genetic disorder.
- Progressive degeneration of the cochlea.
- Starting in late childhood or early adult life.
- Bilateral SNHL with flat or basin-shaped (U-shaped) audiogram.
- Excellent speech discrimination.

3. Ototoxin-Induced Hearing Loss:

- Drugs can damage the inner ear and cause SNHL, tinnitus or vertigo.
- **Cochlear Toxicity:**
 - o Pattern:
 - Acute cochlear damage may present as Tinnitus.
 - Initially, Bilateral HF-SNHL (>4000 Hz).
 - Later, Lower speech frequencies are affected.
 - If drug is stopped early, further loss may be prevented.
 - Partial recovery is possible, but sometimes permanent.
 - o Time of onset:
 - Unpredictable.
 - Occurs even after a single dose or after several weeks after completion of therapy.
- **Vestibular Toxicity:**
 - o Mainly with Aminoglycoside antibiotics.
 - o May appear early with Nystagmus.
 - o If severe, can lead to dysequilibrium and oscillopsia.
- Common ototoxic agents:
 - 1. Antibiotics** (Aminoglycosides, Macrolides, Vancomycin)
 - 2. Anti-neoplastic** (Cisplatin)
 - 3. Loop Diuretics**
 - 4. Salicylates**
 - 5. Antimalarials** (Quinine, Chloroquine)

1. Aminoglycoside Antibiotics:

- 2-3%.
- Aminoglycosides are cleared more slowly from inner ear than from serum and remain in cochlea long time after therapy has ended.
- Results in progression or onset of hearing loss after cessation of the therapy (delayed toxicity).
- **Continuing monitoring up to 6 months after cessation of therapy.**
- **Vestibular Toxicity:**
 - o 1-11%.
 - o The most vestibulotoxic of all ototoxic drugs.
 - o Selectively destroy type I hair cells of the crista ampullaris.
 - o If administered in large doses, cochleotoxic .
 - o Primary Vestibulotoxic Drugs:
 - Gentamicin
 - Streptomycin

- **Cochlear Toxicity:**
 - 3-13%
 - Irreversible destruction of outer hair cells (OHC) mainly at the basal turn of the cochlea causing HF-SNHL.
 - Inner hair cells are more resistant to injury due to the higher concentration of the natural antioxidant, [glutathione](#), in IHC and in the apical turn OHC compared with that in OHC of the basal turn.
 - Primary Cochleotoxic Drugs:
 - Neomycin
 - Amikacin
- Others:
 - **Tobramycin:**
 - Affects vestibular and auditory function equally.
 - **Netilmicin:**
 - Least ototoxic aminoglycosides.
- Risk factors:
 1. Elderly.
 2. Bacteremia.
 3. Hepatic or renal dysfunction.
 4. Concomitantly other ototoxic drugs.
 5. Longer duration of therapy.
 6. Increased serum levels (either peak or trough levels)

2. **Macrolides (Erythromycin/Azithromycin)**

- Sporadic cases of ototoxicity in patients with other risk factors, including:
 - Renal failure
 - Hepatic failure
 - Doses of more than 4 g/d
 - Intravenous administration
- Reversible
- Mainly Cochleotoxic.

3. **Vancomycin**

- No studies demonstrate conclusive evidence of ototoxicity with vancomycin alone and in therapeutic doses.
- Reported ototoxicity (Tinnitus), in patients with high serum concentrations attributed to renal failure or with concomitant

4. Loop Diuretics:

- 6%.
- Furosemide and Ethacrynic acid
- Block NA and Cl channels in the ascending loop of Henle.
- Ototoxicity is dose-dependant and **Reversible** in most patients, although irreversible hearing loss has been reported in patients with renal failure.
- **Mainly cochleotoxic:**
 - o SNHL and Tinnitus.
 - o Affect Stria vascularis in scala media by changes in potassium gradients.
- **Risk factors:**
 1. Renal failure
 2. Rapid infusion
 3. Concomitant aminoglycoside

5. Salicylates:

- Level > 2,700 mg/ day.
- Ototoxicity is **reversible**.
- Recovery usually occurs 24 to 72 hours after cessation of the drug.
- **Mainly cochleotoxic:**
 - o **Tinnitus**
 - o Bilateral SNHL particularly affecting higher frequencies.

6. Anti-neoplastic drugs:

- Cisplatin and carboplatin
- **Mainly cochleotoxic:**
 - o SNHL and Tinnitus.
 - o Damage Stria vascularis in scala media and outer hair cell at basal turn of cochlea.
- Ototoxicity is **irreversible**.
- Risk increase in patients receiving radiation therapy to H&N.

7. Anti-malaria:

- Quinine and Chloroquin.
- **Mainly cochleotoxic:**
 - o SNHL and Tinnitus.
 - o Quinine toxicity can present as a syndrome known as Cinchonism
- **Reversible.**
- Characteristic notch often is present at 4000 Hz.
- If taken during 1st trimester of pregnancy:
 - o Congenital deafness of child
 - o Hypoplasia of cochlea

8. Desferrioxamine:

- Iron-chelating substance used in the treatment of thalassaemic patients who receive repeated blood transfusions.
- Causes high frequency SNHL.
- Onset is sudden or delayed.
- It is permanent but might be reversible when the drug is discontinued.

9. Topical ear drops:

- Any substance enters ME (through TM perforation) can access IE via permeability of Round window membrane into perilymph of Scala Tympani.
- In patients with ear infections, Round window membrane may become thickened secondary to an immune response and the deposition of connective tissue, which renders the membrane less permeable and provide protection from ototoxicity.
- Treatment with drops for prolonged periods of time (>7 days) increases the risk of ototoxicity.
- Examples:
 - o **Aminoglycoside drops**
 - o **Chlorhexidine**

- Ototoxicity monitoring for hearing loss

- Baseline Tests:

- o Should be done prior to any ototoxic drug administration.
- o For Aminoglycosides, tests can be obtained within 72 hours of starting the treatment.
- o Three main approaches:
 - **PTA:**
 - Limited to conventional frequency range of 250-8000 Hz.
 - Does not permit the earliest detection of ototoxic changes in higher frequencies.
 - **High frequency PTA (HF-PTA):**
 - Comprises AC threshold testing for frequencies above 8000 Hz, ranging up to 16 or 20 kHz.
 - **Permit the earliest detection of ototoxic changes in higher frequencies before changes become evident in the conventional range.**
 - Not available in all institutes.
 - **Otoacoustic Emission (OAE):**
 - **Responses tend to change before hearing thresholds in the conventional frequency range, but not before changes in HFA thresholds.**
 - Major limitation is that the results are significantly affected by middle ear pathology such as otitis media.

- **Follow-up Tests:**

- Monitoring should continue during therapy at regular intervals with at least weekly testing.
- Monitoring should continue for at least 6 months following cessation of the potentially ototoxic medication.

- **Management:**

- Prevention.
- Most hearing loss is **Irreversible**.
- No therapy to reverse ototoxic damage.
- Benefits of ototoxic drugs must be weighed against potential risks.
- Alternative medications should be considered when appropriate.
- **For severe hearing loss, Amplification is the only treatment option.**

▪ **Prevention:**

1. Monitor serum drug levels.
2. Monitor renal function.
3. Frequent Hearing evaluation:
 - Baseline pre-treatment hearing evaluation.
 - Before subsequent antineoplastic drug cycles.
 - Periodic evaluation after completion of therapy and up to 6 months post therapy.
4. Avoid noise exposure up to 6 months post therapy.

TABLE 6–1. Common Ototoxic Medications

Category	Representative Examples and Comments
Antibiotics	aminoglycosides exert their ototoxic effects by damaging vestibular and cochlear hair cells, streptomycin and gentamicin affect vestibular function more than auditory function (hearing loss occurs initially with supra-audiological thresholds, >8 kHz), vancomycin causes synergistic ototoxic effects with aminoglycosides
Chemotherapy Agents	cisplatin may cause auditory nerve and vestibular toxicity
Diuretics	furosemide and ethacrynic acid are the most common diuretic ototoxic agents, often potentiate other ototoxic medications
NSAIDs and Salicylates	aspirin is the most common medication to cause tinnitus (reversible with discontinued use)

4. Noise Trauma:

- Occupational hazard.
- Occupational Safety and Health Administration (OSHA) regulations exposure equivalent to > 90 dB time-weighted average for 8 hours requires hearing protection programs (equivalent exposure to 95 dB for 4 hours, 100 dB for 2 hours, or 130 dB for less than 2 minutes)
- Stapedial reflex is protective for sounds < 2 kHz.
- Stapedial reflex has a latency of 10 ms, thus will not protect cochlea from unexpected sounds.
- Hearing loss caused by excessive noise can be divided into two groups:

1. Acoustic trauma:

- o Permanent damage to hearing can be caused by a single brief exposure to very intense sound.
- o E.g. an explosion, gunfire or a powerful cracker.
- o More severe losses, especially in low frequencies.
- o Noise level in rifle or a gun fire may reach 140-170 dB SPL.
- o Damage outer hair cells, mechanical damage to organ of Corti and rupture the Reissner's membrane.
- o Concomitantly rupture TM and disrupt ossicular chain.

2. Noise-induced hearing loss (NIHL)

- o Chronic exposure to less intense sounds than seen in acoustic trauma.
 - o Mainly a hazard of noisy occupations.
 - o NIHL causes damage to hair cells, starting in the basal turn of cochlea.
 - o Outer hair cells are affected before the inner hair cells.
 - o Rate of hearing loss due to chronic noise exposure is greatest during the first 10–15 years of exposure and decreases as the hearing threshold increases (decelerating process).
 - o This is in contrast to age-related hearing loss, which accelerates over time (accelerating process)
- Temporary Threshold Shift (TTS):
 - o Temporary SNHL which occurs immediately after exposure to noise and resolves within 24 hours.
 - Permanent Threshold Shift (PTS):
 - o Permanent SNHL from noise exposure that does not recover.
 - o Does not progress after exposure ceased.
 - Symptoms of NIHL:
 - o High pitched tinnitus
 - o Difficulty in hearing in noisy surroundings
 - o No difficulty in day to day hearing.
 - o Hearing impairment becomes clinically apparent to the patient when the frequencies of 500, 1000 and 2000 Hz (the speech frequencies) are affected.

- Audiogram in NIHL:

- Symmetrical bilateral SNHL.
- Typical notch, at 4 kHz, both for air and bone conduction.
- As the duration of noise exposure increases, the notch deepens and also widens to involve lower and higher frequencies.
- After about 10 years, loss in the high frequencies tends to plateau, but the loss continues to broaden gradually into lower frequencies.
- Notch will disappear as aging changes.

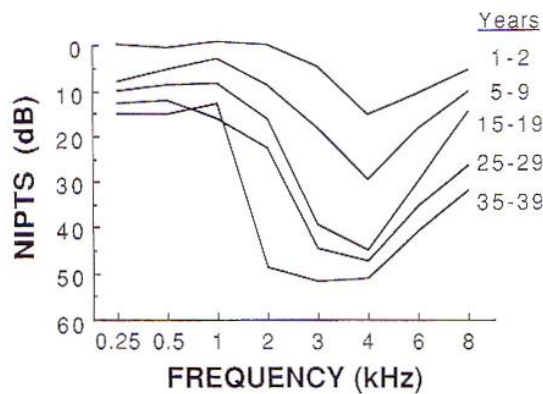


Figure 147.2 Median noise-induced permanent threshold shift as a function of audiometric frequency for different durations of exposure in jute-weaving mills (>100 dBA).

- Factors causing Noise trauma:

- Higher frequency (2000-3000 Hz)
- Higher intensities (dB).
- Continuous.
- Longer duration
- Susceptibility of the individual.
- Pre-existing ear disease.

- Management:

- **Preventable.**
- Persons who have to work at places where noise is above 85 dB (A) should have pre-employment and then annual audiograms for early detection.
- Ear protectors when noise levels exceed 85 dB (Provide protection up to 35 dB).
- Hearing aids.

- **Sudden Hearing Loss:**
- Definition:
 - SNHL of >30 dB in 3 consecutive frequencies within 3 days.
- Median age 40-54 years.
- Mostly unilateral.
- Bilateral in 1-5%
- Acute tinnitus accompanies the hearing loss in most cases.
- Vestibular symptoms are present in 25-50% of patients.

- **Etiology:**
- Divided into categories of
 - **Idiopathic sudden SNHL (90%):**
 - Theories includes:
 - Viral
 - Vascular
 - Intracochlear membrane rupture
 - Autoimmune.
 - **Defined causes that must be exclude (10%):**
 - Infections:
 - Mumps, herpes zoster, meningitis, encephalitis, syphilis, otitis media.
 - Trauma:
 - Head injury, ear operations, noise trauma, barotrauma, spontaneous rupture of cochlear membranes.
 - Large vestibular aqueduct syndrome is associated with SNHL, after minor head trauma.
 - Vascular:
 - Hemorrhage, embolism or thrombosis of labyrinthine or cochlear artery or their vasospasm.
 - They may be associated with diabetes, hypertension, polycythaemia, macroglobinaemia or sickle cell trait.
 - Otologic:
 - Meniere's disease, Cogan's syndrome, Perilymphatic Fistula.
 - Toxic:
 - Ototoxic drugs.
 - Neoplastic:
 - Acoustic neuroma.
 - Responsiveness to steroids is not a reliable indicator that a retrocochlear lesion is not present.
 - Neurologic:
 - Multiple sclerosis
 - Psychogenic.

TABLE 160.1	DIFFERENTIAL DIAGNOSIS IN SUDDEN SENSORINEURAL HEARING LOSS
Category	Etiology
Infection	Viral cochleitis, labyrinthitis, or cranial nerve
	VIII neuropathy
	Herpes simplex
	Mumps
	Varicella zoster (Herpes zoster oticus)
	Rubella
	Influenza
	Epstein-Barr
	Enterovirus
	Cytomegalovirus
	HIV
	Meningitis
	Syphilis
Inflammation	Lyme disease
	Autoimmune
	Cogan syndrome
	Behcet syndrome
Trauma	Systemic lupus erythematosus
	Multiple sclerosis
	Temporal bone fracture
	Acoustic trauma
	Barotrauma
Neoplasm	Perilymph fistula
	Vestibular schwannoma
	Meningioma
	Temporal bone metastasis
Toxins	Carcinomatous meningitis
	Aminoglycosides
	Aspirin
	Cisplatin
	Radiation therapy
Cardiovascular	Thromboembolism
	Macroglobulinemia
	Sickle-cell disease
	S/P coronary bypass graft surgery
	Severe acute hypotension
Otologic	Endolymphatic hydrops
	Meniere disease
Genetic	GJB2 mutation (DNFB1; encoding connexin 26)
	Enlarged vestibular aqueduct in Pendred syndrome
	Branchio-otorenal syndrome
	Renal tubular acidosis
	Fabry disease

- **Evaluation:**
 - Must be done early; faster treatment improves prognosis.
 - Detailed history, physical examination to reach a cause.
 - Initial investigations should include:
 - Baseline hearing assessment
 - MRI or ABR to rule out CPA masses (even if hearing improved).
 - Routine laboratory tests and CT are not recommended in the initial evaluation of patients with ISSNHL.
- **Treatment:**
 - **Oral steroids:**
 - Mainstay treatment for sudden SNHL.
 - Prednisone 1 mg/kg/d (maximal dose is 60 mg/d).
 - Full dose for 7-14 days, then taper over similar time period.
 - **Intra-tympanic steroids:**
 - Indications:
 - Contraindications to oral steroids.
 - Salvage after failed oral steroids.

Table 9. General Guidelines for Corticosteroid Therapy for Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)^a

	Oral Corticosteroids	Intratympanic Corticosteroids
Timing of treatment	Immediate, ideally within first 14 days. Benefit has been reported up to 6 weeks following onset of sudden sensorineural hearing loss (SSNHL)	Immediate Salvage (rescue) after systemic treatment fails
Dose	Prednisone 1 mg/kg/d (usual maximal dose is 60 mg/d) or Methylprednisolone 48 mg/d or Dexamethasone 10 mg/d	Dexamethasone 24 mg/mL or 16 mg/mL (compounded), or 10 mg/mL (stock) Methylprednisolone 40 mg/mL or 30 mg/mL
Duration/frequency	Full dose for 7 to 14 days, then taper over similar time period	Inject 0.4 to 0.8 mL into middle ear space every 3 to 7 days for a total of 3 to 4 sessions
Technique	Do not divide doses	Anterosuperior myringotomy after topical anesthetic Inject solution into the posterior inferior quadrant via narrow-gauge spinal needle to fill middle ear space Keep head in otologic position (one side down, affected ear up) for 15 to 30 minutes
Monitoring	Audiogram at completion of treatment course and at delayed intervals	Audiogram before each subsequent injection, at completion of treatment course, and at delayed intervals Inspect tympanic membrane (TM) to ensure healing at completion of treatment course and at a delayed interval
Modifications	Medically treat significant adverse drug reactions, such as insomnia Monitor for hyperglycemia, hypertension in susceptible patients	May insert pressure-equalizing tube if planning multiple injections, but this increases risk of TM perforation May consider adding round window transport facilitator

^aThis table is designed to provide guidance for systemic and intratympanic steroid treatment for SSNHL. Treatment is routinely individualized by provider and per patient. The most important principles pertain to early institution of high enough dosages of treatment. Prednisone 1 mg/kg/d or its equivalent and/or adequate concentration of intratympanic dexamethasone or solumedrol should be administered.

- **Hyperbaric oxygen:**
 - Younger patients respond better to hyperbaric oxygen therapy (HBOT) than older patients (50-60 years).
 - Early HBOT from 2 weeks to 3 months is better than late HBOT.
 - Patients with moderate to severe hearing loss benefit more from HBOT than those with mild hearing loss.
- **Other treatments:**
 - **Should not routinely prescribed.**
 - Includes:
 - Antivirals
 - Thrombolytics
 - Vasodilators
 - Vasoactive substances
 - Antioxidants
- Monitoring and prognosis:
 - Regular follow-up audiometric evaluation should be done within 6 months of diagnosis.
 - 50% showed recovery within a 10-day course of steroid therapy.
 - Final hearing levels is reached by 1 month in 80% of patients and by 3 months in 97% of patients.
 - 30% of patients return to normal hearing.
 - 30% of patients end up with profound hearing loss.
 - Recovery after treatment:
 - **Complete:**
 - PTA within **10 dB** of initial hearing level or within 10 dB of the hearing level of the unaffected ear.
 - **Partial:**
 - PTA within **50%** of initial hearing level or > 10 dB improvement of hearing level.
 - **No recovery:**
 - < 10 dB improvement in hearing level relative to the initial hearing level.
 - Factors for good prognosis:
 - Younger patients below 40.
 - Minimal hearing loss.
 - Low-frequency loss.
 - No change in ECoG N1 latency.
 - Factors for poor prognosis:
 - Advanced age
 - Total deafness
 - High-frequency loss (down-sloping audiogram)
 - Vertigo
 - Delay in treatment initiation.
 - Associated vascular risk factors

- **Autoimmune Hearing Loss:**
- Most common between ages 20–50 years old.
- Associated with systemic immune diseases (rheumatoid arthritis, lupus erythematosus, etc), allergy, and vasculitides.
- Pathophysiology:
 - may arise from host's defense from infection causing autoimmunity.
 - Cross reactivity from distant antigens
 - Circulating immune complexes affecting circulation in the stria vascularis
- Symptoms:
 - Rapidly progressive (weeks to months) or fluctuating SNHL.
 - Bilateral.
 - Normal otoscopic exam
 - Tinnitus
 - Vestibular symptoms in 50%
- **Serological Evaluation for Autoimmunity in Hearing Loss::**
 - CBC,
 - ESR
 - CRP
 - ANA
 - RF
 - ANCA
 - C1q binding test
 - Raji cell assay (for circulating immune complexes).
 - Cryoglobulins,
 - Complement profiles,
 - Lymphocyte transformation test (exposes patients lymphocytes to known inner ear antigens to evaluate for reactivity)
 - Western blot (looking for antibodies against 68 kD inner ear proteins antigen).
 - Lymphocyte migration inhibition test (targets type II collagen)
- **Management:**
 - High-dose oral corticosteroid trial for at least 30 days with audiogram follow-up to assess response.
 - If an immunological diagnosis is highly suspected (positive serology), Immunosuppressive medications (cyclophosphamide, methotrexate) for nonresponsive cases.
 - Consider adjunctive plasmapheresis.

- **Presbycusis:**
- Progressive SNHL associated with physiological aging process in the ear.
- Most common cause of adult hearing loss.
- Manifests at the age of 65 years.
- 50% of people > 75 years old have hearing impairment.
- Manifests early if there is:
 - Hereditary predisposition
 - Chronic noise exposure
 - Generalised vascular disease.
- **Age-Related Changes in the Auditory Systems:**
 - Cerumen: desquamated epithelium + sebum (from sebaceous glands) + watery secretions (from modified apocrine sweat glands)
 - Cerumen becomes harder, drier, less likely to be cleared due to atrophy of modified apocrine sweat glands with aging
 - Tragi hairs are coarser, larger, more prominent; leads to trapping of cerumen
 - Arthritic changes in the joints of ossicles do not result in hearing loss
- **Pathophysiology: (PASHA)**
 1. Mainly, Loss of sensory hair cells due to age-related degeneration, cumulative noise-induced trauma or cumulative ototoxin-induced injury over life.
 2. Vascular disorder causing atrophy to stria vascularis, loss of ganglion cells or degeneration of nerve fibers.
 3. Stiffing of basilar membrane.
- **Four pathological types: (DHINGRA)**
 1. **Sensory:**
 - **Degeneration of the organ of corti.**
 - Starting at the basal coil and progressing gradually to the apex.
 - Higher frequencies are affected.
 - Speech discrimination remains good.
 2. **Neural:**
 - **Degeneration of the cells of spiral ganglion.**
 - Starting at the basal coil and progressing to the apex.
 - Neurons of higher auditory pathways may also be affected.
 - Higher frequencies are affected.
 - Speech discrimination is poor and out of proportion to the pure tone loss.

3. Strial or metabolic:

- **Atrophy of stria vascularis** in all turns of cochlea..
- Physical and chemical processes of energy production are affected.
- Runs in families.
- Audiogram is flat.
- Speech discrimination is good.

4. Cochlear conductive

- **Stiffening of the basilar membrane** thus affecting its movements.
- Audiogram is sloping type.

- **Symptoms:**

- Progressive symmetric SNHL.
- Initially in High frequency (>2000 Hz)
- Downsloping curve in PTA.
- Hearing difficulty in noisy background.
- Hear well in quiet surroundings.
- Sometimes poor speech discrimination.
- Tinnitus
- Recruitment phenomenon is positive and all the sounds suddenly become intolerable when volume is raised.

- **Management:**

- Hearing aid.
- Cochlear implant if severe-profound.
- Lessons in speech reading through visual cues.
- Curtailment of smoking and stimulants like tea and coffee may help to decrease tinnitus.

Congenital Hearing Loss:

- Congenital Hearing loss that is present AT birth.
- Types:
 - **Non-Genetic HL (50%):**
 - Not caused by genetic mutations.
 - Types:
 - Inner ear malformations.
 - Infections.
 - Trauma.
 - Ototoxicity.
 - Hyperbilirubinemia
 - **Genetic HL (50%):**
 - HL presents At or After birth.
 - Caused by genetic mutations.
 - Classification:
 - **Non-Syndromic (2/3).**
 - Syndromic (**1/3**).
 - Mode of inheritance:
 - **Autosomal Recessive (80%).**
 - Autosomal Dominant (**18%**).
 - X-linked or Mitochondrial (**2%**).
 - Classic Audiogram pattern is "Cookie Bite" (U-shape).

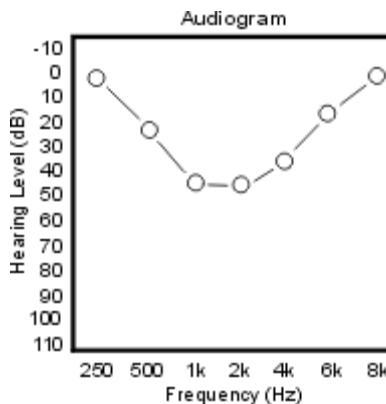


Fig. 9. Cookie-bite loss

Congenital Non-genetic Hearing Loss:

- Congenital hearing loss not caused by genetic mutations.
- Pre-Natal causes:
 - **Infections:**
 - SNHL may be present at birth or later during childhood.
 - Intrauterine infections (TORCHES) are the most common pre-natal cause of congenital hearing loss.
 - **TO**xoplasmosis, **R**ubella, **C**ytomegalovirus, **H**erpes, **S**yphilis.
 - CMV is the most common one.
 - Examples:
 - **Congenital Toxoplasmosis:**
 - Maternal Toxoplasmosis gondii infection crosses the placenta.
 - Clinical picture:
 - Congenital SNHL
 - Chorioretinitis
 - Hydrocephalus
 - Intracranial calcifications.
 - **Congenital Rubella:**
 - Rare since vaccine and prenatal testing.
 - Clinical picture:
 - Congenital SNHL
 - Cardiac malformation
 - Congenital cataracts
 - **Congenital Cytomegalovirus:**
 - Spread from maternal primary CMV.
 - Most common congenital viral infection.
 - Most common viral cause of congenital SNHL.
 - Clinical picture:
 - Congenital SNHL (**UNILATERAL**)
 - Mental retardation
 - Intracranial calcifications
 - Microcephaly
 - Retinitis
 - Hepatosplenomegaly
 - **Congenital Herpes:**
 - HSV-1 and HSV-2.
 - Mainly peripartum transmission.
 - Rare cause of SNHL.

- **Congenital Syphilis:**
 - Maternal Treponema pallidum infection that crosses the placenta.
 - Clinical picture:
 - Congenital SNHL
 - Hennebert's sign
 - Interstitial keratitis
- Peri-Natal causes:
 - Anoxia
 - Prematurity and low birth weight (<1.5 g).
 - Birth trauma
 - Neonatal jaundice (Hyperbilirubinemia):
 - Damages the cochlear nuclei.
 - ECMO
 - Ototoxic drugs
- Post-Natal causes:
 - **Infections:**
 - Meningitis is the most common post-natal cause of congenital hearing loss.
 - Examples:
 - **Bacterial Meningitis:**
 - 30% incidence of hearing loss.
 - Most commonly after pneumococcal meningitis.
 - Pathophysiology:
 - Most commonly secondary to penetration of bacteria and bacterial toxins via cochlear aqueduct or IAC contents resulting neuritis of CN-VIII and/or suppurative labyrinthitis.
 - Acute otitis media with direct spread through labyrinthine may result in labyrinthitis.
 - Dexamethasone was associated with reduction in severe neurologic sequelae and hearing loss.
 - If a child with meningitis has normal audiometric studies after first few days of antibiotic therapy, it is unlikely that significant SNHL will develop later.
 - Some children with initially abnormal hearing may improve, suggesting a resolving serous labyrinthitis.
 - Late progression of post-meningitic SNHL after years of stability has been reported.

- **Mumps:**
 - Paramyxovirus results in acute viral illness. Spread from maternal primary CMV.
 - Clinical picture:
 - Unilateral or bilateral parotitis.
 - Congenital SNHL (**UNILATERAL**).
 - Aseptic meningitis
 - Encephalitis
 - Orchitis
 - Mastitis.

Congenital Genetic Hearing Loss:

- Congenital hearing loss caused by genetic mutations.
- Types:
 - **Non-syndromic (2/3):**
 - More common than syndromic.
 - Congenital hearing loss with no other physical or systemic findings.
 - Most commonly due to **Connexin Mutations**.
 - **Syndromic (1/3):**
 - Congenital hearing loss associated with other physical or systemic findings.
 - Most common inheritance pattern is **Autosomal Recessive**.
 - Examples:
 - **Usher Syndrome:**
 - Most common cause of Autosomal Recessive syndromic HL.
 - Most common cause of deaf-blindness.
 - Clinical picture:
 - Congenital SNHL
 - **Retinitis pigmentosa**
 - Mental retardation
 - Delayed walking
 - **Goldenhar Syndrome (Hemifacial Microsomia/ Oculo-Auriculo-Vertebral syndrome):**
 - Autosomal Recessive syndrome.
 - Typically unilateral and on the right side.
 - Clinical picture:
 - **Hemifacial Microsomia**.
 - **Ocular abnormalities**.
 - **Auricular abnormalities** (SNHL, microtia/atresia, Facial nerve paralysis).
 - **Vertebral abnormalities** (fusion or absence of cervical vertebrae).



- **Pendred Syndrome:**
 - Autosomal Recessive syndrome.
 - Associated with **Mondini deformity** and **enlarged vestibular aqueduct**.
 - Clinical picture:
 - SNHL.
 - **Euthyroid multinodular goiter**
- **Treacher Collins Syndrome (Mandibulofacial Dysostosis):**
 - Autosomal Dominant syndrome.
 - Malformation of the 1st and 2nd branchial arches.
 - Clinical picture:
 - Typically normal intelligence.
 - Auricular deformities (microtia/atresia).
 - Retro-gnathia
 - Macrostomia.
 - Lower eyelid coloboma.



- **Alport Syndrome:**
 - X-linked syndrome.
 - Mutation in type IV collagen gene.
 - Clinical picture:
 - SNHL.
 - Renal dysplasia/agenesis.
 - Progressive nephritis.

Table 19-1. Syndromes associated with hearing loss

Syndrome	Features	Type of hearing loss	Onset (Congenital/Delayed)	Type of inheritance
1. Waardenburg's syndrome (Fig. 19.1)	• White forelock • Heterochromia iridis • Vitiligo • Dystopia canthorum	Unilateral or bilateral SNHL	Congenital	AD
2. Usher syndrome	• Retinitis pigmentosa • Night-blindness	SNHL	Delayed	AR
3. Jervell and Lange-Nielsen's syndrome	• Repeated syncopal attacks • Prolonged QT interval in ECG	SNHL	Congenital	AR
4. Pendred syndrome	• Goitre (non-toxic) usually evident before puberty • Perchlorate discharge test shows defect in organic binding of iodine	SNHL	Congenital	AR
5. Alport syndrome	• Hereditary progressive glomerulonephritis • Corneal dystrophy	Progressive SNHL	Delayed	AD or X-linked
6. Treacher-Collins syndrome (mandibulofacial dysostosis)	• Antimongoloid palpebral fissures • Coloboma of lower lid • Hypoplasia of mandible and malar bones • Malformed pinna and meatal atresia • Malformed malleus and incus (stapes normal)	Conductive	Congenital	AD
7. Crouzon's syndrome (craniofacial dysostosis)	• Frog eyes (exophthalmos with divergent squint) • Hypertelorism • Parrot-beak nose • Mandibular prognathism • Premature closure of cranial sutures with mental retardation	Conductive or mixed	Congenital	AD
8. Apert's syndrome	• Syndactyly • All other features of Crouzon's syndrome	Conductive (Stapes fixation)		AD
9. Klippel-Feil syndrome	• Short neck • Fused cervical vertebrae • Spina bifida • Atresia of ear canal	SNHL or mixed	Congenital	AR
10. Wildervanch syndrome	• Klippel-Feil syndrome • SNHL • CN VI Paralysis	SNHL	Congenital	X-linked
11. Branchio-oto-renal syndrome	• Branchial fistulas/cysts • Malformed pinnae with preauricular pits or sinuses • Renal abnormalities	Conductive or mixed	Congenital	AD
12. Stickler's syndrome	• Small jaw • Cleft palate (Pierre-Robin sequence) • Myopia $\geq$ retinal detachment • Cataract • Juvenile onset arthritis	Conductive or SNHL	Delayed	AD
13. Van der Hoeve's syndrome	• Osteogenesis imperfecta with history of fractures • Blue sclera • Hearing loss (delayed onset)	Conductive, SNHL or mixed (like otosclerosis)	Delayed	AD
14. Pierre-Robin sequence*	• Micrognathia • Glossoptosis • Cleft palate • Often part of Stickler's syndrome	SNHL Conductive loss		AD
15. Goldenhar's syndrome (Facio-auriculo-vertebral dysplasia) or (oculo-auriculo-vertebral [OAV] syndrome)	• Facial asymmetry • Low set ears, atresia of ear canal • Cardiac abnormalities • Preauricular tags/pits • Hemivertebrae in cervical region • Epibulbar dermoid • Coloboma of upper lid	Mixed or Conductive	Congenital	AD or sporadic Extra chromosome Multifactorial (genetic and environmental)
16. Down's syndrome (Trisomy 21)	• Microcephaly • Mental retardation/delayed development • Short stature • Epicanthal folds • Stenosis of ear canal • High incidence of serous otitis media • Atlanto-axial instability	Conductive	Congenital	Extra chromosome

- **Evaluation of HL in Pediatrics:**
- **History:**
- Character of Hearing Loss:
 - o Age of onset
 - o Progression of hearing loss
 - o Communication skills
- Prenatal and Perinatal History:
 - o Term of delivery
 - o Birth weight
 - o Prenatal infections
 - o Bilirubinemia
 - o Apgar score
 - o Maternal drug and Alcohol abuse
 - o Complications (NICU, ECMO)
- Contributing Factors:
 - o Syndromic features
 - o Family history of hearing loss
 - o History of neurologic disease (seizures).
 - o History of cardiac disease (Jervell and Lange-Nielsen).
 - o History of thyroid disease (Pendred).
 - o History of renal disease (Alport's).
 - o History of Sickle cell anemia
 - o History of Infections (Recurrent OM, Meningitis),
 - o History of other congenital disease.
 - o Delayed development (Growth history).
 - o Surgical history (otologic, neurologic).
 - o Medications (ototoxic).
 - o Recent trauma.
- Associated Symptoms:
 - o Delayed speech development.
 - o Imbalance or Gait disturbances
 - o Vision problems
 - o Other neurologic complaints
- **Physical Exam:**
- Inspection and Palpation:
 - o Auricle
 - o Periauricular pits.
 - o Facial malformations.
- Otoscopy/Microscopy:
 - o EAC, TM, Pneumatostcopy.
- Look for Any syndromic features.

Neonatal Hearing Loss and Speech Development:

- Normal hearing threshold in children is from 0-15 dB.
- Children with profound HL (>90 dB HL) fail to develop speech and often been termed as deaf-mute.
- They have never heard speech and therefore do not develop it.
- In mild to moderate degrees of HL, speech does develop but is defective.
- The period from **birth to 5 years of life** is critical for the development of speech and language:
 - o Needs early identification and assessment of hearing loss and early rehabilitation in infants and children.
 - o Children whose hearing loss was observed and managed before **6 months of age** had higher scores of vocabulary, better expressive and comprehensive language skills than those diagnosed and managed after 6 months of age.
- **Suspicion of hearing loss in Children:**
 - o Child sleeps through loud noises unperturbed.
 - o Child fails to startle to loud sound.
 - o Failure to develop speech at 1-2 years.
 - o Examine the child every 6 months until the age of 3 years "critical age for speech/language development" (even if birth screening is normal).
- **Risk Factors for hearing loss in children:**
 1. Family history of hearing loss.
 2. Prenatal intra-uterine infections (TORCHES).
 3. Craniofacial Anomalies including those of Pinna and EAC.
 4. Birth weight < 1.5 g (3.3 lbs).
 5. Hyperbilirubinaemia requiring exchange transfusion.
 6. Ototoxic medications.
 7. Bacterial meningitis.
 8. Apgar score of < 4 at 1 minute < 6 at 5 minutes (Asphyxia).
 9. Prolonged stay at NICU or ECMO.
 10. Syndromic child.
 11. Head trauma.
 12. Chemotherapy.
- 50% of infants with marked hearing loss do not have any risk factors.

Neonatal Hearing Screening:

- Should be done for ALL newborns (preferably) or newborns with high risk factors for hearing loss.
- Joint Committee on Infant Hearing (JCIH) recommends:
 - o Universal hearing screening (**OAE**) by the age of **1 month**, preferably prior to discharge from the hospital.
 - o Diagnostic evaluation (**ABR/OAE/Tympanometry**) before the age of **3 months** for newborn who fail initial screening test.
 - o Intervention for by the age of **6 months**.

1. Otoacoustic Emissions (OAE):

- o Low cost.
- o Non-invasive.
- o Measures cochlear (OHC) Function.
- o Independent of neural and central auditory system.
- o Both DPOAE and TEOAE are used:
 - **Transient Evoked OAE (TEOAE):**
 - Presents in all normal ears.
 - Evoked by clicks or tone bursts.
 - Absent if hearing loss ≥ 30 db.
 - **Distortion Product OAE (DPOAE):**
 - Presents in all normal ears.
 - Evoked by simultaneous 2 pure tone frequencies (F1 and F2).
 - Evaluate higher frequencies than TEOAE.
 - Absent if hearing loss ≥ 50 db.
- o Drawbacks if done alone as screening test:
 - **False Positive results:**
 - Infant with normal cochlear function fails the screening test.
 - External and Middle ear disorders like EAC obstruction or OME.
 - **False Negative results:**
 - Infant with SNHL passes screening test.
 - Normal OHC function.
 - IHC disorders or Auditory Neuropathy.

2. Auditory Brainstem Response (ABR):

- o Objective means of assessing integrity of Peripheral and Central Auditory Systems.
- o Records activity of CN-VIII and central nervous system's response to an Auditory signal.
- o Used as hearing screening test for:
 1. High Risk neonates.
 2. Failed initial screening with OAE.

Congenital Disorders of External Ear:

○ Pre-auricular Skin Tag:

- Accessory auricular remnants.
- Consist of nonfunctional redundant tissue (skin and fat) found anterior to tragus.
- May contain small pieces of cartilage.
- It may be a duplication of one or several of the ectodermal hillocks of His.
- Treatment:
 - Surgical excision.
 - Care should be taken as facial nerve may be very superficial in young children.



○ Pre-auricular Pits and Sinuses:

- Faulty fusion between 1st and 2nd branchial arches tubercles.
- Depressions or sinuses lined with squamous or columnar epithelium.
- Commonly seen between tragus and crus of helix.
- Pathway:
 - Punctum anterior to Root of Helix.
 - Short tract and sinus which ends with an attachment to perichondrium of Root of Helix.
- Clinical Picture:
 - Asymptomatic.
 - Discharging a small amount of mucoid discharge.
 - Repeated infection.
- Management:
 - Complete surgical excision to reduce recurrence.
 - Punctum is excised by Elliptical incision.
 - Tract and sinus are traced to Perichondrium of Root of Helix.
 - Perichondrium is scored sharply and peeled off the cartilage.
 - Full thickness of cartilage is excised to assure complete removal in case of recurrence.
 - No risk for Facial nerve injury since it is deeper and inferior to area of dissection.

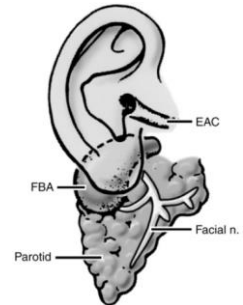


- **1st Branchial Cleft Cyst, Sinuses, and Fistula:**

- Result from anomalous duplication of EAC.
- May occur between EAC and structures anterior or inferior to auricle, including neck.
- Classification:

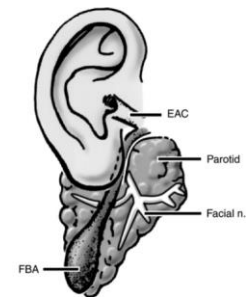
▪ **Work Type I:**

- Less common.
- Duplicated EAC.
- Contains ectodermal elements only.
- Begin periauricularly, pass lateral (superior) to Facial nerve, parallel to EAC, End as a blind sac near mesotympanum.



▪ **Work Type II:**

- Most common.
- Contains ectodermal and mesodermal elements.
- Presents near angle of mandible.
- Passes through Parotid, lateral or medial to Facial nerve, end near or into EAC.



○ Treatment:

▪ **Surgical Excision:**

- These anomalies may contain keratin debris that may become infected which require I & D, but this is not recommended if it can be avoided.
- The cyst, sinus, or fistula may be gently probed with a sterile, malleable lacrimal probe or another suitable instrument to map out its pathway.
- Opening may be gently filled with dye such as methylene blue prior to excision.
- A rim of tissue around the internal opening is resected with the specimen and every attempt is made to excise the tract in toto, leaving none of it behind.
- If the tract is suspected to course in any relation to the extratemporal facial nerve, the excision should be done in a more formal way.
- A modified Blair incision is made, and the parotid is exposed as is the facial nerve at least from the stylomastoid foramen to the pes anserinus and distal branching points, with the use of intraoperative facial nerve monitoring.
- The cyst, sinus, or fistula must be carefully dissected from the intact and stimulated facial nerve.

- **Protruding Ear (Bat Ear):**

- Auriculocephalic angle $>30^\circ$
- Poorly developed Antihelix and scapha.
- Large concha.
- Best age for reconstruction is at 5–6 years old when ear is at 85% of adult size and before the age of social stigmatization.



- Suture Technique:

- Simple technique.
- Mattress sutures placed along scapha through a posterior incision to create an antihelical fold with resulting reduction of auriculocephalic angle.
- Does not address conchal bowl.

- Cartilage Sculpting Technique:

- Reshaping (scoring, thinning) or splitting of auricular cartilage to weaken the cartilage surface to create a convexity for antihelical fold.

- Farrior Technique:

- Combination suture and cartilage sculpting techniques.
- Removes wedges of cartilage from posterior surface to allow bending to supplement the mattress sutures.

- Concha Setback Technique:

- Conchomastoid Suture Technique of Furnas.
- Sutures conchal bowl to mastoid periosteum to reduce the auriculocephalic angle.

- **Cup Ear Deformity:**

- Constriction of helix and scapha.
- Surgical correction requires unfurling of helical rim by dividing the helical-scaphoid cartilage to allow for expansion followed by redraping of skin over excess cartilage.





STAHL'S EAR

The Stahl's ear deformity (Spock Ear, Elf Ear) is characterized by a transverse crus extending outward from the antihelix rather than continuing upward in a gentle bend as the superior limb of the triangular fossa. Stahl's often presents with multiple or combinations of deformities.



HELICAL RIM

Helical rim irregularities or compression may occur anywhere along the entire circumference of the helical rim.



LIDDING

A folding over of the helical rim or upper third of the ear. It occurs when the superior limb of the antihelix or the fossa fails to properly form. (See superior limb of triangular fossa in normal ear.) Lop ear is the severe expression of lidding.



CUP EAR

An advanced form of the prominent ear with an incomplete opening of the ear. Often characterized by very stiff and resistant cartilage around the scapha and helical rim that can feel as though a string envelopes the helical rim.



PROMINENT EAR

Presents as an abnormally protruding ear (Bat Ear, Dumbo Ear). The over-projection is significant and typically exceeds 9mm from the mastoid. Often is under- or undiagnosed until too late for EarWell™ correction.



CONCHAL CRUS

The conchal crus is an abnormal fold of cartilage crossing the mid portion of the concha symba that appears to divide the ear in half. It often results in prominent ear.



CRYPTOTIA

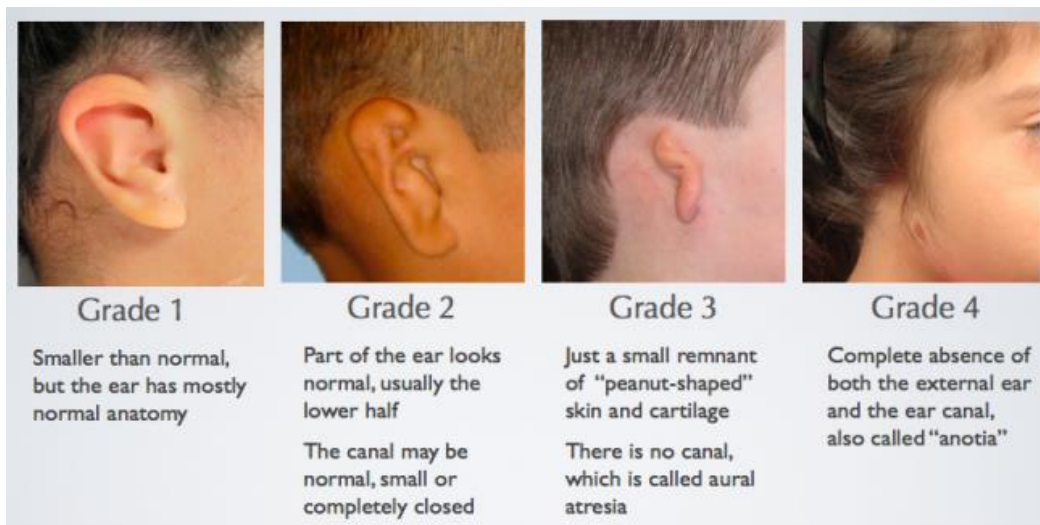
The ear cartilage framework appears buried beneath the skin with no apparent sulcus (where the ear meets the skull) or skin behind the ear.



MIXED

Many ears present multiple deformities in combination. (1) Stahl's, (2) Helical Rim, (3) Conchal Crus deformities.

- **Microtia:**
 - Major auricular malformation.
 - Often associated with EAC atresia.
 - Degree of auricular malformation correlates with degree of middle ear deformity.
 - Associated inner ear abnormalities are rare.
 - Vestibular dysplasia is the most common.
 - Usually unilateral with right side is the most common.
 - 25% of cases are bilateral.
 - M:F ratio of 2.5:1.
 - May have syndromic association (eg, hemifacial microsomia).
 - **Causes:**
 - Genetic
 - Teratogens (vitamin A/isotretinoin, thalidomide)
 - Vascular insult
 - **Classification:**
 - **Grade I:**
 - Small auricle with all subunits are present.
 - **Grade II:**
 - Small auricle with some subunits are severely underdeveloped or absent.
 - **Grade III:**
 - Small remnant of skin and cartilage (Peanut ear) associated with aural atresia.
 - **Grade IV (Anotia):**
 - Complete absence of auricle and lobule associated with aural atresia.
 - Usually forms part of 1st Arch Syndrome.

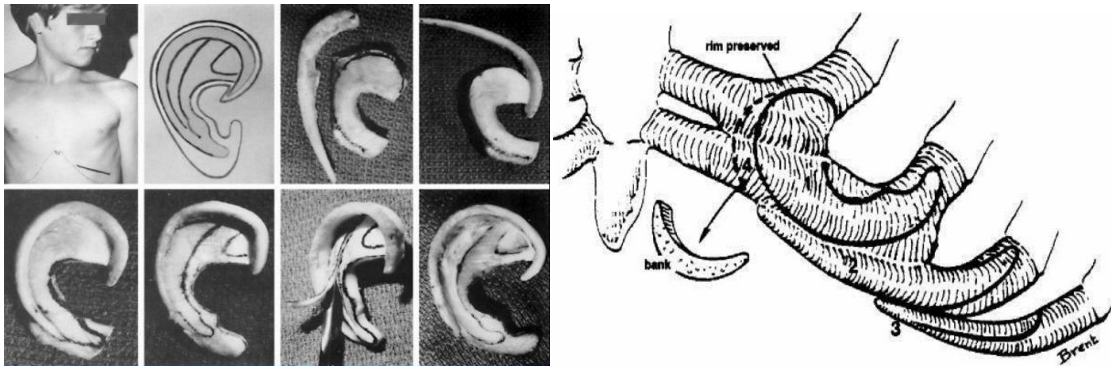


- Evaluation of patient with Microtia or Aural Atresia:
 - **H&P:**
 - Complete H&N examination looking for any other dysmorphic features.
 - Assess facial nerve function:
 - Most common anomaly of facial function is a congenital absence of depressor anguli oris muscle.
 - **Hearing Assessment:**
 - ABR:
 - Usually 40–60 dB CHL
 - 10–15% have SNHL
 - **CT Temporal bone:**
 - Evaluate presence of:
 - Middle and Inner ear anomalies.
 - Congenital cholestatoma.
 - No need to perform CT before age 4 years.
 - Should be performed near the time of operation and must be repeated if done early after birth before ultimate aural atresia repair.
 - Congenital cholesteatoma usually slow growing and not happen before this age.
- Treatment Options for Microtia:
 - 1. Surgical Repair:**
 - Typically at age of 6 years old.
 - Timing also influenced by coexisting atresia.
 - Types of Grafts:
 - **Costal Cartilage Autograft:**
 - Rib is popular cartilage source due to suitable integrity, sufficient quantity, and minimal morbidity with harvest.
 - **Alloplastic Implant:**
 - Medpor (porous polyethylene) implant.
 - Technically easier, does not tolerate trauma well, risk of extrusion.
 - Techniques of Surgical Repair:
 - Brent Cartilage Autograft Technique.
 - Nagata Cartilage Autograft Technique.
 - 2. Cosmetic Auricular Prosthesis:**
 - Prosthesis is anchored by magnets (previously clips) to titanium posts, which are connected to osseointegrated implants placed in the mastoid bone, often very realistic.

3. BAHA.

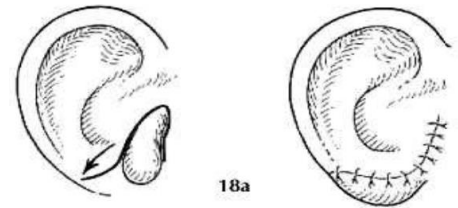
○ Brent Cartilage Autograft Technique:

- 4 stages separated by 3 months.
- Some stages may be combined.
- **1st Stage (Auricular Framework):**
 - Fabrication of the auricular framework from contralateral costal cartilage.
 - Placed in a postauricular subcutaneous pocket (thin overlying skin as much as possible).



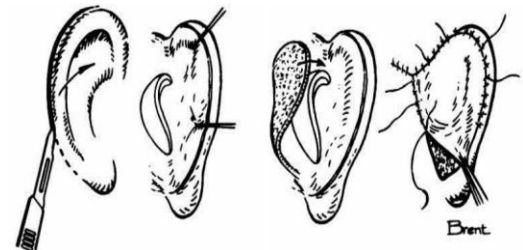
▪ **2nd Stage (Lobule Transposition):**

- Lobule is rotated and often filleted to receive the end of the cartilage framework



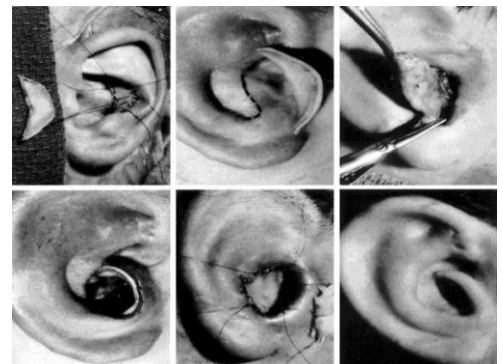
▪ **3rd Stage (Framework Elevation):**

- Framework is elevated to achieve projection of the helical rim.
- Retro-auricular scalp advancement flap and STSG used to close the postauricular defect.



▪ **4th Stage (Tragus Reconstruction):**

- Tragus construction, conchal excavation, symmetry adjustments.

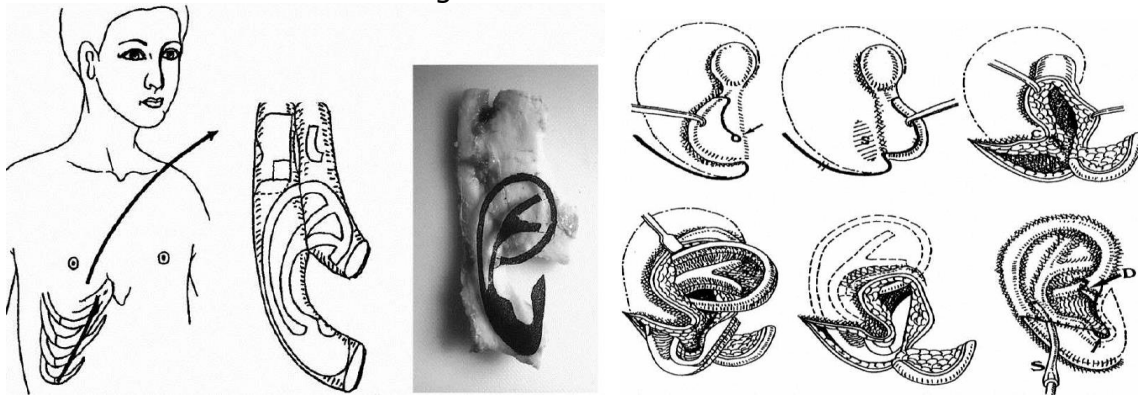


- Nagata Cartilage Autograft Technique:

- 2 stages separated by 6 months.

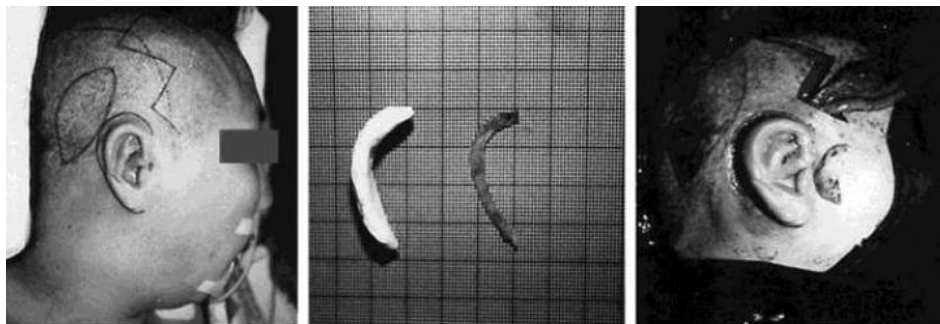
- **1st Stage:**

- Fabrication of the auricular framework from ipsilateral costal cartilage.
- Lobule Transposition.
- Tragus Reconstruction.



- **2nd Stage:**

- Framework is elevated using a crescent-shaped piece of cartilage to achieve projection of the helical rim.
- Temporoparietofascial flap is elevated and tunneled subcutaneously to cover the posterior surface of the cartilage graft, reconstructed auricle, and the mastoid surface.
- STSG may also be used.



- **Aural Atresia:**

- 90% nonsyndromic.
- 10% syndromic.
- 20–30% bilateral
- When it occurs alone, it is due to failure of canalisation of the ectodermal core that fills the dorsal part of first branchial cleft.
- The outer meatus, in these cases, is obliterated with fibrous tissue or bone while the deep meatus and the tympanic membrane are normal.
- Associations:
 - Microtia (55–93%)
 - Malformed SCCs (10%).
 - Malformed cochlea (5%).
 - Cholesteatoma (4–7%).
 - Stapes fixation (4%).
- Fusion of malleus/incus is most common middle ear anomaly.
 - Footplate is usually normal.
- Otitis media should be treated aggressively to preserve hearing in the normal contralateral ear.
- Evaluation of patient with Microtia or Aural Atresia:
 - **H&P:**
 - Complete H&N examination looking for any other dysmorphic features.
 - Asses facial nerve function:
 - Most common anomaly of facial function is a congenital absence of depressor anguli oris muscle.
 - **Hearing Assessment:**
 - ABR:
 - Usually 40–60 dB CHL
 - 10–15% have SNHL
 - **CT Temporal bone:**
 - Evaluate presence of:
 - Middle and Inner ear anomalies.
 - Congenital cholestatoma.
 - No need to perform CT before age 4 years.
 - Should be performed near the time of operation and must be repeated if done early after birth before ultimate aural atresia repair.
 - Congenital cholesteatoma usually slow growing and not happen before this age.



○ Treatment Options for Aural Atresia:

1. Conservative Management:

- No immediate medical intervention is necessary in infant discovered to have unilateral Atresia.
- Early amplification within 1st 3-4 months of life is essential in infants with bilateral atresia.
 - Soft band BAHA must be used.

2. Surgical Management:

- Otologic surgery is generally planned after the auricular reconstruction surgery if needed.
- Definitive treatment is controversial.
 - BAHA vs Surgical repair.
- Candidacy for Surgical Repair Grading Systems:
 - **Jahrsdoerfer Grading:**
 - 10-point system based on CT findings.
 - Better atresiaplasty candidate if ≥ 6 points.

**TABLE
148.1**

**JAHRSDOEFER GRADING SYSTEM
FOR CONGENITAL AURAL ATRESIA**

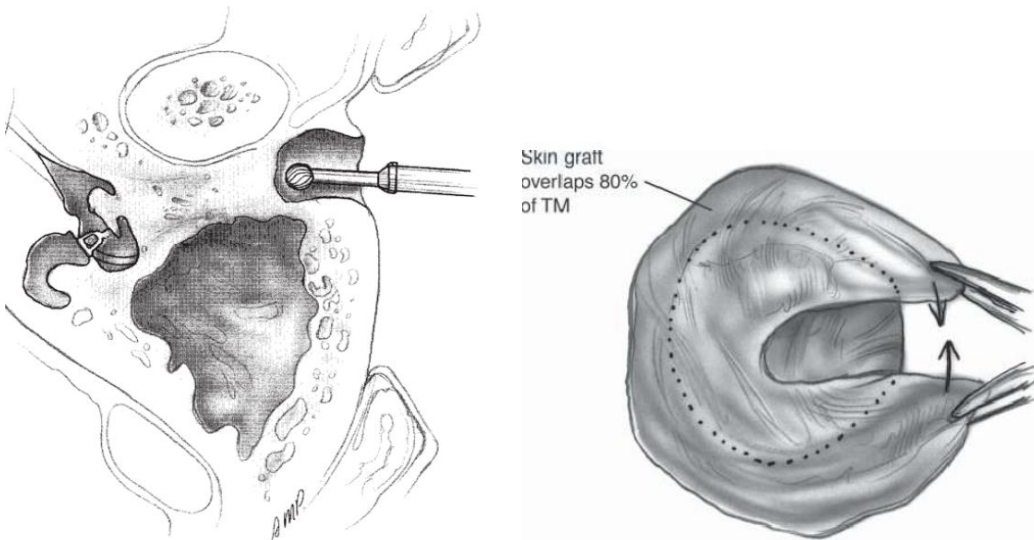
Parameter	Points*
Stapes present	2
Oval window patent	1
Middle ear space	1
Facial nerve	1
Malleus-incus complex	1
Mastoid pneumatization	1
Incus-stapes connection	1
Round window	1
External ear appearance	1

*Total points: 8, good prognosis; 7, fair prognosis; 6, marginal candidate; 5, poor prognosis.

○ **De la Cruz Classification:**

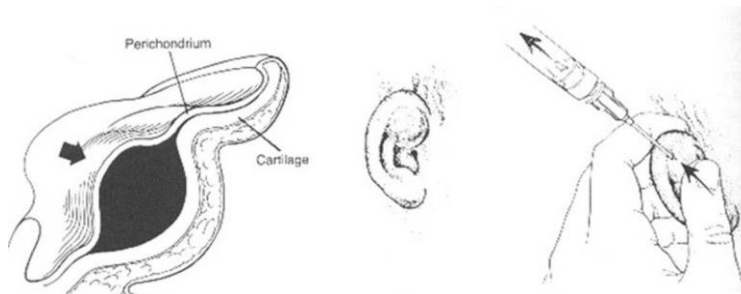
- Minor malformations (better surgical candidate):
 - Normal mastoid pneumatization.
 - Normal oval window/footplate.
 - Reasonable facial nerve-footplate relationship.
 - Normal inner ear.
- Major malformations (poor surgical candidate, HA is better option):
 - Poor pneumatization.
 - Abnormal or absent oval window/footplate.
 - Abnormal facial nerve course.
 - abnormalities of inner ear.

- Surgical approach:
 - Anterior approach with drilling over the atretic EAC.
 - Visualizing the Middle ear and assessing the mobility and integrity of the ossicles.
 - Tympanic membrane grafting with fascia graft.
 - Meatoplasty
 - STSG over the canal.



Disorders of External Ear:

- External ear can be afflicted by:
 1. Congenital disorders
 2. Traumatic disorders
 3. Inflammatory disorders
 4. Non-inflammatory disorders
 5. Neoplastic disorders
- **Traumatic disorders of External Ear:**
 - **Auricular Hematoma:**
 - Collection of blood between auricular cartilage and its perichondrium.
 - Etiology:
 - Blunt trauma as seen in boxers, wrestlers and rugby players.
 - Complications:
 - Perichondritis and Abscess formation:
 - Caused by bacterial superinfection.
 - Cartilage Necrosis and Cauliflower Ear:
 - Extravasated blood may clot and then organize and causing separation of cartilage from overlying perichondrium that supplies its nutrients resulting in permanent cartilage necrosis and formation of fibrous tissue in the overlying skin causing a typical deformity called Cauliflower Ear.
 - Management:
 - Aspiration of the hematoma under strict aseptic precautions and a pressure dressing with dental rolls (aspiration may need to be repeated).
 - If Aspiration is failed, Evacuation with (I&D) and compression dressing applied (in all concavities) to prevent reaccumulation, perichondritis, and cauliflower ear.
 - Oral antibiotics while compression dressing in place (1 week).



- **Auricular Abscess:**

- Collection of abscess between the auricular cartilage and its perichondrium.
- Etiology:
 - Complication of severe or inadequately treated external ear infections.
 - Accidental or surgical trauma to external ear.
 - Hematoma.
 - Cartilage piercing.
 - Exposed cartilage.
 - Infected endaural incision.
- Complications:
 - Cartilage Necrosis and Cauliflower Ear.
 - Cartilage survives only on the blood supply from its perichondrium.
- Management:
 - I&D of abscess under controlled conditions in OR.
 - Affected area is cleansed and injected with LA.
 - Incision is made in the natural folds.
 - Skin flaps are appropriately planned and dissection taken down to the affected cartilage.
 - Abscess should be evacuated and sent for C/S.
 - If the cartilage has lost its normal pearly white appearance, it is necrotic and should be excised.
 - Irrigation of the wound with antibiotic irrigation such as bacitracin (50,000 U of bacitracin dissolved in 250 mL of normal saline).
 - Small drain is placed beneath the flaps and sutured to the skin.
 - Compression dressing applied (in all concavities) to prevent re-accumulation.
 - Aggressive local care
 - IV Antibiotics

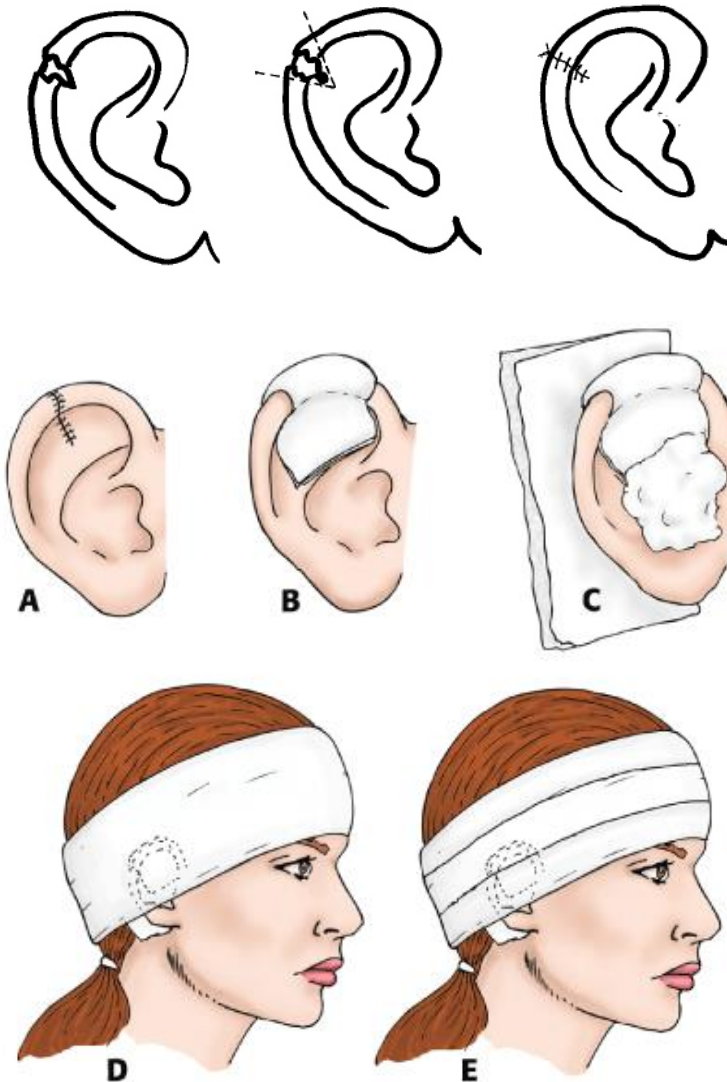


- **Auricular Laceration:**

- Repaired as early as possible.
- Perichondrium is stitched with absorbable sutures.
- Special care is taken to prevent stripping of perichondrium from cartilage for fear of avascular necrosis.
- Exposed cartilage must be either debrided or covered by skin to survive.
- Auricular skin often stretches to allow coverage of most defects.
- If the remaining skin cannot cover the cartilage, the cartilage should be cut away from the wound margin to allow overlying skin closure.



- In the case of a linear laceration to the pinna in which the skin does not approximate, a wedge excision of full thickness triangle from the antihelix can be used.
 - Up to 5 mm of cartilage can be removed without significant deformity.
 - A 1-mm overhang of the skin beyond the cartilage is recommended to allow skin eversion when closing.
- Skin is closed with fine non-absorbable sutures.
- Broad spectrum antibiotics are given for one week.



○ **Auricular Avulsion:**

- Auricle has a plentiful blood supply and will heal if the
- cartilaginous and soft tissue structures remain even partly attached.
- Partial avulsions usually survive with at least a 1–2 mm skin pedicle and primary reattachment should be considered and it is usually successful.
- Complete auricular avulsion requires attempted replantation (microvascular techniques), may require a meatoplasty to avoid external auditory canal stenosis.



○ **Frostbite:**

- Initially, the auricle become pale, hard, and cold due to extreme vasoconstriction.
- Followed by erythema and edema, bullae formation, necrosis of skin and subcutaneous tissue, and complete necrosis with loss of the affected part.
- Treatment:
 - Rewarming with moist cotton pledgets at a temperature of 38-42°C.
 - Application of 0.5% silver nitrate soaks for superficial infection.
 - Analgesics for pain as rapid rewarming of frost bitten ear causes considerable pain.
 - Protection of bullae from rupture.
 - Systemic antibiotics for deep infection.
 - Surgical debridement should wait several months as the true demarcation between the dead and living tissues appears quite late.

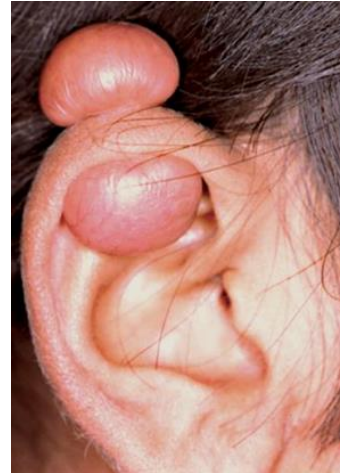


- **Keloid of Auricle:**

- Deposition of collagen that extends beyond the limits of the scar and does not regress over time.
- May follow trauma or piercing of the ear for ornaments.
- Usual sites are the lobule or helix.
- Recurs frequently following single modality treatment, marginal response to steroid injection when used as a single modality of treatment.

- Treatment:

- Gentle massage with intralesional corticosteroid injections (triamcinolone acetonide) with repeat injections every 3 weeks.
- If no response, may consider excision (with corticosteroid injections).
- Pressure delivery devices, silicone gel applications.
- Radiation therapy for severe cases may be considered but must weigh risk of malignant transformations.



- **Trauma to EAC:**

- Minor lacerations:

- Result from Q-tip injury (scratching the ear with hair pins, needles or match stick) or unskilled instrumentation by the physician.
- Usually heal without sequelae.

- Major lacerations:

- Result from gun shot wounds, automobile accidents or fights.
- Condyle of mandible may force through the anterior canal wall.
- Require careful treatment with the aim is to attain a skin-lined meatus of adequate diameter.
- Stenosis of the ear canal is a common complication.

- **Inflammatory disorders of External Ear:**
 - **Perichondritis and Chondritis:**
 - Perichondritis is inflammation of external ear perichondrium.
 - Chondritis is inflammation of external ear cartilage.
 - Etiology:
 - Complication of severe or inadequately treated external ear infections.
 - Accidental or surgical trauma to external ear.
 - Cartilage piercing.
 - Exposed cartilage.
 - Hematoma.
 - Infected endaural incision.
 - Pathogens:
 - Pseudomonas Aeruginosa (most common).
 - Staph. Aureus.
 - Streptococcus
 - Clinical Picture:
 - Painful, warm, erythematous and edematous auricle with involvement of EAC.
 - Involves all the auricle except lobule.
 - Does not contain cartilage.
 - Severe itching deep within the canal.
 - Skin becomes crusted with squamous debris.
 - Involvement of surrounding soft tissues of the face and neck in severe cases.
 - Complications:
 - Abscess formation between the cartilage and perichondrium with necrosis of cartilage and development of cauliflower ear as the cartilage survives only on the blood supply from its perichondrium.
 - Management:
 - **Mild cases:**
 - No abscess formation or involvement of surrounding soft tissues.
 - Thorough ear toilet and debridement.
 - Topical anti- Pseudomonas ear drops.
 - Oral anti- Pseudomonas antibiotics for 10 days.
 - **Severe cases:**
 - Abscess formation
 - Involvement of surrounding soft tissues.
 - Not responding to oral antibiotics.
 - Thorough ear toilet and debridement.
 - Culture and sensitivity.
 - I&D of Abscess if present in OR.
 - Topical anti- Pseudomonas ear drops.
 - IV anti- Pseudomonas antibiotics.



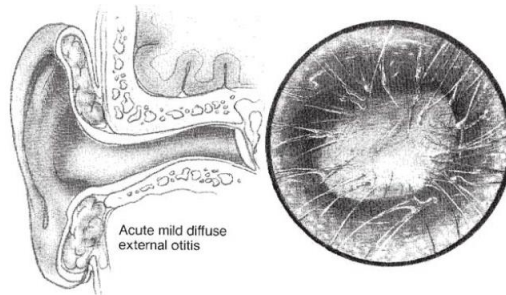
- **Relapsing Polychondritis (Perichondritis):**
 - Rare autoimmune disease of unknown etiology.
 - Episodic and progressive in nature.
 - Involves inflammatory destruction of elastic cartilages.
 - [Clinical picture:](#)
 - Auricular chondritis.
 - Entire auricle except its lobule becomes inflamed and tender.
 - External ear canal becomes stenotic.
 - Cochlear and vestibular injury (vertigo, hearing loss).
 - Respiratory chondritis (laryngeal collapse).
 - Nasal chondritis (saddle-nose deformity).
 - Polyarthritits (nonerosive, migratory).
 - Cardiac valve insufficiency.
 - [Diagnosis:](#)
 - History and physical examination.
 - Labs:
 - Elevated ESR.
 - Elevated IgG.
 - Elevated antibodies to type II and type IV collagen
 - Biopsy of involved cartilage:
 - Perichondrial inflammation with fibrosis.
 - [Management:](#)
 - NSAID.
 - Systemic steroids.



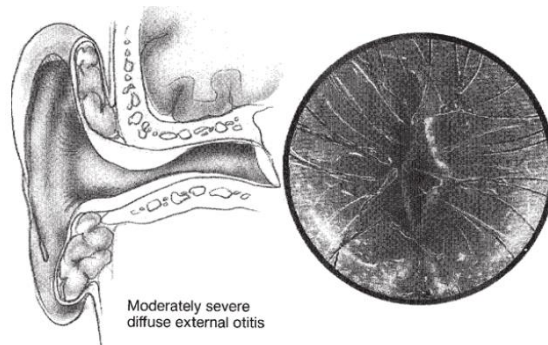
- **Otitis Externa:**
 - Spectrum of inflammatory conditions and infections of the EAC.
 - **Infective Otitis Externa:**
 - **Bacterial:**
 - Localized Otitis Externa (Furuncles).
 - Diffuse Otitis Externa
 - Malignant Otitis Externa
 - **Fungal:**
 - Otomycosis.
 - **Viral:**
 - Herpes Zoster Oticus.
 - Otitis Externa Hemorrhagica.
 - **Reactive Otitis Externa:**
 - Eczematous Otitis Externa.
 - Seborrhoeic Otitis Externa.
 - Neurodermatitis.
 - **Localised Acute Otitis Externa (Furuncle and Carbuncle):**
 - Staphylococcal infection of hair follicle inside cartilaginous EAC, most commonly at junction of concha and canal skin.
 - Hair are confined only to cartilaginous part of EAC.
 - **Clinical picture:**
 - Small well-circumscribed pustule that enlarge to become furuncle or merge with several similar lesions to form carbuncle.
 - Severe pain and tenderness out of proportion to the size of the furuncle.
 - Movements of the pinna are painful.
 - Jaw movements cause pain in the ear.
 - **In case of recurrent furunculosis:**
 - Diabetes should be excluded.
 - Attention paid to patient's nasal vestibules which may harbor staphylococci and then the infection transferred by patient's fingers.
 - **Treatment:**
 - In early cases (without abscess formation):
 - Consists of medicated ear wick, systemic antibiotics, analgesics and local heat.
 - If abscess has formed:
 - Incision and drainage should be done under LA in addition to medicated ear wick, systemic antibiotics, analgesics and local heat.

- **Acute Otitis Externa (AOE):**
- Diffuse bacterial infection of EAC skin caused by a break in normal skin/cerumen protective barrier in presence of elevated humidity and temperature (**swimmer's ear**).
- **Pathophysiology:**
 - Aggressive washing of cerumen or retention of water results in a more alkalotic EAC and decreased production of antibacterial agents (eg, lysozyme), which are permissive for bacterial overgrowth and penetration into the pilosebaceous unit.
 - Begins with itching which is commonly caused by instrumenting EAC with a cotton swab or fingernail.
 - Temporarily relieves itching but allows proliferation of bacteria in locally macerated skin.
 - Itch-scratch cycle.
 - Pain.
 - EAC Soft tissue swelling.
 - Purulent discharge.
 - Involvement of Auricle and periauricular soft tissues.
- **History:**
 - Major symptoms of AOE:
 - Pain
 - Fullness
 - Itching
 - CHL
 - Predisposing factors:
 - Auricular instrumentation or trauma
 - Swimmers.
 - Immunocompromised:
 - DM
 - HIV
 - Radiotherapy
- **Physical Examination:**
 - Edematous, erythematous and tender EAC
 - Purulent discharge.
 - Tragal tenderness confirms the clinical suspicion.
 - Periauricular erythema or cellulitis
 - TM perforation may suggest underlying CSOM.
- **Evaluate presence of signs and symptoms of Malignant Otitis Externa:**
 - Cranial nerve involvement.
 - EAC granulation tissue.

- Staging of Otitis Externa:
 - **Pre-inflammatory stage:**
 - Edema of stratum corneum due to removal of protective lipid layer and acid mantle from EAC.
 - Obstruction of apopilosebaceous unit.
 - Sense of fullness and itching begins.
 - Allows invasion of bacteria into the disrupted epithelial layer.
 - **Acute inflammatory stage:**
 - Mild stage:
 - Mild redness and edema of EAC skin.
 - Small amount of cloudy discharge.



- Moderate stage:
 - More pain and itching.
 - More redness and edema of EAC skin.
 - Thick and profuse exudate.



- Severe stage:
 - Sever pain.
 - Obliteration EAC lumen obscuring TM.
 - Profuse and purulent exudate.
 - Extension of involvement beyond EAC:
 - Periauricular cellulitis.
 - Lymph node enlargement.



- **Chronic inflammatory stage:**

- Secondary to persistent low-grade infection or inflammation
- Less pain but more profound itching.
- EAC skin of thick and hypertrophic causing stenosis and obstruction of EAC in severe cases.
- Auricle and concha often show secondary changes such as eczematization, lichenification, and superficial ulceration.



- Pathogens:

- **Pseudomonas Aeruginosa**

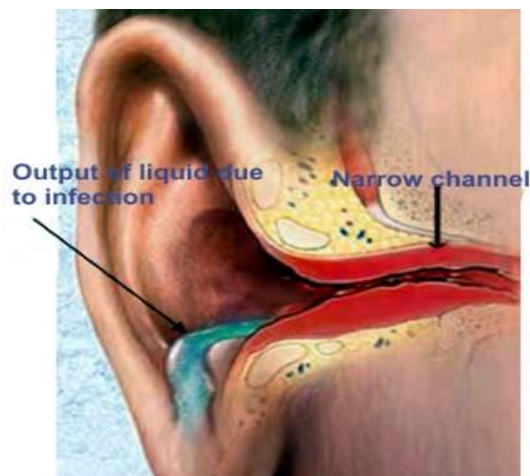
- Most common, opportunist infection.

- Staphylococcus

- Other gram-negative bacilli

- Differential diagnosis of Otitis Externa:

- Malignant otitis externa.
- Bullous and granular external otitis
- Perichondritis, chondritis and relapsing polychondritis
- Furunculosis and carbunculosis
- Psoriasis and seborrheic dermatitis
- Carcinoma may present as infection (SCC most common, can have BCC, melanoma, adenoma or adenoCa, adenoid cystic).
 - Presence of pain in an old previously stable mastoid cavity is the hallmark of carcinoma and must be excluded by biopsy and other investigations.



- Management of Otitis Externa:

▪ **Mild stage AOE:**

1. Frequent and thorough Ear toilet:

- Most important single factor in treatment.
- All exudate and debris should be meticulously and gently removed.
- Done by:
 - Dry mopping
 - Suction clearance
 - Irrigation with warm, sterile normal saline.

2. Antibiotics Drops:

- Anti-Pseudomonas ear drops for 7–10 days (better if combined with steroids to reduce the edema as anti-inflammatory).
- **Fluoroquinolone:**
 - Ofloxacin.
 - Ciprofloxacin.
 - Ciprodex (Ciprofloxacin/ Dexamethasone).
- **Aminoglycosides (Intact TM):**
 - Gentamicin.
 - Garasone (Gentamicin/ Betamethasone).
 - Otosporin (Neomycin/Polymyxin B/Hydrocortisone).
 - Oflox.

3. Oral Pain Analgesia

4. Precautions:

- Avoid EAC trauma or instrumentation.
- Maintain dry ear precautions.

▪ **Moderate stage AOE:**

1. Frequent and thorough Ear toilet:

2. Medicated Ear wick:

- Wick soaked in Antibiotic steroid preparation is inserted in EAC.
- Ichthammol glycerine can be mixed also into the wick which acts as anti-inflammatory to reduce pain and edema.
- Wick is kept moist by instilling the same drops twice or thrice a day.
- Changed daily for 2-3 days when it can be substituted by ear drops after resolution of EAC edema.

3. Oral Pain Analgesia

4. Precautions.

- **Severe stage AOE:**

- 1. Frequent and thorough Ear toilet**

- 2. Medicated Ear wick**

- 3. Oral Antibiotics:**

- Used if infection extended beyond EAC.
 - Continued for 10 to 14 days.
 - If no response, Admission with vigorous daily local care, repeat culturing, and intravenous antibiotics are indicated.
 - Anti-Pseudomonas antibiotics:
 - Ciprofloxacin or levofloxacin
 - Caution in children under 12 years old.
 - Antistaphylococcal penicillins, or cephalosporins.

- 4. Oral Pain Analgesia**

- 5. Precautions.**

- **Chronic Otitis Externa:**

- 1. Frequent and thorough Ear toilet**

- 2. Ear drops:**

- Antibiotic and steroid combination drops.
 - DermOtic (fluocinolone 0.01%).
 - Acidifying drop composed of equal measures of vinegar and water.

- 3. Precautions.**

- 4. Surgical :**

- Indicated only if local measures failed to eradicate the disease and re-establish EAC lumen.
 - Goal of surgery:
 - Excise involved canal skin
 - Perform wide meatoplasty
 - Resurface canal with split-thickness skin graft.

- [Refractory Otitis Externa:](#)
 - Take swab from ear discharge for C/S.
 - Look for the following:
 - Noncompliance or chronic instrumentation of EAC.
 - Otomycosis:
 - Prolonged antibiotic drops suppress EAC normal flora and lead to a fungal superinfection.
 - Suspected if:
 - Grayish matted discharge
 - Precinse of fungal hyphae.
 - Managment:
 - Ear toilet.
 - Canesten (Clotrimazole)
 - CSOM:
 - Presence of TM perforation with active discharge from middle ear.
 - Presence of cholestatoma.
 - Perichondritis.
 - Periauricular cellulitis.
 - Temporal bone CT scan may add additional information.
 - Some patients may need to be admitted for intravenous antibiotics and daily aural toilet.

**TABLE
146.4**



TREATMENT OTITIS EXTERNA

Medical

Frequent thorough cleaning
Antibiotic coverage
Drops
Oral
Intravenous as needed
Treat inflammation, pain
Recommendations for prevention

Surgical

Excise involved canal skin
Perform wide meatoplasty
Resurface canal with split-thickness skin graft

**TABLE
146.3**



**EMERGENCIES OTITIS
EXTERNA**

Unresolving pain, despite local care
Admit to hospital
Analgesia
Control inflammation
Cranial neuropathy with external otitis
Consider necrotizing external otitis

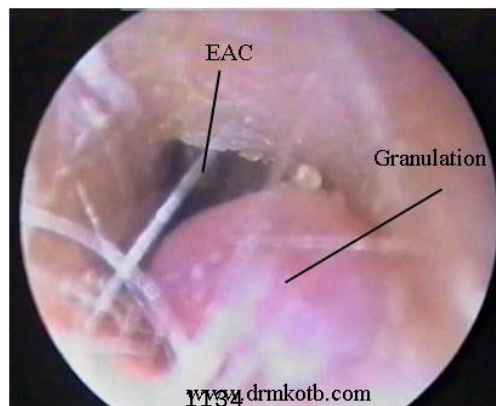
**TABLE
146.2**



**COMPLICATIONS OTITIS
EXTERNA**

Cellulitis
Erysipelas
Perichondritis
Chondritis
Chronic nonresolving rate

- **Skull Base Osteomyelitis (SBO) /Necrotizing (Malignant) Otitis Externa:**
 - Begins as AOE that does not resolve despite medical therapy.
 - Infection extends from EAC into temporal bone, skull base and jugular foramen resulting in severe progressive osteomyelitis with multiple cranial nerve palsies.
 - "Malignant" otitis externa is a misnomer.
 - Caused mainly by Pseudomonas infection.
 - Should be the main differential diagnosis of refractory Acute Otitis Externa in high risk (immunocompromised) patients .
 - Diabetics.
 - Elderly.
 - HIV.
 - Radiation exposure.
- **Diagnosis Criteria of SBO:**
 - **History:**
 1. Persistent deep-seated severe otalgia > 1 month.
 2. Persistent purulent otorrhea with granulation tissue for several weeks
 3. Immunocompromised state.
 4. Cranial nerve involvement
 - CN-VII > CN-X > CN-XI.
 - **Physical Exam:**
 1. Granulation tissue in inferior aspect of EAC at the bony-cartilaginous junction.
 2. Purulent discharge.
 3. Cranial nerve involvement
 - CN-VII > CN-X > CN-XI.
 - **Culture and Biopsy:**
 1. Almost always P. Aeruginosa.
 2. Granulations should be biopsied to rule out carcinoma or another pathologic entity.
 - **Imaging:**
 1. CT scan temporal bone with contrast.
 2. MRI with temporal bone with contrast.
 3. Bone scans:
 - Technetium: used for diagnosis.
 - Gallium: used for follow up.



**TABLE
146.5**



**DIAGNOSIS NECROTIZING
EXTERNAL OTITIS**

History

Persistent otalgia
Persistent purulent otorrhea, granulations
Diabetes mellitus, advanced age, immunocompromised state
Cranial neuropathy(ies)

Physical examination

Granulations in external canal
Purulent discharge seen
± cranial neuropathy, especially cranial nerve VII

Culture

Pseudomonas sp.^a
Pseudomonas aeruginosa^a

Radiology

Nuclear (gallium, technetium)
CT with contrast
MRI with contrast

CT, computed tomography; MRI, magnetic resonance imaging.

^aAlmost always.

- Imaging of SBO:

▪ **CT scan temporal bone with contrast:**

- Initial radiograph to be done.
- Excellent bony detail.
 - Define subtle bony changes such as:
 - Erosion of anterior canal wall with involvement of TMJ.
 - Erosion of tympanic ring and base of skull.
- Less precise information about soft tissue.
 - Demonstrate soft tissue thickening and mastoid clouding



- **MRI temporal bone with contrast:**

- Advantages:
 - Evaluation of medial extent of disease at the skull base.
 - Evaluation of dural enhancement and cerebral involvement.
- Disadvantages:
 - Imprecise information about bone.
 - Changes seen on MRI do not quickly resolve with clinical improvement.
 - MRI is a useful diagnostic tool to assess the extent of disease but less useful to follow the clinical course of SBO.

- **Bone Scan:**

- Base line Tc-99m and Ga-67 Bone Scan are used complementary to each other in evaluation of SBO.
- **Tc-99m Bone Scan:**
 - **Advantages:**
 - Used in initial diagnosis of SBO.
 - Excellent information about bone function.
 - Evaluates osteoblastic activity as little as 10% above normal.
 - Excellent for localization the extent of acute or chronic process
 - **Disadvantages:**
 - Not used in follow up.
 - Stays positive for long period (in acute and chronic osteomyelitis).
 - Positive results in areas of active bone repair without infection as in trauma.
- **Ga-67 Bone Scan:**
 - **Advantages:**
 - Used in follow up of SBO.
 - Gallium is taken up by PMNs and lights up when active inflammation is present.
 - As treatment progresses, Ga-67 scan will revert to normal (negative).
 - **Disadvantages:**
 - Does Not show the extent of osteomyelitis.

- **Indium-111 Bone scan (New):**
 - **Advantages:**
 - Better detection of osteomyelitis than Ga-67 and/or Tc-99m.
 - May replace the former two radionuclide modalities in the evaluation of SBO-suspected patients.
- **Differential diagnosis of SBO:**
 - Severe AOE
 - Squamous cell carcinoma
 - Glomus jugulare tumor
 - Cholesteatoma
 - Nasopharyngeal carcinoma
 - Hand-Schuller-Christian disease
 - Eosinophilic granuloma
 - Wegener granulomatosis
 - Clival chordoma
- **Complication of SBO:**
 - Cranial neuropathy
 - Sinus thrombosis
 - Sepsis
 - Meningitis
 - Intracranial infections
 - High mortality (particularly in immunocompromised).
- **Poor prognostic factors:**
 - Facial paralysis
 - Polyneuropathy
 - Intracranial extension

TABLE 146.7	 COMPLICATIONS NECROTIZING EXTERNAL OTITIS
Cranial neuropathy (VII and lower)	
Progression despite aggressive local care (to mastoid, parotid, lower cranial nerves, base of skull, dural venous sinuses, brain)	
Meningitis	
Brain abscess	
Death	

- [Management of SBO:](#)
 - **Medical Management (Mainstay Therapy):**
 1. Aggressive diabetic control.
 2. Local Ear Care:
 - Frequent and thorough Ear toilet.
 - Medicated Ear wick with Anti-Pseudomonas Antibiotics drops.
 - Water precautions.
 3. Prolonged IV Anti-Pseudomonas Antibiotics:
 - IV Ciprofloxacin.
 - Used for extended period (6 weeks).
 4. Hyperbaric Oxygen:
 - Facilitate osteoneogenesis and to promote repair of diseased bone.
 - Useful in the most severe cases due to its cost and inconvenience.
 - Indications:
 1. Advanced disease with significant skull base or intracranial involvement.
 2. Recurrent disease
 3. Infections refractory to antibiotic treatment.
 - **If the ear remains purulent despite adequate intravenous antibiotics and local care:**
 - Repeat cultures should be taken to look for the emergence of a resistant organism.
 - **Surgical Management:**
 - Controversial.
 - Surgical debridement of devitalized tissue and bone should be done judiciously.
 - Radical resections have been abandoned in favor of prolonged intensive medical therapy.
 - Indicated only for severe disease not responding to aggressive medical therapy.
 - Surgical management includes:
 - Excision of granulations
 - Middle ear exploration
 - Mastoidectomy
 - CN-VII decompression
 - Temporal bone resection

**TABLE
146.8**



EMERGENCIES NECROTIZING EXTERNAL OTITIS

Any complications in Table 146.7 (in living patient)

- Change antibiotic
- Increase level of daily care
- Consider surgery

**TABLE
146.6**



TREATMENT NECROTIZING EXTERNAL OTITIS

Medical

- Hospital admission
- Intravenous antibiotics
- Daily cleaning, debridement

Surgical

- Excise $\pm$ granulations
- $\pm$ middle ear exploration
- $\pm$ mastoidectomy
- $\pm$ facial nerve decompression
- $\pm$ temporal bone resection if no response



Axial CT



Coronal CT

CT petrous bone demonstrating mastoiditis and granulation tissue at the external auditory canal in cases of necrotizing otitis externa

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- **Otomycosis:**
- Fungal infection of EAC skin.
- Primary fungal infection occurs mainly in immunocompromised patients including diabetics.
- Secondary otomycosis occurs in patients with chronic bacterial infection in which prolonged antibiotic drops suppress EAC normal flora and lead to a fungal super-infection.
- Basic growth requirements for fungal infections:
 1. Moisture
 2. Warmth
 3. Darkness
- Most common fungal pathogens:
 - Aspergillus:
 - Most common pathogens.
 - Mainly Aspergillus Niger.
 - If aural culture should grow Aspergillus Fumigatus or Aspergillus Flavus, one would be concerned about a more invasive infection.
 - Candida Albicans:
 - 10% of otitis externa.
- Clinical picture:
 - Pruritus:
 - Intense itching is the primary clinical complaint.
 - Otalgia.
 - Otorrhea.
- Diagnosis:
 - Examination under microscope:
 - Dotted white, black, or gray membrane over EAC.
 - Aspergillus produces distinct small black conidiophores on top of fluffy white filamentous hyphae.
 - Culture and sensitivity.
- Treatment:
 - **Thorough ear toilet:**
 - First and absolutely most important step.
 - Removal of all discharge and epithelial debris.
 - **Precautions:**
 - Avoid EAC trauma or intrumenation.
 - Maintain dry ear precautions.
 - **Antifungal topical drops (Canesten/Clotrimazole):**
 - Most effective and widely used topical azole.
 - It has antibacterial and antifungal effect.
 - Painful in the presence of a TM perforation or patent ventilation tube.
 - No reports of ototoxicity.
 - **Medicated Ear wick:**
 - In severe cases.



- **Herpes Zoster Oticus:**
- Occurs as primary infection or reactivation of latent varicella-zoster virus (VZV) that has remained dormant within sensory ganglia (commonly the geniculate ganglion) of the facial nerve.
- **Risk factors for the reactivation:**
 - Decrease immunity.
 - Physical and emotional stress.
- **Clinical picture:**
 - Burning pain
 - Localized headache
 - Coetaneous vesicular eruption of EAC and pinna.
 - Appear unilaterally in a dermatomic distribution.
 - Involvement of CN-VII may produce paresis or paralysis (**Ramsay Hunt syndrome**):
 - Accounts for 10% of all facial paralyses.
 - More severe paralysis and worse prognosis than Bell palsy.
 - Involvement of CN-VIII may produce SNHL and vertigo.
- **Complications of HZ oticus:**
 - Post-herpetic neuralgia
 - Residual paralysis
 - Herpes zoster encephalitis
- **Treatment of HZ oticus:**
 - Supportive Measures.
 - Warm compresses
 - Good analgesics
 - Anti-viral:
 - Acyclovir, Famciclovir, Valacyclovir
 - Early administration (< 72 h) increases rate of facial nerve function recovery.
 - Decreases severity of post-herpetic neuralgia.
 - Corticosteroids:
 - Used to relieve acute pain, decrease vertigo, and limit the occurrence of postherpetic neuralgia.
 - Treatment with acyclovir and prednisone has more effective return to facial nerve function and prevention of nerve degeneration.

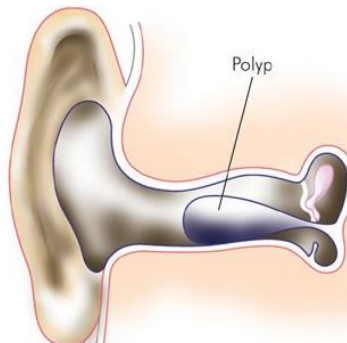
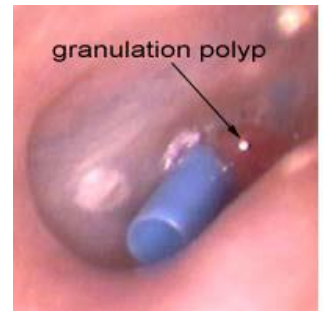
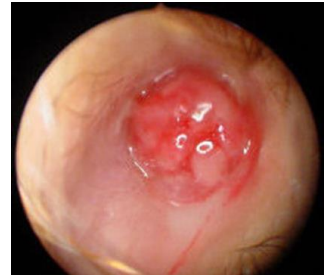


- **Bullous Otitis Externa:**
- Characterized by formation of serous/hemorrhagic bullae on epithelial surface of tympanic membrane and deep meatus.
- If it involves the tympanic membrane only, it is called **Bullous Myringitis**.
- Etiology:
 - Probably viral in origin.
 - Caused by same viruses or bacteria that cause middle ear infections.
 - Despite common belief, Mycoplasma pneumoniae is an extremely rare causative agent.
- Clinical picture:
 - Sudden severe pain in the ear.
 - Blood-stained discharge when the bullae rupture.
 - Hearing loss.
- Treatment:
 - Analgesics is directed to give relief from pain.
 - Antibiotics are given for secondary infection of the ear canal if the bulla has ruptured.
 - Severity of pain may warrant decompression of tense bullae with a sterile straight needle.
- **Eczematous Otitis Externa:**
- Hypersensitivity to infective organisms or topical ear drops.
- Marked by intense irritation, vesicle formation, oozing and crusting in the canal.
- Treatment is withdrawal of topical antibiotic causing sensitivity, and application of steroid cream.
- **Seborrhoeic Otitis Externa:**
- Associated with seborrhoeic dermatitis of the scalp.
- Itching is the main complaint.
- Greasy yellow scales are seen in the external canal, over the lobule and postauricular sulcus.
- Treatment consists of ear toilet, application of a cream containing salicylic acid and sulphur, and attention to the scalp for seborrhoea.
- **Neurodermatitis:**
- Compulsive scratching due to psychological factors.
- Patient's main complaint is intense itching.
- Otitis externa of bacterial type may follow infection of raw area left by scratching.
- Treatment is sympathetic psychotherapy and meant for any secondary infection.
- Ear pack and bandage to the ear are helpful to prevent compulsive scratching.



- **Aural Polyps:**

- Inflammatory tissue of EAC or middle ear mucosa.
- **Etiology:**
 - CSOM
 - Cholestatoma
 - Foreign body reaction to ventilation tube.
 - Malignant tumors
- **Clinical picture:**
 - Well-circumscribed, soft, fleshy mass.
 - Painless.
 - Symptoms related to EAC obstruction or secondary infection.
- **Diagnosis:**
 - History and physical examination.
 - CT/MRI with contrast:
 - Precisely define the anatomic site of origin.
 - Indicated if there is any suspicion that:
 - It could be attached to a deeper structure, such as facial nerve, stapes footplate, and a dehiscence of the vestibular labyrinth.
 - It could be a herniation of meninges and/ or brain (meningoencephalocele, encephalocele).



- **Treatment:**
 - Topical steroid/antibiotic drops.
 - Aural polyp excisional biopsy:
 - Must be done with caution and should never be avulsed.
 - Inflammatory polyp will easily yield to gentle manipulation.
 - Another underlying disease process may be more resistant and should never be removed with force.
 - Middle ear exploration:
 - In patients with aural polyp secondary to cholestatoma.

- **Non-Inflammatory disorders of External Ear:**

○ **Chondrodermatitis nodularis chronica (Winkler nodule):**

- Benign inflammatory condition.
- Caused by breakdown of elastic fibers due to chronic sun exposure.
- Clinical picture:
 - Most commonly located at free rim of helix or on the antihelix.
 - Papules or nodules painful to touch or pressure and patient is unable to sleep on the affected side.
 - Distinguishes it from other painless dermatologic lesions such as:
 - Senile keratosis
 - Keratoacanthoma
 - Cutaneous hom
 - Skin cancer (squamous or basal cell).
 - There is a chronic inflammatory infiltration of the perichondrium with focal degeneration or deformity of the underlying cartilage.
- Treatment:
 - Wide excision with deep shave of the underlying cartilage.
 - Intralesional injections with steroid.



○ **Keratoacanthoma:**

- Self-healing basal cell carcinoma.
- Caused by actinic (sun) exposure.
- May arise from hair follicles.
- Clinical picture:
 - Rapidly growing, raised, and circular with a central crater usually containing a keratin plug.
- Diagnosis:
 - Excisional biopsy for definitive diagnosis.
- Treatment:
 - Observation if the lesion is undergoing gradual resolution.



- **Keratitis Obturans:**

- Acute upon chronic condition of external ear.
- Accumulation of desquamated keratinous material in bony EAC leading to secondary, acute infection.
- Occurs bilaterally in young patients with bronchiectasis and chronic sinusitis.
- Occurs also in elderly patients in a nursing home in which care of the ear is neglected.
- Keratitis Obturans is reversible condition.
 - Once completely cleaned and topically medicated, the ear will gradually revert back toward a healthy appearance.
 - Different than External canal cholesteatoma which is irreversible condition.
- Etiology:
 - Abnormal epithelial migration and/or hyperplastic epithelium with increased desquamation.
 - Normally, epithelium from surface of tympanic membrane migrates onto the posterior meatal wall.
- Clinical picture:
 - Otorrhea with keratin debris inside EAC.
 - Severe otalgia.
 - CHL
 - Widening of bony EAC with ulceration and granuloma formation.
 - In advanced cases, EAC stenosis occurs in response to chronic irritation and infection.
- Diagnosis:
 - History and physical examination.
 - CT scan shows diffuse widening of the EAC.
 - Pathologic examination of keratin materials.
- Treatment:
 - Regular thorough debridement of EAC under microscope in the clinic.
 - Extremely painful to clean and pain must be eliminated prior to aural toilet.
 - May require a four-quadrant block of EAC with LA or may be done in OR under GA.
 - Antibiotics/steroid drops if infected.
 - Routine use of acetic acid solution or hydrogen peroxide solution may reduce accumulation.
 - Canaloplasty with skin graft for recalcitrant cases.

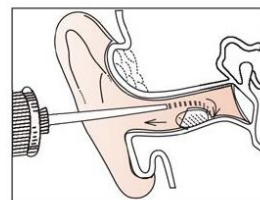
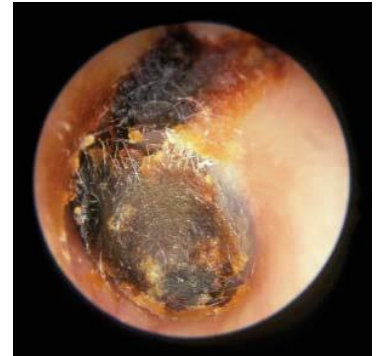


- **External Canal Cholesteatoma:**
 - Localized ulceration in skin of bony EAC with infection and sequestration of the underlying exposed tympanic bone.
 - Caused by blockage of EAC permitting accumulation of epithelial debris causing bone remodeling from pressure of the keratin.
 - Usually unilateral and affects older patients.
 - External Canal Cholesteatoma is irreversible condition.
 - It does not revert back toward a normal state once obstruction and local infection have been relieved and treated.
 - Different than Keratosis Obturans which is reversible condition.
 - External Canal Cholesteatoma has many features in common with primary acquired cholesteatoma including active matrix that elaborates keratin debris which can locally erode bone and soft tissue.
 - Etiology:
 - Acquired:
 - Surgery
 - Trauma
 - Acquired stenosis
 - Chronic inflammation
 - Spontaneous
 - Clinical picture:
 - Tympanic ring especially its anterior and inferior aspects is a favored site for External Canal Cholesteatoma.
 - Persistent dull otalgia.
 - Otorrhea with keratin debris inside EAC.
 - CHL
 - Diagnosis:
 - History and physical examination.
 - CT scan shows focal erosion.
 - Treatment:
 - External canal cholesteatoma is mainly managed medically and not surgically.
 - Regular thorough debridement of EAC under microscope in the clinic.
 - There is really very little need for surgery in even the most advanced cases.
 - Meatoplasty may be indicated to facilitate aural toilet.
 - Canaloplasty with skin graft for recalcitrant cases.



○ **Cerumen impaction:**

- Normally, small amount of wax is secreted which dries up and later expelled from the meatus by movements of the jaw.
- **Wax is composed of:**
 - Sebaceous glands secretions
 - Ceruminous glands secretions
 - Hair
 - Desquamated epithelial debris
 - Keratin
 - Dirt.
- **Risk factors of cerumen impaction:**
 - Higher activity of ceruminous glands
 - Narrow and tortuous ear canal
 - Stiff hair
 - Obstructive lesion of the canal.
 - Using Q-tips
 - Hearing aids
 - Earplugs
- **Clinical picture:**
 - Onset of these symptoms may be sudden when water enters the ear canal during bathing or swimming and the wax swells up.
 - CHL if >95% of canal is occluded
 - Fullness
 - Tinnitus
 - Autophony
 - Vertigo (rarely)
- **Treatment:**
 - Manual removal under magnification.
 - **Techniques:**
 - Instrumental manipulation.
 - Low-pressure irrigation if no perforation
 - Cerumen-softening agents:
 - Waxsol (Docusate Sodium)
 - Sodium Bicarbonate
 - Mineral oil

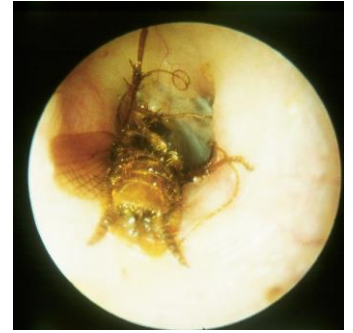


- **EAC foreign body (FB):**

- Classified into:

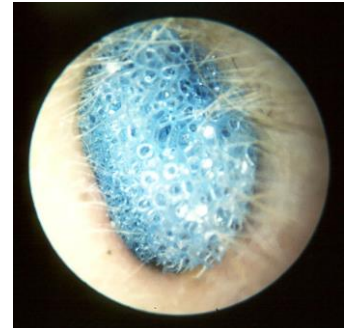
- **Living (Organic):**

- Flying or crawling insects.
 - Cause intense irritation and pain.
 - Patient is bothered by insect's movement.
 - Insects must be killed and then removed.
 - Lidocaine or mineral oil.



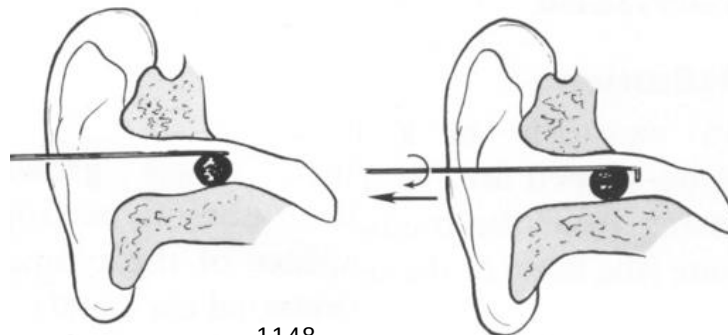
- **Non-living (Inorganic):**

- Piece of paper or sponge, grain seeds (rice, wheat, maize), batteries, slate pencil, piece of chalk or metallic ball bearings, overlooked cotton swab.
 - Batteries
 - Vegetable foreign bodies tend to swell up with time and get tightly impacted in the ear canal or may even suppurate.

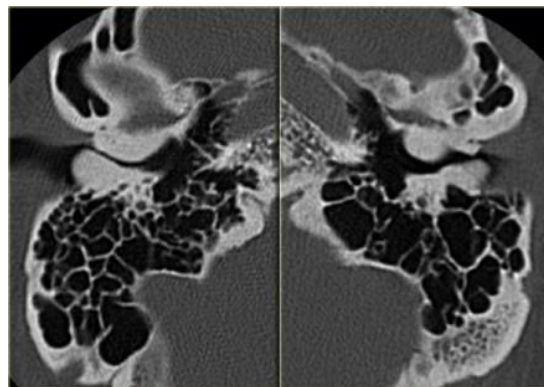


- **Treatment:**

- Manual removal under magnification.
 - Batteries must be removed as soon as possible without delay because they may cause extensive chemical burns to the EAC.
 - For uncooperative patient or impacted FB with failed attempts of removal, best done in OR under GA.
 - **Techniques:**
 - Soft and irregular foreign bodies like a piece of paper, swab or a piece of sponge can be removed with fine crocodile forceps.
 - Most of the seed grains and smooth objects can be removed with syringing.
 - Smooth and hard objects like steel ball bearing should not be grasped with forceps as they tend to move inwards and may injure the tympanic membrane and best removed with a blunt wax hook.
 - Postaural approach is used to FB impacted in deep meatus, medial to the isthmus or those which have been pushed into the middle ear.



- **Neoplastic disorders of External Ear:**
 - **EAC Exostoses (Surfer's ear):**
 - Benign, slowly growing, broad-based bony growth of deep EAC.
 - Not true neoplasms.
 - Localized bony calluses or hyperostoses arising from the medial surface of tympanic bone.
 - Males are affected three times more than females.
 - 70% of exostoses is reported in surfing population.
 - **Etiology:**
 - Acquired, develop as a reactive phenomenon (Refrigeration Periostitis) due to chronic exposure of EAC bone to cold water or air in activities such as swimming, surfing, skiing, or boating.
 - Cold induce vasoconstriction of deep EAC, followed by a reactive hyperaemia and a stimulation of periosteum lining the medial surface of the tympanic bone which lays down consecutive layers of subperiosteal bone.
 - **Clinical picture:**
 - Bilateral, multiple, sessile bony lesion of deep EAC.
 - Covered by normal deep canal skin.
 - Found more medial than Osteoma.
 - Usually asymptomatic, unless it is large or multiple causing trapping of wax and keratin debris in the deep meatus producing:
 - CHL
 - Recurrent otitis externa
 - Acquired external canal cholesteatoma
 - **Diagnosis:**
 - History and physical examination.
 - **CT scan:**
 - Broad based bony overgrowth with no deep extension (no stalk).
 - More dense than osteomas.



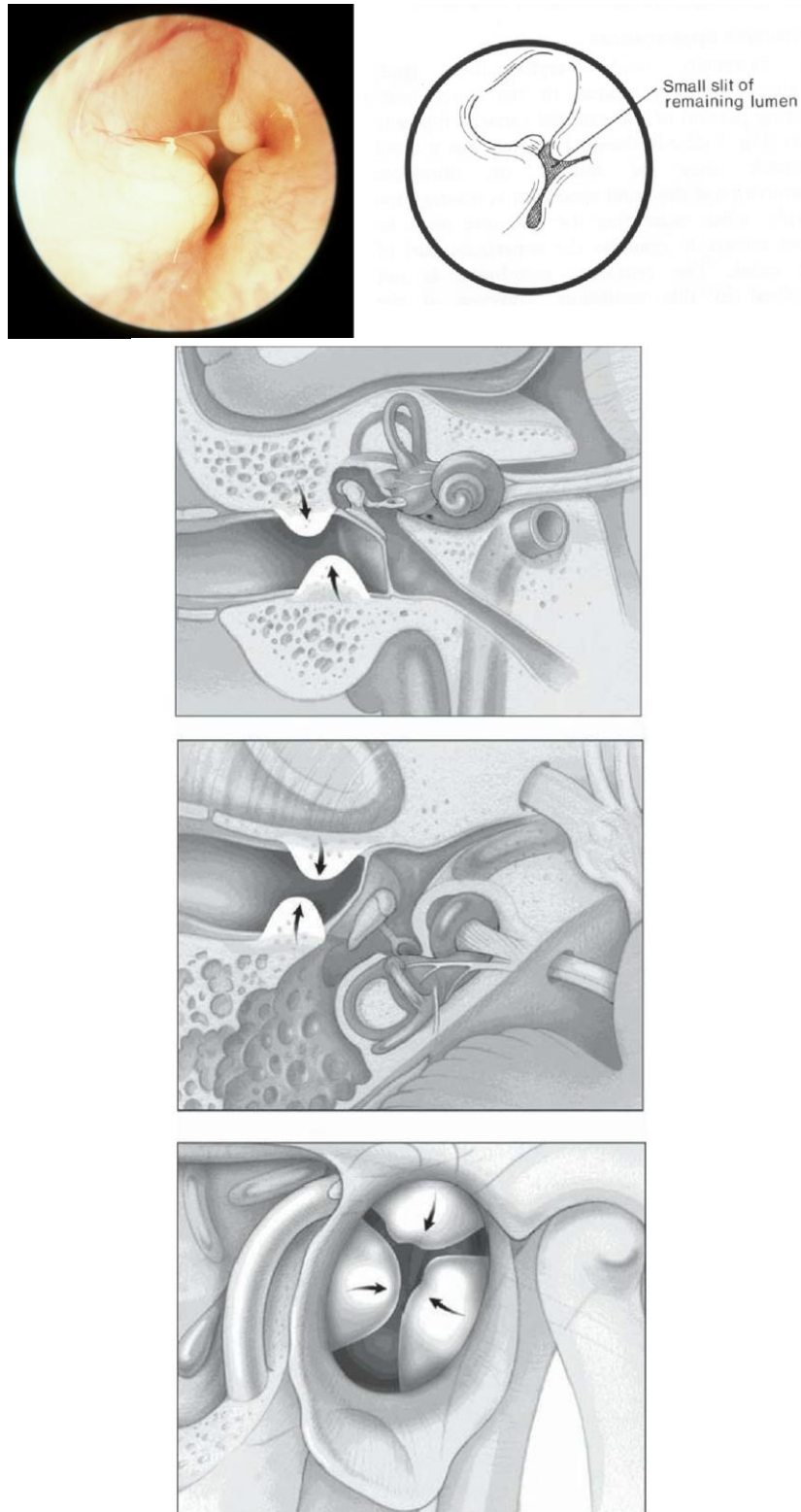


Figure 146.9 A: Coronal section of a right ear showing multiple growth centers of exostoses. B: Axial view of left ear showing multiple growth centers of exostoses. C: Sagittal section of a right ear showing multiple growth centers of exostoses. Figure courtesy of Robert K. Jackler MD.

- **EAC Osteoma:**
 - True benign bony neoplasm.
 - Arises from tympanic bone at tympano-squamous or tympano-mastoid suture lines.
 - **Etiology:**
 - Unknown.
 - Does not develop as a reactive phenomenon.
 - Not related to cold water or air exposure.
 - **Clinical picture:**
 - Unilateral, solitary, pedunculated bony lesion of EAC.
 - Covered by normal deep canal skin.
 - Found more lateral than Exostoses.
 - Usually asymptomatic, unless it is large trapping of wax and keratin debris in the deep meatus producing:
 - CHL
 - Recurrent otitis externa
 - Acquired external canal cholesteatoma
 - **Diagnosis:**
 - History and physical examination.
 - **CT scan:**
 - Osteoma is more heterogeneous with areas of cancellous bone.
 - Less dense than exostosis.
 - Stalk leading to the osteoma can be seen as well.



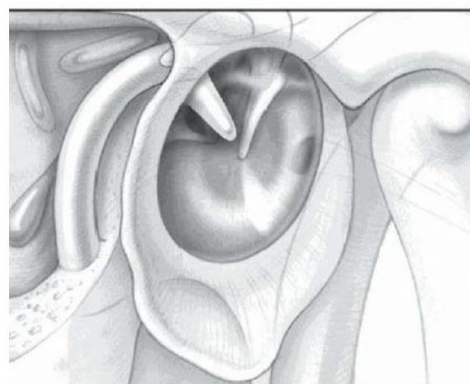
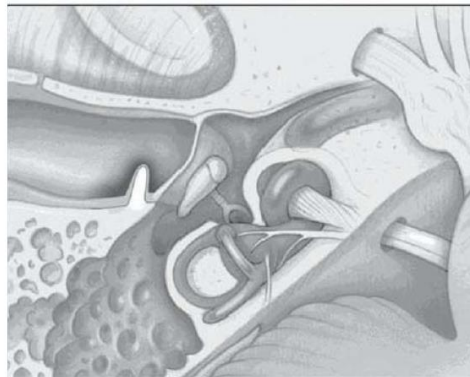
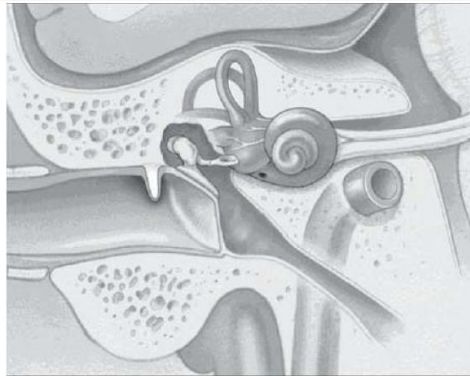
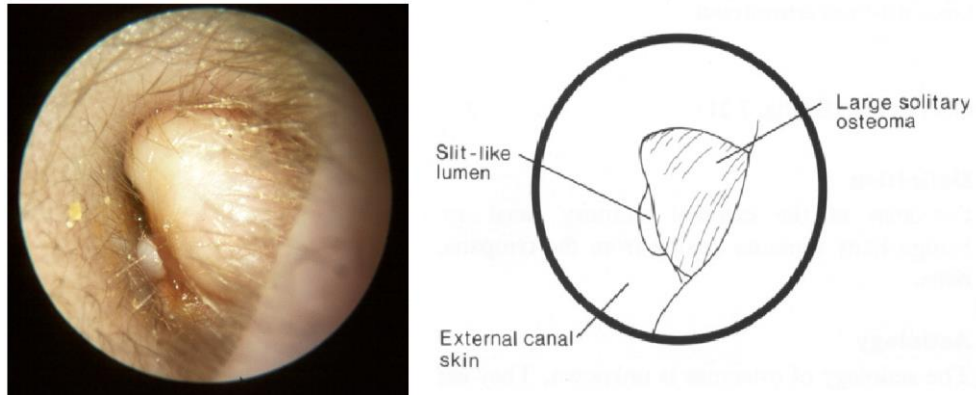


Figure 146.8 A: Coronal section of a right ear showing a single stalk of an osteoma. B: Axial view of left ear showing a single stalk of an osteoma. C: Sagittal section of a right ear showing a single stalk of an osteoma. Figure courtesy of Robert K. Jackler MD.

- [Treatment of Exostoses and Osteoma:](#)
 - **Observation:**
 - For small, asymptomatic lesions and when most of TM is visible.
 - **Surgery:**
 - Indications:
 1. Symptomatic patients
 - CHL
 - Recurrent otitis externa
 - External canal choistatoma.
 2. To allow proper HA fitting.
 - Surgical approach:
 - Trans-canal shaving with facial nerve monitoring and preservation of the skin:
 - The goal is to be able to see most of TM and to enlarge the EAC enough to hold an ITC (in the canal) HA mold.
 - Post-auricular approach is used sometimes.
 - Surgical complications:
 - TMJ violation.
 - Canal stenosis
 - SNHL
 - TM perforation
 - Facial nerve injury

**TABLE
147.1**

**NEOPLASMS OF THE EAR
AND LATERAL SKULL BASE**

Neoplasms of the pinna and external auditory canal

Cutaneous carcinoma

Squamous cell carcinoma

Basal cell carcinoma

Malignant melanoma

Glandular neoplasm

Ceruminous adenoma

Ceruminous adenocarcinoma

Pleomorphic adenoma

Adenoid cystic carcinoma

Osteoma and exostosis

Miscellaneous neoplasm

Merkel cell carcinoma

Squamous papilloma

Pilomatrixoma

Myxoma

Auricular endochondral pseudocyst

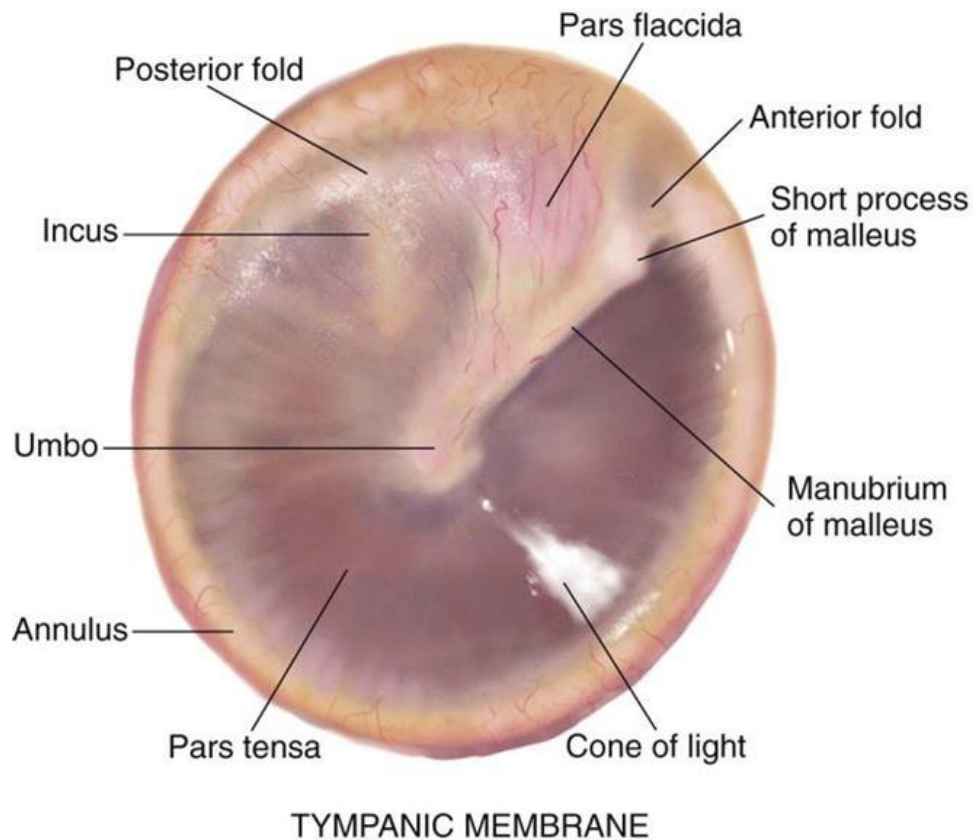
Chondrodermatitis nodularis chronica helices (Winkler disease)

- **Malignant Neoplasms of External Ear:**
 - Cancers of auricle and EAC include more commonly:
 - Basal cell carcinoma
 - Squamous cell carcinoma
 - Melanoma (melanotic and amelanotic).
 - In majority of skin cancers, malignancy is result of chronic sun (actinic) exposure.
 - Risk factors:
 - Individuals with fair skin, light hair and blue eyes.
 - Individuals with genetic defects of skin repair.
 - High amount and long sun exposure.
 - Tight-fitting ear molds leading to chronic irritation of the EAC
 - Treatment:
 - Many malignancies of the auricle may be treated with local, chemo-controlled excision with the defect repaired via skin grafting or local advancement flaps.

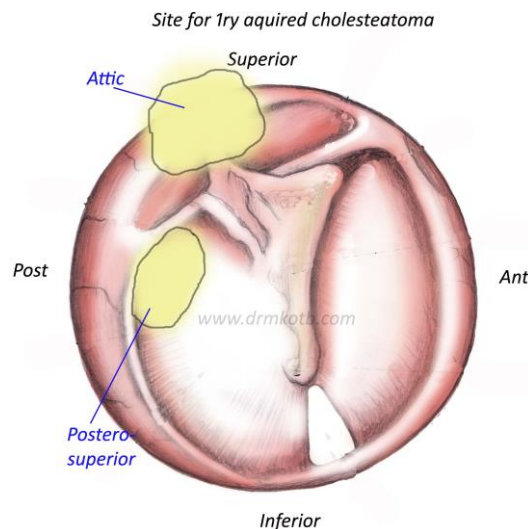


Disorders of Tympanic Membrane:

- Disorders of TM may be primary or secondary to conditions affecting external ear, middle ear or eustachian tube.
- Normal tympanic membrane:
 - o Shiny and pearly-grey in color.
 - o Semitransparent and some middle ear structures can be seen.
 - o Mobile when tested with pneumatic otoscope.
 - o Concave on its lateral surface, more marked at the tip of malleus (umbo).
 - o Bright cone of light can be seen in Antero-inferior quadrant.
 - o Attic area lies above lateral process of malleus and is slightly pinkish.

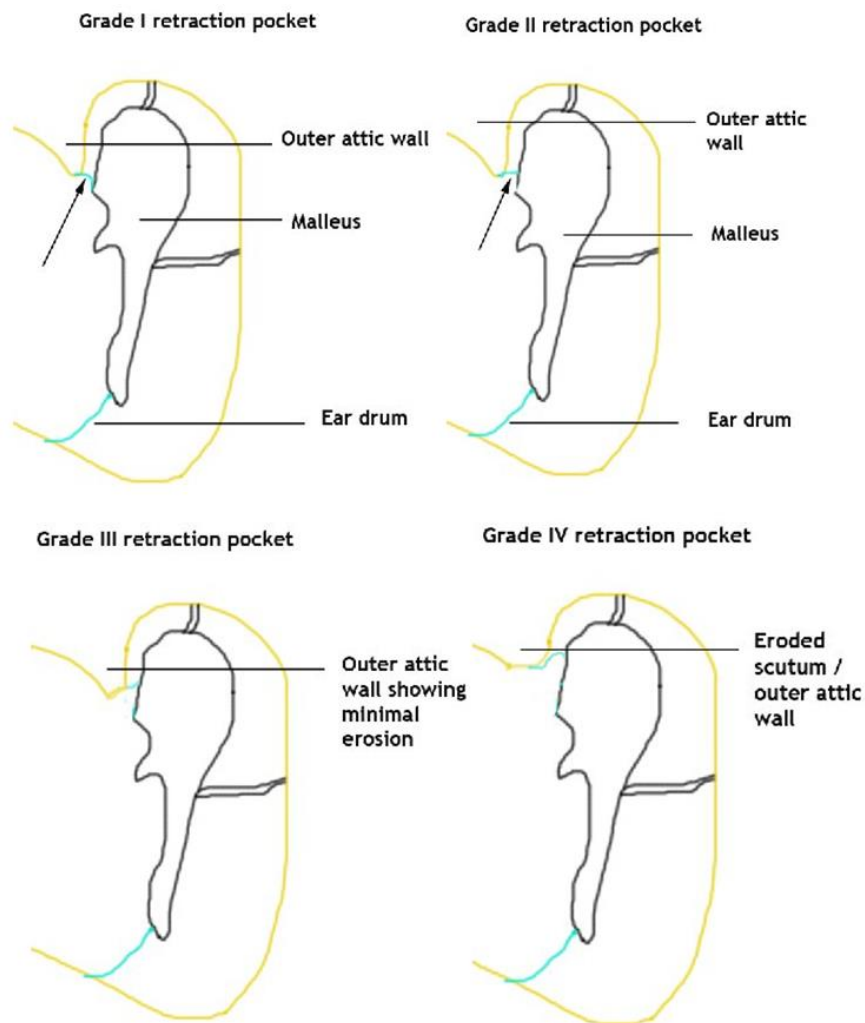


- **Retracted Tympanic Membrane and Retraction Pockets:**
 - Collapsing of a part or entire TM into middle ear.
 - Occurs as a result of negative intratympanic pressure when the eustachian tube is blocked.
 - Prolonged dysfunction of Eustachian tube and excessive negative pressure in the middle ear leads to atrophic changes in middle, fibrous layer of tympanic membrane and to development of localized or generalized TM atelectasis.
- Complications of Retracted TM:
 - 1. Cholestatoma:**
 - Deep retraction pocket impairs the normal migration of epithelial cells leading to accumulate keratin debris and formation of cholesteatoma.
 - 2. Ossicular Necrosis:**
 - Progression of TM retraction causes the atrophic membrane to drape over incus and stapes leading to necrosis of these ossicles.
 - Commonest ossicle eroded being the long process of incus.
 - 3. Adhesive Otitis Media:**
 - Prolonged TM retraction forms adhesions with the surrounded structures, which make changes irreversible.
- Most common sites of retraction pockets:
 - Pars flaccida:
 - Retraction pocket development is quicker, followed by quick bone absorption, and faster formation of cholesteatoma due to lack of fibrous layer of TM.
 - Management should be more aggressive.
 - Postero-superior parts of Pars tensa.



- Tos Classification for Pars Flaccida Retractions:

- **Tos I:**
 - Slight retraction of Pars flaccida that is not in contact with malleus head.
- **Tos II:**
 - Retraction of Pars flaccida that is in contact with malleus head.
 - Full extent of retraction pocket can be clearly seen.
- **Tos III:**
 - Severe retraction of Pars flaccida that is in contact with malleus head.
 - Limited erosion of outer attic wall (scutum).
 - Part of retraction pocket may be hidden.
- **Tos IV:**
 - Severe retraction of Pars flaccida that is in contact with malleus head.
 - Severe erosion of outer attic wall (scutum).
 - Extent of retraction pocket cannot be clearly seen.



- [Sadé Classification for Pars Tensa Retractions:](#)
 - **Sadé I:**
 - Slight retraction of Pars tensa.
 - **Sadé II:**
 - Severe retraction of Pars tensa with contact into incus or stapes.
 - **Sadé III:**
 - Middle ear atelectasis.
 - Severe retraction of Pars tensa with contact into promontory, but mobile with the Valsalva or Toynbee maneuvers.
 - **Sadé IV:**
 - Adhesive otitis media.
 - Severe retraction of Pars tensa with contact into promontory, but fixed.

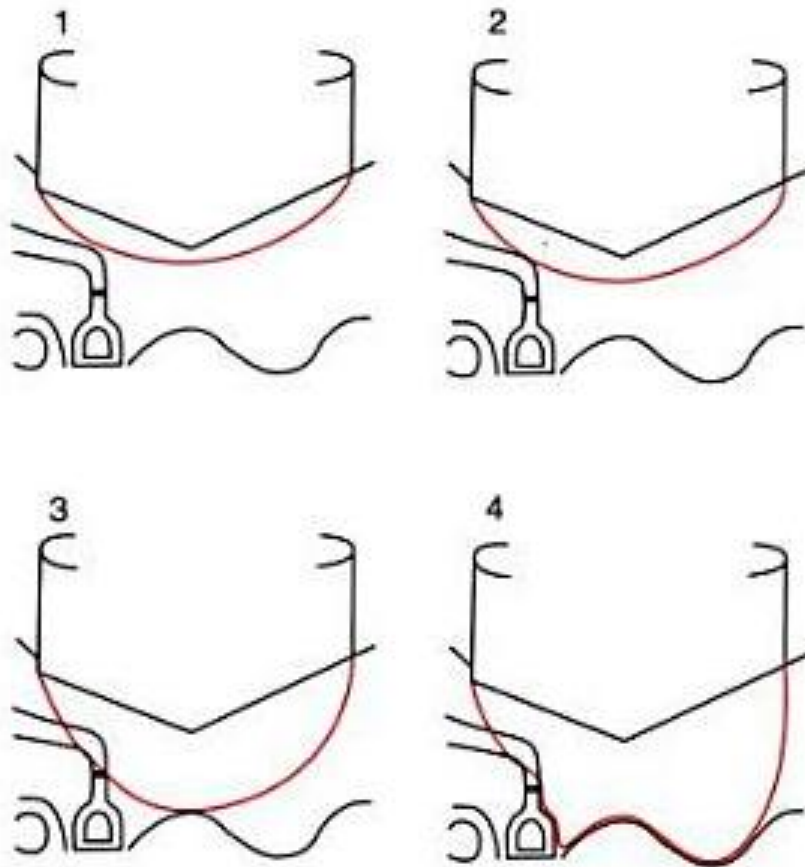
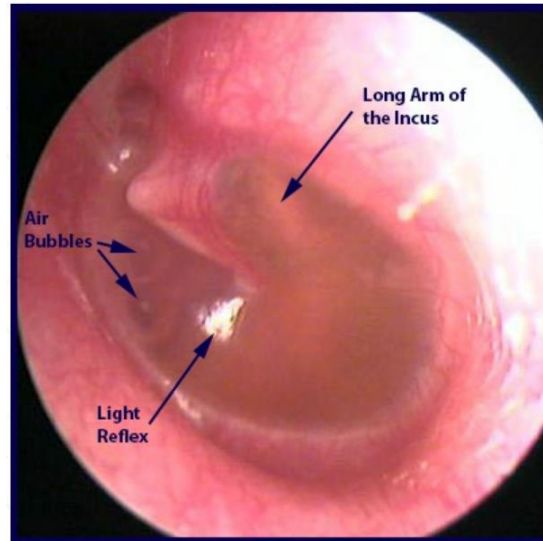


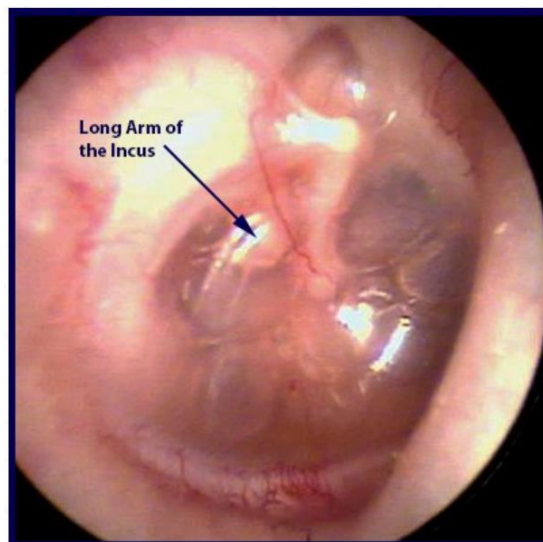
Figure 6.1 Sadé classification of atelectasis (modified) (see text).



14. Retracted Left Eardrum

The picture on the right shows a retracted eardrum with orange serous fluid in the middle ear. There is a near normal light reflex and an anterior superior air bubble. The long arm of the incus can also be seen through the eardrum.

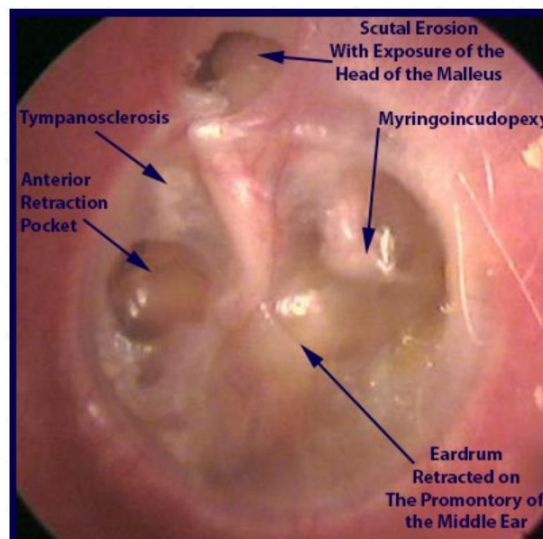
Sadé I



7. Retracted Right Eardrum

The picture on the right shows a retracted eardrum with fluid and multiple bubbles in the middle ear. The eardrum is also draped over the long arm of the incus.

Sadé II



15. Severely Retracted Left Eardrum

The picture on the right shows a severely retracted eardrum. The eardrum has mild tympanosclerosis and is forming three retraction pockets. One anteriorly, a second in the posterior-superior quadrant which is starting to erode the long arm of the incus forming a myringo-incudopexy, and a third attic retraction pocket with scutal erosion and the beginnings of exposure of the head of the malleus.

Sadé III - IV Tos III

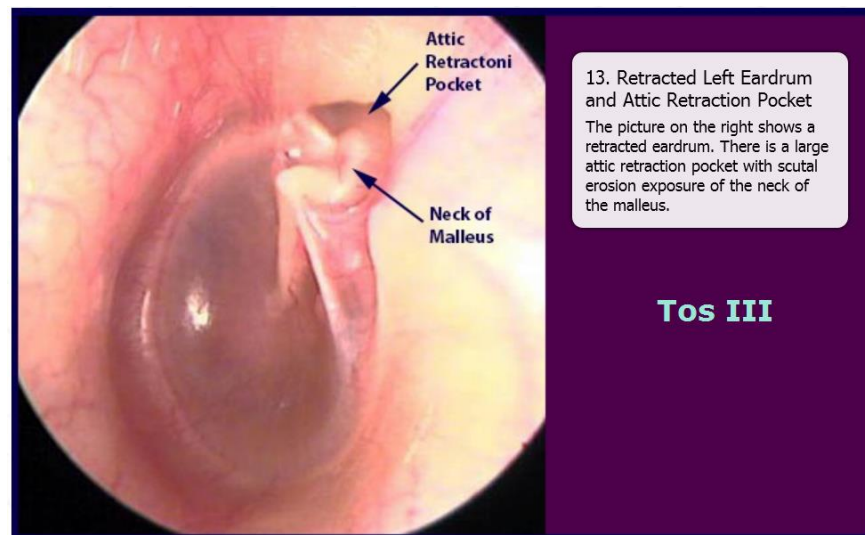
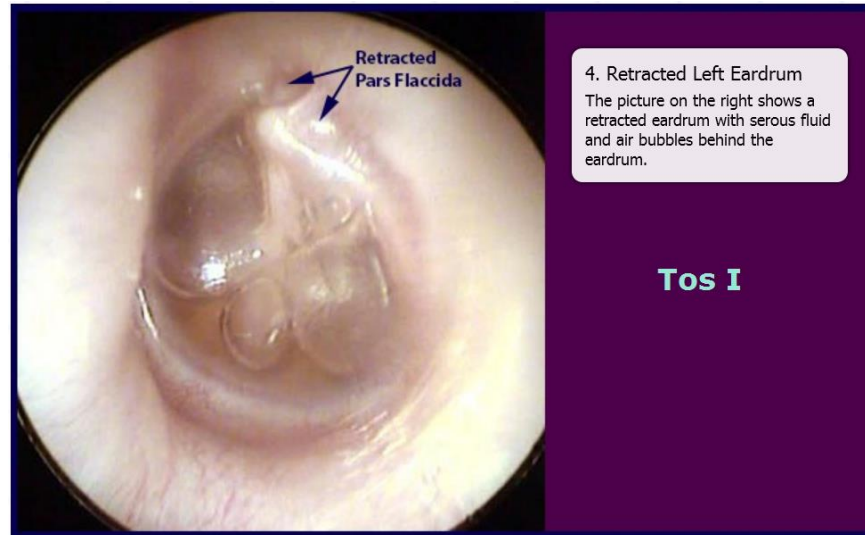


Table 1: Tympanic membrane retraction classification.

Retraction classification	Attic retraction classification (Tos and Poulsen) ⁷	Pars tensa retraction classification (Sade) ⁵
Stage 1	Slight retraction towards neck of malleus but airspace is visible	Slight retraction of the tympanic membrane (TM) over its annular fold
Stage 2	Retraction onto neck of malleus - no airspace is visible behind TM	TM touches ossicular chain (incudostapedial joint)
Stage 3	Retraction extends beyond bony annulus- full extent of retraction pocket is seen	TM touches promontorium
Stage 4	Erosion of outer attic wall	Adhesion of pars tensa to promontorium
Stage 5	Attic cholesteatoma	Atelectatic otitis media

- [Charachon Classification for TM Retractions:](#)
 - **Stage I:**
 - Mobile retraction pocket.
 - **Stage II:**
 - Fixed and controllable retraction pocket.
 - **Stage III:**
 - Fixed and uncontrollable retraction pocket.

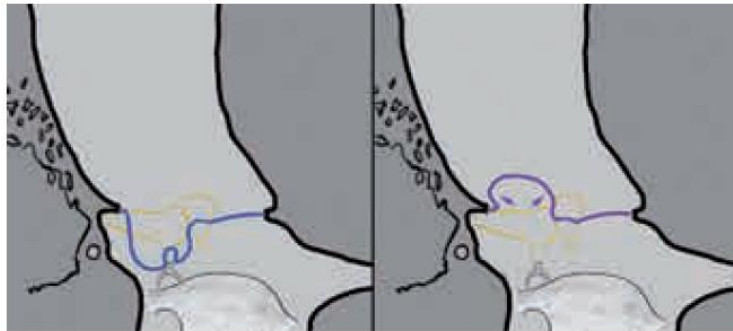


Fig. 2. Retraction pocket type 1 before and after inflation: controllable, not fixed (Fig. A.J. Fishman)



Fig. 3. Retraction pocket type 2 before and after inflation: controllable, fixed (Fig. A.J. Fishman)



Fig. 6. Retraction pocket type 3 before and after inflation: incontrollable, fixed (Fig. A.J. Fishman)

- [Clinical picture:](#)
 - May be asymptomatic.
 - Otolgia due to changes in middle ear pressure or infection.
 - Conductive hearing loss due to middle ear effusion, splinting of the tympanic membrane or erosion of the ossicular chain.
 - Recurrent ear discharge due to cholestatoma formation.
- [Diagnosis:](#)
 - **History and physical examination:**
 - Otological and nasal symptoms including allergy should be reviewed.
 - Thorough examination of Ear, nose and nasopharynx should be done.
 - **Examination under microscope:**
 - Ear should be thoroughly cleaned of cerumen and debris retraction pocket is often hidden behind them.
 - **Pneumatic otoscopy:**
 - Essential in establishing whether pocket is reversible (movable or fixated).
 - Patient may be also asked to perform Valsalva maneuver to inflate middle ear while otoscopy.
 - **Audiometry:**
 - When air bone gap is significant all cases should be considered for treatment.
 - **Tympanometry:**
 - Helpful in establishing whether retraction pocket is accompanied by middle ear fluid.
 - **CT Temporal bone:**
 - Indicated in deep retraction pocket where the bottom cannot be seen.
 - Evaluate presence of cholestatoma and any bony erosions.

- Treatment:

- Regular reassessment visits with audiological examination are important to assess disease progression and monitor for complications, such as cholesteatoma formation.

- Important issues before decision making:

- 1. Anatomic status of the retraction pocket:**

- Pars tensa or pars flaccida RP:
 - Pars flaccida retraction pockets developed quicker followed by quick bone absorption, and faster formation of cholesteatoma due to lack of fibrous layer of TM and management should be more aggressive.
- Controllable or uncontrollable RP:
 - If possible to see under microscope the extent of the pocket or not it is called controllable or incontrollable RP
- Mobile or Fixed RP:
 - If RP adheres to the middle ear structures but cannot be reversed is called fixed.
 - The one which adheres to the ossicles and promontory but can be reversed is called mobile.

- 2. Functional status of the ear (hearing):**

- If the hearing does not exceed 20dB Air Bone Gap (ABG) these RP should not be treated.

- 3. Presence of chronic middle ear effusion:**

- If retraction pocket is accompanied by chronic middle ear effusion it can be regarded as a proof of an active process in the middle ear.
- Long lasting fluid together with negative tympanic cavity pressure causes secondary changes of TM and should be treated without any delay.

- 4. Behavior of retraction pocket over time:**

- In some individuals, RP progressively proceeds to form adhesions and finally cholesteatoma.
- If there is rapid progression over time, RP should to be treated in order to prevent progression to more advanced stages.

- **Management protocol of RP (Charachon Classification):**
 - **Stage I (Mobile RP):**
 - Wait and see policy.
 - If after 3 months no progression is observed, the patient is followed up every three months, for two years.
 - Indications of surgery:
 1. Hearing loss > 20dB ABG.
 2. Presence of chronic middle ear effusion.
 3. Progression of RP overtime.
 - Types of Surgery:
 - Tympanostomy tube placement
 - Excision of RP:
 - It is reasonable not to excise to large areas of TM since large perforation may persist.
 - Excision should better be done if retraction is limited to one quadrant.
 - Pars flacida RP are not excised since there
 - is a risk of in growth of squamous epithelium and cholesteatoma formation at that area.



Fig. 7. Ear after subannular T-tube insertion

- **Stage II (Fixed and Controllable RP):**
 - Wait and see policy.
 - If after 3 months no progression is observed, the patient is followed up every three months, for two years.
 - Indications of surgery:
 1. Hearing loss > 20dB ABG.
 2. Presence of chronic middle ear effusion.
 3. Progression of RP overtime.
 - Types of Surgery:
 - Tympanostomy tube placement.
 - T-tube with cartilage graft:
 - T-tube for ventilation and cartilage for TM reinforcement.
 - Trans-canal approach, elevation of tympanomeatal flap and separation of middle ear adhesions, T-tube with cartilage TM support is inserted.
 - Cartilage tympanoplasty:
 - Atelectatic membrane should be elevated or excised and cartilage tympanoplasty should be performed.



Fig. 8. T-tube at place with cartilage tympanic membrane reinforcement in the place of previous t2 retraction pocket

- **Stage III (Fixed and Uncontrollable RP):**
 - Surgery should be performed to prevent cholesteatoma formation.
 - Temporal bone CT scan should be done to assess the extent of RP before surgery.
 - Types of Surgery:
 - Cartilage tympanoplasty:
 - Most authors advocate Cartilage tympanoplasty.
 - Atelectatic membrane should be elevated or excised and cartilage tympanoplasty should be performed.
 - Tympanomastoidectomy with posterior tympanotomy:
 - For more extent cases.
 - With or without ossicular reconstruction.
 - If RP is ruptured during surgery, second look surgery should be planned in the future to exclude residual cholesteatoma.
- **Tympanosclerosis:**
 - Deposition of hyaline and calcium within submucous layer of tympanic membrane (Myringosclerosis) or middle ear cavity (Intratympanic Tympanosclerosis).
 - **Myringosclerosis:**
 - Hyalinization and calcification of fibrous layer of tympanic membrane only.
 - Clinical picture:
 - Chalky white crescentic shaped plaque.
 - Mostly, it remains asymptomatic and does not affect the hearing.
 - No myringosclerosis will develop in the area of healed TM perforation because it lacks a fibrous middle layer.
 - Causes:
 - TM trauma
 - After grommet insertion
 - Chronic otitis media with effusion
 - Treatment:
 - Observation



- **Intratympanic Tympanosclerosis:**
- Hyalinization and calcification of submucous layer of middle ear cavity.
- Clinical picture:
 - Chalky white bone-like mass located anywhere in middle ear cavity but more commonly in:
 - Epitympanum
 - Promontory
 - Ossicles
 - Associated with marked conductive or mixed hearing loss and the degree of hearing loss depends on the extent of tympanosclerotic involvement of the ossicular chain.
- Causes:
 - Chronic otitis media with effusion
 - Recurrent acute otitis media
 - Autoimmune process
- Tympanosclerosis Vs Otosclerosis:
 - **Pre-op:**
 - Tympanosclerosis
 - History of recurrent otitis media.
 - Slowly progressive CHL.
 - Appears earlier in life.
 - Negative family history of otosclerosis.
 - Myringosclerosis on examination.
 - False cahart notch on PTA.
 - Otosclerosis:
 - No history of recurrent otitis media.
 - Slowly progressive CHL.
 - Appears in 2nd or 3rd decade of life.
 - Positive family history of otosclerosis.
 - Intact TM on examination.
 - Cahart notch on PTA.
 - **Intra-op:**
 - Tympanosclerosis covering footplate of stapes could easily be mistaken for otosclerosis since both fix footplate and look similar.
 - Tympanosclerosis:
 - Confined to mucosa covering the footplate and other parts of the middle ear.
 - Can be peeled off.
 - Otosclerosis:
 - Invades and replaces bone.
 - Requires removal of at least a part of the footplate to restore hearing.

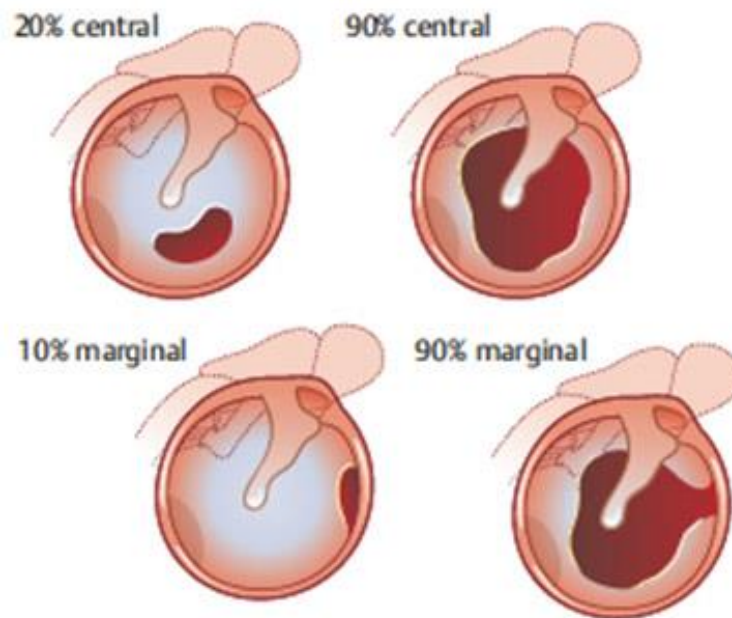


- Staging of Tympanosclerosis:
 1. Myringosclerosis only.
 2. Fixed incudomalleolar joint with mobile stapes.
 3. Fixed stapes with mobile incudomalleolar joint.
 4. Completely fixed ossicular chain.
- Treatment:
 - Management of tympanosclerosis is depending on the staging and degree of involvement:
 - **Stage 1:**
 - Observation.
 - **Stage 2:**
 - Mobilize the incudomalleolar joint if possible.
 - Remove the incus and do either incus transposition or incus replacement prosthesis between stapes and malleus.
 - **Stage 3:**
 - Mobilize the stapes.
 - Stapedectomy.
 - **Stage 4:**
 - Conservative with hearing aids.
 - Important points:
 - Results of surgical treatment of CHL in tympanosclerosis is unpredictable and sometimes unsatisfactory.
 - Tympanosclerotic plaques are difficult to remove.
 - Risk of recurrence and re-fixation of ossicles over time.
 - Risk of inner ear trauma mainly during mobilization or stapedectomy



○ **TM Perforation:**

- Tympanic membrane may be perforated by:
 - Trauma:
 - FB
 - Unskilled instrumentation.
 - Sudden change in air pressure:
 - Slap or a kiss on the ear.
 - Sudden blast.
 - Forceful Valsalva may rupture a thin atrophic membrane.
 - Pressure by a fluid column:
 - Diving, water sports.
 - Fracture of temporal bone.
 - AOM
 - CSOM



D. Examples of central and marginal perforations of the tympanic membrane. The exact size and location should be documented during the initial clinical examination.

- **Atrophic tympanic membrane:**

- A normal tympanic membrane consists of outer epithelial, middle fibrous and inner mucosal layer.
- In serous otitis media, the middle fibrous layer gets absorbed leaving a thin drumhead which easily gets collapsed with eustachian tube insufficiency.
- A perforation of tympanic membrane also heals only by epithelial and mucosal layers without the intervening fibrous layer.



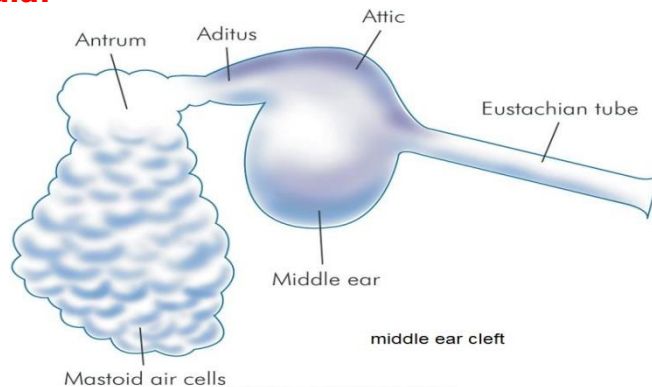
- **Bullous Myringitis:**

- Characterized by formation of serous/hemorrhagic bullae on epithelial surface of tympanic membrane and deep meatus.
- Etiology:
 - Probably viral in origin.
 - Caused by same viruses or bacteria that cause middle ear infections.
 - Despite common belief, Mycoplasma pneumoniae is an extremely rare causative agent.
- Clinical picture:
 - Sudden severe pain in the ear.
 - Blood-stained discharge when the bullae rupture.
 - Hearing loss.
- Treatment:
 - Analgesics is directed to give relief from pain.
 - Antibiotics are given for secondary infection of the ear canal if the bulla has ruptured.
 - Severity of pain may warrant decompression of tense bullae with a sterile straight needle.



Acute Suppurative Otitis Media:

- ✓ It is an acute (<3 weeks) inflammation of middle ear by **pyogenic organisms**.
- ✓ middle ear implies **middle ear cleft**, i.e.
 - eustachian tube
 - middle ear
 - attic,
 - aditus antrum
 - mastoid air cells



- ✓ The highest incidence of otitis media occurs between the ages of 6 and 12 months and decreases with age
- ✓ Most children experience at least one episode of AOM during their childhood

❖ Aetiology

- ✓ follows **viral infection** of upper respiratory tract but soon the pyogenic organisms invade the middle ear.
- ✓ second most common disease in **infants and children** especially in children of lower socio-economic group (upper respiratory infection is the most common)

❖ Routes of Infection

✓ 1. Via eustachian tube

- **most common** route.
- Infection travels via the lumen of the tube or along subepithelial peritubal lymphatics.
- Eustachian tube in infants and young children is **shorter, wider and more horizontal** and thus may account for higher incidence of infections in this age group " **by the age of 7 years, when the tube has a more adult configuration, the prevalence of otitis media is low** "
- Breast or bottle feeding in a young infant in horizontal position may force fluids through the tube into the middle ear and hence the need to keep the infant propped up with head a little higher.
- Swimming and diving can also force water through the tube into the middle ear.

✓ 2. Via external ear

- Traumatic **perforations** of tympanic membrane due to any cause open a route to middle ear infection.

✓ 3. Blood-borne

- This is an uncommon route

❖ Risk Factors

- ✓ Anything that **interferes** with normal functioning of **eustachian tube** results in **negative middle** ear pressures leading to:-

- an influx of bacteria and viruses from the NP when the ET opens
- **transudative fluid** collection in the middle ear space **predisposes to middle ear infection**.

✓ **host risk factors**

1. Age (highest incidence of AOM is between 6 and 11 months of age)
2. Gender (Male)
3. Race (black)
4. Genetic predisposition
5. Ciliary dyskinesia (Kartagener syndrome)
6. Adenoids , tonsil , Chronic rhinitis and sinusitis (reservoir of infection & mechanical ET obstruction)
7. ET dysfunction (short, horizontal, compliant)
8. Cleft palate " **After surgical repair of the palate, some pt the occurrence of otitis media is reduced**" , Craniofacial abnormality, Down's , Apert; mucopolysaccharidoses , Treacher Collins syndrome
9. Immune deficiency (especially IgA and IgG subclass 2 and 3 deficiencies)
10. allergy (disputed) "middle ear mucosa acts as a **shock organ**"
11. +ve family Hx
12. Gastroesophageal reflux

✓ **Environmental risk factors**

1. URTIs (most common)
2. Daycare attendance (2.6x)
3. Season (Fall/Winter)
4. Older siblings
5. Parental history of OM
6. Passive smoking
7. Low S/E status (overcrowding, poor sanitation)
8. Lack of breastfeeding
9. Night-time bottle (horizontal position)
10. Pacifier use "possibly due to the sucking action of the child propelling **nasopharyngeal: secretions into the middle ear or by the pacifier acting as a fomite**"
11. NGT , packing of nose or nasopharynx for epistaxis
12. Obesity.

❖ Bacteriology

- ✓ **Most common organisms in infants and young children are**
 - *Streptococcus pneumoniae* (30%) (Gram-positive, alpha-hemolytic)
 - *Haemophilus influenzae* (20%) (Gram-negative aerobic, coccobacilli)
 - *Moraxella catarrhalis* (12%) (Gram-negative, aerobic)
- ✓ **Other organisms include**
 - *Streptococcus pyogenes* (called Group A (beta-hemolytic) Streptococcus)
 - *Staphylococcus aureus* (anaerobic Gram-positive coccus)
 - *Pseudomonas aeruginosa*. (Gram-negative aerobic, coccobacillus)
- ✓ In about 18-20%, no growth is seen.
- ✓ Many of the strains of *H. influenzae* (34%) and *Moraxella catarrhalis* (100%) are β -lactamase producing.
- ✓ if **immunocompromised**, look for oddities (Mycoplasma, Chlamydia)
- viral pathogens **4 – RRIP!** (RSV “most common”, rhino, influ, parainflu) are often present alone (**sterile otitis media**) or potentiate bacterial pathogens
- ✓ **Pathogen causing bilateral, dull TM, non-mobile, T > 37°C**
 - *Haemophilus influenzae*
- ✓ **Pathogen causing unilateral, bulging TM, T > (38.1)°**
 - *Streptococcus pneumoniae*
- ✓ **Pathogen causing bullous myringitis**
 - *Streptococcus pneumoniae*
 - Others include Mycoplasma, H. flu, Beta-hemolytic strep, M. catarrhalis, Parainfluenza & influenza virus
- ✓ **Pathogen causing spontaneous**
 - *Streptococcus pyogenes*

❖ Pathology and Clinical Features

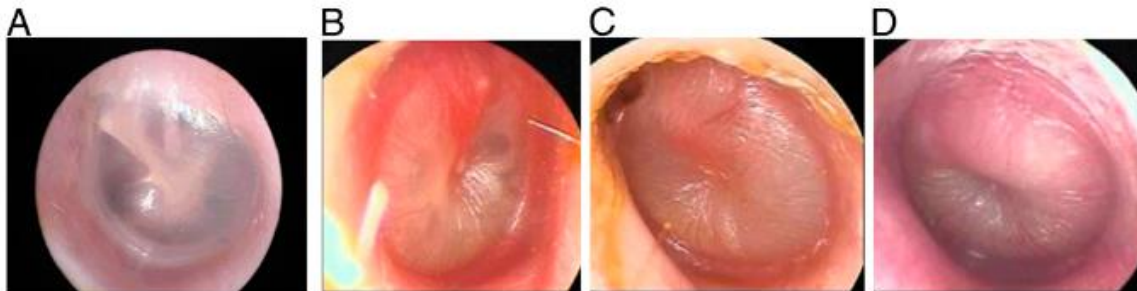
✓ The disease runs through the following stages:

✓ 1. Stage of tubal occlusion

- ✓ **Oedema and hyperaemia** of nasopharyngeal end of eustachian tube **blocks** the tube, leading to absorption of air and **negative intratympanic pressure**.
- ✓ **retraction of tympanic membrane** with some degree of **effusion** in the middle ear but **fluid may not be clinically appreciable**.
- ✓ **Symptoms.**
 - **Deafness and earache** "not marked"
 - generally **no fever**.
- ✓ **Signs.**
 - Tympanic membrane is **retracted** with **handle of malleus assuming a more horizontal position, prominence of lateral process of malleus**
 - loss of light reflex
 - Tuning fork tests show conductive deafness.

✓ 2. Stage of pre-suppurative

- ✓ If tubal occlusion is prolonged
- ✓ pyogenic organisms invade tympanic cavity causing hyperaemia of its lining.
- ✓ Inflammatory exudate appears in the middle ear.
- ✓ Tympanic membrane becomes congested.
- ✓ **Symptoms.**
 - **Earache** marked **disturb sleep** and **throbbing nature**.
 - **Deafness and tinnitus** are also present, only by **adults**.
 - **high degree of fever and is restless** usually, runs in **child**.
- ✓ **Signs.**
 - **begin** with
 - **congestion** of pars tensa.
 - Leash of **blood vessels** radiate from **handle of malleus** to the **periphery of tympanic** membrane imparting it a **cart-wheel appearance**.
 - **Later,**
 - whole of tympanic membrane including pars flaccida becomes **uniformly red**.
 - Tuning fork tests will again show conductive type of hearing loss.



A, Normal TM. B, TM with mild bulging. C, TM with moderate bulging. D, TM with severe bulging.

✓ 3. Stage of suppuration

- ✓ This is marked by formation of **pus** in the **middle ear** and to some extent in **mastoid air cells**.
- ✓ Tympanic membrane starts **bulging** to the point of **rupture**.
- ✓ **Symptoms.**
 - **Earache** becomes excruciating.
 - **Deafness** increases
 - child may run **fever**
 - may be accompanied by **vomiting** and even **convulsions**.
- ✓ **Signs.**
 - Tympanic membrane appears red and bulging with loss of landmarks.
 - Handle of malleus may be engulfed by the swollen and protruding tympanic membrane and may not be discernible.
 - yellow spot (blister) may be seen on the tympanic membrane where rupture is imminent.
 - Tenderness may be elicited over the mastoid antrum.
- ✓ X-rays of mastoid will show clouding of air cells because of exudate.



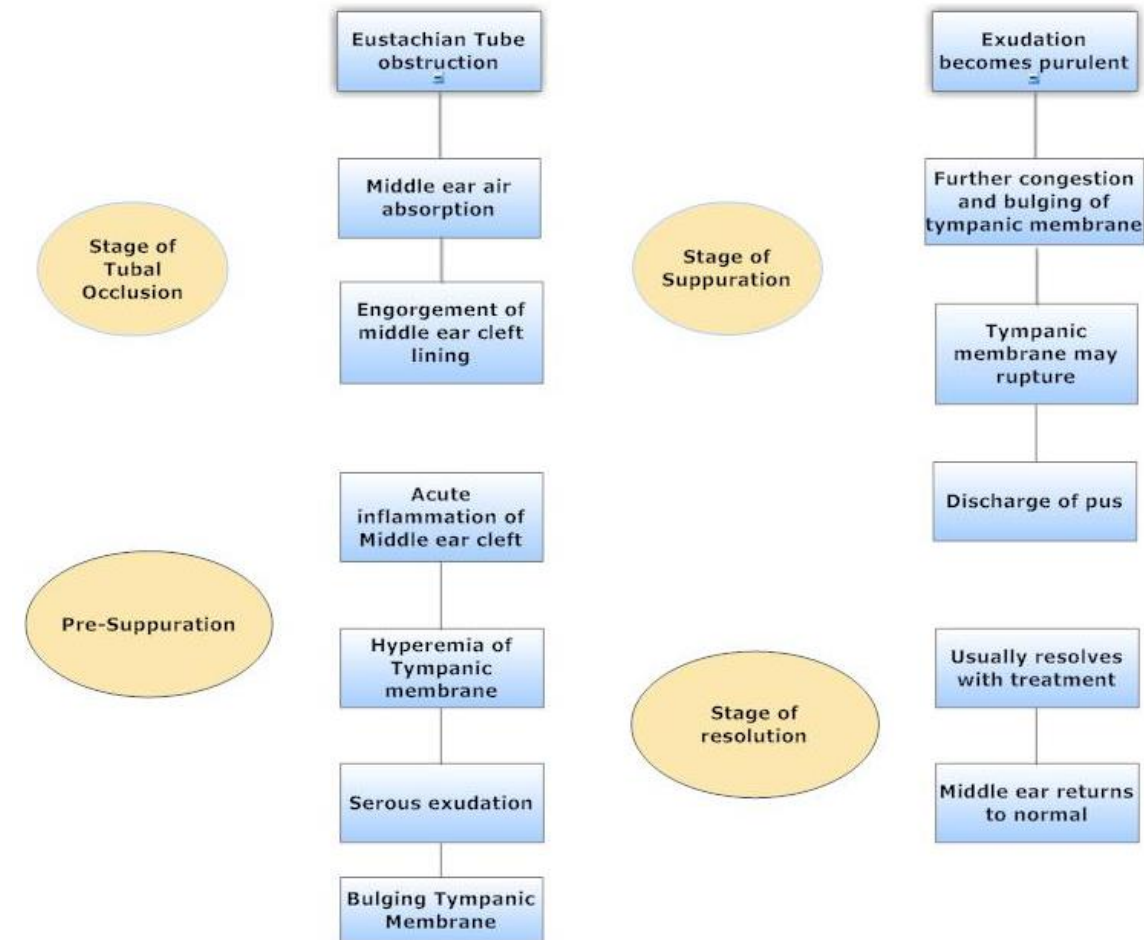
✓ 4. Stage of resolution

- ✓ The tympanic membrane **ruptures** with **release of pus** and **subsidence of symptoms**.
- ✓ Inflammatory process begins to resolve.
- ✓ If proper treatment is started early or if the infection was mild, resolution may start even without rupture of tympanic membrane.
- ✓ **Symptoms.**
 - earache is relieved, fever comes down and pt feels better.
- ✓ **Signs.**
 - External auditory canal may contain **blood-tinged discharge** which later becomes **mucopurulent**.
 - Mucoïd discharge from an ear must mean that there is a perforation of the tympanic membrane. There are no mucous glands in the external canal.
 - Usually small perforation is seen in **antero-inferior** quadrant of pars tensa.
 - Hyperaemia of tympanic membrane begins to subside with return to normal colour and landmarks.

✓ **5. Stage of complication**

- ✓ If virulence of organism is high or resistance of patient poor, resolution may not take place and disease spreads **beyond** the confines of middle ear.
- ✓ It may lead to:-

Intratemporal complications of AOM	Intracranial complications of AOM
○ Chronic suppurative OM ○ Adhesive otitis ○ Tympanic membrane perforation ○ Cholesteatoma ○ Tympanosclerosis ○ Fixation and Discontinuity of ossicular chain ○ Mastoiditis with or without Abscess (Postauricular, Bezold's, Zygomatic, Parapharyngeal, Retropharyngeal) ○ Petrositis ○ Labyrinthitis – Serous or Suppurative ○ Facial palsy ○ Labyrinthine fistula (Pasha) ○ CHL or SNHL	○ Meningitis (#1, HiB > Pneumococcus, Mondini) ○ Epidural/subdural/cerebral abscesses ○ Focal encephalitis ○ Lateral/sigmoid sinus thrombosis (Tobey-Ayer/Quesckenstedt's Test; Griesinger's sign)) ○ Otitic hydrocephalus ○ Blindness with optic neuropathy



❖ **Diagnosis :-**

- ✓ clinical and physical exam "ears, a proper head and neck examination is invaluable, because it may identify condition that may predispose "
- ✓ audiogram (CHL <30 dB) and tympanometry

❖ **PREVENTION OF DISEASE**

1. MANAGEMENT OF RIEK FACTORS

- ✓ Promotion of breastfeeding in the first 6 months of life
- ✓ Avoidance of supine bottle feeding and pacifier use
- ✓ Elimination of passive tobacco smoke .
- ✓ Alteration of child care arrangements so that the child is exposed to fewer children

2. VACCINES

- ✓ **Bacterial Vaccines**
- ✓ Streptococcus pneumoniae Vaccine
- ✓ Haemophilus influenzae Vaccine
- ✓ Moraxella catarrhalis Vaccine
- ✓ **Viral Vaccines**
- ✓ Influenza Vaccine
- ✓ Respiratory Syncytial Virus Vaccine

❖ **Treatment****1. Antibacterial therapy**

- ✓ It is indicated in **all cases** with **fever and severe earache**.
- ✓ As the most common organisms are **Strept. pneumoniae** and **H. influenzae**, the drugs which are effective in acute otitis media are **amoxicillin**.
- ✓ **amoxicillin-clavulanate (augmentin)** Recommended
 - for children who have been treated with amoxicillin in the previous 30 days
 - for those with concurrent purulent conjunctivitis
 - for those with a history of recurrent AOM unresponsive to amoxicillin. "**β-lactamase-producing H. influenzae or Moraxella catarrhalis**"
- ✓ if allergic to these penicillins can be given **Cephalosporins** such as **cefdinir, cefuroxime, cefpodoxime, and ceftriaxone**
- ✓ Antibacterial therapy must be continued for of **7-10 days**, till **tympanic membrane regains normal appearance and hearing returns to normal**.
- ✓ typically resolves infection within 72 hours; if no resolution may consider broader spectrum coverage (augmentin , ceftriaxon+ clindamycin)
- ✓ Early discontinuance of therapy with relief of earache and fever, or therapy given in inadequate doses may lead to secretory otitis media and residual hearing loss.
- ✓ Tympanocentesis should always be considered if the child does not respond to the antibiotic treatment, in order to identify the bacteria in the MEE and to select an appropriate antibiotic.

TABLE 5 Recommended Antibiotics for (Initial or Delayed) Treatment and for Patients Who Have Failed Initial Antibiotic Treatment

Initial Immediate or Delayed Antibiotic Treatment		Antibiotic Treatment After 48–72 h of Failure of Initial Antibiotic Treatment	
Recommended First-line Treatment	Alternative Treatment (if Penicillin Allergy)	Recommended First-line Treatment	Alternative Treatment
Amoxicillin (80–90 mg/ kg per day in 2 divided doses)	Cefdinir (14 mg/kg per day in 1 or 2 doses)	Amoxicillin-clavulanate ^a (90 mg/kg per day of amoxicillin, with 6.4 mg/kg per day of clavulanate in 2 divided doses)	Ceftriaxone, 3 d Clindamycin (30–40 mg/kg per day in 3 divided doses), with or without third-generation cephalosporin
or	Cefuroxime (30 mg/kg per day in 2 divided doses)	or	Failure of second antibiotic
Amoxicillin-clavulanate ^a (90 mg/kg per day of amoxicillin, with 6.4 mg/kg per day of clavulanate [amoxicillin to clavulanate ratio, 14:1] in 2 divided doses)	Cefpodoxime (10 mg/kg per day in 2 divided doses)	Ceftriaxone (50 mg IM or IV for 3 d)	Clindamycin (30–40 mg/kg per day in 3 divided doses) plus third-generation cephalosporin
	Ceftriaxone (50 mg IM or IV per day for 1 or 3 d)		Tympanocentesis ^b Consult specialist ^b

IM, intramuscular; IV, intravenous.

^a May be considered in patients who have received amoxicillin in the previous 30 d or who have the otitis-conjunctivitis syndrome.^b Perform tympanocentesis/drainage if skilled in the procedure, or seek a consultation from an otolaryngologist for tympanocentesis/drainage. If the tympanocentesis reveals

2. Decongestant nasal drops

- ✓ should be used to relieve eustachian tube oedema and promote ventilation of middle ear.
 - Ephedrine nose drops (1% in adults and 0.5% in children)
 - oxymetazoline (Nasivion)
 - xylometazoline (Otrivin)
- ✓ nasal drops is traditional but its value is uncertain in the presence of acute inflammation of the middle ear.

3. Oral nasal decongestants

- ✓ Pseudoephedrine OR combination of decongestant and antihistaminic
- ✓ may achieve the same result without resort to nasal drops which are difficult to administer in children.

4. Analgesics and antipyretics

- ✓ Paracetamol helps to relieve pain and bring down temperature.
- ✓ Avoid the use of aspirin in children because of the risk of Reye's syndrome.

5. Ear toilet

- ✓ If the ear is already discharging when the patient is first seen, a swab should be sent for culture of the organism

6. Dry local heat

- ✓ It helps to relieve pain.

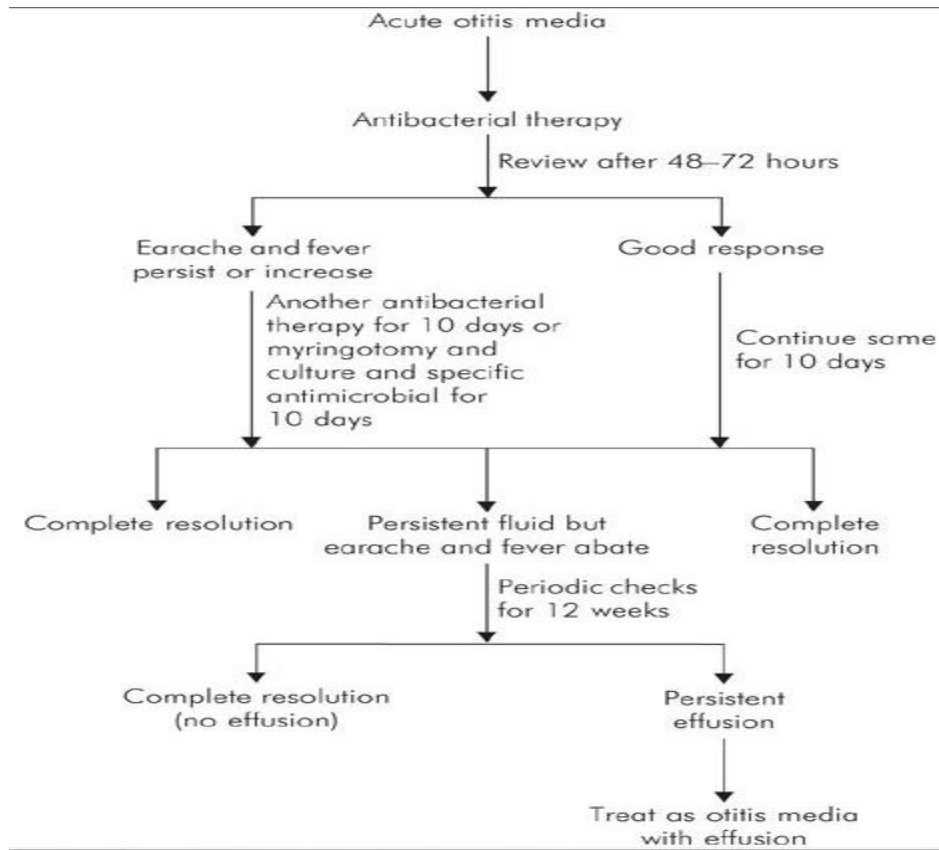
7. Myringotomy

- ✓ It is incising the drum to evacuate pus and is **indicated when**
 - a. drum is bulging and there is severe acute pain or toxic patients
 - b. there is an incomplete resolution despite antibiotics when drum remains full **with persistent conductive deafness**
 - c. there is persistent effusion beyond **12 weeks (3 months)**.
 - d. Complications of acute otitis media, e.g. facial paralysis, labyrinthitis or meningitis with bulging tympanic membrane
- ✓ helpful for relief of pain and allows samples to be obtained for culture to identify the pathogen and to guide in the selection of antibiotics, but provides no advantage in duration of effusion or recurrence of episodes of AOM

All cases of acute suppurative otitis media should be carefully followed till drum membrane returns to its normal appearance and conductive deafness disappears

If resolution does not occur, suspect:

- 1** the nose, sinuses or nasopharynx? Infection may be present;
- 2** the choice or dose of antibiotic;
- 3** low-grade infection in the mastoid cells



❖ **Recurrent Acute Otitis Media**

- ✓ 4 or more AOM in 1 yr, or 3 or more in 6/12
- ✓ Infants and children between the age of **6 months and 6 years** may get recurrent episodes of acute otitis media.
- ✓ Usually, they occur after acute upper respiratory infection, the child being free of symptoms between the episodes
- ✓ Recurrent middle infections may sometimes be **superimposed upon an existing middle ear effusion.**
- ✓ Sometimes, the underlying cause is
 - recurrent sinusitis
 - velopharyngeal insufficiency
 - hypertrophy of adenoids
 - infected tonsils allergy
 - immune deficiency as " IgA deficiency or hypogammaglobulinaemia".
 - Feeding the babies in supine position without propping up the head may cause the milk to enter the middle ear directly that can lead to middle ear infection.

✓ **Management of such children involves:**

- **1. Finding the cause and eliminating it, if possible.**
- **2 Myringotomy and insertion of tympanostomy tube** "guidelines (2013) recommend against insertion of tympanostomy tubes in children with recurrent AOM who do not have MEE "
- **3. Antimicrobial prophylaxis.**
 - disadvantage of creating antimicrobial resistance or hypersensitivity reaction
 - not preferred by many in favour of early insertion of tympanostomy tubes.
- Adenoidectomy may provide modest improvement in children with recurrent AOM but is not recommended as a firstline procedure unless indicated for airway obstruction.
- Tonsillectomy, in conjunction with adenoidectomy, provides no significant advantage over adenoidectomy alone, and the risk outweigh the benefits.

❖ **ACUTE NECROTISING OTITIS MEDIA**

- ✓ It is a **variety** of **acute suppurative otitis media**,
- ✓ often seen in **children** suffering from **measles, scarlet fever or influenza**.
- ✓ Causative organism is **β-haemolytic streptococcus**.
- ✓ There is rapid **destruction** of whole of tympanic membrane with its annulus, mucosa of promontory, ossicular chain and even mastoid air cells.
- ✓ There is profuse otorrhoea.
- ✓ healing is followed by fibrosis or ingrowth of squamous epithelium from the meatus (**secondary acquired cholesteatoma**).
- ✓ Treatment
 - **Antibacterial therapy**. It is continued for at least 7-10 days, even if response is seen early.
 - **Cortical mastoidectomy** may be indicated if medical treatment fails to control or the condition gets complicated by **acute mastoiditis**.

CLINICAL PRACTICE GUIDELINE The Diagnosis and Management of Acute Otitis Media From THE AMERICAN ACADEMY OF PEDIATRICS 2013

- Evidence-based clinical practice guideline is a revision of the 2004 acute otitis media (AOM) guideline from the American Academy of Pediatrics (AAP) and American Academy of Family Physicians.
- It provides recommendations to primary care clinicians for the management of children from 6 months through 12 years of age with uncomplicated AOM.

Statement 1A:

Clinicians should diagnose acute otitis media (AOM) in children who present with moderate to severe bulging of the tympanic membrane (TM) or new onset of otorrhea not due to acute otitis externa.

Evidence Quality: Grade B. Strength: **Recommendation**.

Statement 1B:

Clinicians may diagnose AOM in children who present with mild bulging of the TM and recent (less than 48 hours) onset of ear pain (holding, tugging, rubbing of the ear in a nonverbal child) or intense erythema of the TM.

Evidence Quality: Grade C. Strength: **Recommendation**.

Statement 1C:

Clinicians should not diagnose AOM in children who do not have middle ear effusion (MEE) (based on pneumatic otoscopy and/or tympanometry).

Evidence Quality: Grade B. Strength: **Recommendation**.

.

Statement 2:

The management of AOM should include an assessment of pain. If pain is present, the clinician should recommend treatment to reduce pain.

Evidence Quality: Grade B. Strength: **Strong Recommendation**.

Statement 3A:

Severe AOM: The clinician should prescribe antibiotic therapy for AOM (bilateral or unilateral) in children 6 months and older with severe signs or symptoms (ie, moderate or severe otalgia or otalgia for at least 48 hours or temperature 39°C [102.2°F] or higher).

Evidence Quality: Grade B. Strength: **Strong Recommendation**.

Statement 3B:

Non severe bilateral AOM in young children: The clinician should prescribe antibiotic therapy for bilateral AOM in children 6 months through 23 months of age without severe signs or symptoms (ie, mild otalgia for less than 48 hours and temperature less than 39°C [102.2°F]).

Evidence Quality: Grade B. Strength: **Recommendation**.

Statement 3C:

Non severe unilateral AOM in young children: The clinician should either prescribe antibiotic therapy or offer observation with close follow-up based on joint decision making with the parent(s)/caregiver for unilateral AOM in children 6 months to 23 months of age without severe signs or symptoms (ie, mild otalgia for less than 48 hours and temperature less than 39°C [102.2°F]).

When observation is used, a mechanism must be in place to ensure follow-up and begin antibiotic therapy if the child worsens or fails to improve within 48 to 72 hours of onset of symptoms.

Evidence Quality: Grade B. Strength: **Recommendation.**

Statement 3D:

Non severe AOM in older children: The clinician should either prescribe antibiotic therapy or offer observation with close follow-up based on joint decision-making with the parent(s)/ caregiver for AOM (bilateral or unilateral) in children 24 months or older without severe signs or symptoms (ie, mild otalgia for less than 48 hours and temperature less than 39°C [102.2°F]).

When observation is used, a mechanism must be in place to ensure follow-up and begin antibiotic therapy if the child worsens or fails to improve within 48 to 72 hours of onset of symptoms. Evidence Quality: Grade B. Strength:

Recommendation.

Statement 4A:

Clinicians should prescribe amoxicillin for AOM when a decision to treat with antibiotics has been made and the child has not received amoxicillin in the past 30 days or the child does not have concurrent purulent conjunctivitis (otitis-conjunctivitis syndrome) or the child is not allergic to penicillin.

Evidence Quality: Grade B. Strength: **Recommendation.**

Statement 4B:

Clinicians should prescribe an antibiotic with additional β -lactamase coverage for AOM when a decision to treat with antibiotics has been made, and the child has received amoxicillin in the last 30 days or has concurrent purulent conjunctivitis, or has a history of recurrent AOM unresponsive to amoxicillin.

Evidence Quality: Grade C. Strength: **Recommendation.**

Statement 4C:

Clinicians should reassess the patient if the caregiver reports that the child's symptoms have worsened or failed to respond to the initial antibiotic treatment within 48 to 72 hours

and determine whether a change in therapy is needed.

Evidence Quality: Grade B. Strength: **Recommendation.**

Statement 5A:

Clinicians should not prescribe prophylactic antibiotics to reduce the frequency of episodes of AOM in children with recurrent AOM.

Evidence Quality: Grade B. Strength: **Recommendation**.

Statement 5B:

Clinicians may offer tympanostomy tubes for recurrent AOM (3 episodes in 6 months or 4 episodes in 1 year with 1 episode in the preceding 6 months).

Evidence Quality: Grade B. Strength: **Option**.

Statement 6A:

Clinicians should recommend pneumococcal conjugate vaccine to all children

Evidence Quality: Grade B. Strength: **Strong Recommendation**.

Statement 6B:

Clinicians should recommend annual influenza vaccine to all children

Evidence Quality: Grade B. Strength: **Recommendation**.

Statement 6C:

Clinicians should encourage exclusive breastfeeding for at least 6 months.

Evidence Quality: Grade B. Strength: **Recommendation**.

Statement 6D:

Clinicians should encourage avoidance of tobacco smoke exposure.

Evidence Quality: Grade C. Strength: **Recommendation**.

Otitis Media With Effusion:

- ✓ Also known
 - Serous Otitis Media
 - Secretory Otitis Media
 - Mucoïd Otitis Media
 - "Glue Ear"
- ✓ persistence of fluid in the middle ear space without evidence of infection" **non-purulent** " effusion in the middle ear cleft.
- ✓ Often the effusion is thick and viscid but sometimes it may be thin and serous.
- ✓ The fluid is **nearly sterile**.
- ✓ **Time that fluid has to be present for the condition to be chronic is usually taken as 12 weeks**
- ✓ patients with a history of chronic OME have more sclerotic mastoids with decreased pneumatization compared with healthy subjects.
- ✓ Two theories
 - hereditary theory states that children with hypoaeration of the mastoid are prone to OME,
 - environmental theory states that chronic OME results in hypopneumatization of the mastoid.

❖ Epidemiology

- First Episode
 - 50% of all children- before the first birthday
 - 80% of all children - before the third birthday
- Prevalence bimodal at 2 & 5 yrs when child first attends playgroup school & when goes to primary school
- Above 15 yrs → prevalence 0.6%
- More during winters
- More than a third of consultations to pediatricians each year
- M > F

❖ Pathogenesis

- **Two main mechanisms are thought to be responsible:**
- ✓ **1. Malfunctioning of eustachian tube**
Eustachian tube fails to aerate the middle ear and is also unable to drain the fluid.
- ✓ **2. Increased secretory activity of middle ear mucosa**
Biopsies of middle ear mucosa in these cases have confirmed increase in number of mucus or serous-secreting cells.

❖ Aetiology

✓ 1. Malfunctioning of eustachian tube, the causes are:

- (i) Adenoid hyperplasia.
- (ii) Chronic rhinitis and sinusitis.
- (iii) Chronic tonsillitis. "Enlarged tonsils mechanically obstruct the movements of soft palate and interfere with the physiological opening of eustachian tube".
- (iv) Benign and malignant tumours of nasopharynx. This cause should always be excluded in unilateral serous otitis media in an adult.
- (v) Palatal defects, e.g. cleft palate, palatal paralysis. (Although some studies have shown that the incidence of middle-ear disease decreases after surgical repair of the cleft palate, many children with a cleft palate continue to have middle-ear problems)
- (vi) Oedema during radiation therapy
- (vii) Syndromes: Down's; Apert; mucopolysaccharidoses

✓ 2. Allergy AND IMMUNOLOGY

- Seasonal or perennial allergy to inhalants
- foodstuff :- milk
- not only obstructs eustachian tube by oedema but may also lead to **increased secretory activity** as middle ear mucosa acts as a **shock organ** in such cases.
- History of immunological disorders (especially IgA and IgG subclass 2 and 3 deficiencies) **Recurrence of bilateral OME**

✓ 3. Unresolved otitis media

- **≈10%** of acute otitis media cases have persistent fluid **3 months** after resolution of the infection
- Inadequate antibiotic therapy in acute suppurative otitis media may inactivate infection but fail to resolve it completely "**Low grade infection**".
- **Biofilms** also have been identified in the nasopharynx of children with otitis media, and it was suggested that the biofilm may act as a reservoir for bacterial pathogens resistant to antibiotics "**Mechanical debridement of nasopharyngeal biofilms may explain the observed clinical benefit associated with adenoidectomy in subsets of pediatric patients**"
- **Low grade infection , biofilm**, acts as stimulus for mucosa to secrete more fluid & number of **goblet cells and mucous glands** also increase.

✓ 4. bacterial , Viral infections

- Various "**syncytial virus (RSV), influenzavirus, adenovirus, parainfluenza virus, and rhinoviruses**" of upper respiratory tract may invade middle ear mucosa and stimulate it to increased secretory activity.
- H. influenzae was the single most common bacterial pathogen, with S.

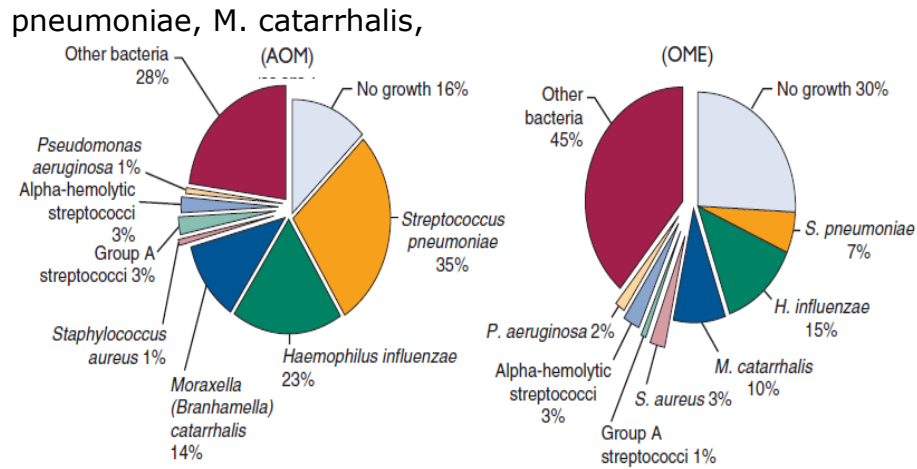


FIGURE 195-3. Comparison of distribution of isolates in 2807 effusions from patients with acute otitis media (AOM) and 4589 effusions from patients with otitis media with effusion (OME) at the Pittsburgh Otitis Media Research Center, 1980 to 1989. Total percentages are greater than 100% because of multiple organisms. (Modified from Bluestone CD, Klein JO: *Otitis media in infants and children*, ed 4, Hamilton, Ontario, 2007, BC Decker, p 103.)

✓ 5.gastroesophageal reflux

- may be a causative factor in otitis media,
- potential role for antireflux therapy in the treatment of otitis media in some children.
- Adequately controlled trials have not yet been done.

❖ Risk Factors

✓ host risk factors

1. Age (< 2 year)
2. Gender (Male)
3. Race (Caucasian)
4. Genetic predisposition
5. Ciliary dyskinesia (Kartagener syndrome)
6. Cystic fibrosis
7. Adenoids , tonsil , Chronic rhinitis and sinusitis (reservoir of infection & mechanical ET obstruction)
8. ET dysfunction (short, horizontal, compliant)
9. Cleft palate " After surgical repair of the palate, some pt the occurrence of otitis media is reduced" , Craniofacial abnormality, Down's , Apert; mucopolysaccharidoses , Treacher Collins syndrome
10. Immune deficiency (especially IgA and IgG subclass 2 and 3 deficiencies)
11. allergy (disputed) "middle ear mucosa acts as a shock organ"
12. +ve family Hx
13. Gastroesophageal reflux
14. Poor mastoid pneumatization

✓ Environmental risk factors

1. URTIs , post AOM
2. Daycare attendance with > 4 children
3. Season (Fall/Winter)
4. Passive smoking
5. Low S/E status (overcrowding, poor sanitation)
6. Lack of breastfeeding
7. Night-time bottle (horizontal position)
8. Pacifier use "possibly due to the sucking action of the child propelling nasopharyngeal: secretions into the middle ear or by the pacifier acting as a fomite"

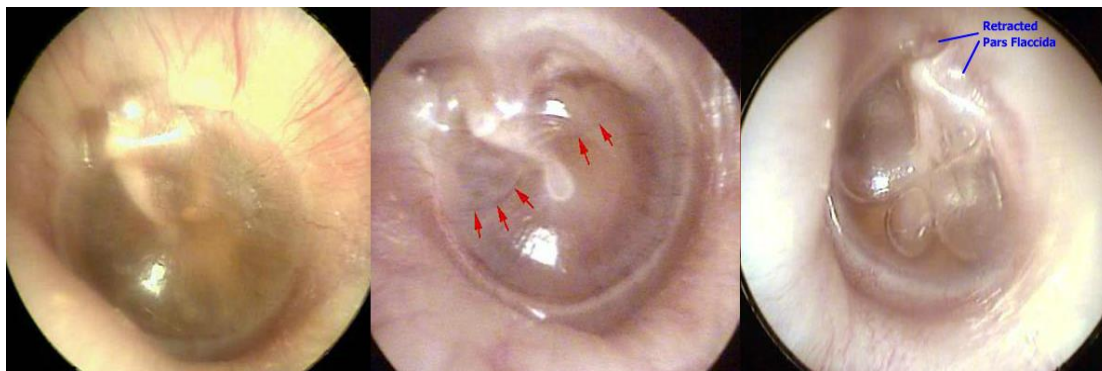
❖ Clinical Features

✓ Symptoms

- The disease affects children of 5-8 years of age. The symptoms include:
 - i. **Hearing loss.**
 - sometimes the only symptom.
 - insidious in onset and rarely exceeds 40 dB.
 - Deafness may pass unnoticed by the parents and may be accidentally discovered during audiometric screening tests.
 - i. **Delayed and defective speech Poor Academics in children.**
 - ii. **Mild earaches & tinnitus.**
 - There may be history of upper respiratory tract infections with mild earaches (fullness) .

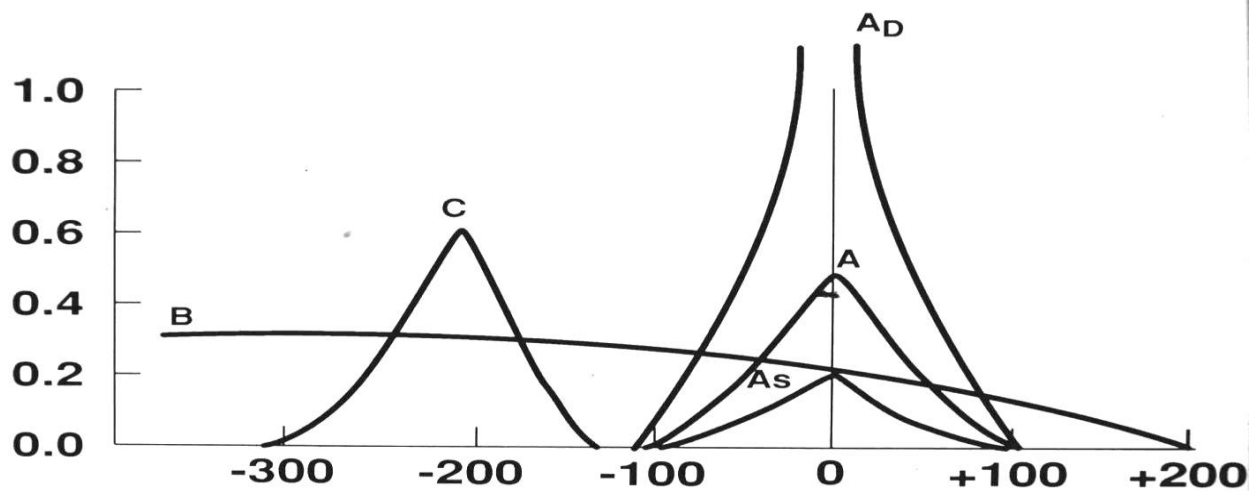
✓ Otoscopic findings (sensitivity 85 – 93%)

- Tympanic membrane is often **dull** and **opaque** with **loss of light reflex**.
- It may appear **yellow**, **grey** or **bluish** in colour.
- **Thin leash of blood vessels** may be seen **along the handle of malleus or at the periphery of tympanic membrane** and differs from marked congestion of acute suppurative otitis media.
- Tympanic membrane may show varying degree of **retraction**.
- may appear full or slightly bulging in its posterior part due to effusion.
- **Fluid level** and **air bubbles** may be seen when fluid is thin and tympanic membrane transparent
- Mobility of the tympanic membrane is restricted.



✓ Hearing Tests

1. **Tuning fork tests** show conductive hearing loss.
2. **Audiometry**. (sensitivity 92 %)
 - ✓ **conductive hearing loss of 20-30 dB** (rarely exceeds 40 dB).
 - ✓ Sometimes, there is associated sensorineural hearing loss due to fluid pressing on the round window membrane.
 - ✓ This disappears with evacuation of fluid.
3. **Impedance audiometry (Tympanometry)** (sensitivity 96 %)
 - ✓ It is an objective test useful in infants and children.
 - ✓ Presence of fluid is indicated by reduced compliance and **flat curve with a shift to negative side**.



4. Effusion is the main cause of failed newborn OAE
5. **ABR** is excellent methods for testing children who do not cooperate with behavioral hearing evaluation because of very young age or developmental delay

✓ Radiology

- ✓ X ray Skull Lateral View
 - Adenoid Hyperplasia
- ✓ Xray Mastoid Schuller's View
 - Clouding
- ✓ MRI
 - Absence of fluid does not imply an absence of OME, as one-third of patients in MRI study had fluid in mastoid, but not in the mesotympanum (Kew et al)

✓ Nasopharynx evaluation

❖ Treatment

- The aim of treatment is **removal of fluid** and **prevention of its recurrence**.
 - MEDICAL
 - SURICAL

A. Medical

- ✓ mainly **observation** until fluid resolved, or until hearing compromised For children **not** at risk for speech and language or learning disabilities
- ✓ For children not at risk & asymptomatic , examination at 3- to 6-month intervals is recommended until
 - fluid has resolved
 - hearing loss or language or learning delays are identified;
 - structural abnormalities of the eardrum are suspected
- ✓ Who at risk?

Table 3. Risk factors for developmental difficulties*

Permanent hearing loss independent of otitis media with effusion
Suspected or diagnosed speech and language delay or disorder
Autism-spectrum disorder and other pervasive developmental disorders
Syndromes (eg, Down) or craniofacial disorders that include cognitive, speech, and language delays
Blindness or uncorrectable visual impairment
Cleft palate, with or without associated syndrome
Developmental delay

*Sensory, physical cognitive, or behavioral factors that place children who have otitis media with effusion at increased risk for developmental difficulties (delay or disorder).

1. Decongestants & Antihistamin , intranasal steroid

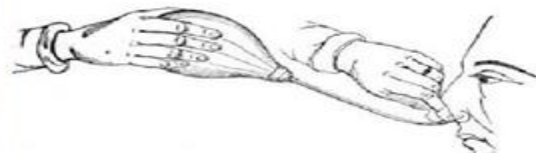
- popular treatment for OME, but In studies of OME at the Children's Hospital of Pittsburgh, **no efficacy** was found for an oral decongestant-antihistamine combination given either alone or with an antimicrobial agent
- randomized doubleblind, placebo-controlled trial of intranasal mometasone furoate spray in 217 children 4 to 11 years of age with bilateral OME found **no efficacy** for clearance of effusion

2. Antibiotics

- antibiotics are not recommended for routine treatment of OME, due to
 - lack of long-term efficacy" most pt had recurrence "
 - high spontaneous cure rate
 - overuse of antibiotics
- although effusions were thought to be sterile, studies showed that specimens from asymptomatic children with MEE contained bacteria
- Mandel and colleagues reported the results of their doubleblind, randomized trial in which 518 children with OME of varying durations were divided into three treatment groups
 - amoxicillin (40 mg/kg/day) for 14 days plus a decongestantantihistamine combination for 28 days
 - amoxicillin for 14 days plus placebo for decongestant-antihistamine for 28 days,
 - placebo for both amoxicillin and decongestant/ antihistamine.
- 4 weeks, the rate of resolution of MEE was twice as high in those treated with amoxicillin with or without a decongestant-antihistamine agent31.6%,, as those who received placebo 14.1%
- decongestant-antihistamine made no difference
- Recurrence of effusion occurred in most subjects within 3 months after completion of treatment
- Other study show all antibiotics had the same efficacy , none has been clearly shown to have any long-term advantage over the others
- Antimicrobials may be considered in some cases of parental refuse to surgery
 - 10-14 day course
 - Unlikely to provide long term benefit
 - Multiple courses not recommended (only one course)

3. 4. Middle ear aeration (Autoinflation.)

- Patient should repeatedly perform **Valsalva manoeuvre**.
- This helps to ventilate middle ear and promote drainage of fluid.
- Children can be given chewing gum to encourage repeated swallowing which opens the tube.
- Sometimes, politzerisation or eustachian tube catheterisation has to be done ??
- Autoinflation by devices is not recommended for routine use at this time

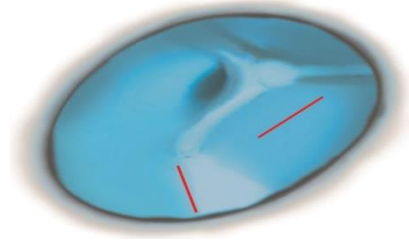


B. Surgical

- When fluid persists > 3/12 or if HL,,, language delay suspected
- When fluid is thick and medical treatment alone does not help, fluid must be surgically removed.

1. Myringotomy and aspiration of fluid (Tympanocentesis)

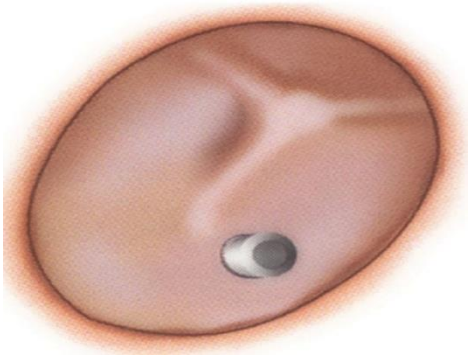
- ✓ Myringotomy alone not recommended for chronic OME
- ✓ An incision is made in tympanic membrane and fluid aspirated with suction.
- ✓ Thick mucus may require installation of saline or a mucolytic agent like chymotrypsin solution to liquefy mucus before it can be aspirated.
- ✓ Sometimes, two incisions are made in the tympanic membrane, one in the antero-inferior and the other in antero-superior quadrant, to aspirate thick "beer-can" principle
- ✓ The mean time to closure of the perforation is only 2 to 3 weeks



✓ **Indication** for Myringotomy

1. **Acute suppurative otitis media** in a seriously-ill , sever pain , toxic, newborn or immune deficient patient
2. Complications of acute otitis media, e.g. facial paralysis, labyrinthitis or meningitis with bulging tympanic membrane.
3. Unsatisfactory response to antibiotics drum remains full with persistent conductive deafness
4. Suspected unusual pathogen (newborns, immunodeficiency) for aspiration and culture

2. Ventelation tube insertion

- (improves hearing by 12 dB)
 - If myringotomy and aspiration combined with medical measures has not helped and fluid recurs (malfunctioning eustachian tube), a grommet is inserted to provide continued aeration of middle ear.
 - It is left in place for weeks or months or till it is spontaneously extruded.
- 
- Grommet Stay up to 6 mths to 1 year
 - T tubes stay upto 1-2 yrs
 - The time to extrusion is a function of :-
 - Size
 - shape of the medial flange
 - absence of a lateral flange
 - material of the tube
 - Materials for tubes
 - Silicone
 - Teflon
 - Stainless steel
 - Titanium
 - Gold
 - T-tubes recommended for :-
 - Adult & older children with persistent problems due to poor eustachian tube function.
 - for atrophic TM after going through multiple sets of tympanostomy tubes, because a regular grommet tube may be very quickly extruded.



- **Indication.**

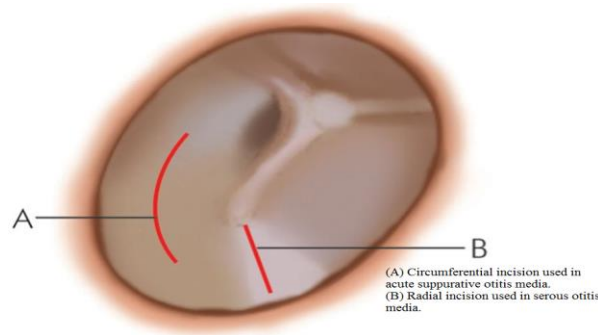
- Recurrent AOM (>3 in 6 month , >4 in 1 year, with failed medical management)
(OPTION)
- OME
 - bilateral >3 month or unilateral >6 month
 - earlier when >40 db CHL
 - speech/language delay
 - severe retraction pocket,
 - disequilibrium/vertigo, or tinnitus present
- Eustachian tube dysfunction with autophony, disequilibrium or
 - vertigo, tinnitus, or Atelectatic TM/severe retraction pocket, unrelieved by medical management
- Patulous eustachian tube
- Barotitis media(Aero-otitis media) /Hyperbaric oxygen therapy
- Suspected unusual pathogen
- Suppurative complication, present or suspected
- Unsatisfactory response to antibiotics

- **Steps of Operation**

1. Ear canal is cleaned of wax and debris.
2. Operation is ideally performed under operating microscope using a sharp myringotome and a good suction apparatus.
3. In acute suppurative otitis media, a circumferential incision is made in the **posteroinferior quadrant** of tympanic membrane, midway between handle of malleus and tympanic annulus, avoiding injury to incudostapedial joint
4. In serous otitis media, a small radial incision is given in the **posteroinferior , anteroinferior or anterosuperior "controversy regarding the best place"** quadrant and all the effusion sucked out "**anterosuperior quadrant is associated with a longer clinical tube life; however, a persistent perforation I that area is somewhat more difficult to repair "**

- A. Pitfalls of Myringotomy**

1. When tympanic membrane is **thick** (incision may remain only in the superficial layers of drumhead without cutting through its entire thickness)
2. Incision in the posterior meatal wall. (This may happen when distinction between drum-head and posterior meatal wall is lost, when both are inflamed.)



✓ **Complications**

• **Intra op**

- Displacement into middle ear
- Damage to **incudostapedial joint or stapes**
- Injury to jugular bulb with profuse bleeding, if jugular bulb is high and floor of the middle ear dehiscent.
- Injury to external ear

• **Early post op**

- Blockage of tube by blood
- Granulation around tube "act as a foreign body"
- Ear infection
- Otorrhea (3.5% rate of persistent drainage)
 - If untreated, **acute** otorrhea can develop into CSOM
 - ototopical agents such as ofloxacin and ciprofloxacin-dexamethasone " ciprodex" are effective
 - child who has severe systemic symptoms, a systemic antibiotic
 - If the drainage does not resolve in 7 to 10 days, suctioning and sent to culture
 - If treatment fail to produce improvement and the organisms are not sensitive to oral antibiotics, then:-
 1. intravenous antibiotics
 2. removal of the tube(s)
 3. rarely a simple mastoidectomy should be considered.
 - In older children with **recurrent** episodes of otorrhea, removal of the tubes is the treatment of choice
- Early extrusion "3.9%"

• **Late post op**

- Permanent perforation "3%"
- Tympanosclerosis "40%"
- TM atrophy & retraction "67%"
- Cholesteatoma "0.7%, for short-term tubes, 1.4% for long-term tubes"

✓ **Post-operative Care**

- antimicrobial drops educe early postoperative otorrhea and tube blockage
- Drum incisions usually heal rapidly. No water should be permitted to enter the ear canal for at least **2 week for margnotomy**
- if a grommet has been inserted, entry of water is prevented so long as grommet is in position.
 - Several studies have been published, including two meta-analyses demonstrating no increase in episodes of otorrhea in patients with tympanostomy tubes not using water precaution
 - water precautions may be prudent for some children such as:-
 - ✓ those with recurrent episodes of otorrhea, particularly with *Pseudomonas* or *S. aureus*,
 - ✓ and those with risk factors for infections and complications.
 - ✓ heavily contaminated water (lakes) or nonchlorinated swimming pools, for deep diving, dunking the head in the bathtub with soapy water
 - ✓ children who experience ear discomfort during swimming.
- follow-up visit a few weeks after the surgery for
 - an otoscopic examination to assess the status of the tympanostomy tube
 - repeat hearing evaluation postoperatively
- F/U every 6 months to assess the status of the tubes and the TM.

- ✓ **Contraindications** :- intratympanic glomus tumour. Myringotomy in these cases can cause profuse bleeding.

3. Tympanotomy or cortical mastoidectomy

- ✓ It is sometimes required for removal of loculated thick fluid or other associated pathology such as cholesterol granuloma.

4. Surgical treatment of causative factor

- ✓ Adenoidectomy, tonsillectomy may be required. This is usually done at the time of myringotomy.
- ✓ Adenoidectomy is **not recommended** for the initial surgical treatment of otitis media **unless nasal obstruction** is present
- ✓ **recommended for** children in whom a repeat surgical procedure is needed

❖ **Sequelae of Chronic Secretory Otitis Media**

1. Atrophic tympanic membrane and atelectasis of the middle ear

- In prolonged effusions, **fibrous layer** of tympanic membrane dissolution becomes **thin and atrophic and retracts** into the middle ear.

2. Ossicular necrosis

- Most commonly, **long process of incus gets necrosed**.
- Sometimes, **stapes superstructure also gets necrosed**.
- This increases the conductive hearing loss to more than 50 dB.

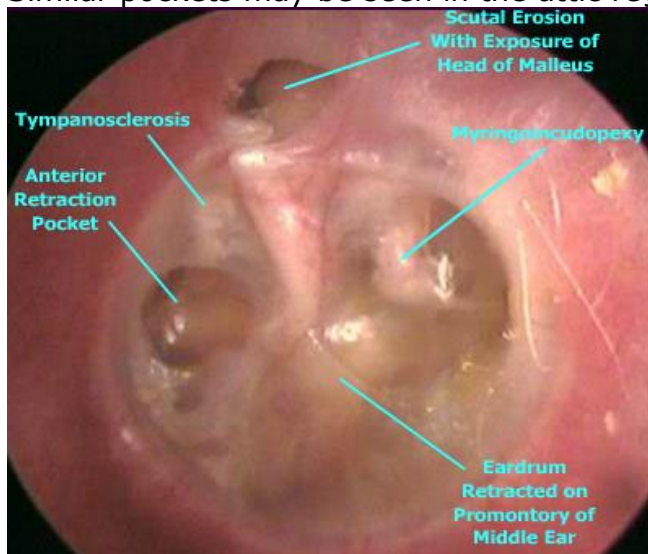
3. Tympanosclerosis

- Hyalinised collagen with chalky deposits may be seen in
 - **tympanic membrane,**
 - **around the ossicles or their joints**
- leading to their fixation.



4. Retraction pockets and cholesteatoma

- Thin atrophic part of pars tensa may get invaginated to form retraction pockets or cholesteatoma.
- Similar pockets may be seen in the attic region.



5. Cholesterol granuloma

- This is due to stasis of secretions in middle ear and mastoid.

Clinical Practice Guideline: Otitis Media with Effusion

Clinical Practice Guideline

Clinical Practice Guideline: Otitis Media with Effusion (Update)

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SAGE

- ✓ Applicable to all children ages 2 months to 12 years with or without disability

(1) Pneumatic otoscopy

- A. The clinician should document the presence of middle ear effusion with pneumatic otoscopy when diagnosing otitis media with effusion (OME) in a child. (Strong recommendation)
- B. Pneumatic otoscopy The clinician should perform pneumatic otoscopy to assess for OME in a child with otalgia, hearing loss, or both. (Strong recommendation)

(2) Tympanometry

Clinicians should obtain tympanometry in children with suspected OME for whom the diagnosis is uncertain after performing (or attempting) pneumatic otoscopy.
(Strong recommendation)

(3) Failed newborn hearing screen

Clinicians should document in the medical record counseling of parents of infants with OME who fail a newborn hearing screen regarding the importance of follow-up to ensure that hearing is normal when OME resolves and to exclude an underlying sensorineural hearing loss.
(Recommendation)

(4) Children at-risk

A. Identifying at-risk children

Clinicians should determine if a child with OME is at increased risk for speech, language, or learning problems from middle ear effusion because of baseline sensory, physical, cognitive, or behavioral factors

(Recommendation)

B. Evaluating at-risk children

Clinicians should evaluate at-risk children for OME at the time of diagnosis of an at-risk condition and at 12 to 18 mo of age (if diagnosed as being at risk prior to this time)

(Recommendation)

Table 3. Risk Factors for Developmental Difficulties in Children with Otitis Media with Effusion.^a

Permanent hearing loss independent of otitis media with effusion
Suspected or confirmed speech and language delay or disorder
Autism spectrum disorder and other pervasive developmental disorders
Syndromes (eg, Down) or craniofacial disorders that include cognitive, speech, or language delays
Blindness or uncorrectable visual impairment
Cleft palate, with or without associated syndrome
Developmental delay

(5) Screening healthy children

Clinicians should not routinely screen children for OME who are not at risk and do not have symptoms that may be attributable to OME, such as hearing difficulties, balance (vestibular) problems, poor school performance, behavioral problems, or ear discomfort.

(Recommendation (against))

(6) Patient education

Clinicians should educate families of children with OME regarding the natural history of OME, need for follow-up, and the possible sequelae.

(Recommendation)

(7) Watchful waiting

Clinicians should **manage** the child with OME who is not at risk with **watchful waiting for 3 mo from the date of effusion onset** (if known)
or **3 mo from the date of diagnosis (if onset is unknown)**
(Strong recommendation)

- Likelihood of resolution determined by cause and duration of effusion
 - 75-90% of episodes following AOM resolve by 3 months
 - 55% of children newly diagnosed with OME with a flat tympanogram will *improvement (not resolution)* change to a non-type B tympanogram within 3 months of onset. One third relapse in next 3 months
 - 25% of newly detected OME of unknown duration in children age 2-4 years *resolves* by 3 months based on tympanogram
 - Resolution rates may be higher for infant and young children
 - Documented bilateral OME of 3 months duration or longer resolves in 30% of children 2 or older after 6-12 months observation with only marginal benefits if observed longer

(8) Medication

A. Steroids

Clinicians should recommend **against** using intranasal steroids or systemic steroids for treating OME.

Strong recommendation (against)

B. Antibiotics

Clinicians should recommend **against** using systemic antibiotics for treating OME.

(Strong recommendation (against))

C. Antihistamines or decongestants

Clinicians should recommend **against** using antihistamines, decongestants, or both for treating OME.

(Strong recommendation (against))

- ✓ No benefit for antihistamines and decongestants vs. placebo
- ✓ Long-term benefits of antimicrobials unproven despite modest short-term benefit for 2-8 weeks in randomized trials.
- ✓ later metaanalysis show a short-term benefit for oral steroid plus antimicrobial versus antimicrobial alone in 1 of 3 children treated
- ✓ Benefits of abx become non-significant within 2 weeks of stopping the medication
- ✓ Adverse effects of antimicrobials are significant including rash, vomiting, diarrhea, allergic reaction, alteration of nasopharyngeal flora, bacterial resistance, and cost.
- ✓ Side effects of oral steroids: weight gain, adrenal suppression, avascular necrosis of femoral head
- Antimicrobials with or without steroids may be considered in some cases of parental refuse to surgery
 - 10-14 day course
 - Unlikely to provide long term benefit
 - Multiple courses not recommended

(9) Hearing test

Clinicians should obtain an age-appropriate hearing **test if OME persists for > 3 mo or for OME of any duration in an at-risk child.**

(Recommendation)

(10) Speech and language

Clinicians should counsel families of children with **bilateral OME and documented hearing loss** about the **potential impact** on speech and language development.

(Recommendation)

(11) Surveillance of chronic OME

Clinicians should reevaluate, at 3- to 6-mo intervals, children with chronic OME until the effusion is **no longer present, significant hearing loss is identified, or structural abnormalities of the eardrum or middle ear are suspected.**

(Recommendation)

- “If OME is asymptomatic and is likely to resolve spontaneously, intervention is unnecessary even if OME persists for more than 3 months”

(12) Surgery

A. children < 4 y old

Clinicians should recommend tympanostomy tubes when surgery is performed for OME in a child less than 4 years old; **adenoidectomy should not be performed unless a distinct indication** (eg, nasal obstruction, chronic adenoiditis) exists other than OME.

(Recommendation)

B. children ≥ 4 y old

Clinicians should recommend tympanostomy tubes, adenoidectomy, or both when surgery is performed for OME in a child 4 years old or older.

(Recommendation)

(13) Outcome assessment

When managing a child with OME, clinicians should document in the medical record resolution of OME, improved hearing, or improved quality of life.

(Recommendation)

❖ **AERO-OTITIS MEDIA (OTITIC BAROTRAUMA)**

- ✓ It is a non-suppurative condition resulting from failure of eustachian tube to maintain middle ear pressure at ambient atmospheric level.
- ✓ The usual cause is rapid descent during air flight, underwater diving or compression in pressure chamber

❖ **Mechanism**

- ✓ Eustachian tube allows easy and passive egress of air from middle ear to the pharynx if middle ear pressure is high.
- ✓ In the reverse situation, where nasopharyngeal air pressure is high, air cannot enter the middle ear unless tube is **actively opened** by the contraction of muscles as in **swallowing, yawning or Valsalva manoeuvre**.
- ✓ When atmospheric pressure is higher than that of middle ear by critical level of 90 mm of Hg, eustachian tube gets "locked", i.e. soft tissues of pharyngeal end of the tube are forced into its lumen.
- ✓ In the presence of eustachian tube oedema, even smaller pressure differentials cause "locking" of the tube.
- ✓ Sudden negative pressure in the middle ear causes retraction of tympanic membrane, hyperaemia and engorgement of vessels, transudation and haemorrhages.
- ✓ Sometimes, though rarely, there is rupture of labyrinthine membranes with vertigo and sensorineural hearing loss.

❖ **Clinical Features**

- ✓ **Severe earache, hearing loss and tinnitus** are common complaints.
- ✓ Vertigo is uncommon. Tympanic membrane appears retracted and congested. It may get ruptured.
- ✓ Middle ear may show air bubbles or haemorrhagic effusion.
- ✓ Hearing loss is usually conductive but sensorineural type of loss may also be seen.

❖ **Treatment**

- ✓ **The aim is to restore middle ear aeration.**
- ✓ This is done by **Valsalva manoeuvre**
- ✓ In mild cases, **decongestant nasal drops or oral nasal decongestant with antihistaminics** are helpful.
- ✓ In the presence of fluid or failure of the above methods, myringotomy may be performed to "unlock" the tube and aspirate the fluid.

❖ **Prevention**

- **Aero-otitis can be prevented by the following measures:**

1. Avoid air travel in the presence of upper respiratory infection or allergy.
2. Swallow repeatedly during descent. Sucking sweets or chewing gum is useful.
3. Do not permit sleep during descent as number of swallows normally decrease during sleep.
4. Autoinflation of the tube by Valsalva should be performed intermittently during descent.
5. Use vasoconstrictor nasal spray and a tablet of antihistaminic and systemic decongestant, half an hour before descent in persons with previous history of this episode.
6. In recurrent barotrauma, attention should be paid to [nasal polyps, septal deviation, nasal allergy and chronic sinus infections](#).

OME 2016 Guidelines

- **OME is diagnosed (or R/O) by either:**
 - Pneumatic otoscope **OR** Tympanometry
- **New child with OME and HL + NO RISK FACTORS:**
 - 1. Watchful waiting for 3M from Dx:**
 - **NO** routine use of steroid, anti-histamine or decongestant
 - Except if having concurrent AR.
 - **No** routine use of Abx.
 - **May** use Autoinflation technique.
 - 2. Persistent OME after 3M F/U:**
 - Request Hearing Test:
 - **IF Bilateral OME with HL > 20 dB:**
 - Surgery is RECOMMENDED:
 - IF Age < 4 Years:
 - **VT only**
 - Adenoidectomy (for patients with OME and Adenoid Hypertrophy)
 - IF Age > 4 Years:
 - **VT + Adenoidectomy** (Regardless Adenoid size):
 - Low rate of future revision surgery compared to VT alone (2% vs 20%).
 - **IF OME with symptoms other than HL (Vestibular problems, poor school performance, behavioral problems, ear discomfort):**
 - Surgery is OPTION.
 - **IF Unilateral OME or Bilateral OME with HL < 20 dB:**
 - Continuous observation with hearing testing every 3-6 M until either:
 - **NO more OME.**
 - **Presence of an indication for surgery:**
 1. At-Risk patient.
 2. Bilateral OME with > 20 dB.
 3. Structural abnormality of TM or ME:
 - Retraction pockets
 - Ossicular erosion
 - Areas of atelectasis or atrophy.
 - Granulations or aural polyp.

- **New child with OME and HL + RISK FACTORS:**

1. Request Hearing Test at initial visit.
2. **Surgery is RECOMMENDED without watchful observation (Same previous surgical approach).**

Table 3. Risk Factors for Developmental Difficulties in Children with Otitis Media with Effusion.^a

Permanent hearing loss independent of otitis media with effusion
Suspected or confirmed speech and language delay or disorder
Autism spectrum disorder and other pervasive developmental disorders
Syndromes (eg, Down) or craniofacial disorders that include cognitive, speech, or language delays
Blindness or uncorrectable visual impairment
Cleft palate, with or without associated syndrome
Developmental delay

^aSensory, physical, cognitive, or behavioral factors that place children who have otitis media with effusion at increased risk for developmental difficulties (delay or disorder).¹

- **New child with RECURRENT AOM WITH OME at time of evaluation:**

1. **VT is RECOMMENDED without watchful observation.**

- **New child with RECURRENT AOM WITHOUT OME at time of evaluation:**

1. **VT is NOT recommended.**

Chronic Suppurative Otitis Media (CSOM):

- Long-standing infection of a part or whole of the middle ear cleft characterised by **ear discharge** and a **permanent perforation**.
 - Persistent (>6 weeks) or recurrent drainage from infection of the middle ear and/or mastoid in the presence of a TM perforation (or ventilation tube).
- ❖ **Pathophysiology:**
- Chronically inflamed or infected middle ear space or mastoid secondary to:
 - Poor aeration (chronic eustachian tube dysfunction),
 - Chronic perforation,
 - Presence of a Cholesteatoma.
 - A perforation becomes permanent when its edges are covered by **Squamous epithelium** and it does not heal spontaneously.
 - A permanent perforation can be likened to an epithelium-lined fistulous track.

❖ **Epidemiology:**

- Incidence of CSOM is higher in Developing countries due to:
 - Poor socio-economic standards
 - Poor nutrition and lack of health education.
- Affects both sexes and all age groups.

❖ **Pathogens:**

- **Mixed infections:**
 - Gram-negative bacilli (Pseudomonas, Klebsiella, Proteus, E. coli,). **Pseudomonas (most common aerobe)**
 - Staphylococcus aureus
 - Anaerobes "anaerobic Streptococci (**most common anaerobe**) and Bacteroides fragilis "

❖ **Risk Factors:**

1. Abnormal Eustachian tube function:
 - Cleft palate
 - Down syndrome
2. Immune deficiency
3. Ciliary dysfunction:
 - Kartagener syndrome.
4. Gastroesophageal reflux

❖ **Clinical presentation:**

- Otorrhea (mucopurulent, odorous)
- TM perforation
- Inflamed middle ear mucosa
- Conductive hearing loss

❖ **Types of CSOM:**

- Clinically, it is divided into two types:

1. Tubo-tympanic TT:

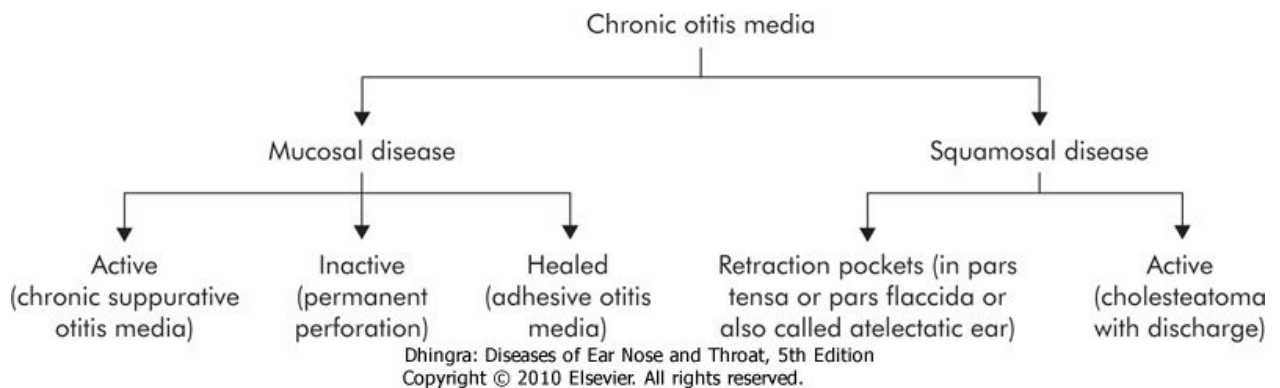
- Safe type.
- Involves Antero-inferior part of middle ear cleft (Eustachian tube and Mesotympanum).
- Central perforation.
- No risk of serious complications.

2. Attico-antral AA:

- Unsafe type
- Involves Postero-superior part of middle ear cleft (Attic, Antrum and mastoid).
- Attic or Marginal perforation.
- Associated with a bone-eroding process such as Cholesteatoma, Granulations or Osteitis.
- High risk of serious complications.

Table 11-1. Differences between atticoantral and tubotympanic type of CSOM

	Tubotympanic or safe type	Atticoantral or unsafe type
Discharge	Profuse, mucoid, odourless	Scanty, purulent, foul-smelling
Perforation	Central	Attic or marginal
Granulations	Uncommon	Common
Polyp	Pale	Red and fleshy
Cholesteatoma	Absent	Present
Complications	Rare	Common
Audiogram	Mild to moderate conductive deafness	Conductive or mixed deafness



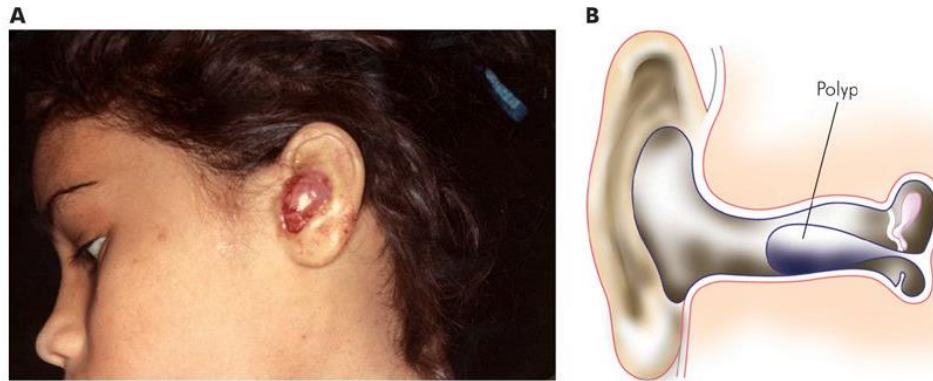
➤ **Tubotympanic CSOM:**

- Mucosal disease of middle ear cleft with no evidence of invasion of squamous epithelium.
- Active disease:
 - Perforation of pars tensa with inflammation of mucosa and mucopurulent discharge.
- Inactive disease:
 - Permanent perforation of pars tensa but middle ear mucosa is not inflamed and there is no discharge.
 - Permanent perforation implies that squamous epithelium on the external surface of pars tensa and mucosa lining its inner surface have fused across its edge.
- Healed chronic otitis media:
 - **Adhesive otitis media.**
 - Healed TM (usually by two layers), atrophic and easily retracted if there is negative pressure in the middle ear.
 - Also have patches of Tympanosclerosis in tympanic membrane, or in middle ear involving promontory, ossicles, tendons of stapedius and tensor tympanic.
 - **Fibrotic** tissue may appear in middle ear.
 - Always associated with some degree of conductive hearing loss.



- **Etiology:**
 - Starts in childhood and is therefore common in that age group.
 - 1. **Sequel of Acute otitis media:**
 - Following fever and leaving behind a large central perforation.
 - Perforation becomes permanent and permits repeated infection from the external ear.
 - Middle ear mucosa is exposed to the environment and gets sensitised to dust, pollen and other aeroallergens causing persistent otorrhoea.
 - 2. **Ascending infections via the eustachian tube:**
 - Infection from tonsils, adenoids and infected sinuses may be responsible for persistent or recurring otorrhoea.
 - Ascending infection to middle ear occur more easily in the presence of infection.
 - 3. **Persistent mucoid otorrhoea is sometimes the result of allergy to ingestants.**
- **Pathology:**
 - Disease localized to the mucosa.
 - Mostly involves **Antero-inferior** part of the middle ear cleft.
 - Processes of healing and destruction, depending on the virulence of organism and resistance of the patient.
 - Acute exacerbations are common.
- **Pathological changes seen in Tubotympanic CSOM:**
 - 1. Perforation of pars tensa:**
 - Central perforation
 - Size and position varies.
 - 2. Middle ear mucosa:**
 - Normal when disease is inactive.
 - Edematous when disease is active.
 - 3. Polyp:**
 - Smooth mass of edematous and inflamed mucosa protruded through a perforation and presents in the external canal.
 - Pale (Pink, fleshy polyp seen in Attico-antral CSOM).





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Figure 11.4 (A) Polyp in the ear canal. (B) Schematic illustration of a polyp arising from the promontory passing through the perforation and presenting in the ear canal.

4. Ossicular chain:

- Intact and mobile.
- May show some degree of necrosis (particularly long process of incus).

5. Tympanosclerosis:

- Results from chronic inflammation or trauma
- Hyalinisation and calcification of subepithelial connective tissue.
- Seen in remnants of TM or under the mucosa of middle ear.
- White chalky deposit on the promontory, ossicles, joints, tendons and oval and round windows.
- Tympanosclerotic masses may interfere with the mobility of these structures and cause CHL.

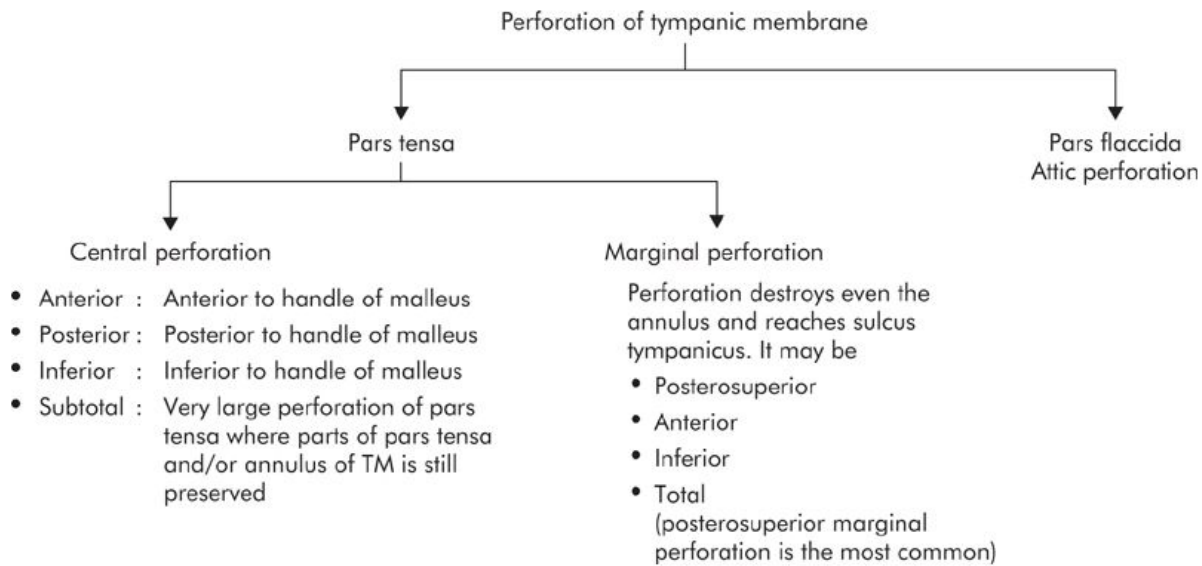
6. granulation tissue

7. Fibrosis and Adhesions:

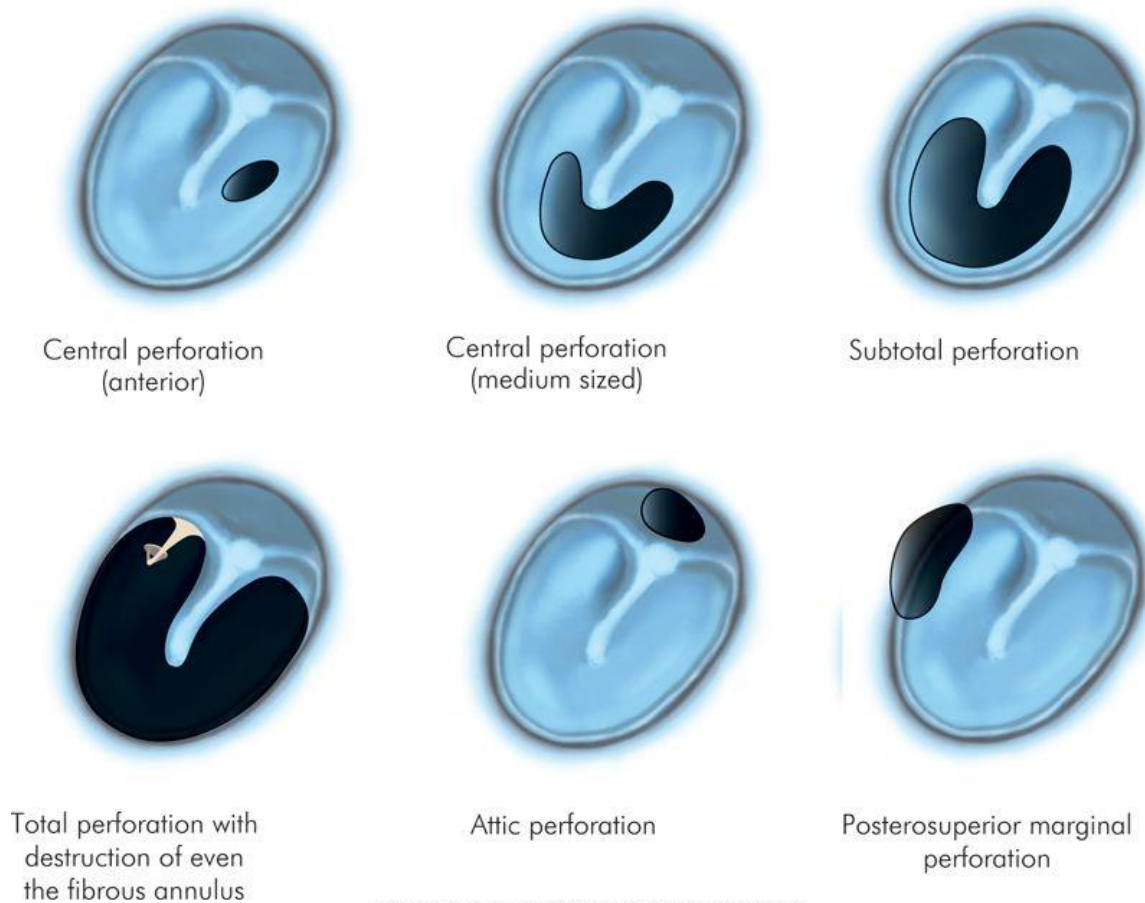
- Long-standing eustachian tube dysfunction Result of healing process ,
- May impair mobility of ossicular chain or block the eustachian tube erosion of the long process of the incus and the stapes suprastructure.



FIGURE 139-2. Middle ear atelectasis with ossicular erosion.



Note: Attic and posterosuperior marginal perforation are seen in dangerous type of CSOM and are often associated with cholesteatoma. Stratified squamous epithelium from the external auditory canal can grow into the middle ear in any type of marginal perforation by immigration and form a cholesteatoma. Therefore, all marginal perforations are considered dangerous. Central perforations are considered safe as cholesteatoma is usually not associated with them.



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- **Clinical Features:**

1. Ear discharge

- Non-offensive, mucoid or mucopurulent, constant or intermittent.
- Appears mostly at time of URTI or on accidental entry of water into the ear.

2. Hearing loss

- CHL
- Severity varies but rarely exceeds 50 dB.
- Patient reports of a paradoxical effect (hears better in the presence of discharge than when the ear is dry) due to "Round window shielding effect" produced by discharge which helps to maintain phase differential.
- In the dry ear with perforation, sound waves strike both the oval and round windows simultaneously, thus cancelling each other's effect.
- In long standing cases, cochlea may suffer damage due to absorption of toxins from the oval and round windows and hearing loss becomes Mixed type.

3. Perforation

- Always central, it may lie anterior, posterior or inferior to the handle of malleus.
- Small, medium or large or extending up to the annulus, i.e. subtotal.

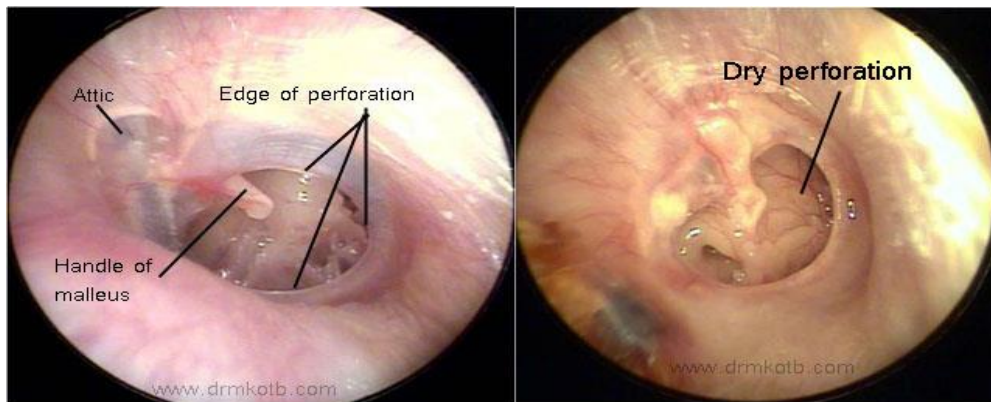
4. Middle ear mucosa

- Seen through large perforation.
- Normally, it is pale pink and moist.
- When inflamed it looks red, oedematous and swollen.
- Polyp may be seen.

- **Investigations:**

1. Examination under Microscope:

- Essential in every case
- Provides useful information regarding :-
 - **Granulations**
 - Growth of squamous epithelium from the edges of perforation
 - Status of ossicular chain
 - Tympanosclerosis
 - Adhesions.
- An ear which appears dry may show hidden discharge under the microscope.
- Rarely, cholesteatoma may co-exist with a central perforation and can be seen under a microscope.



Chronic suppurative otitis media -
safe type = tubo-tympanic

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2. Audiogram:

- Assess degree of hearing loss and its type.
- Usually, CHL but a sensorineural element may be present.
- Decreased in the low frequencies in a small perforation and in the high and low frequencies in a large perforation.

3. Culture and sensitivity of ear discharge:

- To select proper antibiotic ear drops.

4. CT scan temporal bone:

- Mastoid is usually sclerotic but may be pneumatised with clouding of air cells.
- **No evidence of bone destruction.**
- Presence of bone destruction is feature of Attico-antral disease.

- **Treatment of Tubotympanic CSOM:**
- Aim of the management:
 - o Control infection
 - o Eliminate ear discharge
 - o Correct the hearing loss by surgical means.

1. Aural toilet:

- o Remove all discharge and debris from the ear.
- o Done by dry mopping with absorbent cotton buds, suction clearance under microscope or irrigation (not forceful syringing) with sterile normal saline.
- o Ear must be dried after irrigation.

2. Ear drops:

- o Ofloxacin ,Neomycin, polymyxin, chloromycetin or gentamicin are used.
- o Combined with steroids which have local anti-inflammatory effect.
- o To use ear drops, patient lies down with the diseased ear up, antibiotic drops are instilled and then intermittent pressure applied on the tragus for antibiotic solution to reach the middle ear.
- o This should be done three or four times a day.
- o Acid pH helps to eliminate pseudomonas infection, and irrigations with 1.5% acetic acid are useful.
- o Care should be taken as ear drops are likely to cause maceration of canal skin, local allergy, growth of fungus or resistance of organisms.
- o Some ear drops are potentially ototoxic ' **aminoglycoside antibiotics and propylene glycol**'

3. Systemic antibiotics:

- o Role of systemic antibiotics in the treatment of CSOM is limited.
- o Useful in **Acute exacerbation** of chronically infected ear "refractory cases"
- o When specific pathogens are found on culture

4. Precautions:

- o Keep water out of the ear during bathing, swimming and hair wash.
- o Rubber inserts can be used.
- o Hard nose-blowing can also push the infection from nasopharynx to middle ear and should be avoided.

5. Treatment of contributory causes:

- o Treat concomitantly infected tonsils, adenoids, sinuses and nasal allergy.

6. Surgical treatment:

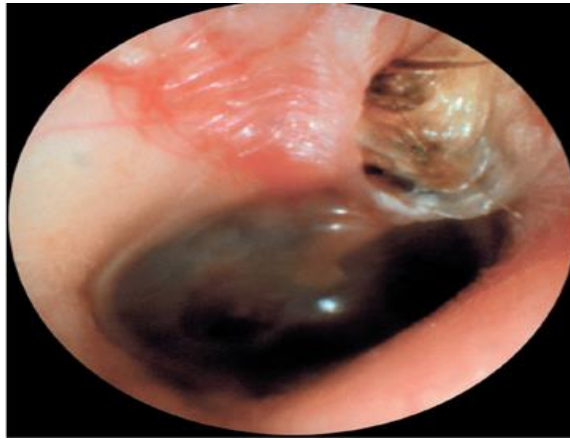
- Aural polyp or granulations, if present, should be removed before local treatment with antibiotics.
- It will facilitate ear toilet and permit ear drops to be used effectively.
- Should NEVER be avulsed as it may be arising from the stapes, facial nerve or horizontal canal and thus lead to facial paralysis or labyrinthitis.

7. Reconstructive surgery:

- Once ear is dry, [Tympanoplasty with or without ossicular reconstruction](#) can be done to restore hearing.
- Closure of perforation will also check repeated infection from the external canal.
- Ideally, an ear with a tympanic membrane perforation should be free from infection for 3 months before tympanoplasty.

➤ **Atticoantral CSOM:**

- Squamosal disease of middle ear.
- Active disease:
 - Presence of cholesteatoma of posterosuperior region of pars tensa or in the pars flaccida.
 - Erodes bone, forms granulation tissue and has purulent offensive discharge.
- Inactive disease:
 - Atelactatic ear.
 - Retraction pockets in pars tensa (usually the posterosuperior region) or pars flaccida.
 - No discharge but there is a possibility of squamous debris in retraction pockets to become infected and start discharging.
 - Some retraction pockets are shallow and self cleansing.



- Atticoantral diseases is associated with the following pathological processes:
 - 1. Cholesteatoma**
 - 2. Osteitis and granulation tissue**
 - Osteitis involves outer attic wall and Postero-superior margin of tympanic ring.
 - Granulation tissue surrounds the area of osteitis and may even fill the attic, antrum, posterior tympanum and mastoid can lead to bone erosion "**Collagenase**".
 - A fleshy red polypus may be seen filling the meatus.
 - 3. Aural polyp**
 - Infected cholesteatoma sometimes manifests as an "aural polyp".
 - These "polyps" are actually granulation tissue at the junction between an eroding cholesteatoma and bone.
 - The presence of an aural polyp in a chronically infected ear should be considered to be a cholesteatoma until proven otherwise

4. Ossicular necrosis:

- Common in Atticoantral disease.
- Destruction may be limited to long process of incus or may also involve stapes superstructure, handle of malleus or the entire ossicular chain.
- Hearing loss is always greater than in disease of tubotympanic type.
- Cholesteatoma may bridges the gap caused by the destroyed ossicles, and hearing loss is not apparent ([cholesteatoma hearer](#)).

5. Cholesterol granuloma:

- Mass of granulation tissue with [foreign body giant cells](#) surrounding the [cholesterol crystals](#).
- A reaction to long-standing retention of secretions or haemorrhage, and may or may not co-exist with [cholesteatoma](#).
- When present in the mesotympanum, behind an intact drum, the TM appears [blue](#).



- **Symptoms**

1. **Ear discharge:**

- Usually scanty, patient may not even be aware of it.
- Always foul-smelling due to bone destruction.
- Total cessation of discharge from an ear which has been active till recently should be viewed seriously, as perforation in these cases might be sealed by crusted discharge, inflammatory mucosa or a polyp, obstructing the free flow of discharge.
- Pus, in these cases, may find its way internally and cause complications.

2. **Hearing loss:**

- Hearing is normal when ossicular chain is intact or when cholesteatoma, having destroyed the ossicles, bridges the gap caused by destroyed ossicles (cholesteatoma hearer).
- Mostly CHL
- Evidence suggests that sensorineural hearing loss (SNHL) can result from chronic otitis media with or without cholesteatoma

3. **Bleeding:**

- From granulations or the polyp when cleaning the ear.

- **Signs:**

1. **Perforation:**

- Either Attic or Postero-superior marginal type.
- Small attic perforation may be missed due to presence of a small amount of crusted discharge.
- Sometimes, the area of perforation is masked by a small granuloma.

2. **Retraction pocket:**

- Invagination of tympanic membrane is seen in the attic or Postero-superior area of pars tensa.
- Degree of retraction and invagination varies.
- In early stages, pocket is shallow and self-cleansing.
- Later, pocket is deep and accumulates keratin mass and gets infected.

3. **Cholesteatoma:**

- **Pearly-white flakes** of cholesteatoma can be sucked from the retraction pockets.

- **Investigations:**

1. Examination under microscope:

- All patients of chronic middle early disease should be examined under microscope.
- Suction clearance and examination under Microscope forms an important part of clinical examination of any type of CSOM.
- May reveal & assess
 - Cholesteatoma "site and extent"
 - Evidence of bone destruction
 - Granuloma
 - Condition of ossicles
 - Pockets of discharge.

2. Tuning fork tests and audiogram:

- Pre-operative assessment and to confirm degree and type of HL.

3. CT scan temporal bone:

- Indicate **extent of bone destruction** and **degree of mastoid pneumatisation**.
- Useful to indicate a **low-lying dura or an anteposed sigmoid sinus** when operation is being contemplated on a sclerotic mastoid.
- Cholesteatoma causes destruction in the area of attic and antrum (key area).

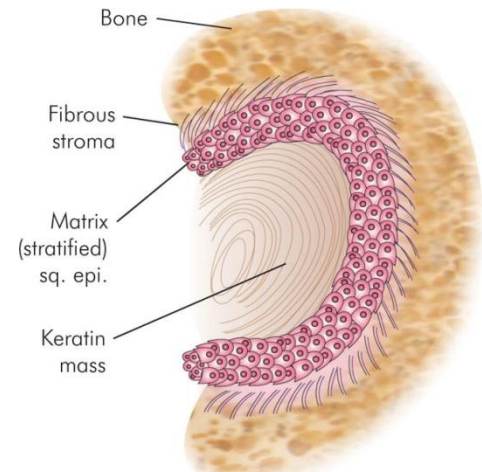
4. Culture and sensitivity of ear discharge:

- Helps to select proper antibiotic for local or systemic use

- **Complications of COM with cholesteatoma in order of frequency:**
 1. HL "conductive, sensorineural, mixed type" – Ossicular chain disruption in up to 30%
 2. Inner Ear Fistula – 10% of cases, mainly HSCC, rarely Cochlea
 3. Extradural or Perisinus Abscess
 4. Labyrinthitis – Serous or Suppurative
 5. Facial Nerve Paralysis – Acute (infection) or Chronic (slow expansion)
 6. Meningitis secondary to Tegmen Erosion
 7. Subdural or Intraparenchymal Brain Abscess
 8. Sigmoid Sinus Thrombosis/Phlebitis
 9. Subperiosteal Abscess/Bezold's Abscess due to erosion of Mastoid Cortex
 10. Recurrent Cholesteatoma
- **Features Indicating Complications in CSOM**
 1. Pain:
 - Uncommon in uncomplicated CSOM.
 - Considered serious as it may indicate extradural, perisinus or brain abscess.
 - Otitis externa associated with a discharging ear.
 2. Vertigo:
 - Erosion of lateral semicircular canal which may progress to labyrinthitis or meningitis.
 - Fistula test should be performed in all cases.
 3. Persistent headache:
 - Intracranial complication.
 4. Facial weakness:
 - Erosion of facial canal.
 5. A listless child refusing to take feeds and easily going to sleep:
 - Extradural abscess.
 6. Fever, nausea and vomiting:
 - Intracranial infection.
 7. Irritability and neck rigidity:
 - Meningitis.
 8. Diplopia:
 - Gradenigo's syndrome.
 9. Ataxia:
 - Labyrinthitis or cerebellar abscess.
 10. Abscess around the ear:
 - Mastoiditis.

❖ Cholestatoma:

- Cystlike, “**epidermal inclusion cysts**” expansile lesions of the temporal bone lined by **stratified squamous epithelium** “**matrix**” that contain desquamated keratin.
- Presence of keratinising squamous epithelium in the middle ear or mastoid (Skin in the wrong place) .
- Normally, middle ear cleft is lined by different types of epithelium in different regions.
 - Ciliated columnar in the anterior and inferior part.
 - Cuboidal in the middle part
 - Pavement-like in the attic.
 - No where lined by keratinising squamous epithelium.
- The term cholesteatoma is a misnomer:
 - It neither contains cholesterol crystals nor is it a tumour to merit the suffix "oma".
 - The term has been retained because of its wider usage.
 - It has also been named **Epidermosis** or **Keratoma**.
- Cholesteatoma consists of two parts:
 1. Matrix:
 - Made up of keratinising squamous epithelium resting on a thin stroma of fibrous tissues.
 2. Keratin mass:
 - Central **white mass** consisting of keratin debris produced by the matrix.



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- [Pathology](#)
- Cholesteatomeas may develop anywhere within pneumatized portions of the temporal bone, with the most frequent locations being the **middle ear** and the **mastoid**.
- Matrix composed of fully differentiated squamous epithelium resting on connective tissue
- Deep layers: Down-growth into connective tissue
- Granulation tissue [contacts bone](#); elaborates **collagenase** causing:-
 - **bone destruction**
 - Infection
 - otorrhea,
 - hearing loss
 - facial nerve paralysis,
 - labyrinthine fistula
 - intracranial complications "epidural and subdural abscesses, parenchymal brain abscesses, meningitis, and thrombophlebitis of the dural ,venous sinuses"
- **Only cholesteatoma and TM epithelium migrate**
- In human temporal bones with chronic otitis media, cholesteatoma was observed in 36% of ears with perforations and in 4% of ears without the perforated tympanic membranes
- Cholesteatoma debris is a favourable medium for bacteria and Aspergillus.
 - Most common aerobic bacteria
 - **Pseudomonas aeruginosa.**
 - **Staphylococcus aureus**
 - Most common anaerobic microorganisms are anaerobic cocci
 - **Peptococcus/Peptostreptococcus**
 - **Bacteroides**
 - **Proteus,**
 - **Enterobacter**
 - **aerobic and anaerobic nonhemolytic streptococci,.**
 - **diphtheroid bacilli**

- Mechanisms of Bone erosion (faster in active infection):
 1. **Mechanical:** Pressure from expansile mass.
 2. **Biochemical:**
 - Bacterial (Endotoxins)
 - Granulation tissue Products (**Collagenase** and Acid hydrolase)
 - Substance related to cholesteatoma itself (Growth factors, Cytokines).
 3. **Cellular:**
 - Osteoclastic activity (Acid phosphate, Acid Proteases, **Collagenase** other protolytic enzymes)
 - Osteocytes – BMP-2, TGF beta
 - Macrophages – ILs -1, -6, -11; TNF- α , TGF- α , PGs, LTs, PTHrP, CSF-1, OPF

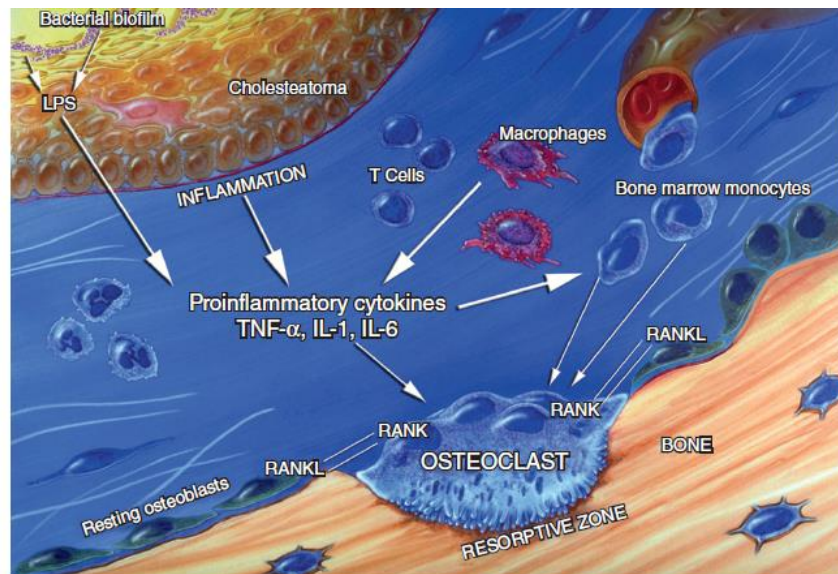


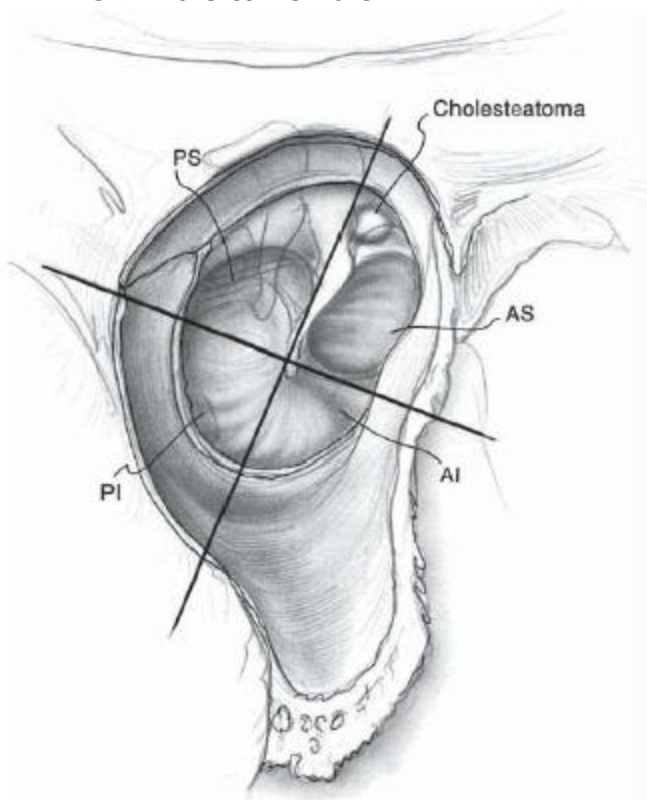
FIGURE 139-14. The bone resorption associated with cholesteatomas occurs as a result of the recruitment of osteoclasts derived from bone marrow mononuclear cells. The receptor activator of nuclear factor (NF)- κ B ligand (RANKL) on osteoblasts or T cells binds to receptor activator of NF- κ B (RANK) on monocytes, forming osteoblasts. Osteoclast activation is potentiated further by numerous proinflammatory cytokines (tumor necrosis factor- α [TNF- α], interleukins [ILs] 1 and 6) that are produced in response to factors produced by the cholesteatoma or bacterial products such as lipopolysaccharide (LPS).

- Classification of Cholesteatoma:

1. Congenital
2. Acquired, Primary.
3. Acquired, Secondary.

Congenital Cholesteatoma:

- Theory:
 - o Arises from Embryonic epithelial cells rests in the middle ear cleft or temporal bone.
 - o Failure of involution and continued growth of the Epidermoid Formation, derived from **1st branchial groove ectoderm**
 - o Found at junction of Eustachian tube orifice and middle ear near anterior tympanic annulus
 - o Normally disappears in **33rd week** of gestation
 - o Other theories:- include Ectodermal **migration**, and **Metaplasia** of the middle ear mucosa
- Occurs at three important sites and produces symptomatology depending on its location.
 - o Middle ear
 - o Petrous apex
 - o Cerebellopontine angle.
- 2/3 of cases are seen as white mass in **Anterior-superior quadrant.**
- Mean age 4.5 years.
- 3:1 Male to Female



- [Levenson Criteria for Congenital Cholesteatoma](#)
 1. White mass Medial to Intact TM
 2. Normal pars flaccida & tensa.
 3. No History of TM Perforation or Discharge.
 4. No History of Trauma or Surgery
 5. **Prior otitis media is NOT an exclusion**
- [Congenital cholesteatomas staging system 'Potsic and others'](#)
 - 1. Stage I:** Limited to one quadrant
 - 2. Stage II:** Involving multiple quadrants without ossicular involvement
 - 3. Stage III:** Ossicular involvement without mastoid extension
 - 4. Stage IV:** Mastoid involvement.
 - There is a correlation between stage and risk of residual disease
 - IV 67% risk of residual cholesteatoma after surgical resection
- It causes CHL.
- Sometimes discovered on routine examination of children or at the time of myringotomy.
- It may also spontaneously rupture through the tympanic membrane and present with a discharging ear indistinguishable from a case of chronic suppurative otitis media (CSOM).

Acquired Cholesteatoma:

- The common factor of all acquired cholesteatomas is that the keratinizing squamous epithelium has grown beyond its normal limits.

✓ **Two types of Acquired Cholesteatoma**

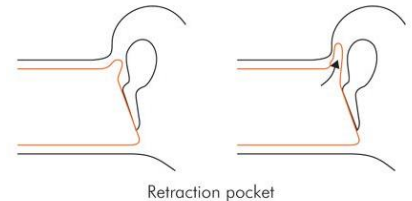
- **Primary Acquired:**
 - AKA Retraction Pocket cholesteatoma
 - NOT secondary to infection, develop within a retraction pocket
- **Secondary Acquired:**
 - Secondary to perforation from infection, surgery or trauma

- **Acquired Primary Cholesteatoma:**

- NOT associated with pre-existing perforation.

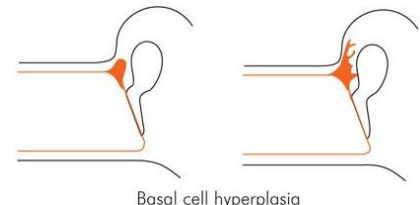
1. Invagination Theory (Retraction pocket):

- Prolonged Eustachian tube obstruction.
- Persistent Negative pressure in Middle ear.
- Poor pneumatization of Epitympanum and Attic.
- Invagination of TM from the Pars Flaccida (due to lack of fibrous layer) or **Postero-Superior** part of pars tensa in the form of Retraction pockets.
- Once a retraction pocket develops, the normal migratory pattern of the tympanic membrane epithelium is altered, enhancing the potential accumulation of **keratin**.
- As the retraction pocket deepens, desquamated keratin cannot be cleared from the recess, and a Cholesteatoma results
- **Infection** has no role in the development of cholesteatoma.



2. Basal cell hyperplasia Theory:

- Modification of invagination theory.
- Cone-like extension of the basal layer of epidermis can become invasive as a result of **infections**.
- Microcholestatoma arise as prickles cells invade the subepithelial layer through breaks in the basal lamina.
- Expanding cholesteatoma then breaks through pars flaccida forming an attic perforation.



- **Patients with cleft palate , long stand OME tend to develop Primary Acquired Cholesteatoma due to ET Dysfunction**



Primary acquired cholesteatoma

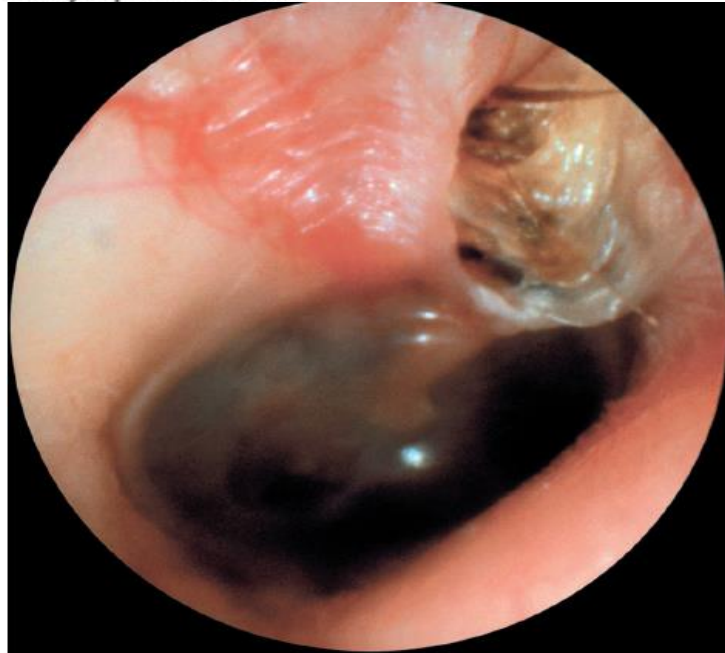
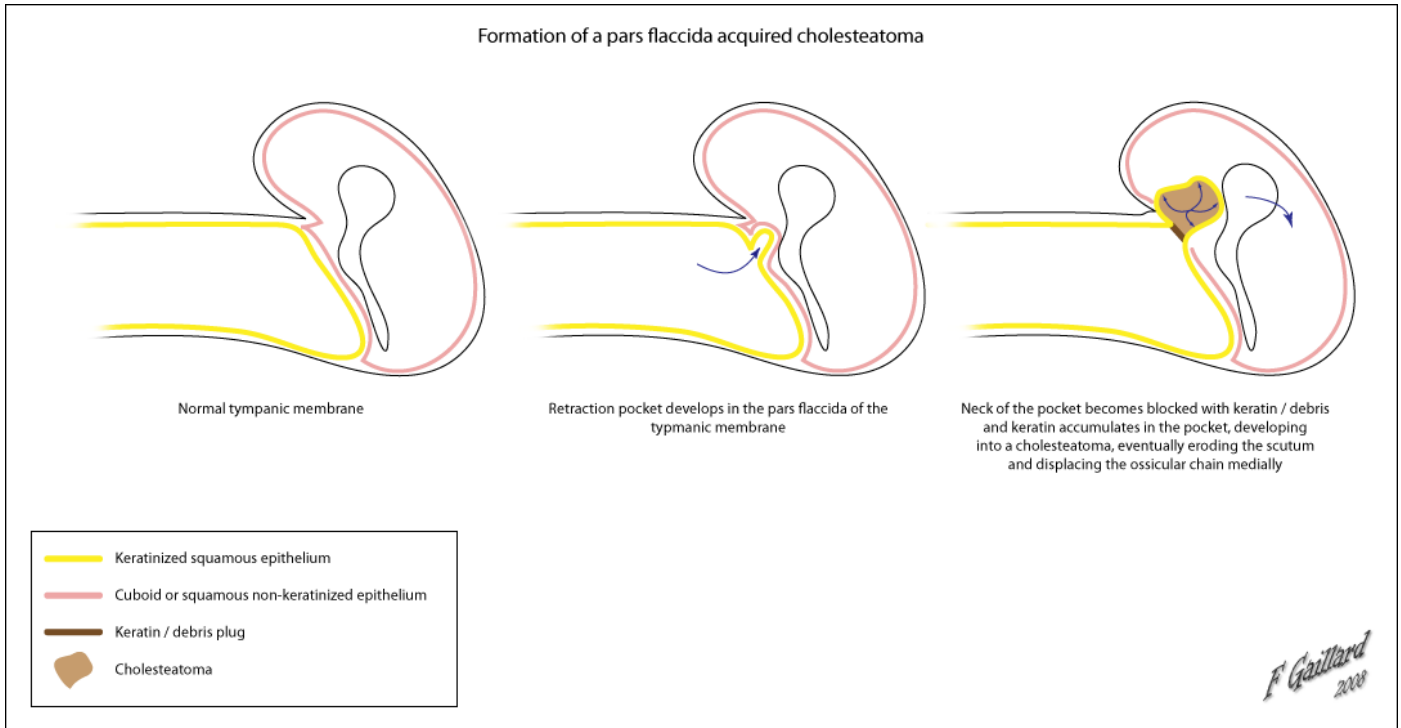


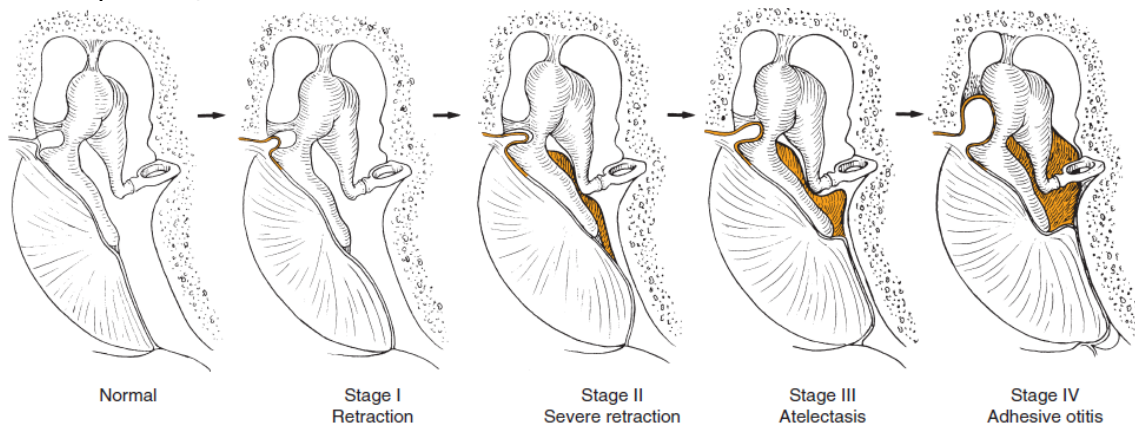
FIGURE 139-5. Primary acquired cholesteatoma in the region of the pars flaccida with scutum erosion.



✓ **Sadý & Berco stages of middle ear atelectasis**

- Stage I – Retraction
- Stage II – Severe retraction (against ISJ)
- Stage III – Atelectasis (against Promontory)
- Stage IV – Adhesive Otitis media

'Nonpneumatized mastoids may have a limited ability to buffer pressure changes and can manifest as an atelectasis, a retraction pocket, or a cholesteatoma'



Acquired Secondary Cholesteatoma:

- There is already a pre-existing perforation in pars tensa from infection, surgery or trauma

1. Epithelial invasion (Migration) Theory:

- Most accepted theory.
- Keratinized stratified squamous epithelium from the meatus or outer drum surface grows into the middle ear through a pre-existing perforation **especially of the marginal type** where part of annulus tympanicus has already been destroyed.

2. Implantation Theory:

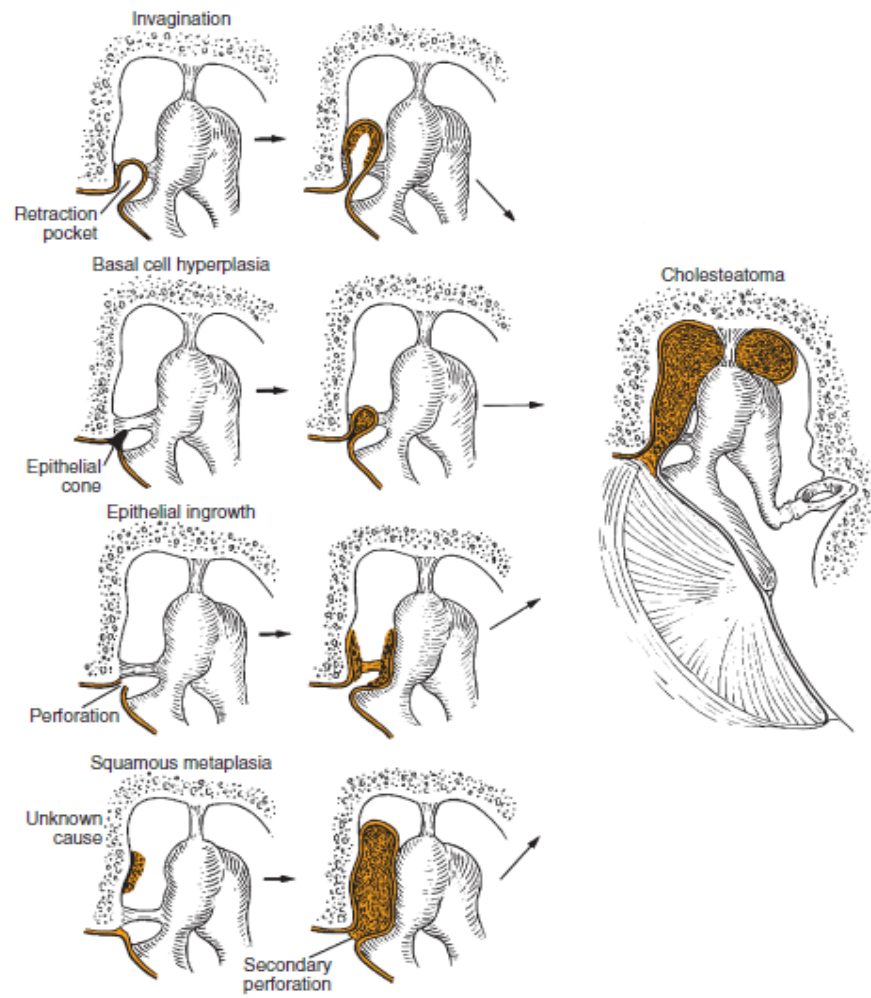
- Iatrogenic implantation of skin into TM or middle ear.
- Caused by Surgery "myringotomy for ventilating tube," FB or Blast injury
- Or arise from a perforation as a result of Acute Necrotic OM in childhood.

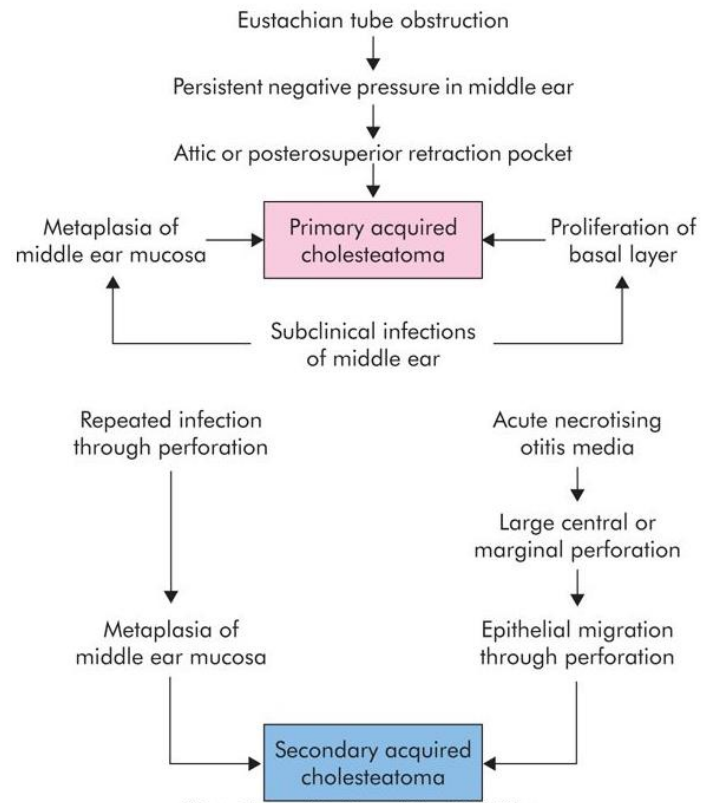
3. Metaplasia Theory:

- Transformation of columnar epithelium to keratinized stratified squamous epithelium.
- Secondary to Chronic or recurrent OM.
- Not believed to be an explanation for a significant cause of cholesteatoma formation in humans.

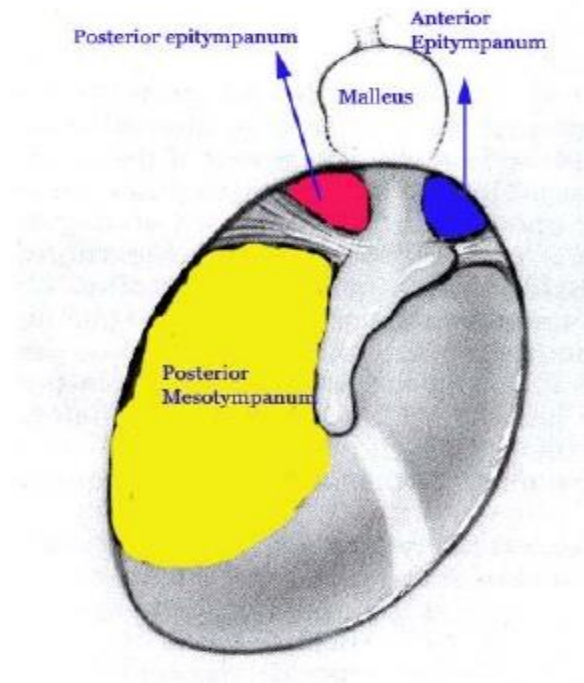


FIGURE 139-6. Cholesteatoma developing at the margin of perforation (secondary acquired cholesteatoma) with secondary infection.





- **Surgical Anatomy:**
- Cholesteatomas of the epitympanum start in **Prussak's space**.
 - o Small pocket bounded by:
 - o Laterally: Pars Flaccida.
 - o Medially: Neck of the Malleus.
 - o Superiorly: Lateral malleolar fold.
 - o Inferiorly: Lateral process of Malleus.
- **Most common locations of origin of Primary Acquired Cholesteatoma**
 1. Posterior Epitympanum. "commonest"
 2. Posterior Mesotympanum.
 3. Anterior Epitympanum.
- **(Hypotympanum is rarely involved by Cholesteatoma)**



- [Posterior Epitympanic route:](#)
- Most common spread pattern.
- Originates **in Prussak's space**, breaks out by either penetrating
 - 1) **Posterior** into Superior Incudal space and into [Aditus and Mastoid](#)
 - 2) **Inferiorly** into Posterior Pouch of von Troltsch (Between TM and post mal fold)
 - 3) **Anteriorly** into Anterior Pouch of von Troltsch

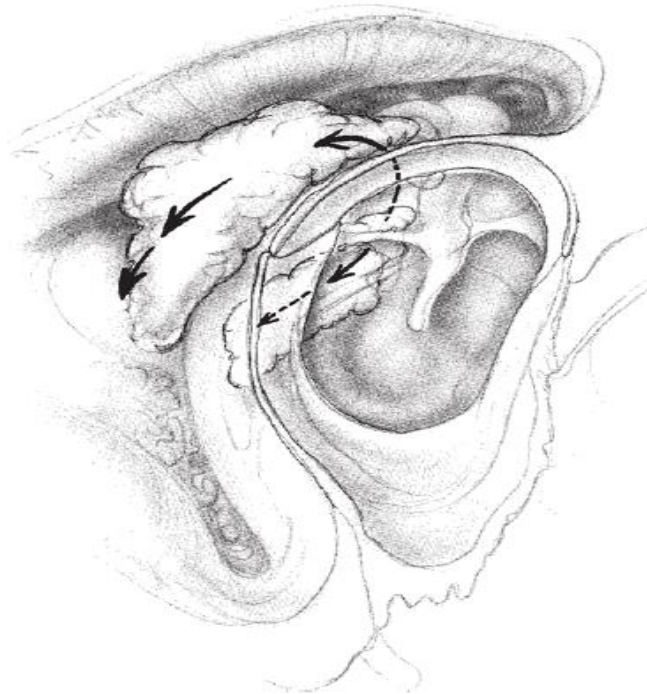
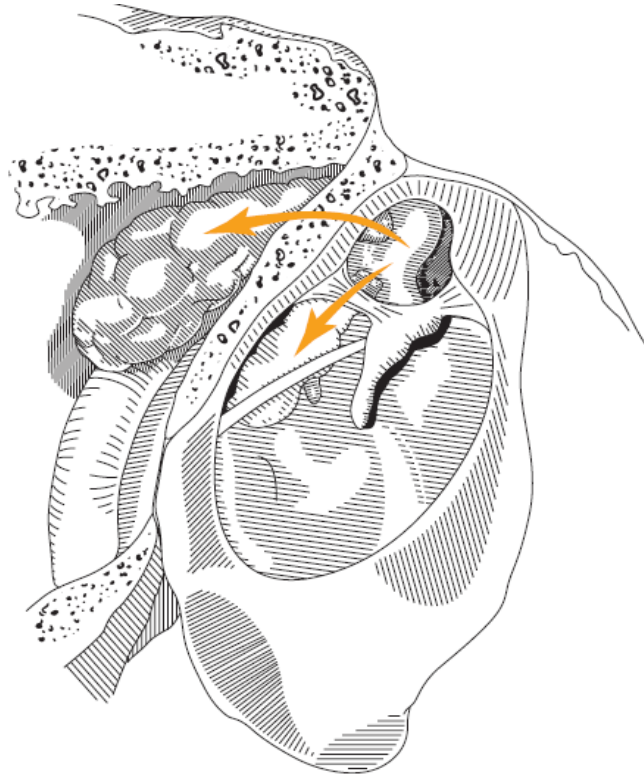


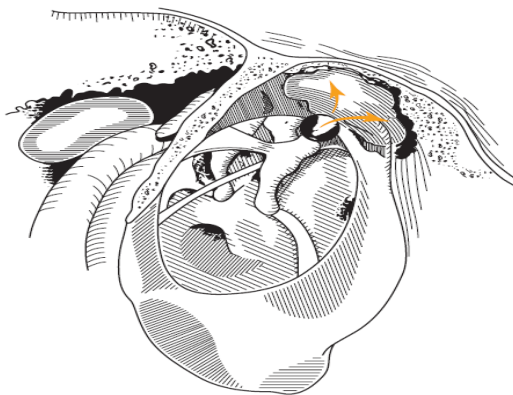
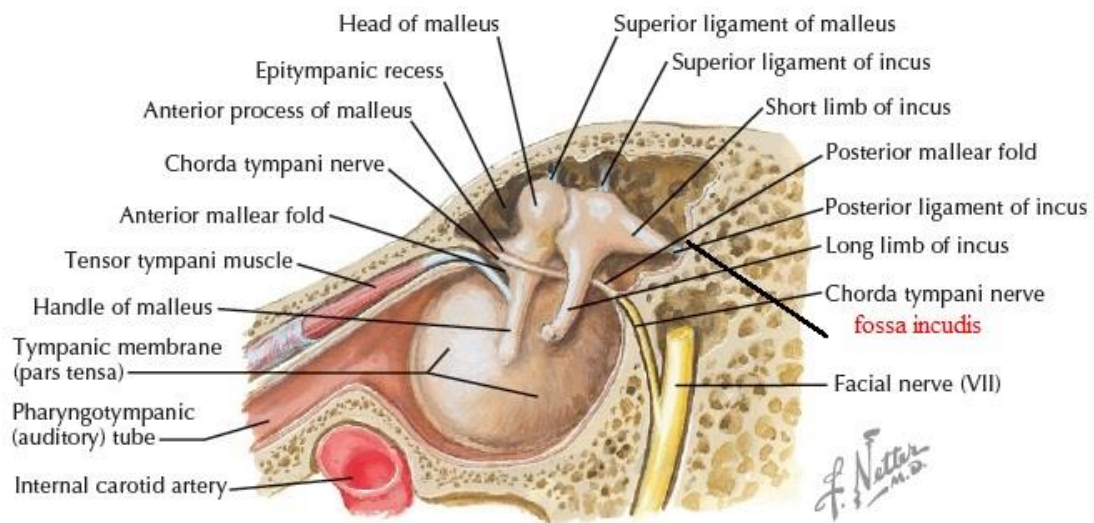
Figure 151.5 Posterior epitympanic cholesteatoma passing through the superior incudal space and the aditus ad antrum.

- [Posterior Mesotympanic route:](#)
- Second most common spread pattern.
- Pars tensa retracts
- Allows cholesteatoma to gain access to the regions of the
 - **Stapes**
 - **Round window**
 - **Sinus tympani**
 - **Facial recess**

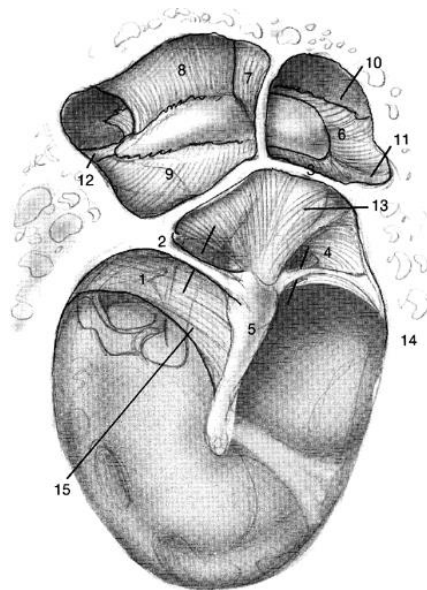
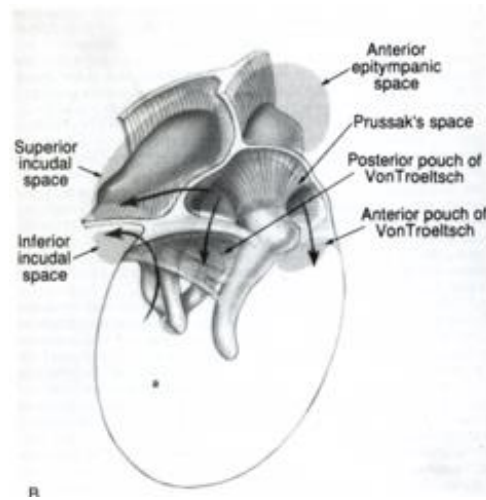


- Anterior Epitympanic route:
- Cholesteatomas form Anterior to the malleus head.
- Easily overlooked during tympanomastoidectomy if the area is not explored.
- Cholesteatomas may involve the **Facial nerve**.
- Extend into **Anterior pouch of von Troeltsch** "Between TM and anterior malleolar fold"
- Into **Supratubal recess**.
 - Also called Anterior Epitympanic recess.
 - Anteriorly: Middle cranial fossa, Petrous tip, Root of zygoma
 - Posteriorly: Cog, extending to the cochleariform process
 - Superiorly: Middle cranial fossa
 - Floor: Associated with the horizontal portion of the facial nerve.
 - Laterally: Tympanic bone and chorda tympani nerve.

Lateral wall of tympanic cavity: medial (internal) view

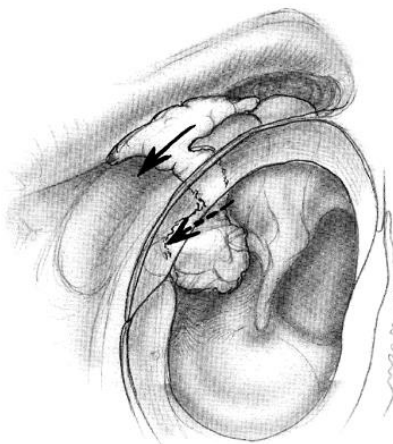
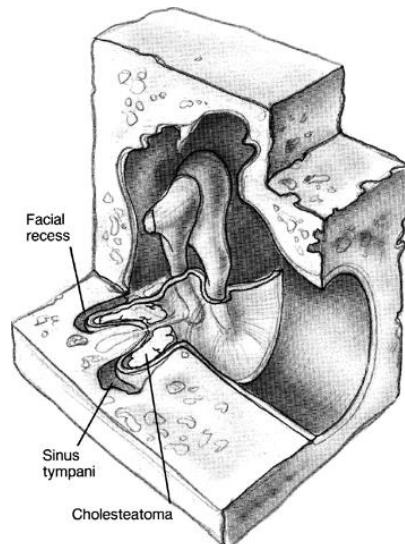


- Most common location for residual cholesteatoma after surgery:
 1. Sinus tympani.
 2. Facial recess.
 3. Anterior Epitympanium(protympanum)
 4. Sinodural angle / mastoid tip (check list)



Spaces and pouches on Middle ear.

- 1, Posterior malleal fold
- 2, Posterior tympanic stria
- 3, Lateral malleal fold
- 4, Anterior tympanic membrane stria
- 5, Malleus (short process)
- 6, Tensor fold
- 7, Superior malleal fold
- 8, Superior incudal fold
- 9, Lateral incudal fold
- 10, Anterior epitympanic space
- 11, Anterior malleal ligament
- 12, Postincudal ligament
- 13, Prussak's space
- 14, Anterior pouch of von Troeltsch.
- 15, posterior pouch of von Troeltsch.

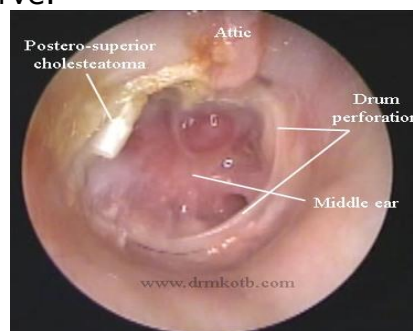


❖ **Prevention of Cholesteatoma Formation**

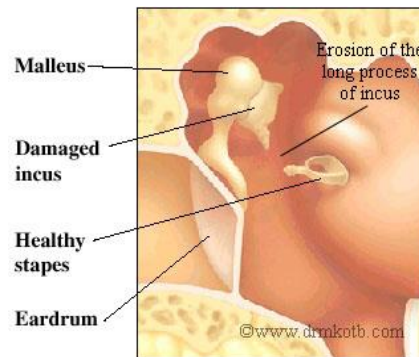
- Retraction pocket is due to Prolonged Eustachian tube dysfunction.
 - Precedes the development of acquired cholesteatoma
 - Most pockets extend into epitympanum or sinus tympani.
 - Good practice to aggressively manage such retraction pockets
- Long-term tympanostomy tube should be placed to
 - Resolve the negative middle ear pressure.
 - Allows the tympanic membrane to revert to a neutral position.
- TM has been retracted for a long time, loses all its elasticity, it will not revert to a normal appearance.
- If the retraction pocket is adherent to the ossicles or folds or if it has been present for an extended period of time, the retraction pocket will persist.
- If the retraction pocket persists, surgical exploration may be indicated.

❖ Patient Evaluation

- History:
- Early symptoms of cholesteatoma:
 - Hearing loss (first)
 - Otorrhea
 - Otalgia
 - Nasal obstruction
 - Tinnitus
 - Vertigo.
- A previous history of middle ear disease, CSOM or tympanic membrane perforation may be revealed.
- Progressive unilateral CHL with a chronic foul smelling otorrhea should raise suspicion.
- some are asymptomatic
- Examination:
- Diagnosis of cholesteatoma is made on otoscopic examination that includes:
 - Endoscopic
 - Microscopic evaluation
 - Surgical exploration
- Microscope:
 - Important for evaluating the presence of cholesteatoma.
 - Ear should be thoroughly cleaned of otorrhea and debris.
 - Attic or Postero-superior quadrant Retraction pocket.
 - Granulation tissue from diseased bone of the scutum or posterior bony wall.
 - Polyp may protrude through an Attic defect.
 - If the disease is very extensive, the entire attic and mastoid antrum will be filled with **granulation tissue**, and the underlying bone will become necrotic and friable over a wide area.
 - Extreme caution should be used with polyp removal as it may be adherent to important underlying structures such as the ossicles or facial nerve.



- Pneumatic Otoscopy:
 - Positive fistula (pneumatic otoscopy will result in nystagmus and vertigo) response suggests erosion of the semicircular canals or cochlea.
- Cultures should be obtained with wet, infected ears.
 - Topical and/or oral antibiotics should be administered in these cases.
- Audiological evaluation:
- PTA with air and bone conduction, speech reception thresholds, and word recognition:
 - CHL in the affected ear varies depending on extent of disease.
 - Moderate CHL deficit in excess of 40 dB indicates ossicular discontinuity, usually from erosion of the long process of the incus or capitulum of the stapes.
 - Mild CHL may be present with extensive disease if the cholesteatoma sac transmits sound directly to the stapes or footplate.
 - Small perforation: CHL in the low frequencies.
 - Large perforation: CHL in both high and low frequencies.
- Tympanometry results will vary and may suggest decreased compliance or perforation of the tympanic membrane.
- Audiometry results should always be correlated with the 512Hz tuning fork exam.



- Radiological investigations:
- **CT Temporal bone:**
 - 1mm -section without contrast in axial and coronal planes.
 - Allows for evaluation of anatomy, extent of the disease and as screen for complications.
 - Scutum erosion, Antrum expansion, Ossicular destruction, Tegmen, HSCC or CN VII dehiscence
-
- **Diffusion-weighted MRI Temporal bone:**
 - T1: HYPOintense.
 - T2: HYPERintense.
 - FLAIR: Intermediate in signal.

❖ **Management:**

- Cholesteatoma must come out surgically unless:
 - Too old or too sick " unfit"
 - Small and easily accessible not infected cholesteatoma to suction clearance under operating microscope
 - Risks of surgery may not outweigh the benefits in some patients with only-hearing ears.
 - Patients refusing surgery.
- Manage medically first in the clinic:
 - Serial Meticulous suction cleaning
 - Remove All attic crusts
 - Remove polyps clinic (never avulse them out)
 - Topical Antibiotics drops
 - Irrigation with 2% acetic acid in 20% isopropyl alcohol may keep some cholesteatomas stable
- Surgical intervention:
- Dry ears are much easier to operate on than wet, infected ears.
- While it is not always possible to dry a chronically infected ear with medical therapy
- **Preoperative counseling:**
- Absolute necessity prior to surgery.
- Primary objective of surgery is **Safe dry ear**
- Which is accomplished by:
 - Aeration of the middle ear
 - Treating all complications
 - Removing diseased bone, mucosa, granulation polyps, and cholesteatoma
 - Preserving as much normal anatomy as possible (posterior canal wall) & closure of the middle ear
- Secondary objective of surgery is **Hearing improvement.**
- Possible adverse outcomes must be discussed including:
 - Facial paralysis
 - Vertigo
 - Further hearing loss
 - Tinnitus.
 - Recurrent and residual cholesteatoma
 - CSF leak and meningitis.
 - Mastoid cavity: Need for cleaning and water precautions.
- Patient should understand that long-term follow-up debridement every 6 to 12 months will be necessary and that they may need additional surgeries "second-stage procedures for residual cholesteatoma or ossicular chain reconstruction".

**TABLE
151.2**

**SURGICAL GOALS FOR
CHOLESTEATOMA**

Treat complications
Remove diseased tissue
Obtain a dry "safe" ear
Preserve normal anatomy
Improve hearing

- **Surgical Management:**
- Mainstay of treatment.
- Factors affecting choice of surgery
 - **Local factor**
 - Extent of disease
 - Fistula
 - ET function
 - Pneumatization of mastoid
 - Hearing level in both ears
 - **General factor**
 - General medical condition
 - Occupation
 - Reliability
 - **Skill and experience of the surgeon**
- The surgical procedure to be used should be designed for each individual case according to the extent of disease.
- More extensive disease will usually dictate a more aggressive surgical approach.

❖ Types of Mastoidectomy are done to deal with cholesteatoma:

- Cholesteatoma limited to the middle ear can be managed with a tympanoplasty.
- The status of the ossicular chain must be meticulously evaluated
- Cholesteatoma can be removed without disrupting the ossicular chain.
- If the lateral chain, malleus and incus, are significantly involved with cholesteatoma, the surgeon should consider separating the incus from the stapes and remove the incus.
- If cholesteatoma medial to the head of the malleus, the surgeon should also consider removing the head of the malleus.
- If Cholesteatoma that is adherent to the stapes can be meticulously removed with microinstrumentation or laser.
- Extensive granulation tissue, significant bleeding, or an exposed facial nerve in the tympanic segment near the stapes, the surgeon should consider leaving some cholesteatoma on the stapes and attempt to remove it at a second look procedure.
- Exteriorization of cholesteatomas without complete removal lead to:
 - Progressive hearing loss
 - Chronically draining ears

○ **Types of Mastoidectomy**

- **Canal-wall-up (Intact wall):**

- Simple mastoidectomy
- Complete mastoidectomy **with** and **without** a facial recess approach "Posterior tympanotomy"

- **Canal-wall-down:**

- Radical mastoidectomy
- Modified radical mastoidectomy (MRM)
- Bondy MRM

- **Other:**

- Atticotomy (transcanal)
- Canal wall reconstruction
- Mastoid obliteration

Surgical Approach

Atticotomy: transcanal

Simple mastoidectomy

Canal wall-up procedure (intact wall) with or without facial recess approach

Canal wall-down procedure (canal down): radical or modified radical mastoidectomy, Bondy procedure

- **Indications Mastoid Surgery**
 1. Persistent or recurrent otorrhea.
 2. Persistent or recurrent ear pain.
 3. Conductive hearing loss
 4. TM perforation and/or cholesteatoma.
 5. Acute mastoiditis with osteitis.
 6. Neoplasm of temporal bone.
 7. Fracture of temporal bone with CSF leak.
 8. Facial nerve paralysis requiring decompression of the facial nerve.
 9. Various other surgical procedures such as:
 - Cochlear implantation
 - Labyrinthectomy
 - Endolymphatic sac decompression
 - Accessing the cerebellopontine angle, skull base, and petrous apex
- **Cortical Mastoidectomy (Simple Mastoidectomy):**
 - Sometimes called a Transmastoid antrotomy
 - A CWU approach to exenterate:
 - Mastoid cortex
 - All accessible mastoid air cells
 - Entering the antrum
 - Good for drain acute mastoid infections that are refractory to ABx "mastoiditis with subperiosteal abscess".

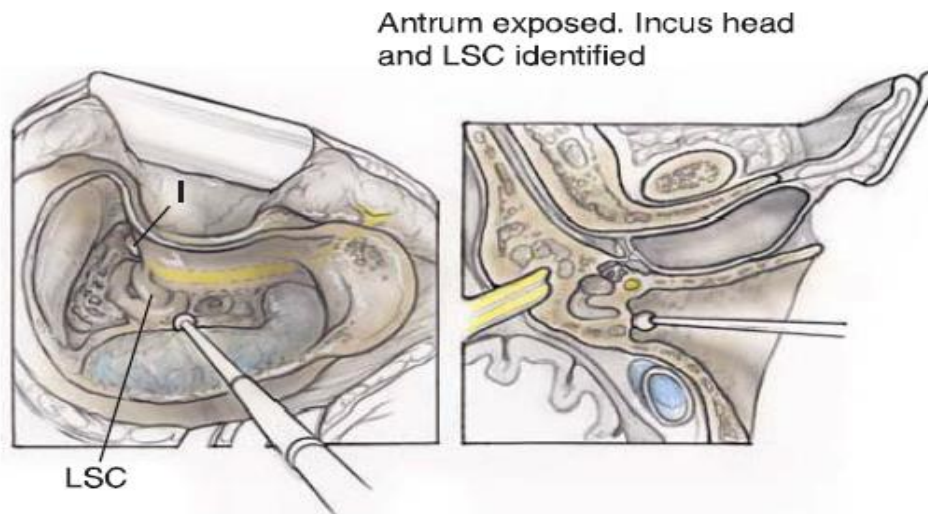
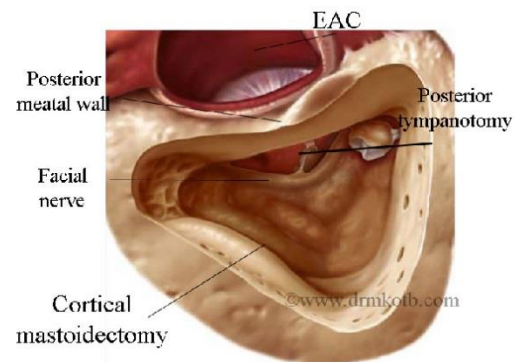
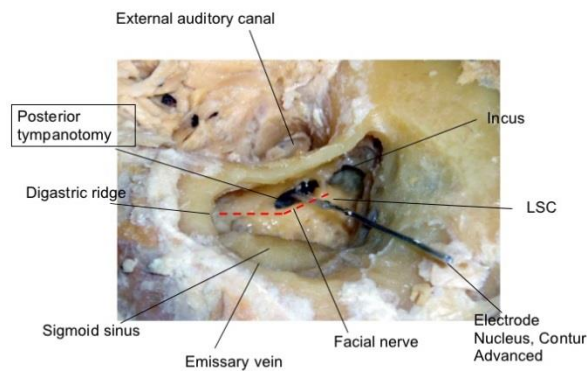
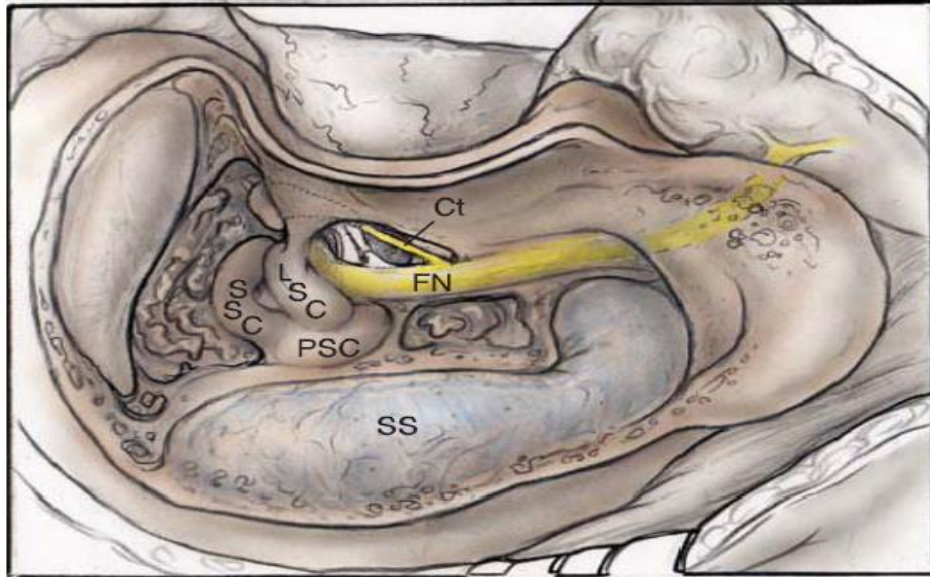


Figure 152.8 Simple mastoidectomy. **Left:** Lateral surgical view of mastoid cavity. The air cells of the mastoid have been removed back to the bone over the sigmoid sinus and the otic capsule of the bone over the lateral semicircular canal can be seen. The posterior bony canal wall and bone overlying the facial nerve (in yellow) are intact. **Right:** An axial section view of mastoid cavity.

- **Canal wall up (CWU) Mastoidectomy (Intact canal mastoidectomy):**
 - More complete removal of the air cell system than simple mastoidectomy
 - Avoid the problems and maintenance necessary of CWD procedures.
 - Consists of preservation of the posterior bony external auditory canal wall during simple mastoidectomy:
 - Without posterior tympanotomy (Complete Mastoidectomy)
 - With posterior tympanotomy (Facial recess approach) performed through a triangle bounded by the fossa incudis, facial nerve, and chorda tympani nerve.
 - A staged procedure is often necessary with a scheduled **second look operation at 6 to 18 months** for removal of residual cholesteatoma and ossicular chain reconstruction if necessary.
 - Recent variations on canal-wall-up mastoidectomy include removing a portion of the canal wall then reconstructing the defect with bone, cartilage, or an alloplastic material.





- This approach may be indicated in patients with:
 - Large pneumatized mastoid
 - Well aerated middle ear space,
 - Good eustachian tube function.
- Contraindications of CWU: (FUSION ET)
 1. Labyrinthine **F**istula & other complication
 2. **U**nfit for second look , **U**nsuccessful previous mastoidectomy (residual/recurrent cholesteatoma)
 3. **S**clerotic mastoid
 4. **I**nvolve ment Posterior canal wall (Automastoidectomy)
 5. **O**nly hearing ear.
 6. **N**on-compliant to follow-up.
 7. Poor **ET** function.
- Advantages
 - Maintaining normal anatomy (Physiologic TM and deep middle ear space)
 - Rapid healing
 - Avoidance of mastoid cavity. (No need for frequent care and water precautions)
 - Hearing aids are easier to fit and wear.
- Disadvantages
 - Incomplete exteriorization of facial recess.
 - **R**esidual disease more likley
 - **R**ecurrent Cholesteatoma may occur in Attic
 - Need for **m**andatory Second look
 - Delayed canal breakdown

- Rates of cholesteatoma recurrence after CWU mastoidectomy:
 - Residual: 20-35%
 - Recurrence: 5-20%
 - Just remember "30 & 15%"
- **Canal wall down (CWD) Mastoidectomy:**
 - Taking down the superior & posterior canal wall to the level of the vertical facial nerve and exteriorizing the mastoid into the external ear canal (one cavity).
 - Epitympanum is obliterated with removal of the
 - Scutum
 - Head of the malleus
 - Incus.
 - Meatoplasty should be large enough to allow good aeration of the mastoid cavity and permit easy visualization to facilitate postoperative care and self cleaning.
- **Modified Radical Mastoidectomy:**
 - A classic CWD operation in which the middle ear space is preserved.
 - To eradicate disease of attic and mastoid.
 - TM remnant, functioning ossicles and the reversible mucosa and function of the eustachian tube are preserved.
 - Ossicular reconstruction , tympanoplasty can also be performed
- **Indications:**
 1. Cholesteatoma confined to the attic and antrum.
 2. Localised chronic otitis media.
- Postoperatively, cavity is fully epithelialised in 2-3 months.
- Should be periodically checked (every 4-6 months) in the first year and then annually for removal of any debris or infection.
- Any granulation tissue which delays epithelialisation is removed or cauterised.
- **Radical mastoidectomy:**
 - CWD operation in which the middle ear space is eliminated and the eustachian tube is plugged.
 - To eradicate disease of the middle ear and mastoid with complete removal of TM, annulus, malleus, incus and middle ear mucosa "stapes is usually preserved"
 - Opening of eustachian tube is plugged with piece of muscle or cartilage aiming for a dry "open" cavity with no secreting epithelium "entire cavity becomes lined with squamous epithelium"
 - Rarely required these days.

- Indicated in these situations:
 1. When all cholesteatoma cannot be safely removed, e.g. that invading eustachian tube, round window niche, perilabyrinthine or hypotympanic cells.
 2. Disease tracking into the petrous apex
 3. Removal of glomus tumour.
 4. Carcinoma middle ear. Radical mastoidectomy followed by radiotherapy is an alternative to en bloc removal of temporal bone in carcinoma middle ear.
- **Bondy Modified Radical Mastoidectomy**
- Rarely used today.
- Bondy procedure is performed like the modern modified radical mastoidectomy with the exception that the middle ear space is **not entered**.
- **Removal of the scutum and portion of the posterior canal wall** to exteriorize the antrum and epitympanum
- **Preservation of the ossicles and middle ear space**
- **The bony defect is not reconstructed; rather, the cholesteatoma matrix is exteriorized.**
- Indication:
 - o Attic cholesteatoma that does not involve the middle ear space and is **lateral** to the ossicles , accompanied by a sclerotic mastoid
- **Indications of CWD: (FUSION ET)**
 1. Labyrinthine **Fistula** & other complicaion "cholesteatoma over the labyrinth simply becomes part of the epithelial coverage of the mastoid cavity"
 2. **Unfit** for second look , **Unsuccessful** previous mastoidectomy (residual/recurrent cholesteatoma)
 3. **Sclerotic** mastoid
 4. **Involvement** Posterior canal wall (Automastoidectomy)
 5. **Only** hearing ear.
 6. **Non-compliant** to follow-up.
 7. Poor **ET** function.
- Advantages of CWD:
 - o Easy to assess for residual disease
 - o Lower incidence of recurrent disease
 - o Total exteriorization of facial recess.
 - o Second look surgery is not mandatory.
 - o Cost effective

- Disadvantages of CWD:
 - Delayed healing (Risk of failure to epithelize)
 - Altered anatomy
 - Implication of Mastoid cavity "frequent episodes Discharge" (requires periodic microdebridement and water precautions for rest of patient's life)
 - Shallow middle ear space is difficult to reconstruct.
 - Greater difficulty achieving Hearing Improvement
- Rates of cholesteatoma recurrence after CWD mastoidectomy:
 - Residual: 2-17%
 - Recurrence: 0-10%
 - **Just remember "10 & 5%"**
- **Five things to do in surgery during Modified Radical Mastoidectomy to ensure Dry ear:**
 1. Complete Saucerization of mastoid cavity (remove all mucosa)
 2. Lower the Facial Ridge to the level of the facial nerve "remove any Dependent Spaces"
 3. Take down the Mastoid Tip
 4. Cavity Obliteration with Fat or Muscle (Palva flap or Temporalis Muscle)
 5. Wide Meatoplasty

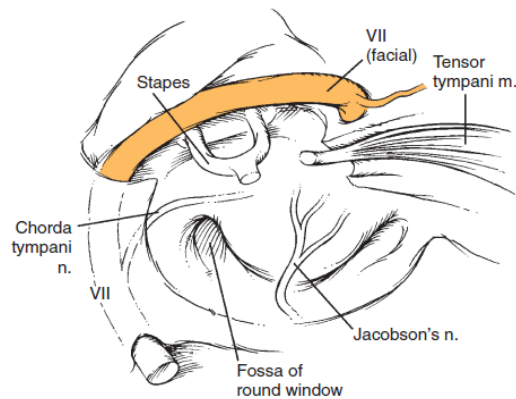


FIGURE 142-8. Course of the tympanic segment of the facial nerve. (From Donaldson JA, Duckert LG, Lambert PR, Rubel EW, eds. *Surgical anatomy of the temporal bone*, ed 4. New York: Raven Press; 1992.)

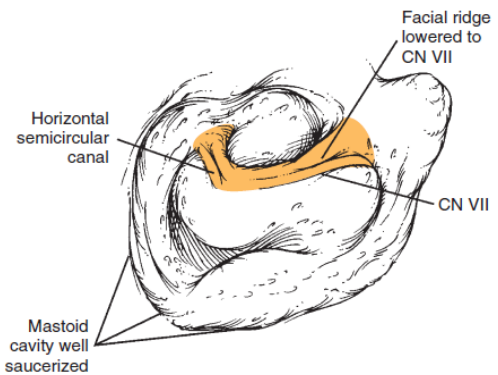


FIGURE 142-10. Canal-wall-down mastoidectomy. CN, cranial nerve. (From Brackmann DE, Shelton C, Arriaga MA, eds. *Otologic surgery*. Philadelphia: Saunders; 1994.)

Table 11-2. Comparison of canal wall up and canal wall down procedures

	Canal wall up procedure	Canal wall down procedure
Meatus	Normal appearance	Widely open meatus communicating with mastoid
Dependence	Does not require routine cleaning	Dependence on doctor for cleaning mastoid cavity once or twice a year
Recurrence or residual disease	High rate of recurrent or residual cholesteatoma	Low rate of recurrence or residual disease and thus a safe procedure
Second look surgery	Requires second look surgery after 6 months or so to rule out cholesteatoma	Not required
Patients limitations	No limitation. Patient allowed swimming	Swimming can lead to infection of mastoid cavity and it is thus curtailed
Auditory rehabilitation	Easy to wear a hearing aid if needed	Problems in fitting a hearing aid due to large meatus and mastoid cavity which sometimes gets infected

Procedure	Advantages	Disadvantages
Canal wall up	Physiologic position of tympanic membrane Enough middle ear space No mastoid cavity problem	Residual and recurrent cholesteatoma may occur Incomplete exteriorization of facial recess Second-stage operation often required delayed breakdown of the bone of posterior canal wall
Canal wall down	Residual cholesteatoma easily found on follow-up evaluation Recurrent cholesteatoma rare Total exteriorization of facial recess	Mastoid cavity problem (frequent) Middle ear shallow and difficult to reconstruct Position of pinna may be altered; second-stage operation sometimes required

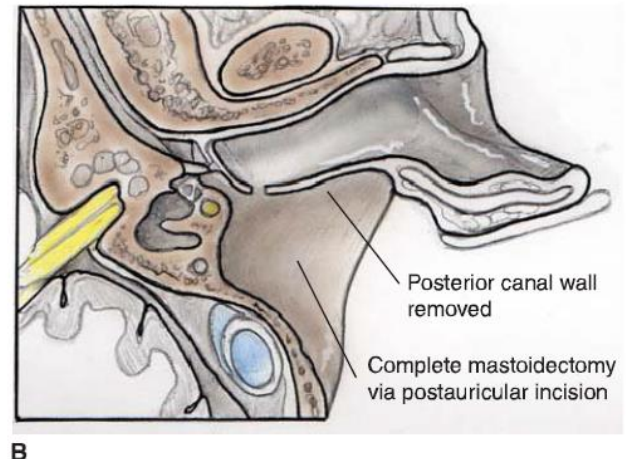
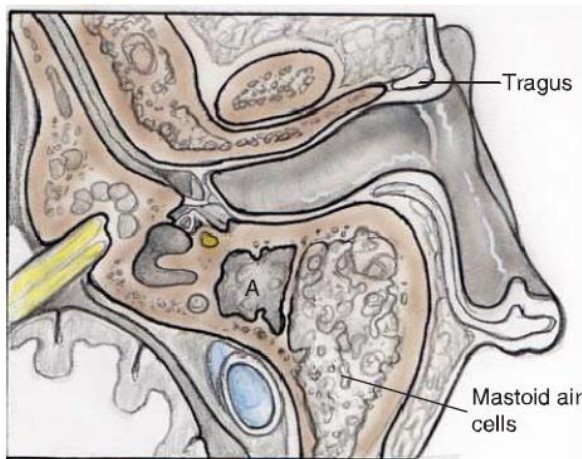
- **Transcanal Anterior Atticotomy**

- Elevation of a tympanomeatal flap via an **endaural incision**
- **Removal of the scutum** to the **limits of the cholesteatoma**.
- After removal of the disease, the aditus is obliterated with muscle, fascia, cartilage or bone prior to reconstruction of the middle ear space.
- Some advocate reconstruction of the lateral attic wall "scutum site" with bone or cartilage, however, this may lead to retraction disease and possible recurrence in patients with poor eustachian tube function.
- Indication:
 - Limited cholesteatoma involving the middle ear, ossicular chain, and epitympanum.

- **MASTOID OBLITERATION**
- Mastoid obliteration is typically used when the canal wall has been removed to decrease the size of the mastoid cavity and make it as close free as possible.
- Extent of mastoid air cell obliteration vary considerably from surgeon to surgeon.
- Various materials are used that include :-
 - **Autogenous** bone paté, bone chips and cartilage, free or vascularized soft tissue,
 - **"Paiva flap"** postauricular musculoperiosteal flap that was rotated inward to obliterate the mastoid cavity , can use of bone chips and bone paté in combination with the flap
 - **"Hong Kong flap"** temporalis muscle flap or temporalis fascia flap based on a superficial temporal artery pedicle
 - **Alloplastic** hydroxylapatite , tricalcium phosphate , ceramic materials " more infection "
- In rare cases, the eustachian tube and external ear canal are closed to completely isolate the mastoid from the exterior " Blind sac"
- The radiographic features suggestive of disease recurrence after mastoid obliteration have not been well studied



- **Canal-wall reconstruction (CWR)**
- Developed to improve exposure and removal of cholesteatoma as in a CWD approach while retaining the benefits of an intact canal wall
 - improved hearing
 - avoidance of the mastoid cavity
- Complete mastoidectomy including a facial recess is performed and the posterior canal is removed.
- The cholesteatoma, the ossicles, and the tympanic membrane are addressed, and the posterior canal wall is replaced.
- Can be done with or without mastoid obliteration
- **Meatoplasty**
- Enlarging the external auditory meatus is a necessary part of canal-wall-down procedures.
 - Promotes aeration and epithelialization of the canal and cavity
 - Facilitates effective postoperative caree debridement much easier.
 - Reduces the depth of the cavity
- Several techniques to meatoplasty have been advised "All involves removing some concha cartilage and draping the posterior meatal skin into the mastoid cavity"



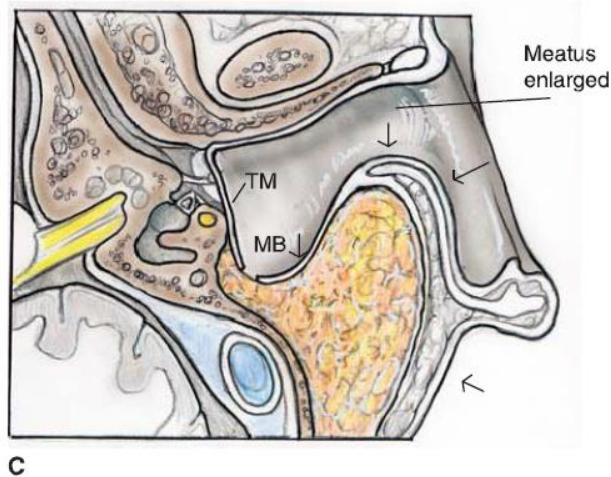


Figure 152.11 Meatoplasty following canal-wall-down mastoidectomy. **A:** Axial view of unoperated temporal bone demonstrating the physical relationships between the external auditory canal and the mastoid for comparison to **(B)** and **(C)**. **A**, Antrum. **B:** Axial view of temporal bone after canal-wall-down mastoidectomy and removal of some of the bone of the conchal bowl. **C:** Axial view after meatoplasty and partial obliteration of mastoid bowl with muscle-periosteal flap. TM, tympanic membrane on facial ridge; MB, mastoid bowl.

- Meatoplasty can be performed by:-
- Connecting superior and inferior Lempert endaural incisions with the postauricular incision.
- The superior cut is brought out laterally into the tragal incisura while the inferior cut is curved just medial to the antitragus.
- Varying amounts of this cartilage can be removed (through the postauricular incision), thin concha skin that drapes into the mastoid bowl)

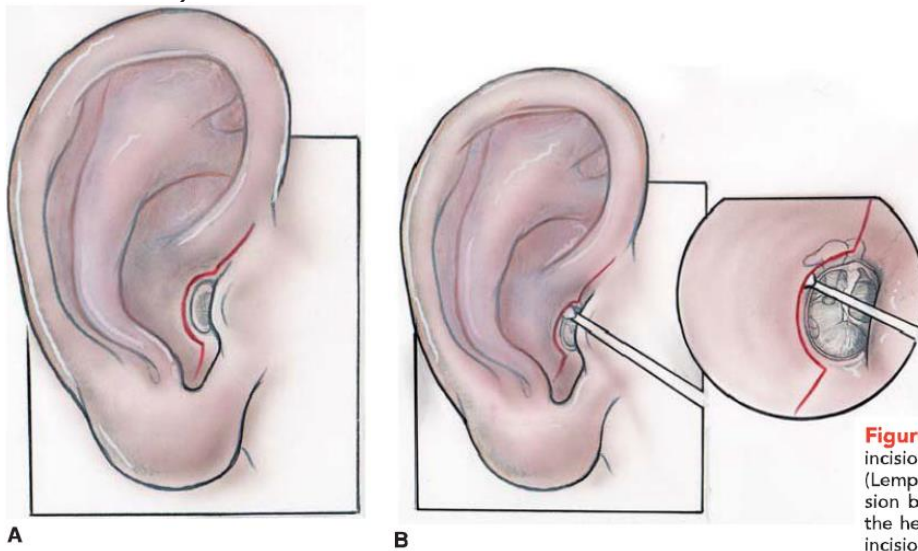


Figure 152.3 A: Endaural incisions. The incision of the posterior bony canal (Lempert I) continues superiorly to an incision between the tragus and the root of the helix (Lempert II). An inferior relaxing incision is often needed. **B:** Separation of

- Causes of a Draining Cavity Post Mastoidectomy
 - Surgical factors:
 1. **Recurrent** or **residual** disease
 2. Inadequate meatoplasty
 3. Defect in tympanic membrane remnant
 4. Poor mastoid shape
 5. High facial ridge
 6. Nooks and crannies
 7. Deep mastoid sump
 - Patient factors:
 1. Poor mastoid care (poor compliance with follow-up appointments)
 2. Inadequate water precautions
 3. Inadequate toilet
 4. Hearing aid wear
 - Monitoring the mastoid for cholesteatoma extremely important.
 - If cholesteatoma is left behind in the mastoid cavity, or trapped underneath a mastoid obliteration, intracranial or vascular complications can occur even many years after the initial procedure.
 - Diffusion-weighted MRI scans have given the otologic surgeon a **noninvasive** mechanism to evaluate a patient's mastoid for recidivistic cholesteatoma with fairly high accuracy

- **SURGICAL TECHNIQUE: MASTOIDECTOMY**

- **Incisions**

- The two principal incisions used:

1. Postauricular incision of Wilde
2. Endaural incision of Lempert.

- Postauricular incision provides better overall exposure and allows complete access to the mastoid tip.

- **In adults**

- Incision is placed 8 to 10 mm posterior to the postauricular sulcus
- It can be placed more posteriorly for wider exposure as might be necessary during translabyrinthine access to the cerebellopontine angle.
- Should not be placed directly in the postauricular crease because such placement creates a deep, difficult clean

- **In children younger than 2 years**

- The inferior portion of this incision must be placed more posteriorly than in adults
- Because the tympanic ring in children is underdeveloped, mastoid pneumatization is incomplete, and the stylomastoid foramen is shallow. Avoid facial nerve injury.

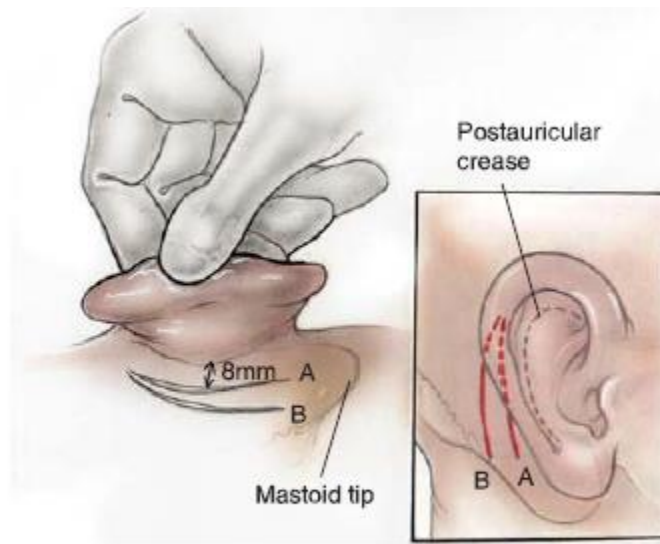


Figure 152.1 Placement of postauricular incisions in adults (A) and infants (B).



FIGURE 142-3. Postauricular incision is placed about 1 cm behind the postauricular crease to facilitate skin closure. (From Sheehy JL. Surgery of

- Postauricular incision is first outlined with a marking pen and 5 to 10 ml infiltrated with a mixture of local anesthetic and epinephrine "postauricularly and within the external ear canal"
- Skin and subcutaneous tissues are incised sharply down to the **temporalis fascia** (superior to the inferior temporal line) and down to the **periosteum** overlying the mastoid cortex (inferior to the inferior temporal line).

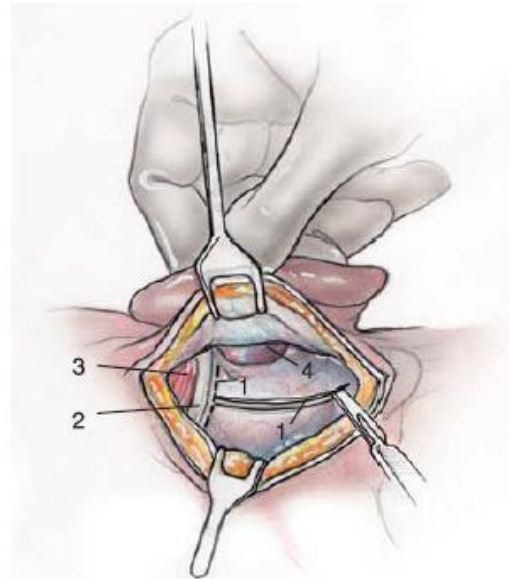


Figure 152.2 After exposure, the periosteum of the mastoid cortex is incised with a "T"-shaped or an "L"-shaped incision. The periosteum is then elevated off the mastoid cortex, exposing the posterior wall of the external auditory canal. 1, Periosteal incisions; 2, Temporal line; 3, Temporalis m.; 4, Posterior canal skin.

- Elevated anteriorly to identify the posterior edge of the external ear canal.
- Elevation superior to the ear canal exposes the root of the zygoma.
- Dissection is carried to the mastoid tip "not to dissect anterior to the tip because this endangers the facial nerve in the stylomastoid foramen".
- Periosteum of the mastoid cortex is Incised with a "T" -shaped or an "L" shaped Incision.
- The periosteum is elevated off the mastoid cortex toward the posterior margin of the ear canal now exposed to start the drilling process

- Self-retaining retractors should now be placed to hold the auricle forward
- Suprameatal spine of Henle marks the lateral extent of the posterior-superior bony ear canal.
- Two anteriorly flaps:
- (a) The pinna and subcutaneous tissues
- (b) The deeper musculo-periosteal tissues.
- Both layers should be closed to maintain a patent meatus and a properly positioned auricle.
- **Endaural incisions**
- Expose a limited portion of the mastoid cortex.
- First, a posterior canal wall incision is made from the 12-o'clock to the 6-o'clock position just medial to the bony cartilaginous junction ([Lempert I incision](#)).
- From the 12-o'clock position of the Lempert I incision, a medial-to-lateral incision is made into the incisura between the tragus and root of the helix ([Lempert II incision](#)).
- A relaxing incision is then made at the inferior margin of the Lempert I incision (in a medial-to-lateral direction)
- This allows the posterior ear canal and conchal skin to be mobilized
- Skin, soft tissues, muscle, and periosteum over the mastoid cortex are elevated using Lempert elevators, and a self-retaining retractor is placed.

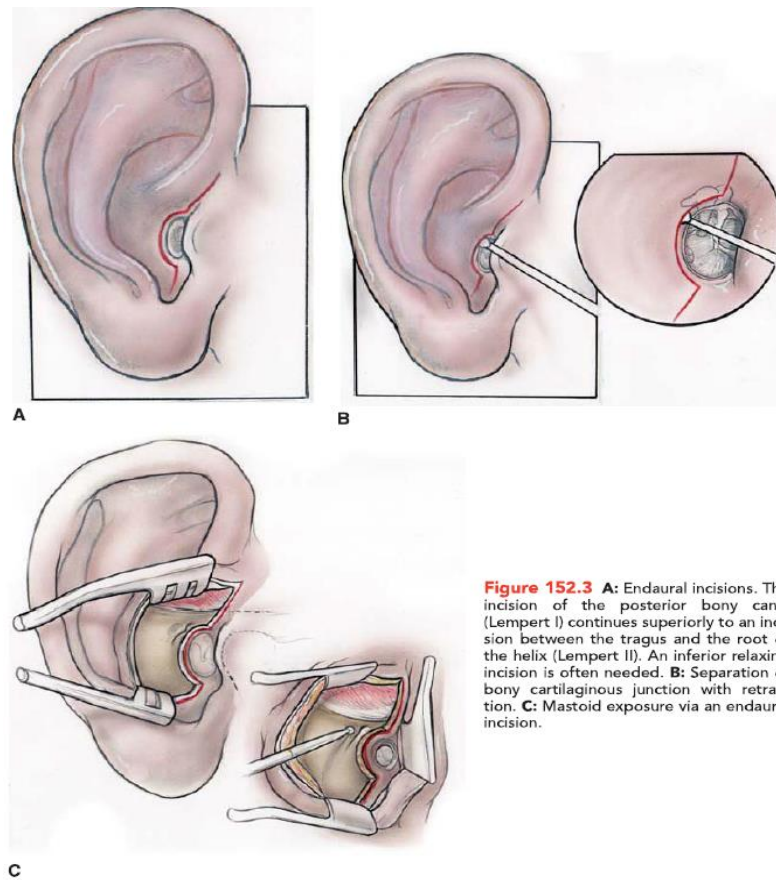
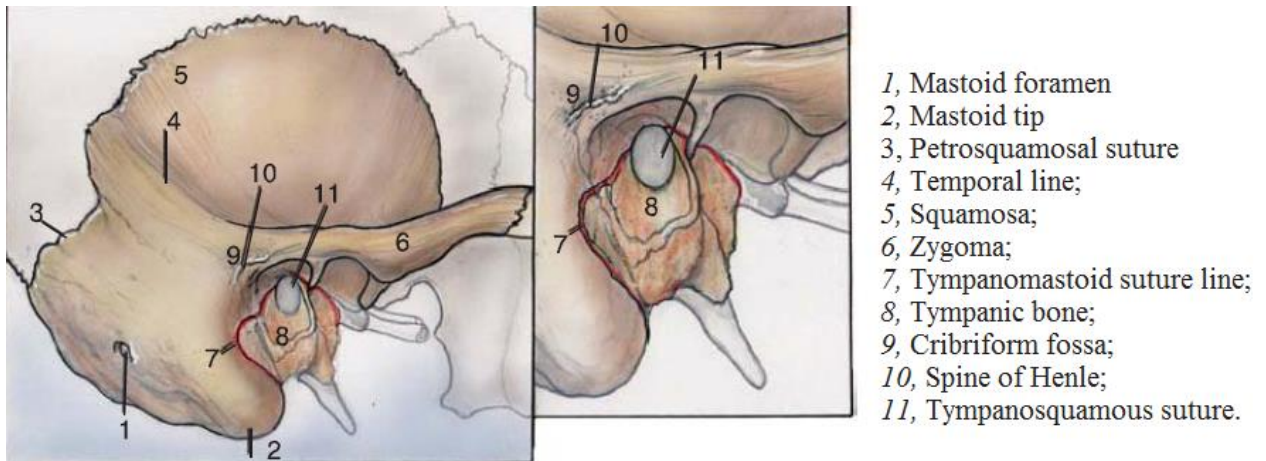


Figure 152.3 A: Endaural incisions. The incision of the posterior bony canal (Lempert I) continues superiorly to an incision between the tragus and the root of the helix (Lempert II). An inferior relaxing incision is often needed. B: Separation of bony cartilaginous junction with retraction. C: Mastoid exposure via an endaural incision.

- **Indications** of this incision:
 - Simple mastoidectomy in very poorly pneumatized temporal bones
 - Atticotomies,
 - Canaloplasty
 - Some tympanoplasty
- **Surface Landmarks**
 - Inferior temporal line (linea temporal is) defines the inferior limit of the temporalis muscle "level of the floor of the middle cranial fossa".
 - Suprameatal spine of Henle is Inferior to the temporal line
 - Macewen triangle (cribrose area) is a depressed pit just posterior to the spine of Henle "landmark for the underlying mastoid antrum"
 - Antrum ia typically located 15 mm medial to the cribrose area
"dome of the horizontal semicircular canal (HSCC) along its floor"
 - Zygomatic root is palpable anterior-superior to the ear canal.
 - The anterior- inferior, and posterior-inferior walls of the atemal auditory canal are formed by the tympanic bone
 - Posterior-superior bony ear canal "between the tympanasquamous and tympanomastoid suture lines" made of squamous bone "thicker and more vascular skin "

- Children younger than 2 years have :-
 - Immature tympanic rings
 - Poorly pneumatization mastoids ,stylomastoid foramen is shallow
 - In children or adults with canal atresia, mal-development of the tympanic bone may result in facial nerve exit directly from the mastoid cortex



- When the mastoid cortex fully exposed:
 - **First bur** cut is made along the temporal line "which approximates the level of the middle cranial fossa dural plate".
 - **Second bur** cut is made perpendicular to this and tangential to the external bony canal; it should be carried inferiorly to the mastoid tip

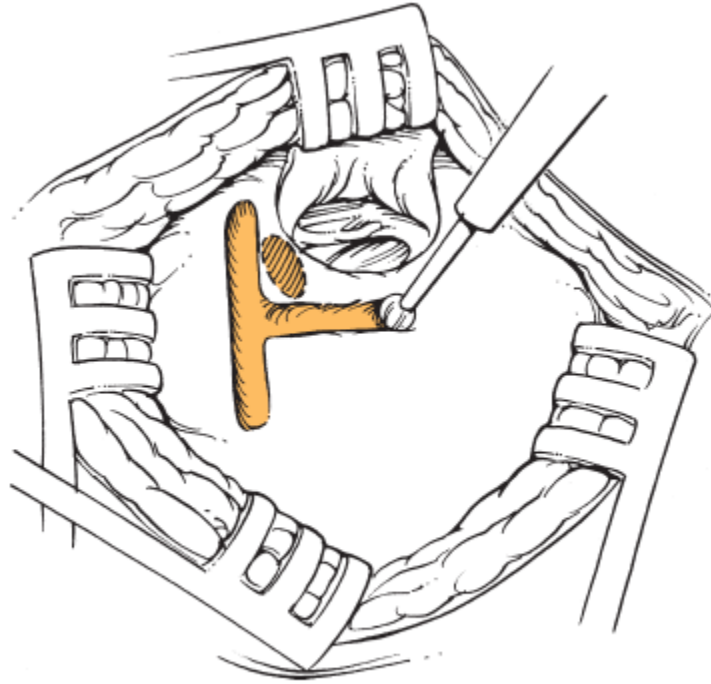
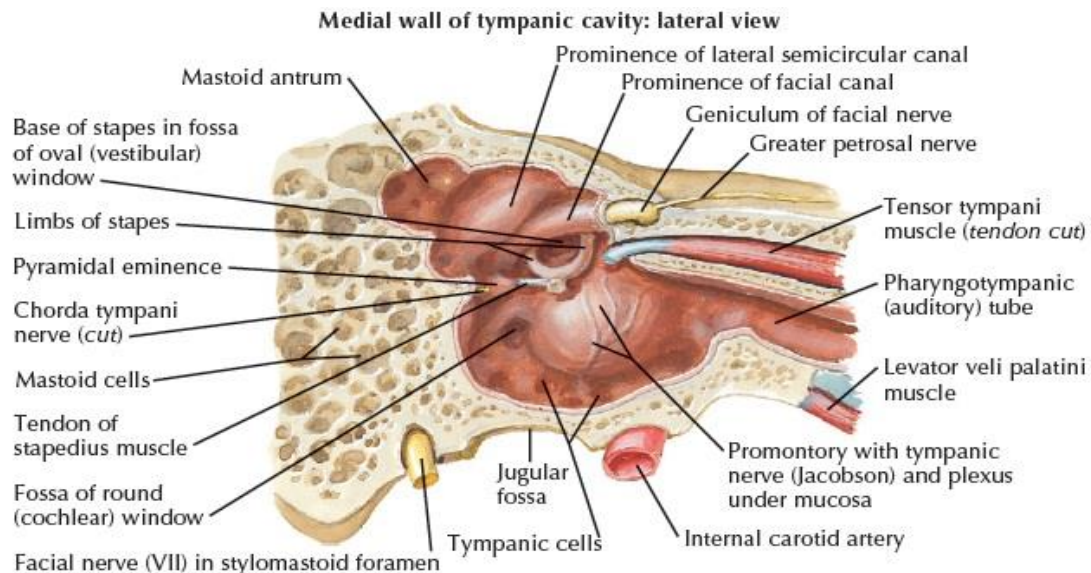


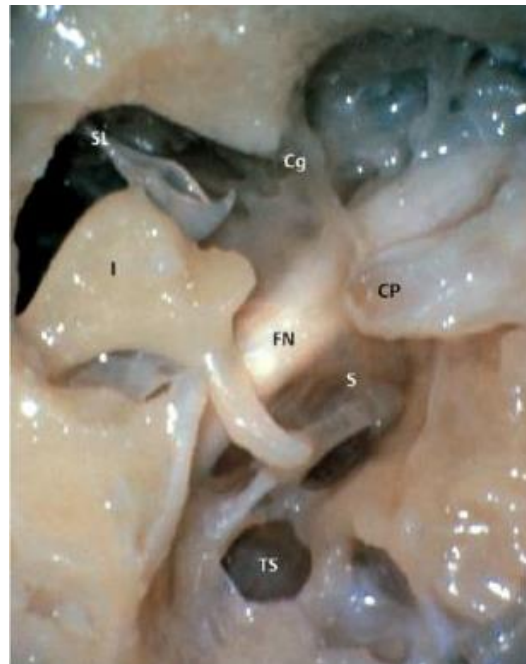
FIGURE 142-4. Initial bur cuts are made along the temporal line and tangential to the bony canal. (From Sheehy JL. Mastoidectomy: the intact canal)

- As the antrum is approached, it is essential to widely saucerize toward the tegmen and especially posteriorly, from the sinodural angle to the mastoid tip.
- Posteriorly, consider the sigmoid sinus, which could be far forward in a poorly pneumatized mastoid.
- Dissection is carried medially, and the antrum is approached, a bony septum (Körner septum)
- Opening the antrum too inferiorly, can injure the HSCC, the genu of the facial nerve, or both.
- Identifying the short process of the incus provides an important landmark for facial nerve will be 1 to 2 mm deeper.

- **Surgical landmarks of CN VII through the Middle Ear**
 - Horizontal SCCs
 - Cochleariform process lie just anterior – VII runs posterior & above this
 - Short process of the Incus – Marks the beginning of the Second Genu & VII runs medial to it
 - Fossa incudis
 - Digastric Ridge – Anterior extent of ridge leads to Stylomastoid foramen
 - Chorda Tympani
 - Oval Window
 - Cog
 - Pyramidal eminence

“Always safer to define the location of this structure than simply to avoid it”





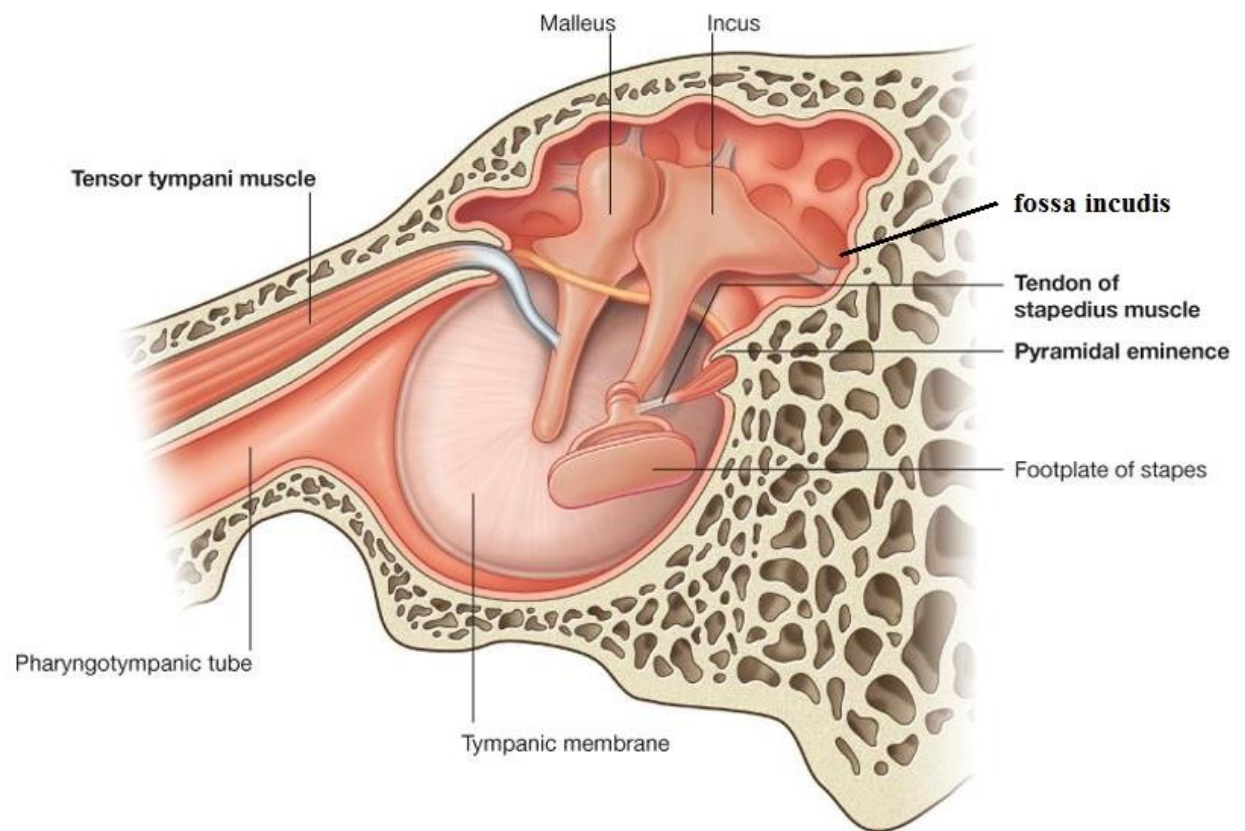
Cg cog

Cp processus
cochleariformis

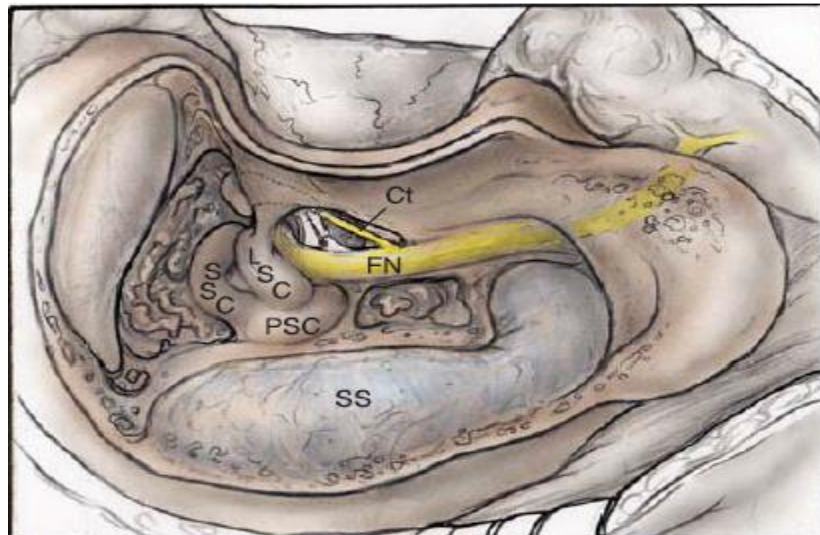
I incus

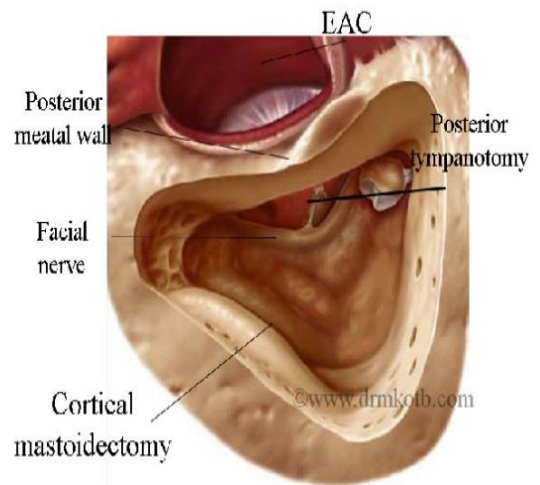
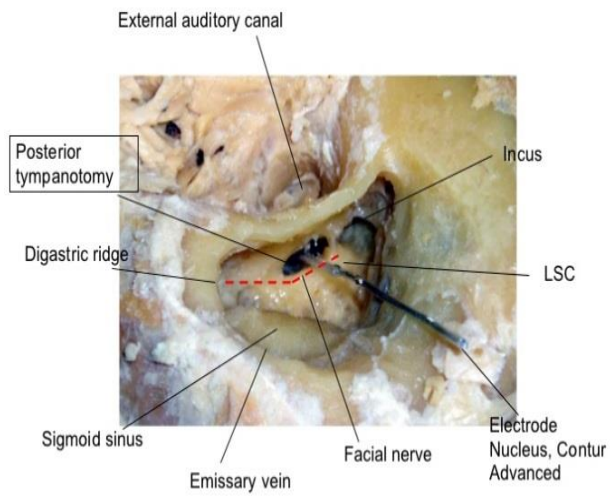
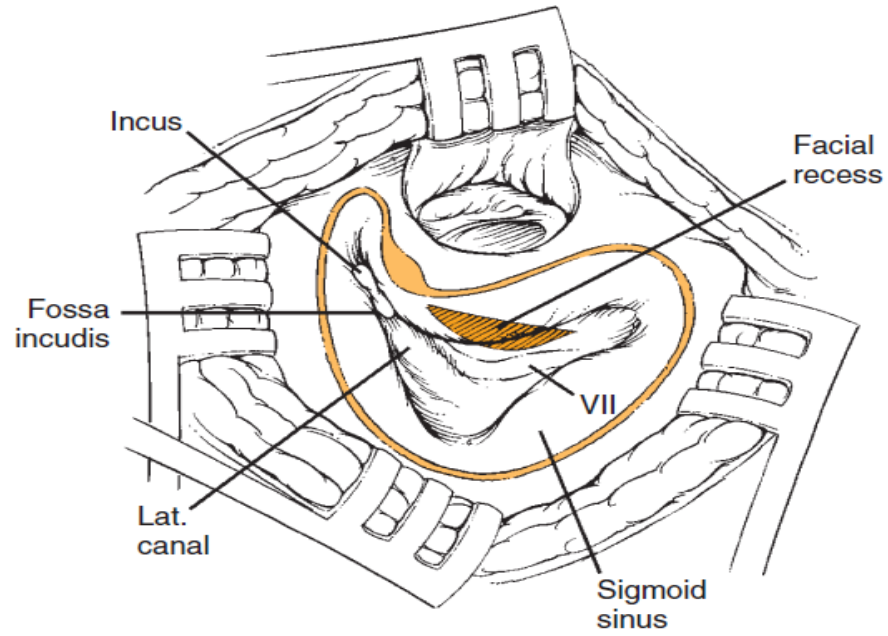
S stapes

TS tympanic sinus



- **OPENING THE FACIAL RECESS performed through a triangle bounded by the**
 1. **Fossa incudes** "superiorly" "small depression ,below the aditus is a, the, which houses the short process of the incus and its suspensory ligament "
 2. **Facial nerve** " medially "
 3. **Chorda tympani nerve** "laterally"
 - Recess provides access to the middle ear from the mastoid "promontory, round window niche, stapes, long process of the incus, cochleariform process, medial side of the tympanic membrane and malleus handle, and Eustachian tube all are well visualized."
 - Facial recess can be extended superiorly and inferiorly to provide a large "posterior tympanotomy."





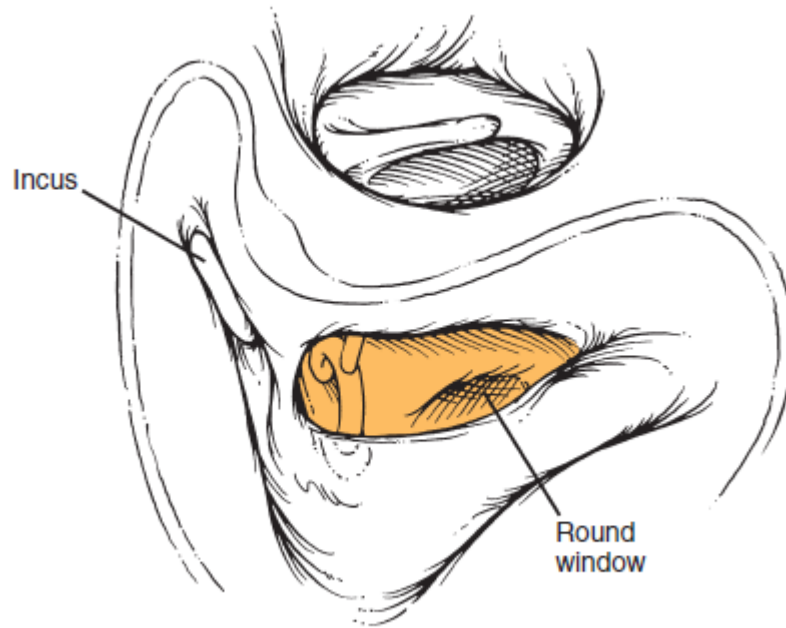


FIGURE 142-6. The facial recess is opened. (From Sheehy JL. Mastoidectomy: the intact canal wall procedure.

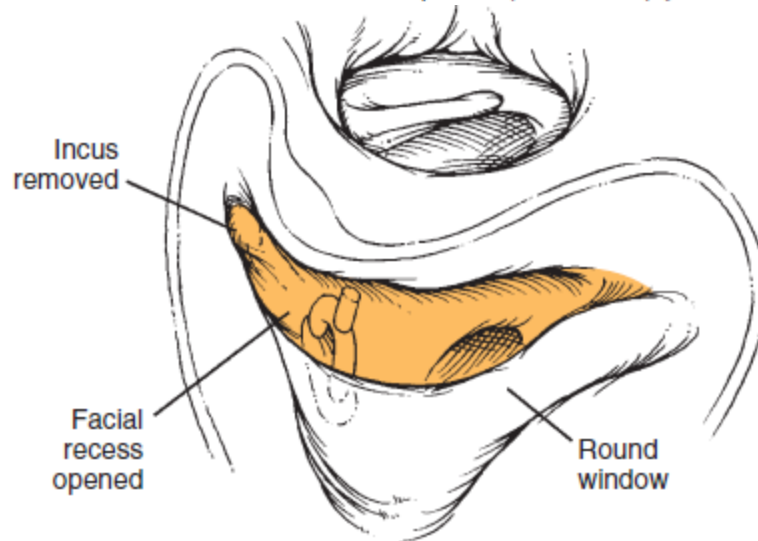
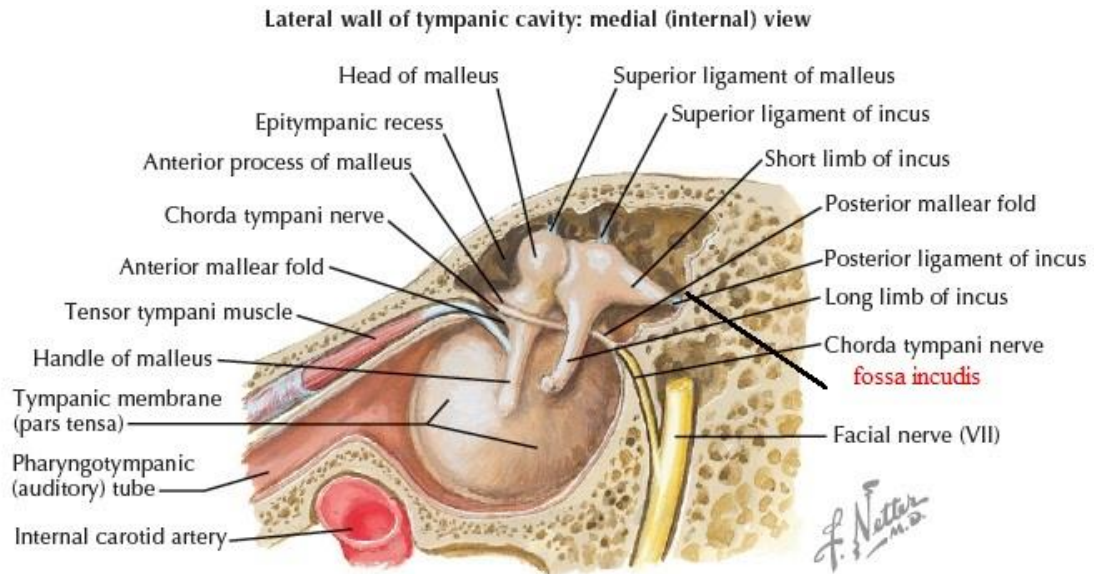


FIGURE 142-7. The facial recess is enlarged by removing the incus and fossa incudis. (From Sheehy JL. Mastoidectomy: the intact canal wall procedure.

- OPENING THE EPITYMPANUM
 - In CWU surgery, it is often necessary to expose the epitympanum.
 - Cholesteatoma can track medially to the heads of the ossicles and can extend into the anterior epitympanic space "epitympanic recess" "supratubal recess"
 - **Incus remnant and the head of the malleus** are removed to provide good access into the anterior aspect of the epitympa



- **CANAL-WALL-DOWN MASTOIDECTOMY**
 - Saucerization cortical edges of the cavity are taken down to the approximate level of the tegmen superiorly, sigmoid sinus posteriorly, and digastric ridge inferiorly
 - The facial nerve is positively identified by removing the posterior bony canal until only a thin shell of bone remains over the nerve.

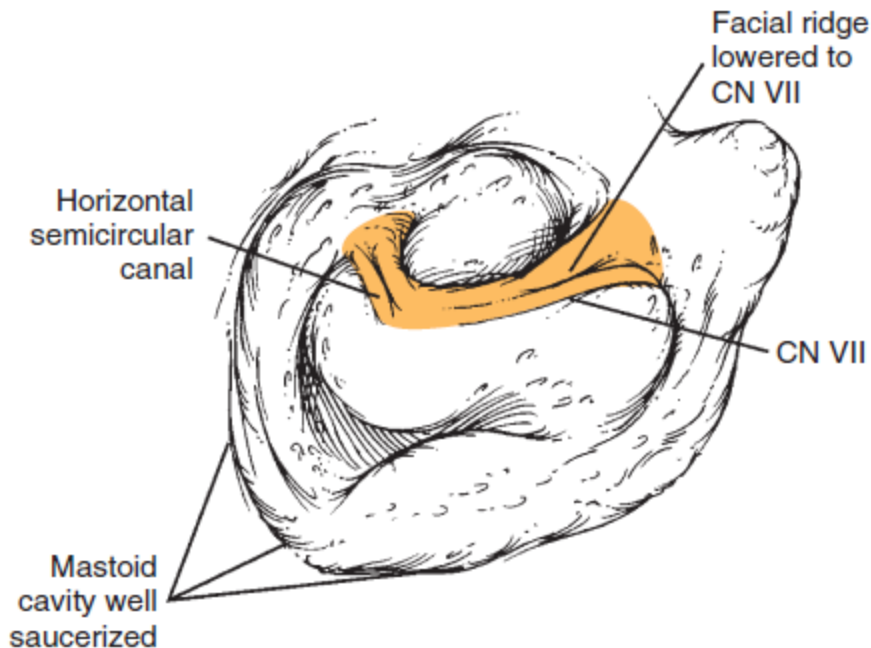


FIGURE 142-10. Canal-wall-down mastoidectomy. CN, cranial nerve.

- This dissection is continued toward the stylomastoid foramen until no bony spur remains (inferior or posterior buttress) between the floor of the external bony canal and the mastoid cavity.
- Anterior extent of the superior canal wall (anterior buttress) is completely removed to create a smooth, gently curving transition from the anterior epitympanum to the anterior canal wall.
- Ideally, the contours of the cavity itself should be smooth, without bony recesses or overhangs.
- Small amounts of bone plate collected during saucerization of the cortical edges can be used to fill irregularities within the cavity.
- Obliteration of these areas, however, should not be performed if the surgeon is not confident that all cholesteatoma remnants have been thoroughly removed.
- Last, a large meatus ensures adequate ventilation of the mastoid cavity and appropriate access for postoperative cleaning.
- Meatoplasty to create a properly sized meatal opening.

**TABLE
152.3**

**COMPLICATIONS
MASTOID SURGERY**

Perioperative complications

Facial nerve injury
 Sensorineural hearing loss
 Postoperative infection
 Dysgeusia
 Brain herniation
 CSF leakage
 Bleeding

Delayed complications

Posterior canal breakdown
 Perichondritis
 Blue-domed cyst
 Mucosalization of mastoid bowl
 Stenosis of external canal

- **Follow-up**

- Healing of mastoid cavity if present
- Healing of surgical wound
- Resolution of presenting symptoms

- **ENDOSCOPY**

- Endoscopes often see where the microscope cannot
- 1.7- to 2.8-mm, 0 or 30-degree rigid telescope,
- Can visualize the facial recess, sinus tympani, or epitympanum
- Can be used to assess the depth of retraction pockets and determine the extent of cholesteatomas.
- Some surgeons use it for second-look procedures.

- **ENDOLYMPHATIC SHUNT**

- Endolymph in the inner ear may flow from the cochlea to the endolymphatic sac.
- "shunt" or "decompress" endolymphatic sac for the treatment of intractable Meniere disease
- Successful control of vertigo has been reported in a majority of patients
- Exposure of the endolymphatic sac require CWU mastoidectomy & open of facial recess
- Endolymphatic sac comes into view just posteroinferior to the posterior semicircular canal.
- Place a sickle knife or similar instrument into the sac and palpate the operculum

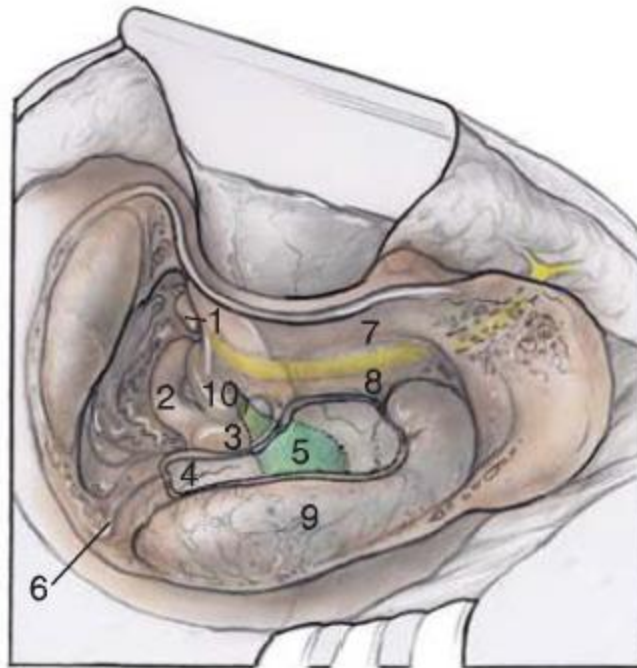
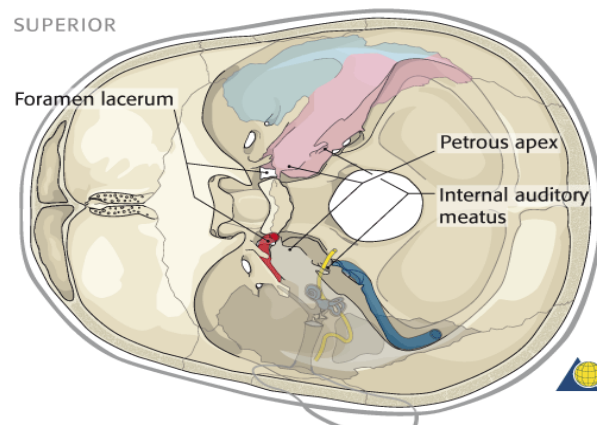


Figure 152.12 Transmastoid exposure of endolymphatic sac. The endolymphatic sac is within the posterior fossa dura generally inferior to the labyrinth and often in contact with the sigmoid sinus. 1, Incus; 2, SSC; 3, PSC; 4, Posterior fossa dura; 5, ELS; 6, SDA—sinodural angle; 7, PCW—posterior canal wall; 8, Retrofacial air cells; 9, Sigmoid sinus skeletonized; 10, LSC.

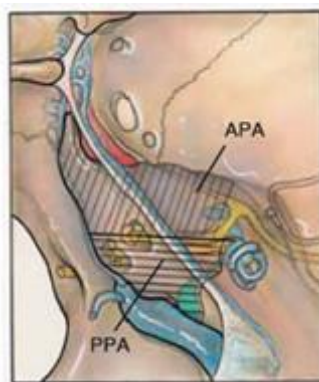
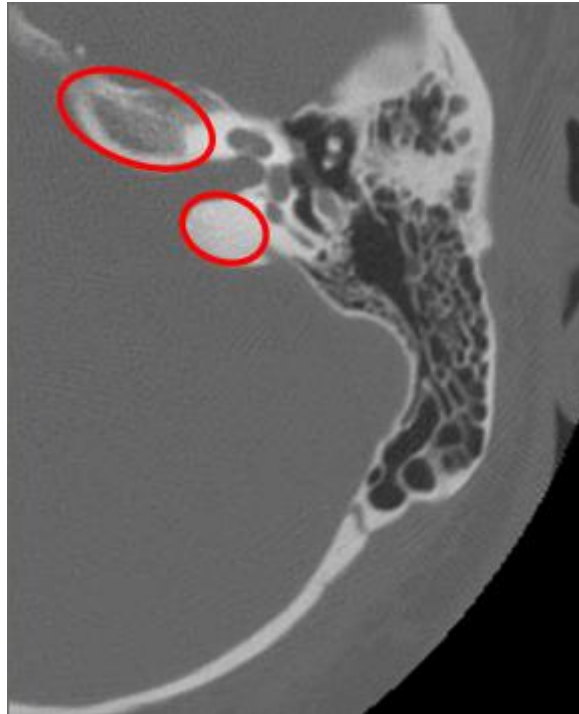
- **PETROUS APICECTOMY**

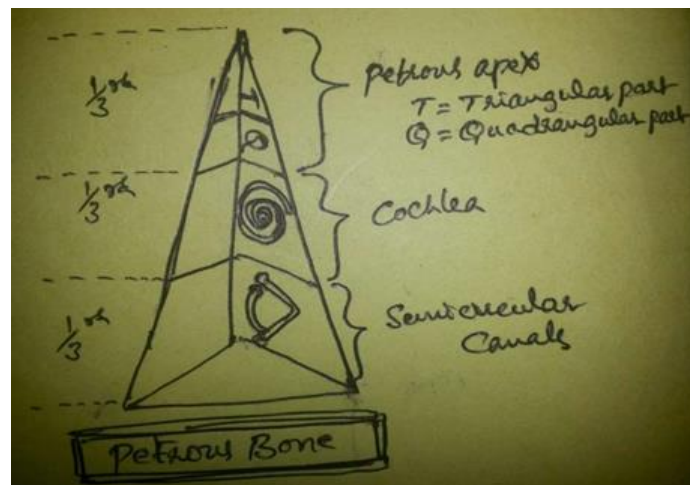
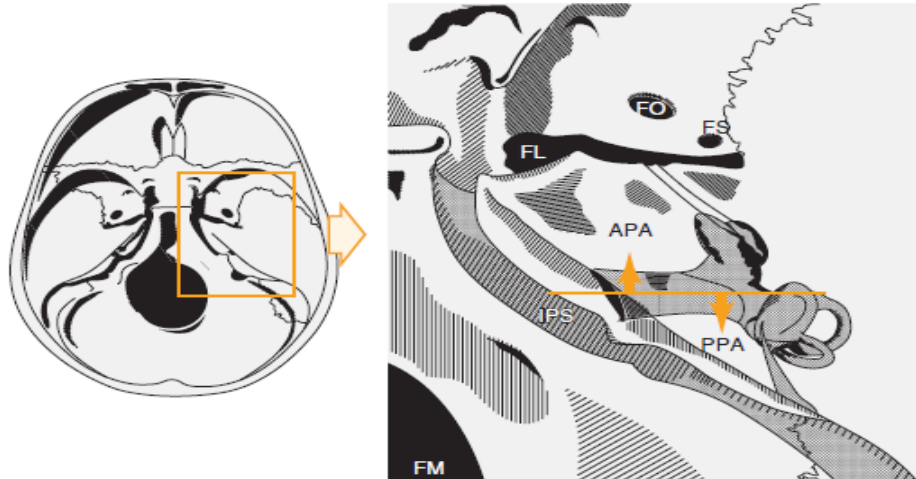
- Surgical access to the petrous apex.
- Becomes necessary for:
 - Drainage of infected air cells "Petrositis (petrous apicitis) "Petrus Apex Syndrome"
 - Cholesteatomas (congenital)
 - Cholesterol granulomas and mucosal cysts
 - Biopsy of various mass lesions
 - **Benign tumors**
 - Meningioma
 - Schwannoma
 - **Malignant tumors**
 - Chondrosarcoma / Chondroma
 - Chordoma
 - Plasmacytoma
 - Metastatic lesion
 - Langerhans cell histiocytosis
- The petrous apex takes the form of a truncated pyramid in the posterior skull base
- Oriented anteromedially between occipital and sphenoid bones
- Forms part of the **foramen lacerum**



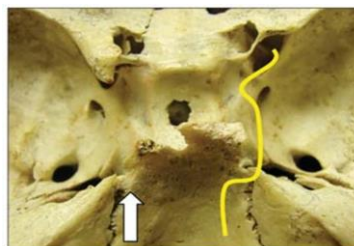
- **Petrous bone** may be of three types (like mastoid),:
 1. **Pneumatized** with air cells extending to the petrous apex "Posterior Petrous Apex pneumatized in 30%, Anterior Petrous Apex pneumatized in 10%"
 2. **Diploic** containing only marrow space
 3. **Sclerotic**.

- Petrous apex divided into Anterior/Posterior by plane running through IAC or Cochlea:
 1. **Anterior Petrous apex**: medial to cochlea, Larger, Consists of bone marrow or air cells.
 2. **Posterior Petrous apex**: medial to SCC, Smaller, dense bone





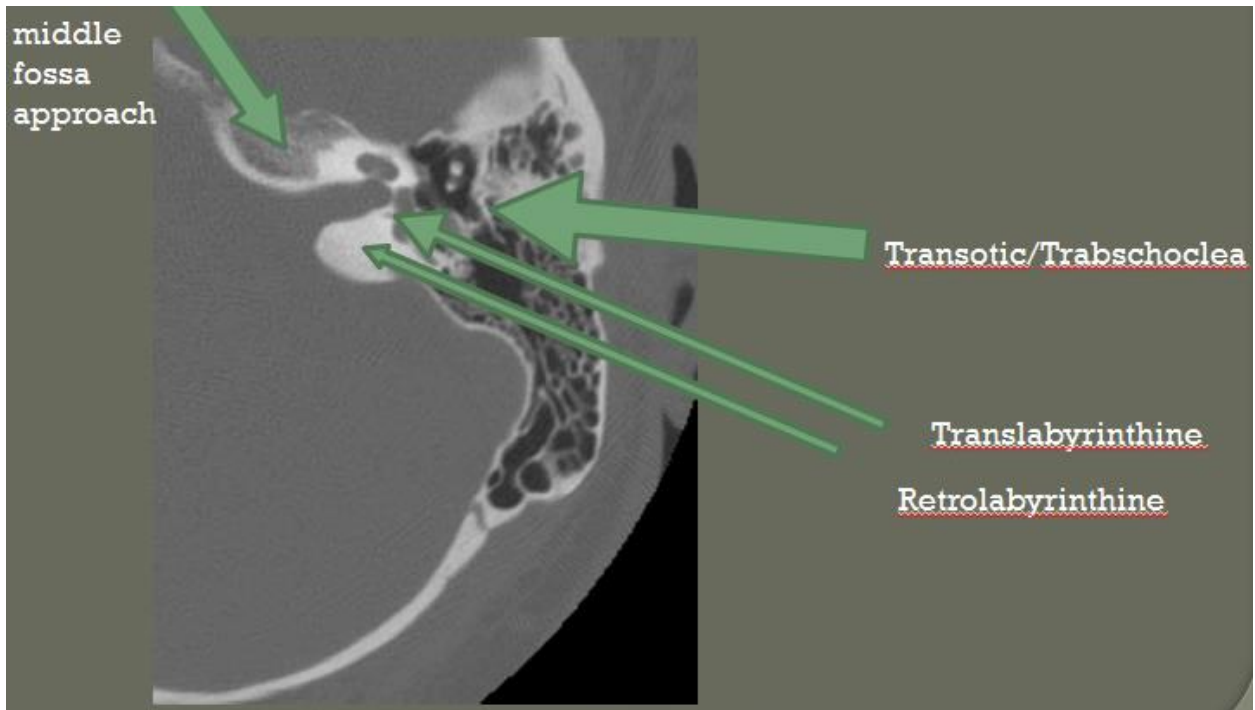
- Petroclinoid ligament Extending from the tip of the petrous apex to the clinoid.
- The abducens nerve travels below the petroclinoid ligament in a small canal called the **Dorello canal**.

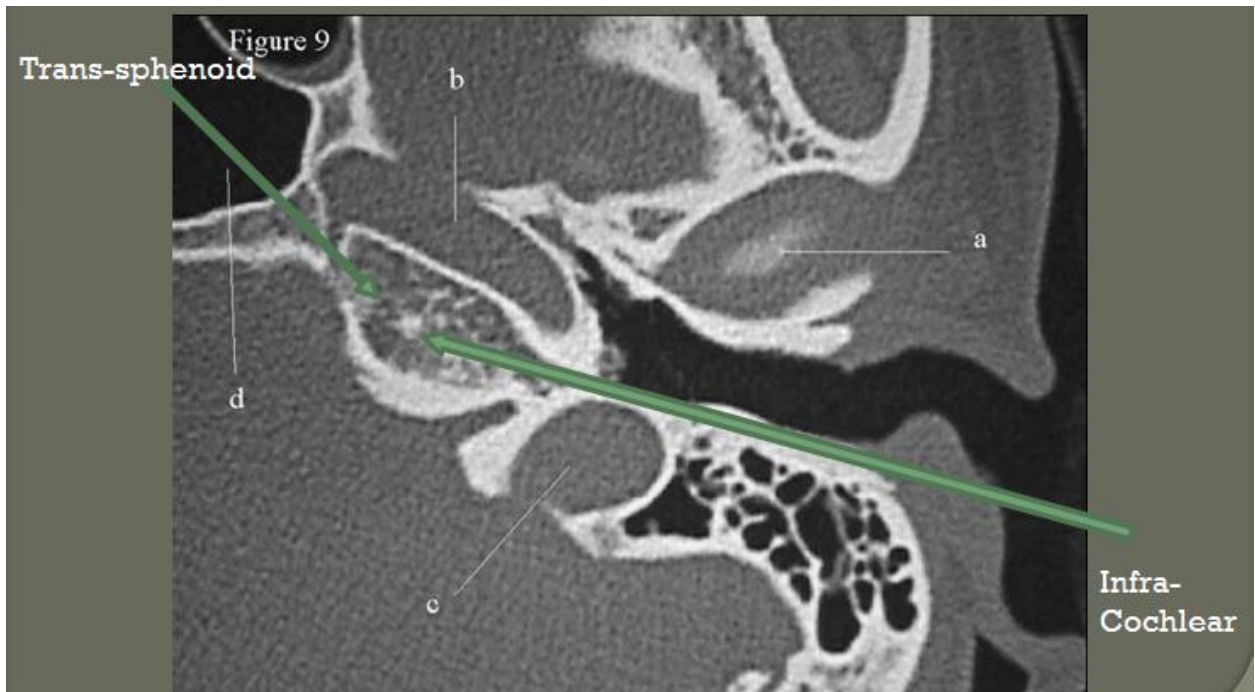
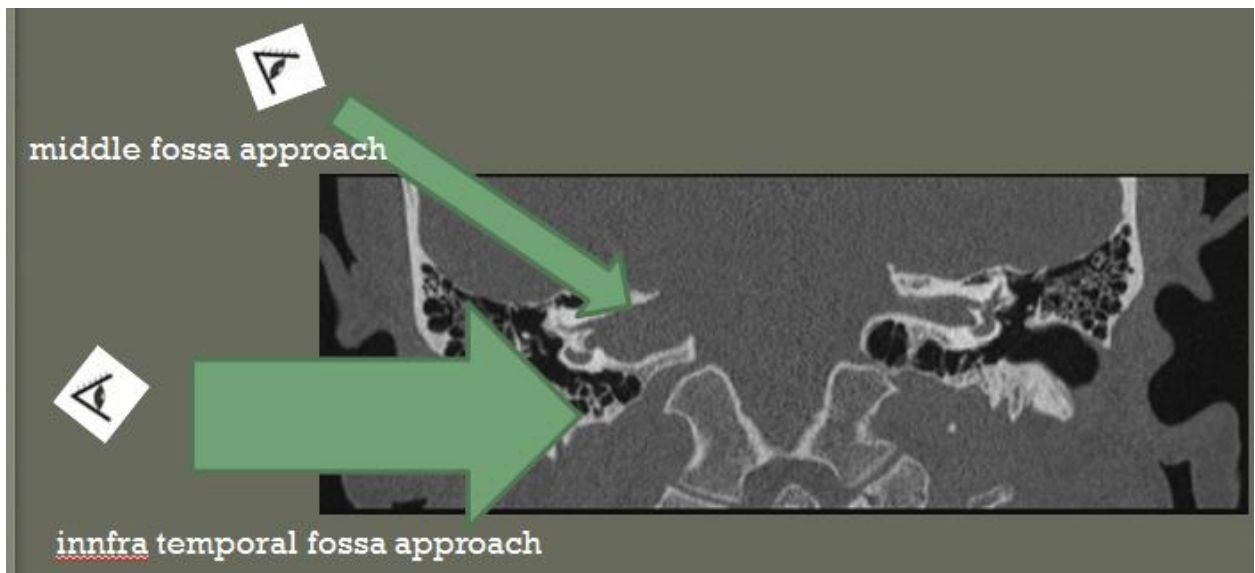


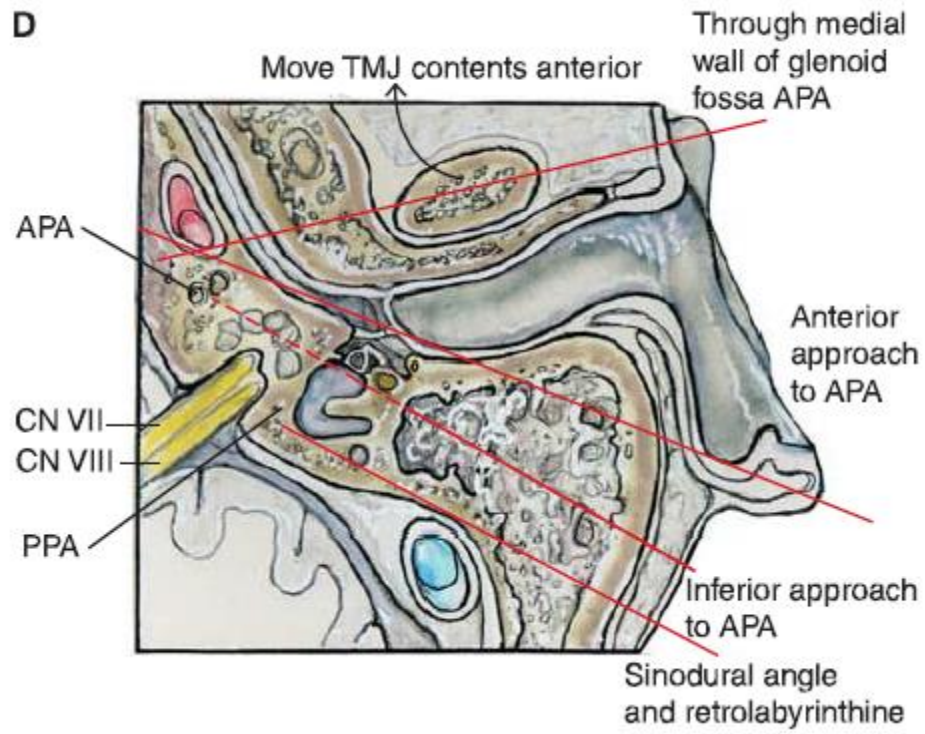
- Air cell tracts extend to the apex **below, above, posterior, and anterior** to the labyrinth
- Direct extension of infection from the mastoid and middle ear through pneumatized air-cell tracts into the petrous apex

- Access to the petrous apex depends on the specific anatomy:
 - May be obtained via the retrolabyrinthine infracochlear, or subarcuate **air cell tracts**.
 - Most patients, the petrous apex can be drained via a CWU approach to the temporal bone.
 - Some may be necessary to perform a canal wall down mastoidectomy or possibly a middle cranial fossa approach to access this area
 - Typically, the margins of the exposure include the cochlea superiorly, the carotid artery anteriorly, and the internal jugular vein and bulb posteriorly.
- **Surgical approach to the petrous apex** depends on:
 1. Available air-cell pathways "below, above, posterior, and anterior to the labyrinth"
 2. The portion of the apex involved "posterior, and anterior"
 3. Hearing , non-hearing ear "Sparing the otic capsule surgically is preferred in patients with serviceable hearing "
- The classic procedures designed to access the **posterior petrous apex** include:
 1. Infralabyrinthine approach
 2. Translabyrinthine approach "Sinodural"
 3. Retrolabyrinthine approach
 4. Subarcuate "provide drainage via cells that extend over the superior semicircular canal and through the "hole in the doughnut," working through the center of the superior semicircular canal"
- Procedures used to access the **anterior petrous apex** include:
 1. Infracochlear approach.
 2. Transotic approach " FN not mobilized"
 3. Transcochlear approach " transposition of FN"
 4. Middle fossa approach: Used when the disease involves the anterior petrous apex but spares the middle ear and mastoid.
 5. Anterior approach through the glenoid fossa: Described by Ramadier and was popularized by Lempert. The exposure involves removal of the anterior canal wall and condyle of the mandible; exposure of the epitympanum; avulsion of the tensor tympani; opening of the tensor semicanal; and then dissection in the triangle between the carotid artery (posterior), the cochlea (superior), and the middle fossa dura (anterior). This approach can be modified by preserving the anterior canal wall and condyle.))

6. Endoscopic Trans-sphenoidal: the presence of venous sinuses between the petrous apex and sphenoid, such as the cavernous sinus, can make this approach challenging. It can be considered for lesions located in the medial section of the petrous apex abutting and/or prolapsing into the posterior wall of the sphenoid sinus.
- In a non-hearing ear (Passing through Otic capsule):
 - Translabyrinthine
 - Transcochlear (Transotic) approach
 - Provides **superior** access for complete removal of mass lesions
 - One disadvantage of these two approaches is that they could potentially expose the cerebrospinal fluid (CSF) to the infectious process.







Reconstructive Surgeries:

1. Myringoplasty:

- Surgical procedure limited to TM reconstruction only.
- **Simple Closure (Paper Patch) Technique:**
 - o Considered for small perforations.
 - o Requires "Rimming" the perforation (Freshening the edges) to stimulate regrowth.
 - o Subsequent placement of a scaffold material to encourage cellular migration (eg, Cotton disk, onion skin paper, cigarette paper, silastic film, or collagen film).
- **Graft Technique:**
 - o Repair of TM utilizing a tissue graft.
 - o Graft material of choice:
 1. Temporalis fascia (Most common).
 2. Perichondrium.
 3. Periosteum.
 4. Cartilage.
 5. Dura.
 6. Vein.
 7. Fat

- [Graft repair can be done by two techniques:](#)

1. Medial (Underlay) Technique:

- o Margins of perforation are freshened.
- o Graft placed under Annulus and remnant TM and either over or under malleus.
- o Supported by gelfoam in the middle ear.
- o Used for Most TM perforations.
- o Difficult with large anterior perforation.
- o High graft take rate.
- o Less risk of graft blunting
- o Easy technique.
- o Short OR time.
- o Less complication

2. Lateral (Overlay) technique:

- o Margins of perforation are freshened.
- o Graft is placed lateral to Annulus and medial to malleus.
- o Used for Larger perforations (especially Ant. perforation).
- o Gives Excellent exposure.
- o High graft take rate.
- o Longer OR time.
- o Longer healing process.
- o Risk of lateralization and anterior blunting of graft.
- o Risk of entering glenoid fossa.
- o Greater postoperative CHL

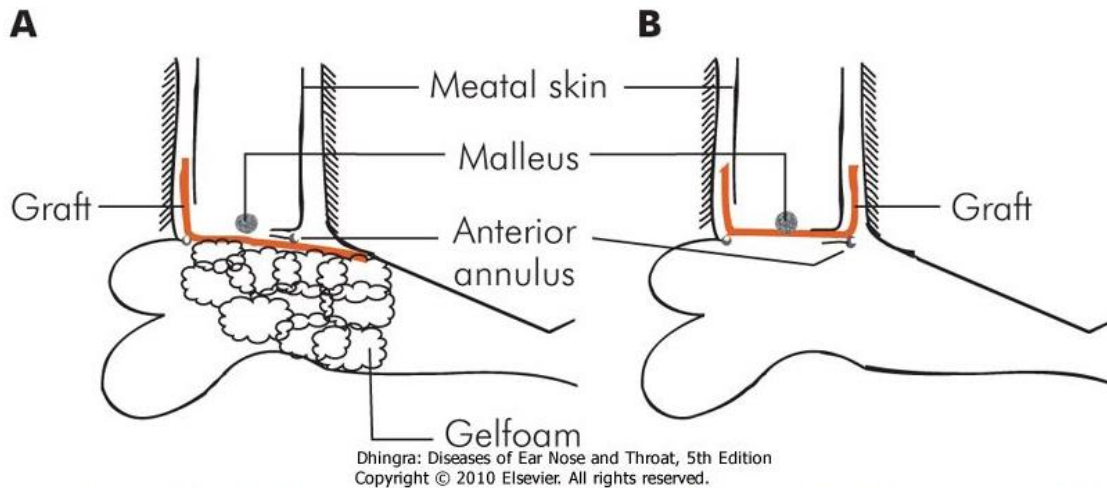


Figure 5.3 Myringoplasty. (A) Underlay technique-fascia graft is under the anterior annulus. It is supported by gelfoam in the middle ear to prevent medial displacement. (B) Overlay technique-fascia graft lies lateral to anterior annulus onto the anterior bony canal wall. It is placed medial to malleus handle to prevent lateralization.

2. Tympanoplasty:

- Operation to:

1. Exploration of Middle ear and eradication of disease.
2. Reconstruction of middle ear.

- TM epithelium shed in a centrifugal direction from the umbo.

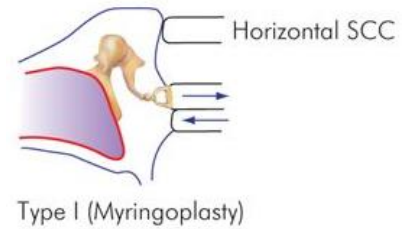
- From the physiology of hearing mechanism, the following principles can be deduced to restore hearing surgically:

1. Intact tympanic membrane:
 - Provides large hydraulic ratio between the tympanic membrane and stapes footplate.
2. Ossicular chain:
 - Conducts sound from tympanic membrane to the oval window.
3. Two functioning windows:
 - Oval window on the scala vestibuli (to receive sound vibrations).
 - Round window on the scala tympani (to act as a relief window).
 - If it is only one window, as in stapes fixation or closure of round window, there will be no movement of cochlear fluids resulting in conductive hearing loss.
4. Acoustic separation of two windows:
 - Sound does not reach both the windows simultaneously.
 - Achieved by providing an intact tympanic membrane, preferential pathway to one window (usually the oval) by providing ossicular chain and by the presence of air in the middle ear.
5. Functioning eustachian tube:
 - Provides aeration to the middle ear.
6. Functioning sensorineural apparatus (cochlea and CN-VIII).

- **Wullstein Classification of Tympanoplasty:**

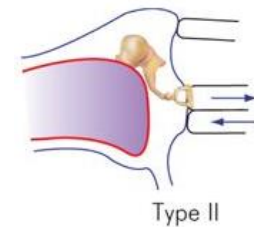
1. Type I:

- Perforation of TM only.
- Repaired with a graft to intact ossicular chain.
- Also called **Myringoplasty**.



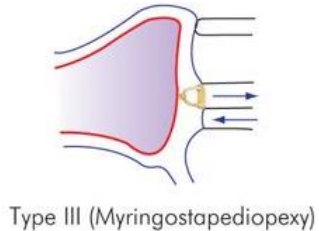
2. Type II:

- Perforation of TM + Erosion of Malleus.
- Repaired with a graft placed on Incus or remnant of malleus.



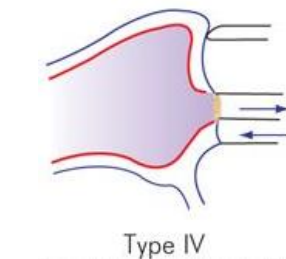
3. Type III:

- Perforation of TM + Erosion of Malleus + Incus.
- Repaired with a graft placed on Stapes Suprastructure.
- Also called Myringostapediopexy or Columella Tympanoplasty.



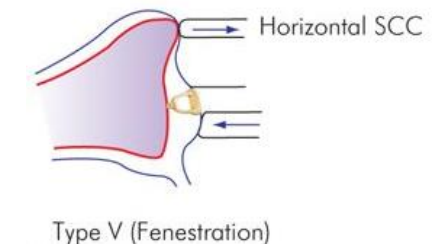
4. Type IV:

- Perforation of TM + Erosion of Malleus + Incus + Stapes Suprastructures.
- Only mobile footplate of Stapes is present.
- Repaired with a graft placed on mobile stapes footplate.



5. Type V:

- Stapes footplate is fixed.
- **Type A:**
 - Graft to Horizontal SCC after fenestration.
- **Type B:**
 - Graft to Oval window after Stapedectomy.



3. Ossiculoplasty:

- Ossicular Chain Reconstruction (OCR).
- Required when there is destruction or fixation of ossicular chain.
- Reestablishes sound conduction mechanism from TM to inner ear fluids
- Ossicular disruption most commonly occurs at [Incudostapedial joint](#) secondary to necrosis of the lenticular process (most susceptible site of avascular necrosis).

- **Most common causes of Malleus Head Fixation**

- Otosclerosis
- Tympanosclerosis
- Postinfectious
- Congenital fixation
- Trauma?

- **Incus Replacment Prosthesis:**

- Short prosthesis with notch.
- **Malleus (+)**
- **Stapes (+)**

- **Incus-Stapes Replacment Prosthesis:**

- Long prosthesis with notch.
- **Malleus (+)**
- **Stapes (-)**

- **Partial Ossicular Replacement Prosthesis (PORP):**

- Short prosthesis without notch.
- **Malleus (-)**
- **Stapes (+)**

- **Total Ossicular Replacement Prosthesis (TORP):**

- Long prosthesis without notch.
- **Malleus (-)**
- **Stapes (-)**

- [Incus Replacement Techniques](#)

1. Transposed or Sculptured Incus Autograft:

- Incus is removed, resculptured, and placed between the malleus and the stapes suprastructure

2. Homograft:

- Well tolerated.
- Provides excellent sound conduction.
- May be presculpted
- Requires storage
- Risk of disease transfer.

3. Synthetic Incus Strut:

- Constructed from a variety of materials (eg, titanium, hydroxyapatite, porous polyethylene).
- Recreate the connection from the malleus to the stapes suprastructure.

4. Partial Ossicular Replacement Prosthesis (PORP):

- Replaces malleus and incus.
- Connect the TM to the stapes capitulum.
- Constructed from a variety of material (eg titanium, hydroxyapatite, porous polyethylene with a cartilagenous cap)

- [Incus-Stapes Replacement Techniques](#)

1. Transposed or Sculptured Incus Autograft:

- Incus is removed, resculptured, and placed between the malleus and the stapes footplate.

2. Homograft:

- Well tolerated.
- Provides excellent sound conduction.
- May be presculpted
- Requires storage
- Risk of disease transfer.

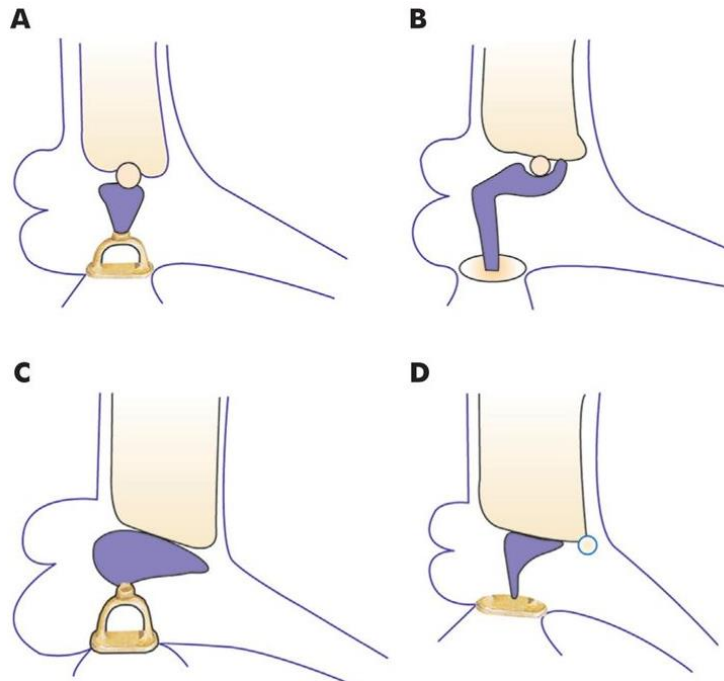
3. Synthetic Incus-Stapes Strut:

- Constructed from a variety of materials (eg, titanium, hydroxyapatite, porous polyethylene).
- Recreate the connection from malleus to the stapes footplate.

4. Total Ossicular Replacement Prosthesis (TORP):

- Replaces malleus, incus and staped suprastructures
- Connect the TM to the stapes footplate.
- Constructed from a variety of material (eg titanium, hydroxyapatite, porous polyethylene with a cartilagenous cap).

- [Most common ossicular fixations:](#)
- **Ankylosis of stapes footplate:**
 - Otosclerosis.
 - Corrected by removal of the fixed stapes and its replacement by a prosthesis.
- **Fixation of head of malleus in attic:**
 - Congenital or acquired.
 - Corrected by removal of the head of malleus and entire incus and then establishing contact between handle of malleus and the stapes.



Dhingra: Diseases of Ear Nose and Throat, 5th Edition
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Figure 5.4 Ossicular reconstruction. Sculptured autograft or homograft ossicles have been used. (A) Malleus-Stapes assembly. Modified incus graft connecting malleus handle with stapes head. (B) Malleus-Footplate assembly. Modified malleus connecting malleus handle with stapes footplate. (C) Modified incus connecting TM to stapes head. Malleus is missing. (D) Modified incus connecting TM to stapes footplate.

Ossiculoplasty

Ossicular Chain Reconstruction (OCR)

- Required when there is **Discontinuity** or **Fixation** of ossicular chain.
- Ossicular disruption most commonly occurs at **Incudostapedial joint** secondary to necrosis of the lenticular process (most susceptible site of avascular necrosis).

➤ **Austine classification:**

Group A

- **Malleus (+)**
- **Stapes (+)**

Group B

- **Malleus (+)**
- **Stapes (-)**

Group C

- **Malleus (-)**
- **Stapes (+)**

Group D

- **Malleus (-)**
- **Stapes (-)**

- #### ➤ **Kartush added three more classes as a modification of this scheme in include ossicular fixity even when all three ossicles are present.**

O - **Intact** ossicular chain

E - Ossicular **head** fixation

F - Stapes **fixation**

➤ **The Middle ear Risk index (Kartush)** “determined parameters for OCR outcome”

✓ **Otorrhea**

- Dry 0
- Occasionally wet 1
- Persistently wet 2
- Wet, cleft palate 3

✓ **Perforation**

- Absent 0
- Present 1

○ **Cholesteatoma**

- Absent 0
- Present 1

○ **Ossicular status**

- M+ I+ S+ 0
- M+ S+ 1
- M+ S- 2
- M- S+ 3
- M- S- 4

○ **Malleus head fixation**

- Absent 0
- Present 1

○ **Stapes fixation**

- Absent 0
- Present 3

○ **Granulations or effusion**

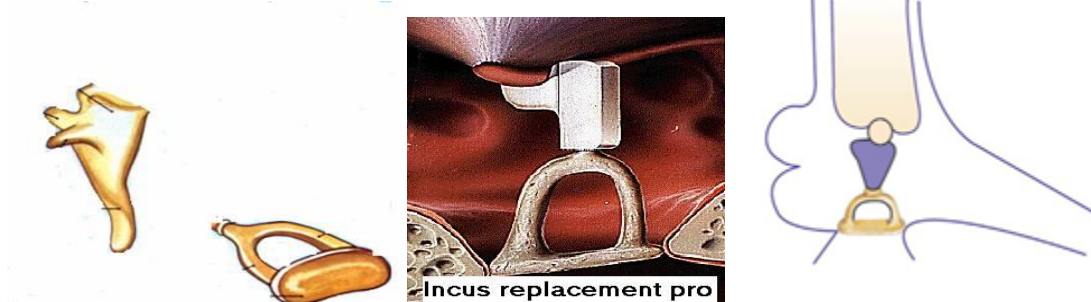
- No 0
- Yes 1

○ **Previous surgery**

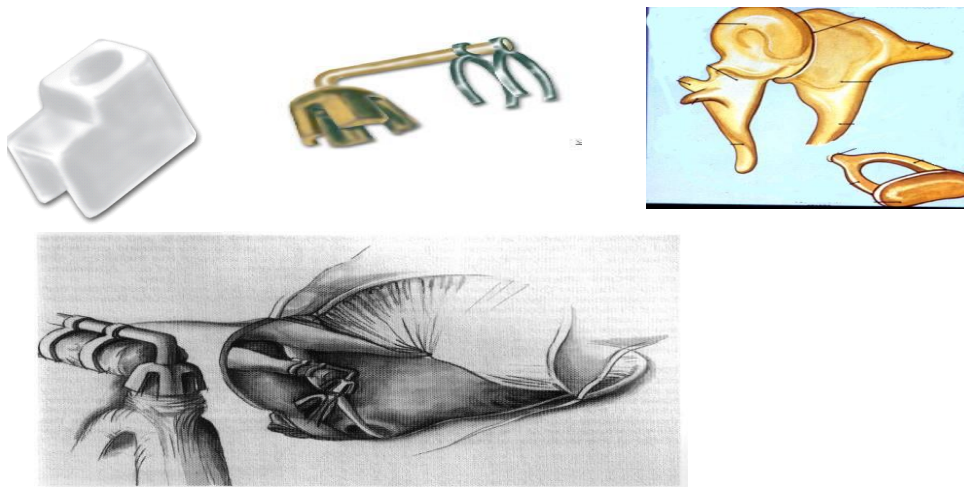
- None 0
- Staged 1
- Revision 2

- **Incus Replacment Prosthesis:**

- Short prosthesis with notch.
- **Malleus (+)**
- **Stapes (+)**

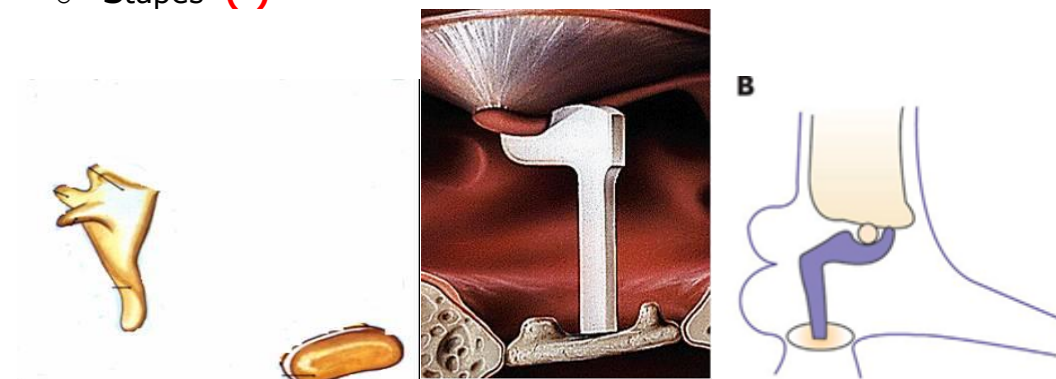


- In case Erosion of incudo-stapedial joint only
 - Can use Applebaum or Titanium prosthesis
 - Or
 - Remove the incus and treat as M+S+ defect



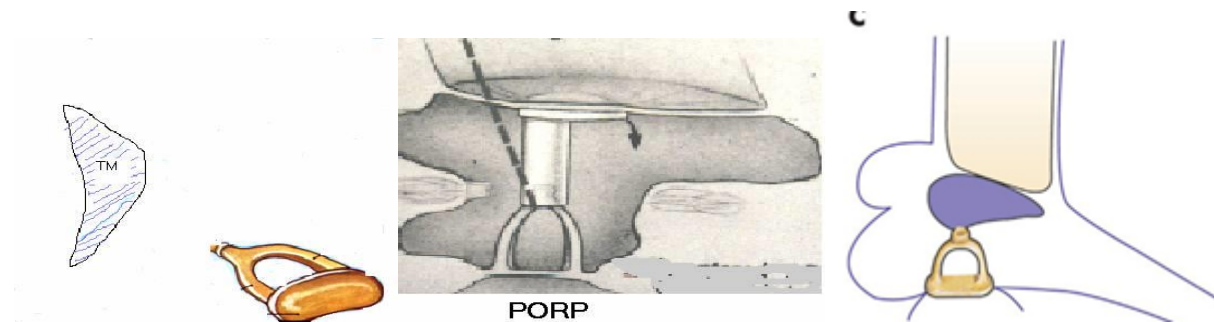
- **Incus-Stapes Replacment Prosthesis:**

- Long prosthesis with notch.
- **Malleus (+)**
- **Stapes (-)**



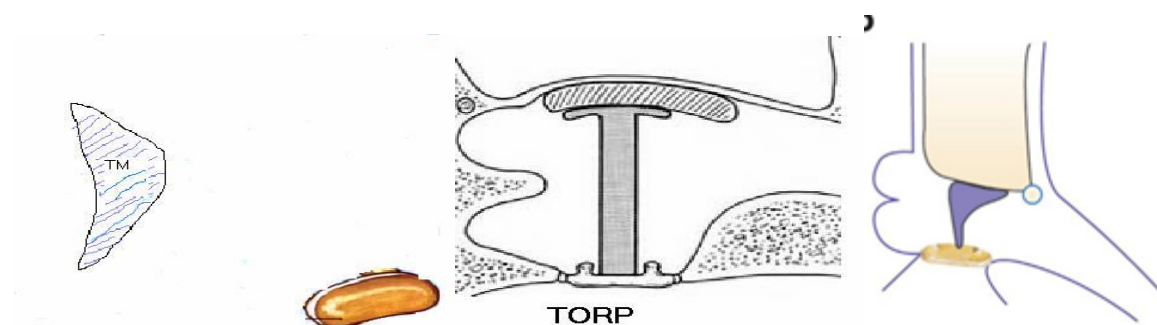
- **Partial Ossicular Replacement Prosthesis (PORP):**

- Short prosthesis without notch.
- Used when the stapes **superstructure** is present
- Connect the TM to the **stapes capitulum**
- **Malleus (-)**
- **Stapes (+)**



- **Total Ossicular Replacement Prosthesis (TORP):**

- Long prosthesis without notch.
- Used when the stapes **superstructure** is absent
- Connect the TM to the **footplate**
- **Malleus (-)**
- **Stapes (-)**



- [Incus Replacement Techniques](#)

1. Transposed or Reshape Incus Autograft:

- Incus is removed, reshape, and placed between the malleus and the stapes suprastructure

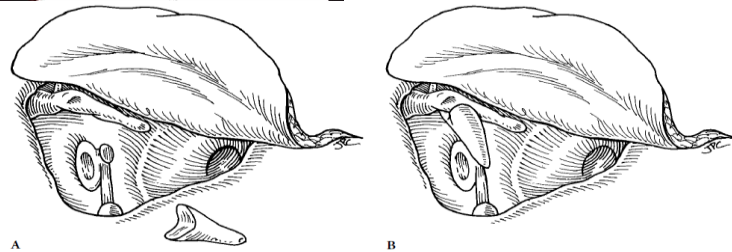
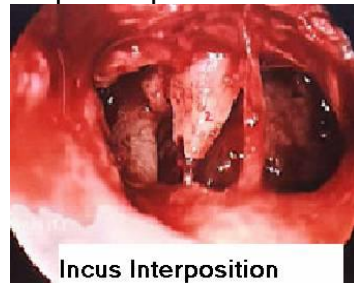


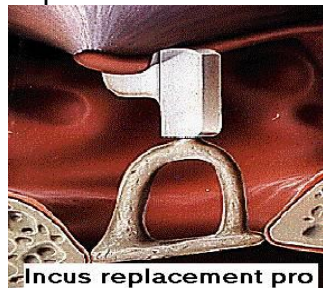
Figure 4-6 Incus interposition. A. The incus is removed and sculpted. B. The sculpted incus is inserted between the malleus and head of the stapes.

2. Homograft:

- Well tolerated.
- Provides excellent sound conduction.
- Requires storage
- Risk of disease transfer.

3. Synthetic Incus Strut:

- Constructed from a variety of materials (eg, titanium, hydroxyapatite, porous polyethylene).
- Recreate the connection from the malleus to the stapes suprastructure.



4. Partial Ossicular Replacement Prosthesis (PORP):

- Replaces malleus and incus.
- Connect the TM to the **stapes capitulum**.
- Cartilage graft is placed between the PORP and tympanic membrane to reduce the chance of extrusion (except [hydroxyapatite](#))
- Constructed from a variety of material (eg titanium, hydroxyapatite, polyethylene ,plastipore with a cartilagenous cap)

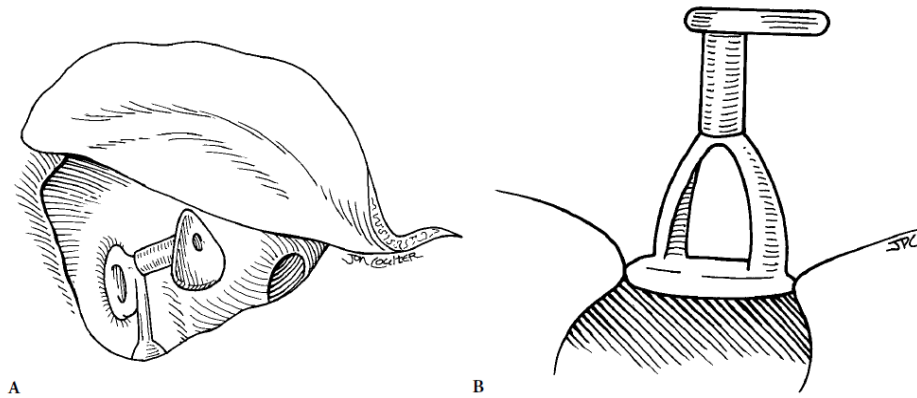
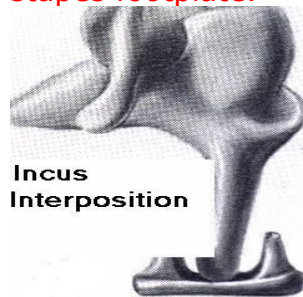


Figure 4-7 Placement of a partial ossicular replacement prosthesis (PORP). A, Surgeon's view of PORP in place. B, Lateral view of the PORP positioned on the stapes head.

- Incus-Stapes Replacement Techniques

1. **Transposed or Reshape Incus Autograft:**

- Incus is removed, reshape, and placed between the malleus and the stapes footplate.



2. **Homograft:**

- Well tolerated.
- Provides excellent sound conduction.
- Requires storage
- Risk of disease transfer.

3. **Synthetic Incus-Stapes Strut:**

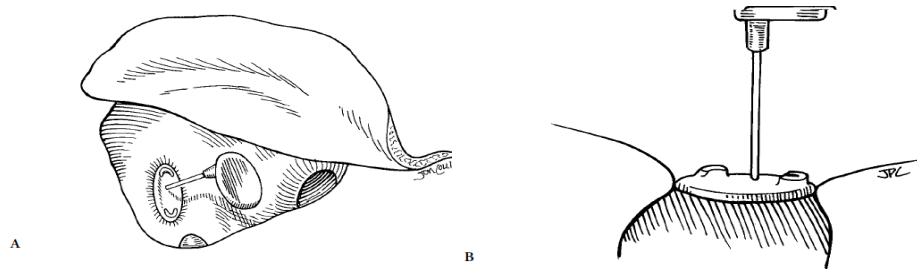
- Constructed from a variety of materials (eg, titanium, hydroxyapatite, polyethylene).
- Recreate the connection from malleus to the stapes footplate.



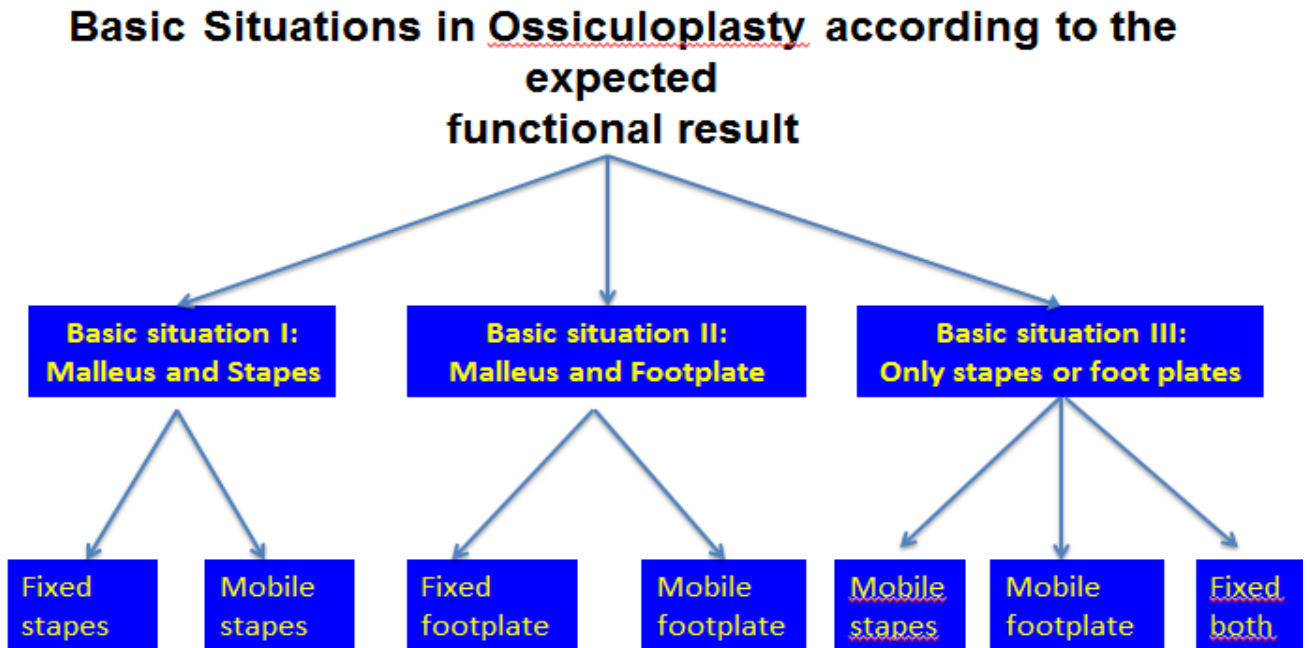
4. Total Ossicular Replacement Prosthesis (TORP):

- Replaces malleus, incus and staped suprastructures
- Connect the TM to the stapes footplate.
- cartilage graft is placed between the TORP and tympanic membrane to reduce the chance of extrusion (except [hydroxyapatite](#))
- Constructed from a variety of material (eg titanium, hydroxyapatite, polyethylene ,plastipore with a cartilagenous cap)

Figure 4-8 Placement of a total ossicular replacement prosthesis (TORP). A, Surgeon's view of TORP in place. B, Lateral view of the TORP positioned on the stapes footplate.



- [Most common ossicular fixations:](#)
 - **Ankylosis of stapes footplate:**
 - Otosclerosis.
 - **Corrected by stapedectomy ,stapedectomy and its replacement by a prosthesis.**
 - **Fixation of head of malleus in attic:** most common
 - Congenital fixation
 - Acquired (Otosclerosis ,, Tympanosclerosis ,, Postinfectious ,, surgical Trauma temporal bone fracture)
 - **Corrected by removal of the head of malleus and entire incus and then establishing contact between handle of malleus and the stapes.**
 - **Isolated Fixation of the incus**
 - **Combined fixation of the malleus and incus**



➤ **Situation 1**

✓ **Mobile stapes**

- Removal and reshaped incus
- Synthetics (Alloplast) incus Strut

✓ **Fixed stapes**

- Incus Replacement with Stapedotomy (IRS)
 - Single stage if tympanic membrane
 - 2nd stage if perforated

➤ **Situation 2**

✓ **Mobile stapes**

- Removal and reshaped incus
- Synthetic Incus-Stapes Strut

✓ **Fixed stapes**

- Incus - Stapes Replacement with Stapedotomy
 - Single stage if tympanic membrane
 - 2nd stage if perforated

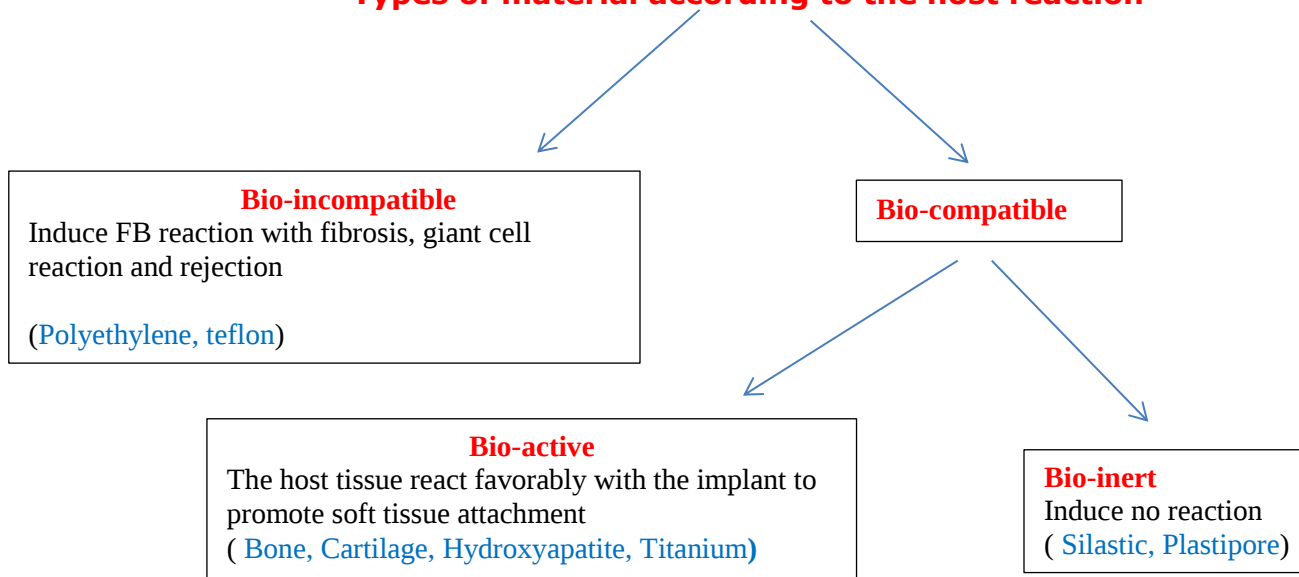
➤ **Situation 3**

✓ **Mobile stapes** : PORP

✓ **Mobile footplate** :- TORP

✓ **Fixed:- Stapedotomy** + TORP

Types of material according to the host reaction



➤ **Prosthetic materials**

- Natural Grafts "bone " (Autograft, allograft):
 - Advantages
 - Biocompatible
 - Good sound conduction
 - Cheap
 - Available
 - Disadvantages
 - May harbor disease
 - Osseous fixation (can placed in direct contact with cochlea)
 - Requires time & skill
 - Transmits disease , Storage requirement (allograft)
- Synthetics (Alloplast):
 - Advantages
 - Ready made (save time)
 - Easy storage
 - Disadvantages
 - Variable bio-compatibility
- **Plastipore®**
 - Easy to trim
 - Bio-Inert unless placed in direct contact with TM
 - No osseous fixation "
- **Hydroxyapatite**
 - Compatible if placed against TM with no cartilage
 - Osseous fixation
 - Not easy to trim

- **Titanium**

- Osseous fixation
- Light and strong
- Easy to handle
- Needs cartilage interposition

➤ **Criteria of the ideal material**

- Bio-compatible
- Shape – size ,weight
- Efficient sound conduction
- Maintains results over time
- Resists and tolerates infection
- Easy to use
- cost

➤ **What are the reasons for OCR failure ?**

- ✓ **Disease :-** recurrent , residual
- ✓ **Prosthesis failure :-**
 - Extrusion (improper size , design flow)
 - Movement (at lateral or medial connection)
- ✓ **Surgeon factor**

Complications of Otitis Media

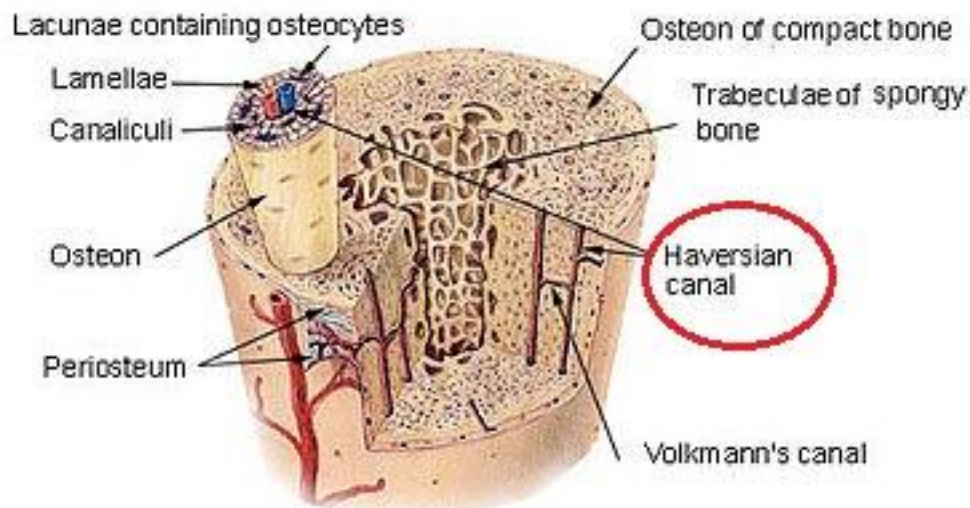
❖ Factors Influencing Development of Complications

- **1. Age**
 - ✓ Most of the complications occur in the **first decade of life** or in the **elderly** when the patient's resistance is low.
- **2. Poor socio-economic group**
 - ✓ Several factors play an important part. such as :
 - a. overcrowding
 - b. poor health education (lack of awareness about middle ear discharge is still being considered merely a nuisance rather than a potentially dangerous condition)
 - c. poor personal hygiene
 - d. limited access to health care
- **3. Virulence of organisms**
 - ✓ Many organisms are developing resistance to antibiotics
 - ✓ Acute infections are either **not controlled** or progress to **subacute or chronic otitis media**.
 - ✓ **Insufficient dose, less effective drug** or **insufficient period** of administration of antibiotic can cause complications.
 - ✓ **Streptococcus pneumoniae** type III (earlier called pneumococcus type III) is very virulent due to production of autolysin and pneumolysin.
 - ✓ **H. influenzae ,Pseudomonas , MRSA** is developing resistance to β -lactam antibiotics .
- **4. Immune-compromised host**
 - ✓ Patients suffering from **AIDS, uncontrolled diabetes, transplant patients receiving immunosuppressive drugs, cancer patients receiving chemotherapy** are more prone to develop complications.
- **5. Preformed pathways**
 - ✓ **Infection** can easily travel beyond the middle ear cleft **if preformed pathways exist** like :-
 - Dehiscence of bony facial canal,
 - previous ear surgery
 - fracture of temporal bone
 - stapedectomy,
 - perilymph fistula
 - congenitally enlarged aqueduct of vestibule (as in Mondini's abnormality of inner ear)
 - dehiscence in the floor of middle ear.
- **6. Cholesteatoma**
 - ✓ Osteitis or granulation tissue in chronic otitis media **destroy the bone and help infection to penetrate deeper**.
 - ✓ In acute and chronic middle ear infection, disease process is limited only to the **mucoperiosteal lining of the cleft** but **if it spreads into the bony walls of the cleft or beyond it, various complications can arise**.

❖ Pathways of Spread of Infection

- ✓ The most common pathway for infection to extend beyond the mastoid is through the lateral cortex behind the ear.
- ✓ Less commonly, it can extend to the soft tissues in the upper portion of the neck (see the section on [Bezold abscess](#))
- ✓ rarely, to the soft tissue anterior and superior to the auricle
- **1. Direct bone erosion**
 - ✓ **In acute infections**, it is the process of **hyperaemic of bone** causes **decalcification**.
 - ✓ In **chronic infection**, it may be **osteitis**, **erosion by cholesteatoma** or **granulation tissue**.
- **2. Venous thrombophlebitis**
 - ✓ **Veins of Haversian canals** are connected with **dural veins** which in turn connect with **dural venous sinuses** and **superficial veins of brain**.

Compact Bone & Spongy (Cancellous Bone)



- ✓ Thus, infection from the mastoid bone can cause **thrombophlebitis of venous sinuses** and even **cortical vein thrombosis**.
- ✓ This mode of spread is common in acute infections.
- **3. Preformed pathways**
 - **(i) Congenital dehiscences**, e.g. in bony facial canal, floor of middle ear over the jugular bulb.
 - **(ii) Patent sutures**, e.g. petrosquamous suture.
 - **(iii) Previous skull fractures**. The fracture sites heal only by **fibrous scar** which permits infection.
 - **(iv) Surgical defects**, e.g. stapedectomy, fenestration and mastoidectomy with exposure of dura.
 - **(v) Oval and round windows**.
 - **(vi) Infection from labyrinth** can travel along **internal acoustic meatus**, **aqueducts of the vestibule** and **that of the cochlea** to the **meninges**.

❖ **Classification**

Complications of otitis media are classified into two main groups	
Extracranial	Intracranial
Intratemporal 1. Mastoiditis a) Acute Mastoiditis b) Coalescent c) Masked (Latent) Mastoiditis 2. Petrositis 3. Facial Paralysis 4. Labyrinthitis 5. Labyrinthine fistula Extratemporal 1. Subperiosteal abscess associate with mastoiditis 2. Inferior deep neck abscess "Bezold's abscess" associate with mastoiditis	1. Extradural abscess or granulation tissue 2. Subdural abscess 3. Meningitis 4. Brain abscess 5. Lateral " sigmoid" sinus thrombophlebitis (Occluding ,Non occluding) 6. Otitic hydrocephalus.

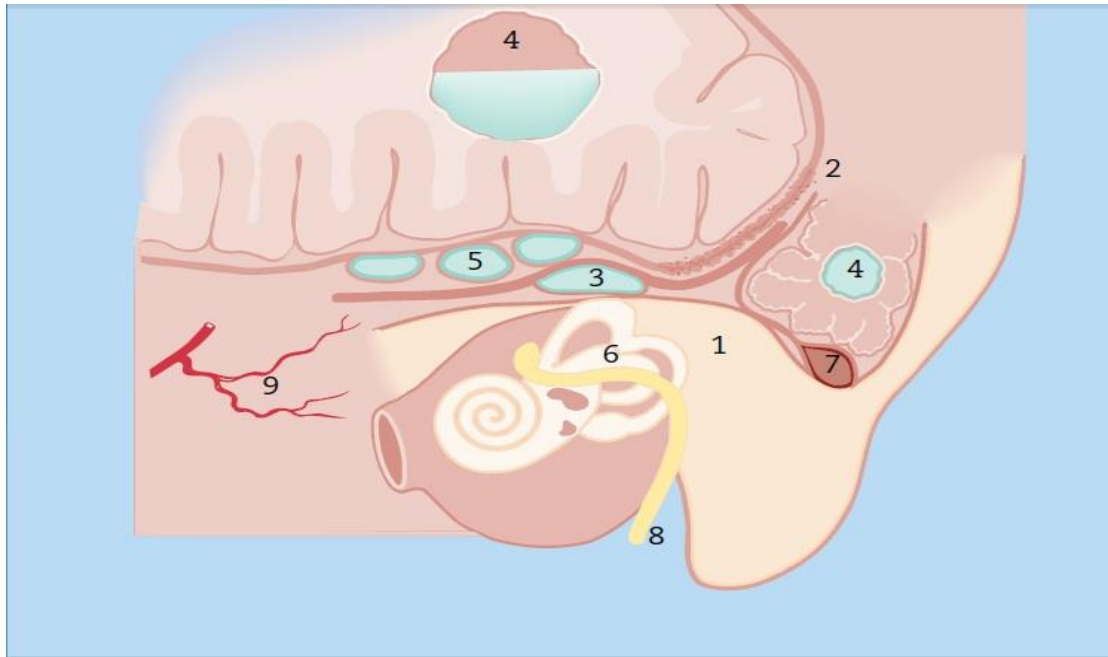


Fig. 10.1 Complications of chronic otitis media. 1, Acute mastoiditis; 2, Meningitis; 3, Extradural abscess; 4, Brain abscess (temporal lobe and cerebellum); 5, Subdural abscess; 6, Labyrinthitis; 7, Lateral sinus thrombosis; 8, Facial nerve paralysis; 9, Petrositis.

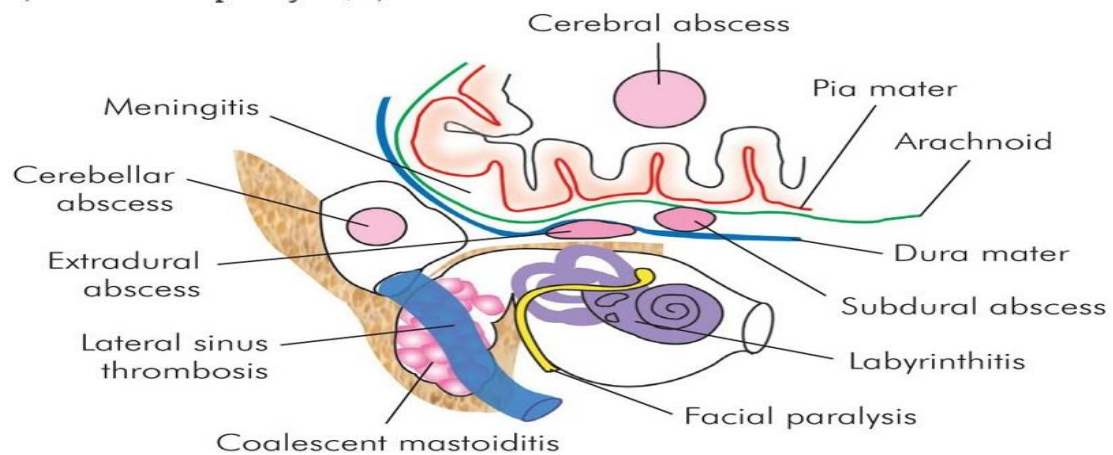


TABLE 149.1	COMPLICATIONS OTITIS MEDIA
Intratemporal (extracranial)	
Mastoiditis	
Acute	
Coalescent	
Chronic	
"Masked"	
Associated with subperiosteal abscess	
Associated with deep neck abscess (Bezold abscess)	
Petrositis	
Labyrinthitis	
Serous or toxic	
Suppurative	
Otogenic	
Meningogenic	
Facial paralysis	
Labyrinthine fistula	
Intracranial	
Extradural granulation tissue or abscess	
Sigmoid sinus thrombophlebitis	
Occluding	
Nonoccluding	
Brain abscess	
Otitic hydrocephalus	
Meningitis	
Subdural abscess	

- Complications secondary AOM are more common in **young children**
- Complications secondary to COM with and without cholesteatoma are more common in **older children and adults**
- 78% of subjects who had complications secondary to COM were found to have cholesteatoma
- Most common extracranial complication from COM & AOM "acute mastoiditis with post auricular abscess" "**Subperiosteal abscess**" "
- 2nd most common extracranial complication from AOM facial palsy
- Most common intracranial complication from AOM & COM meningitis
- 2nd most common intracranial complication of otitis media Brain abscess
- Typically, mastoiditis is the initial complication, with more severe complications developing secondarily

- **Five early Signs of Impending Complication**

1. Persistence of acute infection for 2 weeks
2. Recurrence of symptoms within 2 weeks
3. Bad smell discharge during treatment
4. Acute exacerbation of chronic infection, especially if bad smell
5. Haemophilus influenzae type b or Anaerobes

- **Once a complication has begun, signs and symptoms typically progress rapidly:**

1. **Fever** associated with a chronic perforation (intracranial infection or extracranial cellulitis)
2. **Pinna displaced** inferolaterally and/or edema of the posterosuperior ear canal wall skin (subperiosteal abscess)
3. **Retro-orbital pain** (petrositis)
4. **Vertigo** in a patient with an infected ear (labyrinthitis or labyrinthine fistula)
5. **Facial paralysis** ipsilateral to an infected ear (facial paralysis)
6. **Headache and lethargy** (intracranial complication of any sort)
7. **Meningismus** (meningitis or subdural abscess)
8. **Focal neurologic signs or seizure** (brain abscess)
9. **Global neurologic signs** (subdural abscess or meningitis)

**TABLE
149.2**

**BACTERIOLOGY OF OM
(PATHOGENS LISTED IN ORDER
OF PREVALENCE) (6,7)**

Acute suppurative OM

Streptococcus pneumoniae
Haemophilus influenza (nontypeable)
Moraxella catarrhalis
 Others (far less common)
 Group A Streptococci
 Staphylococcus aureus (infrequent)
 Gram-negative bacilli (infrequent)

Acute mastoiditis

Streptococcus pneumoniae
Pseudomonas aeruginosa
 Group A beta-hemolytic *Streptococci* (e.g., *Streptococcus pyogenes*)
 Coagulase-negative *Staphylococcus* sp.
 Others (far less common)
 Staphylococcus aureus
 Proteus sp.
 Bacteroides sp.

Chronic otorrhea with or without cholesteatoma

Mixed aerobic (also occasionally including *S. pneumoniae*) and anaerobic organisms
 Foul-smelling ears (especially with cholesteatoma) may grow 5–11 organisms, always mixed aerobic and anaerobic
Pseudomonas aeruginosa (most common aerobe)
Staphylococcus aureus and *Staphylococcus epidermidis*
 Other aerobic organisms including *Proteus* sp., *Klebsiella* sp., *Escherichia coli*
 Various anaerobic organisms including *Bacteroides fragilis*

**Intracranial abscesses (brain and subdural) of otogenic origin
(mixed cultures)**

Streptococcus sp.
Staphylococcus sp.
Proteus sp.
 Anaerobic organisms (*Peptococcus*, *Peptostreptococcus*, *Bacteroides fragilis*)

Bacterial meningitis in children

Streptococcus pneumoniae
Haemophilus influenzae, type B
Neisseria meningitidis

A. EXTRACRANIAL COMPLICATIONS OF OTITIS MEDIA

➤ **Intratemporal**

1. Mastoiditis
 - a. Acute Mastoiditis
 - b. Coalescent
 - c. Masked (Latent) Mastoiditis
2. Petrositis
3. Facial Paralysis
4. Labyrinthitis
5. Labyrinthine fistula

➤ **Extratemporal**

1. **Subperiosteal abscess** associate with mastoiditis
2. **Inferior deep neck abscess "Bezold's abscess"** associate with mastoiditis

1. (a) Acute Mastoiditis

- ✓ Inflammation of mucosal lining of **antrum and mastoid air cell system** is **always** accompaniment of acute otitis media and forms a part of it.
- ✓ The term "mastoiditis" is used when infection spreads from the **mucosa lining the mastoid air cells**, to involve **bony walls** of the mastoid air cell system result in breakdown of bony septa and coalescence of air cells (**acute coalescent mastoiditis**)

❖ **Aetiology**

- ✓ Acute mastoiditis usually accompanies or follows **acute suppurative otitis media**
- ✓ Determining factors being:
 - High virulence of organisms
 - Lowered resistance of the patient due poor nutrition or associated systemic disease such as diabetes.
- ✓ Acute mastoiditis is often seen in mastoids with **well-developed air cell system**.
- ✓ **Children** are affected more.
- ✓ **Streptococcus pneumoniae** is the most common offending pathogen
- ✓ Other bacteria frequently implicated in acute mastoiditis include **Streptococcus pyogenes, Staphylococcus aureus, and Haemophilus influenzae**.
- ✓ Anaerobic organisms are also associated with mastoiditis and need antibacterial therapy against them.

❖ Pathology

- ✓ Two main pathological processes are responsible:
 - **1.** Production of pus under tension.
 - **2.** Hyperaemic decalcification and osteoclastic resorption of bony walls.
- Extension of inflammatory process to **mucoperiosteal lining** of air cell system increases the amount of **pus produced** due to large surface area involved.
- Drainage of this pus, through a **small perforation of tympanic membrane and/or eustachian tube**, cannot keep pace with the amount being produced.
- **Swollen mucosa of the antrum and attic** also **impede** the drainage system resulting in **accumulation of pus under tension**.
- Hyperaemia of mucosa causes dissolution of calcium from the bony walls of the mastoid air cells (hyperaemic decalcification).
- ✓ Cause **destruction and coalescence** of mastoid air cells, converting them into a single irregular cavity filled with pus (**Empyema of mastoid**).
- ✓ Pus may break through mastoid cortex leading to **sub-periosteal abscess** which may even burst on surface leading to a **discharging fistula**



❖ **Clinical Features**

➤ **Symptoms** "similar to that of acute suppurative otitis media"

i. **Pain behind the ear.**

- Persistence & throbbing not subsides with establishment of perforation or treatment with antibiotics.

ii. **Fever.**

- **Persistence or recurrence** of fever in a case of acute otitis media, in spite of adequate antibiotic treatment that points to the development of **mastoiditis**.

iii. **Ear discharge**

- **Profuse and increases in purulence.**
- In some cases, discharge may stop due to obstruction to its drainage but other symptoms would worsen
- Any persistence of discharge beyond 2 weeks, in a case of acute otitis media, points to mastoiditis.

➤ **Signs**

i. **Mastoid tenderness.**

- Over the **middle of mastoid process, at its tip, posterior border or the root of zygoma important sign.**
- Tenderness elicited over the **suprameatal triangle** may not be diagnostic of acute mastoiditis as it is seen even in cases of the acute otitis media due to inflammation of mastoid antrum (**antritis**).
- Tenderness should always be compared with that of the healthy side.

ii. **Ear discharge.**

- **Creamy Mucopurulent or purulent discharge**, may be seen coming through a central perforation of pars tensa.

iii. **Sagging of posterosuperior meatal wall.**

- **It is due to periosteitis of bony party wall between the antrum and deeper posterosuperior part of bony canal.**

iv. **Perforation of tympanic membrane.**

- Usually, a **small perforation is seen in pars tensa**
- Sometimes, tympanic membrane is intact but **dull and opaque** especially in those who have received inadequate antibiotics.
- If the tympanic membrane is **normal**, the patient does not have acute mastoiditis.

v. **Swelling over the mastoid.**

- **Initially**, there is oedema of periosteum, imparting a smooth.
- **Later** retroauricular sulcus becomes obliterated and pinna is pushed forward and downwards.
- When pus bursts through bony cortex, a subperiosteal fluctuant abscess is formed



vi. **Hearing loss.** Conductive type of hearing loss is always present.

vii. **General findings.**

- **Ill and toxic** with low-grade fever.
- In children, **fever is high with a rise in pulse rate.**

❖ **Dx**

- Diagnosis often is made **clinical**
- **Contrasted CT of temporal bone**
 - ✓ Coalescence or lack of septations between air cells in mastoid with the presence of fluid or soft tissue
 - ✓ Should be done for patients presenting with mastoiditis symptoms, to aid in therapeutic planning and to rule out other possible complication
- **Blood counts** show polymorphonuclear leucocytosis
- **ESR** is usually raised.
- **Ear swab** for culture and sensitivity.

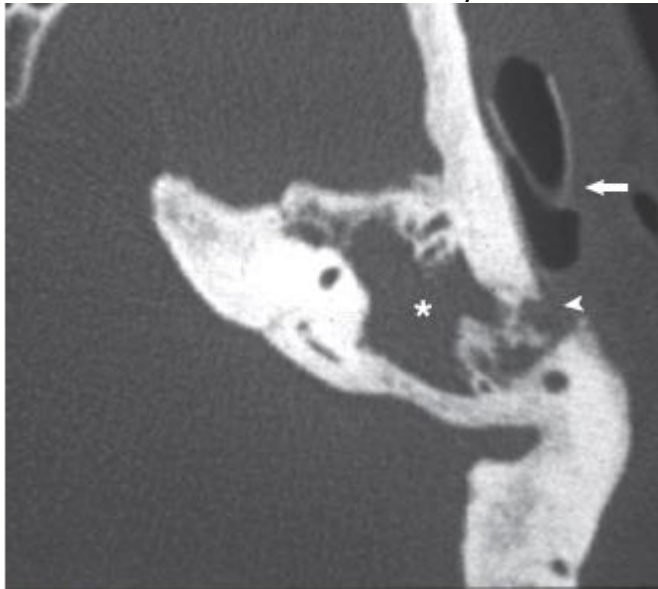


FIGURE 140-5. Axial temporal bone computed tomography scan (soft tissue) shows abscess cavity with gas (arrow). Note cortical defect (arrow-head), mastoid opacification, and loss of bony septa (asterisk).

❖ **Differential Diagnosis**

- i. **Suppuration of mastoid lymph nodes**
 - Scalp infection may cause mastoid lymph node enlargement and then suppuration leading to abscess formation
 - In such cases there is no history of preceding otitis media, ear discharge or deafness.
 - Abscess is usually superficial.
- ii. **Furunculosis of meatus**
- iii. **Infected sebaceous cyst**

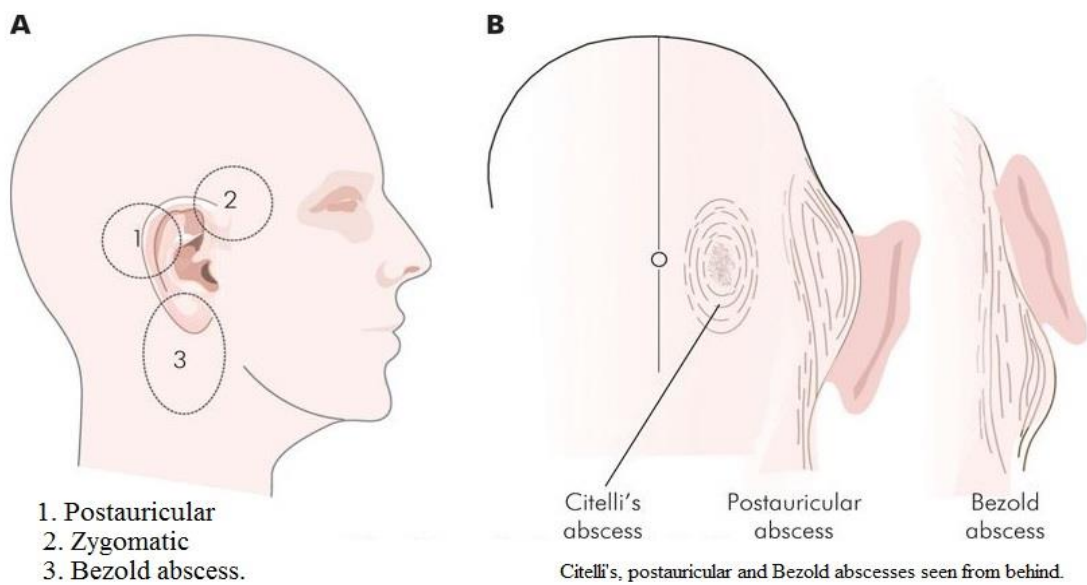
❖ **Treatment**

- **(a) Hospitalization of the patient**
- **(b) Antibiotics**
 - ✓ Start with IV amoxicillin or ampicillin (in the absence of culture and sensitivity).
 - ✓ Specific antimicrobial is started on the receipt of sensitivity report.
 - ✓ Since anaerobic organisms are often present, metronidazole is added.
- **(c) Myringotomy**
 - ✓ Relieved by wide myringotomy tubes & needle aspiration of the abscess without formal drainage
 - ✓ Early cases of acute mastoiditis respond to conservative treatment with antibiotics alone or combined with myringotomy.
 - ✓ Less invasive than mastoidectomy
"experience, 14 of the 17 subjects treated in this manner resolved their abscesses without the need for further intervention, and were discharged home significantly sooner than those subjects who were managed with a mastoidectomy"
- **(d) Cortical mastoidectomy**
 - ✓ Aim of cortical mastoidectomy is to exenterate all the mastoid air cells and remove any pockets of pus granulations
It is indicated when there is:
 - ✓ The object of this operation is to drain the mastoid antrum and air cells but leave the middle ear, the ossicles and the external meatus untouched.
 - ✓ **Indication:**
 1. Abscess resulting from chronic otitis in the presence of a cholesteatoma "bcz no conservative alternatives"
 2. Sagging of posterosuperior meatal wall.
 3. Positive reservoir sign, i.e. meatus immediately fills with pus after it has been mopped out.
 4. No change in condition of patient or it worsens in spite of adequate medical treatment for 48 hours.
 5. Mastoiditis, leading to complications, e.g. facial paralysis, labyrinthitis, intracranial complications.
 - ✓ Antibiotic treatment must be continued at least for 5 days following mastoidectomy.

❖ **Complications of Acute Mastoiditis**

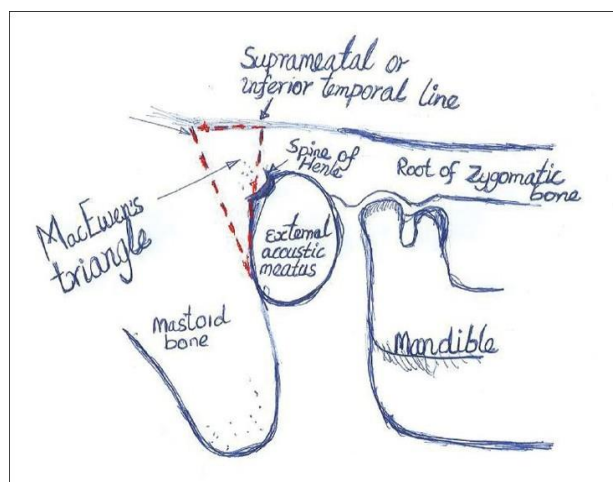
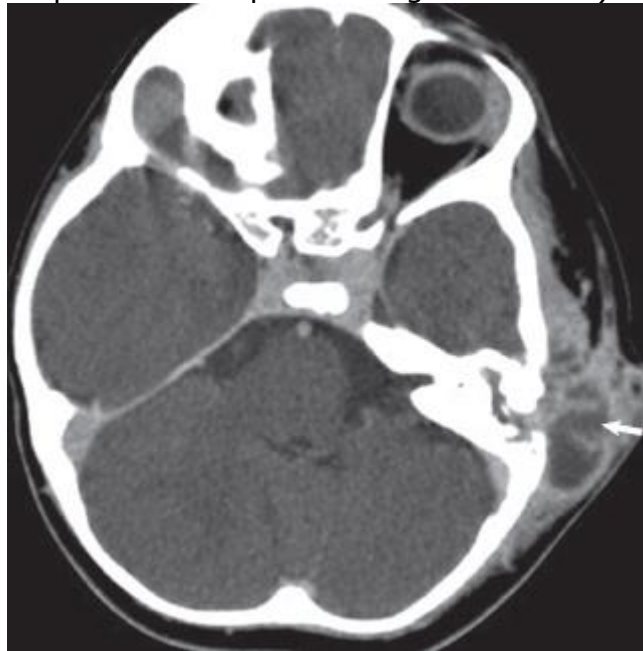
- **1.** Subperiosteal abscess " most common"
- **2.** Labyrinthitis
- **3.** Facial paralysis
- **4.** Petrositis
- **5.** Extradural abscess
- **6.** Subdural abscess
- **7.** Meningitis
- **8.** Brain abscess
- **9.** Lateral sinus thrombophlebitis
- **10.** Otitic hydrocephalous.

❖ **Abscesses in Relation to Mastoid Infection**



○ (a) **Postauricular abscess**

- ✓ **Commonest** abscess that forms over the mastoid "extratemporal complication"
- ✓ Break through mastoid cortex leading to sub-periosteal abscess which may even burst on surface leading to a **discharging fistula**
- ✓ Can also occur as a result of vascular extension secondary to phlebitis of the mastoid veins
- ✓ More commonly in young children with AOM, also found in COM with and without cholesteatoma
- ✓ Pinna is displaced **forwards, outwards and downwards**.
- ✓ In **infants and children**, abscess forms over the **MacEwen's triangle**; pus in these cases travels along the vascular channels of **lamina cribrosa** (mesh-like structures which allows nerve fibres of the optic nerve to pass through the sclera).

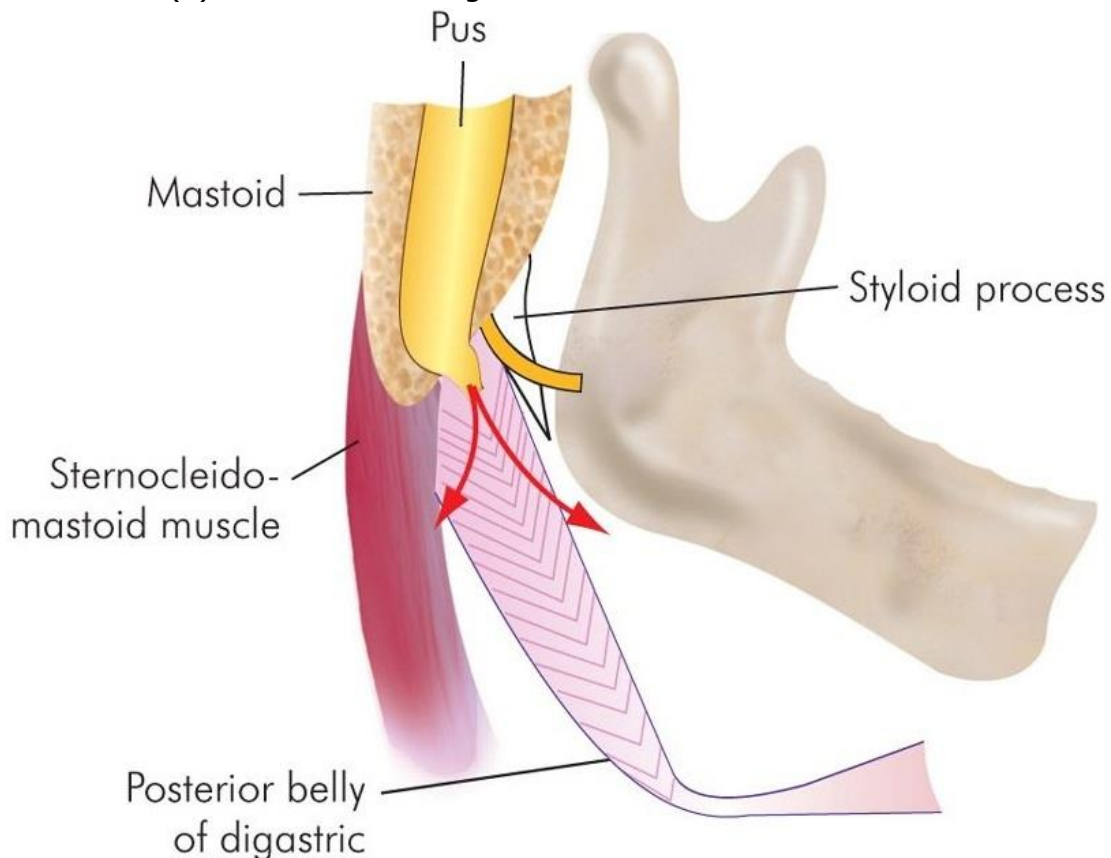


- **(b) Zygomatic abscess**

- ✓ Occurs due to infection of **zygomatic air cells** situated at the **posterior root of zygoma**.
- ✓ Swelling appears in **front of and above the pinna**
- ✓ There is associated **oedema of the upper eyelid**.
- ✓ In these cases, pus collects either **superficial or deep to the temporalis muscle**

- **(c) Bezold abscess**

- ✓ Occur following acute coalescent mastoiditis when **pus breaks through the thin medial side of the tip** of the mastoid and presents as a swelling in the **upper part of neck**.
- ✓ The abscess may:
 - (i) Lie deep to sternocleidomastoid, pushing the muscle outwards
 - (ii) Follow the posterior belly of digastric and present as a swelling between the tip of mastoid and angle of jaw
 - (iii) Present in upper part of posterior triangle
 - (iv) Reach the parapharyngeal space
 - (v) Track down along the carotid vessels

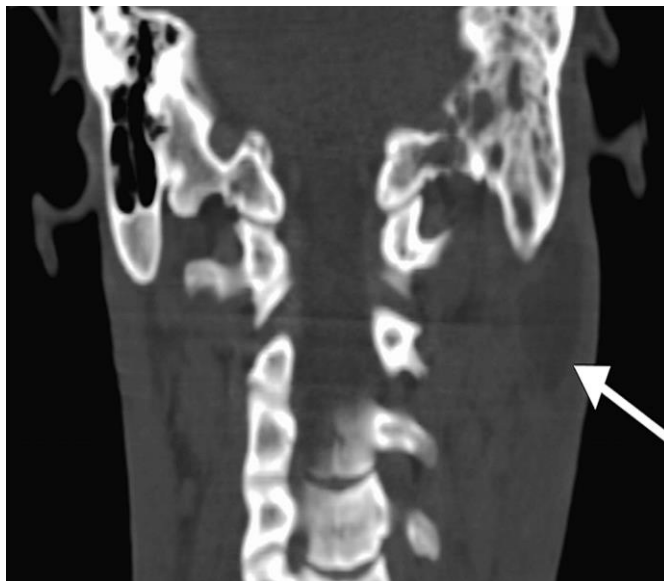


Pus bursting through the medial side of the tip of mastoid and collecting under the sternomastoid or digastric triangle.

✓ **Clinical features of Bezold abscess:**

- Onset is sudden.
- Present as a tender, fever, deep, poorly defined mass in level two of the neck & torticollis.
- **History of purulent otorrhoea.**
- Because the abscess develops from air cells at the tip of the mastoid, it is found in older children and adults
- A Bezold abscess should be differentiated from:
 - Acute upper jugular lymphadenitis.
 - Abscess or a mass in the lower part of the parotid gland.
 - Infected branchial cyst.
 - Parapharyngeal abscess.
 - jugular vein thrombosis.

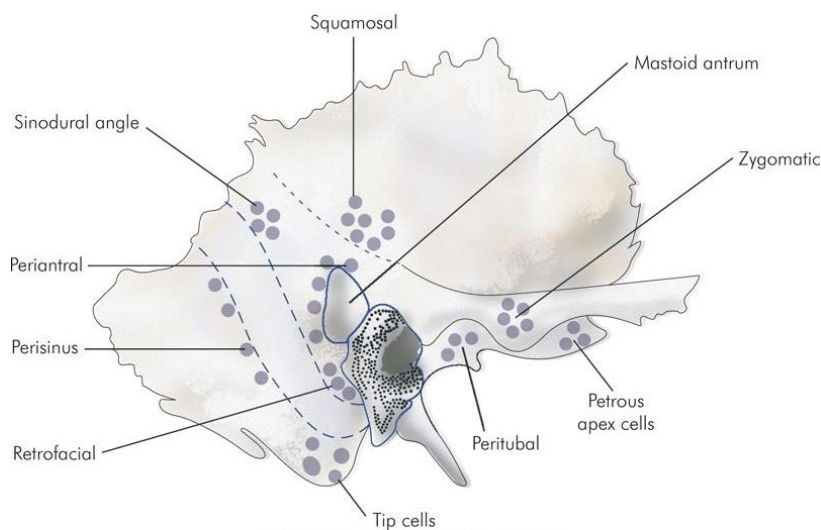
A CT scan of the mastoid and swelling of the neck may establish the diagnosis” abscesses show a rim-enhancing abscess with surrounding inflammation, may demonstrate the bony dehiscence in the tip of the mastoid, and can help in operative planning” .



✓ **Treatment of Bezold abscess**

1. **Cortical mastoidectomy** for coalescent mastoiditis with careful exploration of the tip for a fistulous opening into the soft tissues of the neck.
2. **Drainage of the neck abscess** “transcervical approach”
3. **Intravenous antibiotics** guided by the culture and sensitivity report of the pus taken at the time of surgery.

- **(d) Meatal abscess (Luc's abscess)**
 - ✓ Pus breaks through the bony wall between the **antrum and external osseous meatus**.
 - ✓ **Swelling** is seen in deep part of bony meatus.
 - ✓ **Abscess** may burst into the meatus.
- **(e) Behind the mastoid (Citelli's abscess)**
 - ✓ Abscess is formed behind the mastoid more towards the occipital bone (compare postauricular mastoid abscess which forms over the mastoid).
- **(f) Parapharyngeal or retropharyngeal abscess**
 - ✓ This results from infection of the **peritubal cells** due to acute coalescent mastoiditis.



(b) Coalescent Mastoiditis

- ✓ Implies breakdown and decalcification of the bony septa within the mastoid,
- ✓ Progressing to bony destruction of the cortex or other aspects of the mastoid bone
- ✓ If AOM and mastoiditis that persist for 2 to 4 weeks , coalescent mastoiditis develops.
- ✓ Typically affects boys, 4 years or younger with AOM. who have previously **well-aerated mastoids**
- ✓ Rarely occurs in the setting of COM , **OR** in children with poorly pneumatized mastoids "sclerotic"
- ✓ **Up to 25%** of patients presenting with coalescent mastoiditis will have a concomitant intracranial complication

❖ Aetiology

- ✓ *S. pneumoniae* is the most common offending Organism
- ✓ 30% will culture positive for anaerobic organisms

❖ Pathophysiology

- ✓ Same above " acute mastoiditis"

❖ Clinical Features

- ✓ Fever
- ✓ Otalgia
- ✓ purulent otorrhea
- ✓ mastoid pain
- ✓ tenderness
- ✓ erythema, and/ or edema

❖ Diagnosis.

- ✓ **CT:** breakdown of bony septa. loss of cortical bone, and opacification of the air cell system.
- ✓ **MRI:** should be obtained if there is any suspicion of intracranial complication.

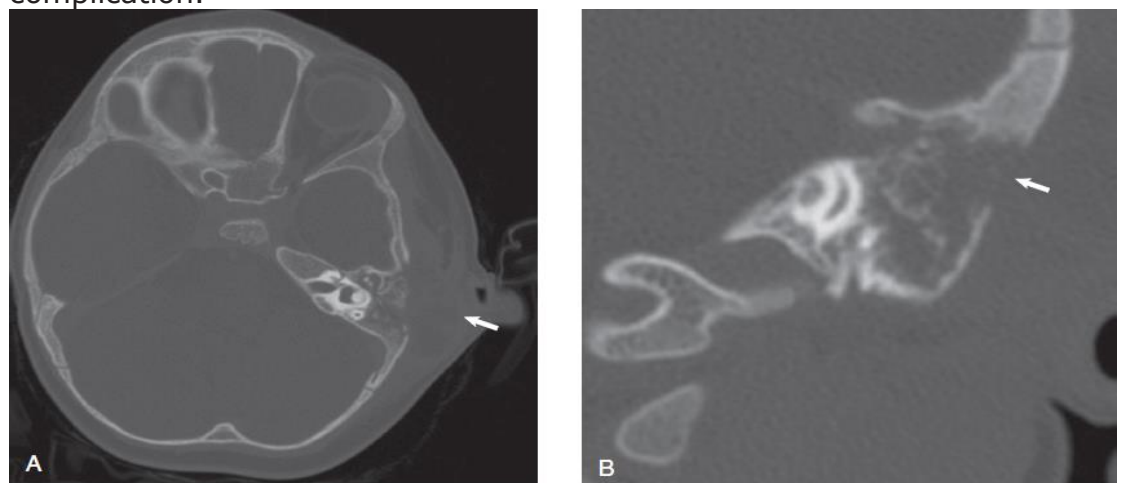


FIGURE 140-2. Axial (A) and coronal (B) temporal bone computed tomography scan shows opacification of the mastoid with loss of bony septa. Note the defect in cortical bone and postauricular fluid collection (arrows).

❖ **Treatment**

- ✓ It is serious medical problem that requires aggressive
- ✓ Treatment, either surgical or medical.

- ✓ Classically treatment "IV antibiotics + cortical mastoidectomy with removal of necrotic, devitalized bone "

- ✓ In recent years, "myringotomy + IV antibiotics 3 o 6 week"
 - Has been advocated as an alternative for surgery
 - This medical management requires a CT scan to confirm resolution of the infection and aeration of the mastoid before stop Abx

- ✓ Cortical mastoidectomy + ventilating tube placement + IV antibiotics
 - Most conservative management of this potentially serious complication
 - Provides prompt, precise eradication of all infected tissue in an expeditious
 - Cost-effective manner.

(c) Masked (Latent) Mastoiditis

- ✓ Rare condition of **slow destruction** of mastoid air cells but **without** the acute signs and symptoms often seen in acute mastoiditis.
- ✓ There is **no pain, no discharge, no fever and no mastoid swelling**
- ✓ **Mastoidectomy may show extensive destruction of the air cells with granulation** tissue and dark gelatinous material filling the some mastoid air cell.
- ✓ May cause erosion of the **tegmen tympani and sinus plate with an extradural or perisinus abscess.**

❖ Aetiology

- ✓ The condition often results from **inadequate antibiotic therapy** in cases of acute otitis media when acute symptoms subside but smouldering infection continues in the mastoid.
- ✓ Middle ear & much of the mastoid respond to the antibiotics, but a focal area in the mastoid persistent infection
- ✓ Granulation or mucosal disease involves some mastoid air cells

❖ Clinical Features

- ✓ Patient is often a child
- ✓ Not feeling well, with mild pain behind the ear but with persistent hearing loss.
- ✓ Tympanic membrane **appears normal** OR thick with loss of translucency.
- ✓ Slight tenderness may be elicited over the mastoid.
- ✓ Audiometry shows conductive hearing loss of variable degree.
- ✓ CT shows a localized area of opacification in an otherwise normal mastoid & tympanic cavity.

❖ Treatment

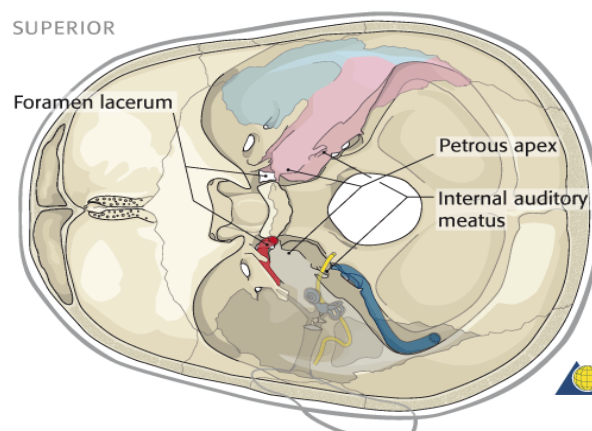
- ✓ Cortical mastoidectomy with antibiotics
- ✓ No different from the diagnosis and management of mastoiditis with otorrhea that emanates from other causes

(d) Chronic Mastoiditis

- ✓ Implies chronic inflammation within the mastoid air cell system.
- ✓ It can occur in typical COM with longstanding tympanic membrane perforation, with **cholesteatoma** or as a complication from an infection after placement of a middle ear **ventilating tube**.
- ✓ Results in mucosal disease and/ or granulation tissue that causes otorrhea to persist in spite of antibiotic therapy
- ✓ Chronic mastoiditis usually results in sclerosis of the mastoid air cell system commonly seen in CSOM
- ✓ **Mastoidectomy** in either of these settings can be helpful in eradicating otorrhea. ,, complete resolution with antibiotics is unlikely
- ✓ Differential diagnosis of chronic mastoiditis can be skull base osteomyelitis "malignant otitis externa"
- ✓ Complications in patients with chronic mastoiditis with tympanic membrane perforation can develop at any time but often occur only after weeks or months of otorrhea.
- ✓ In contrast, cholesteatoma typically requires months or years to produce complications.

2. Petrositis (petrous apicitis) "Petrus Apex Syndrome"

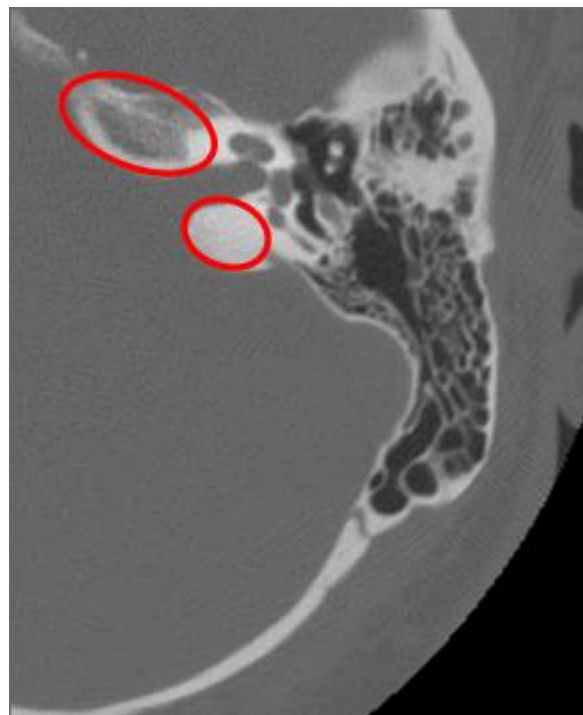
- ✓ Very rarely Spread of infection from middle ear and mastoid to the **petrous part of temporal bone** "anterior, medial portion of the temporal bone "is called petrositis.
- ✓ The petrous apex takes the form of a truncated pyramid in the posterior skull base
- ✓ Anteromedially between occipital and sphenoid bones
- ✓ Forms part of the **foramen lacerum**
- ✓ Air cell tracts extend to the apex in continuity with the middle ear and mastoid through well-described cell tracts **below, above, posterior, and anterior** to the labyrinth, allowing for infection involving the mastoid and middle ear cleft to extend into the petrous apex

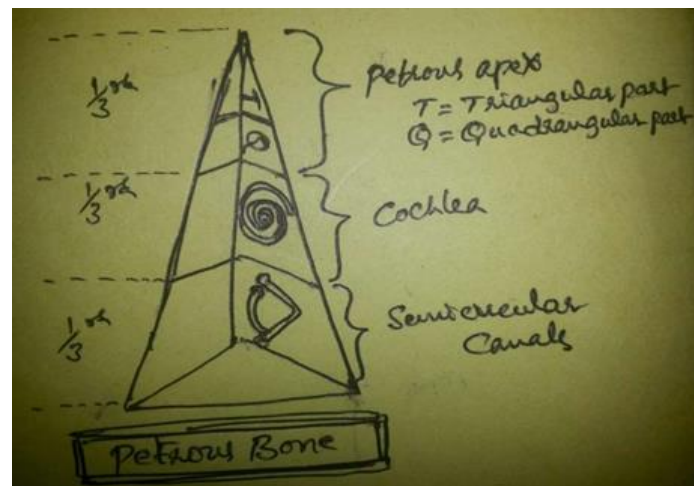
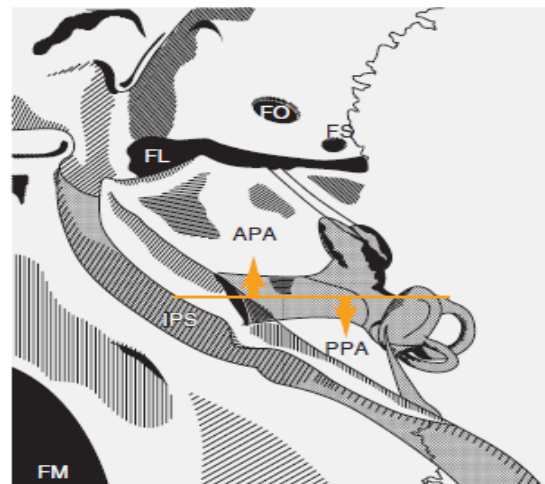
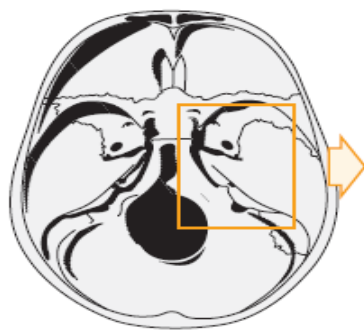
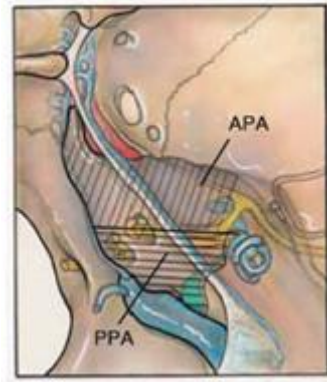


- ✓ It may be associated with:
- ✓ **Acute coalescent mastoiditis**
- ✓ **Latent mastoiditis**
- ✓ **Chronic middle ear infections.**

❖ **Pathology**

- ✓ **Petrous bone** may be of three types (like mastoid),:
"Petrous apex divided into Anterior/Posterior by plane running through IAC or Cochlea"
 - 1. **Pneumatized** with air cells extending to the petrous apex "Posterior Petrous Apex pneumatized in 30%, Anterior Petrous Apex pneumatized in 10%"
 - 2. **Diploic** containing only marrow space
 - 3. **Sclerotic**.
- Petrous apex divided into Anterior/Posterior by plane running through IAC or Cochlea"
 - 1. **Anterior Petrous apex**: medial to cochlea ,,, Larger ,,Consists of bone marrow or air cells.
 - 2. **Posterior Petrous apex**: medial to SCC ,, Smaller ,, dense bone

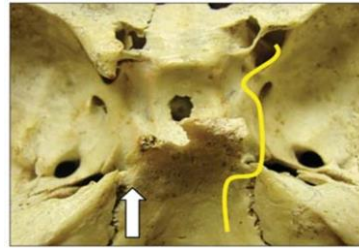




❖ Clinical Features

- ✓ **Gradenigo's syndrome** (secondary to forming epidural abscess at the petrous apex)
- ✓ Today, more commonly caused by Tumor at the Petrous Apex (Cholesteatoma, Meningioma, etc)
- ✓ The classical presentation, and consists of a **triad** of:

A. (Diplopia) external rectus palsy (VIth nerve palsy) occurs from involvement of Dorello's canal osteo fibrous canal situated at the petrous apex containing the abducent nerve & Inferior Petrosal sinus"



B. Deep-seated facial or retro-orbital pain (Trigeminal pain) (Vth nerve involvement) "Trigeminal ganglion in the Meckel cave"

C. Persistent ear discharge.

- ✓ **Persistent ear discharge** with or without deep-seated pain in spite of an adequate cortical or modified radical mastoidectomy also points to petrositis.
- ✓ **Fever, headache, vomiting and sometimes neck rigidity may also be associated.**
- ✓ **Some patients may get facial paralysis and recurrent vertigo , sensory hearing loss due to involvement CN VII, VIII, IX**
- ✓ **Few patients had full triad today**

TABLE 139-4. Symptoms Found in 22 Patients with Petrous Apicitis from 1976-1995

Symptom	No. Patients (%)
Deep pain and headache	13 (59)
Otalgia	16 (73)
Otorrhea	13 (59)
Fever	5 (22)
Coma	2 (9)
Paralysis of cranial nerve	
V	15 (68)
VI	4 (18)
VII	6 (27)
VIII	9 (41)
IX	1 (5)
X	1 (5)

- ❖ **Diagnosis** of petrous apicitis requires both **CT** scan and **MRI**
- ✓ **CT scan** of temporal bone will show:
 - **Bony details** of the petrous apex and the septa of air cells & evaluate surrounding anatomy
 - Provides important details about potential surgical routes
 - Aid in the diagnosis of intracranial
 - Asymmetry of the petrous apex is not diagnostic for apicitis, because asymmetric pneumatization of the apex can occur in healthy subjects.
- ✓ **MRI** helps to **differentiate diploic marrow containing apex from fluid or pus, mucous**.
- ✓ **Gallium bone scan** may provide additional information, showing increased uptake on the side of the apicitis

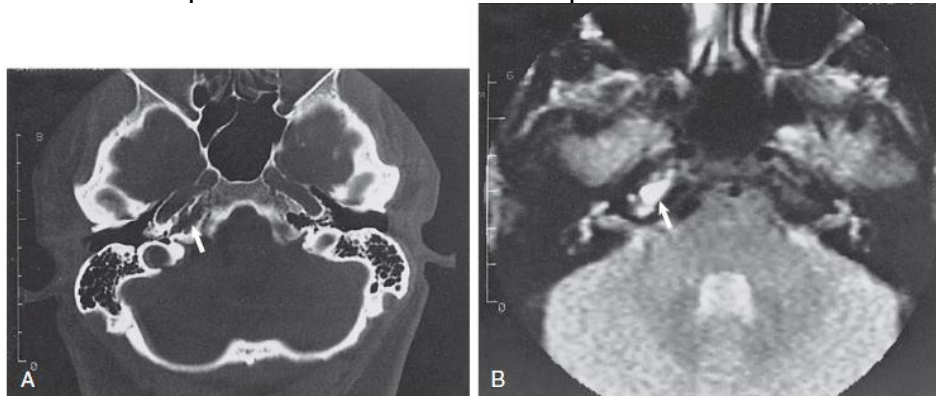
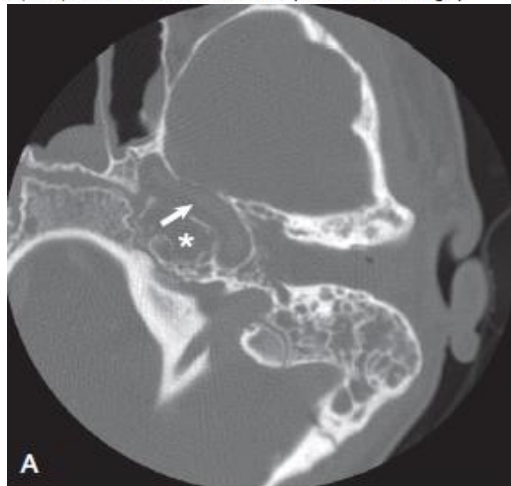


FIGURE 139-20. Computed tomography (CT) and magnetic resonance imaging (MRI) are helpful adjuncts in the diagnosis of petrous apicitis. **A**, In the CT scan, the fluid-filled petrous apex (arrow) can be compared with the opposite air-filled apex. **B**, In the MRI scan, the area of increased signal is present in involved apex (arrow). An amber-colored, fluid-filled cyst was found at surgery.



left petrous apicitis. Note opacification of the petrous apex with destruction of bony septa (asterisk). A dehiscence of the carotid canal and narrowing of the petrous carotid artery (arrow)

✓ **Treatment**

- ✓ Petrous apex is an area that is not easily approached surgically because of its relationship with the otic capsule and carotid artery.
- ✓ Because of the difficult surgical approach and the response rate to antibiotics

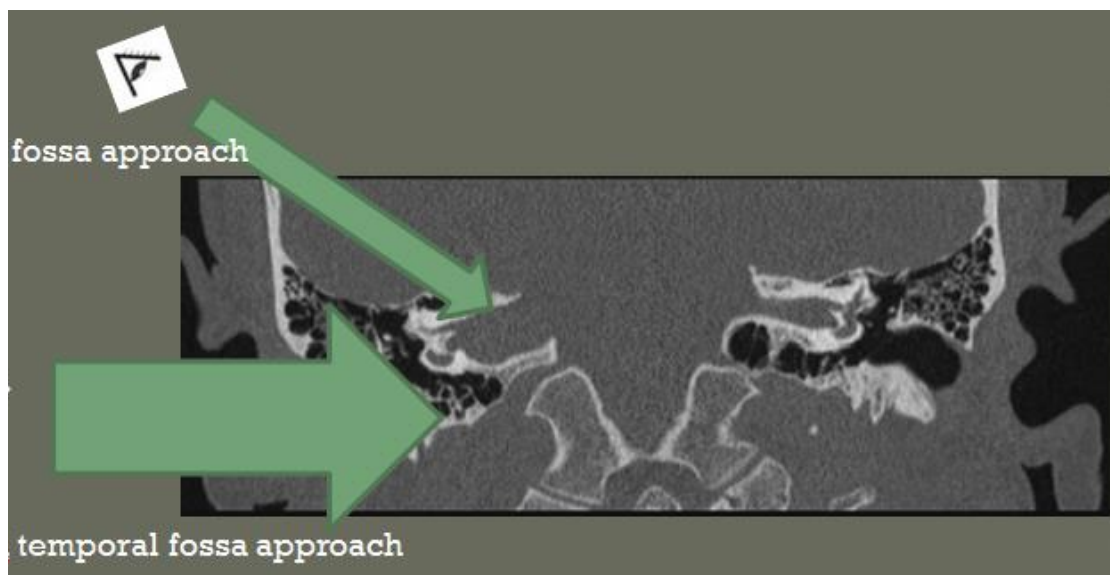
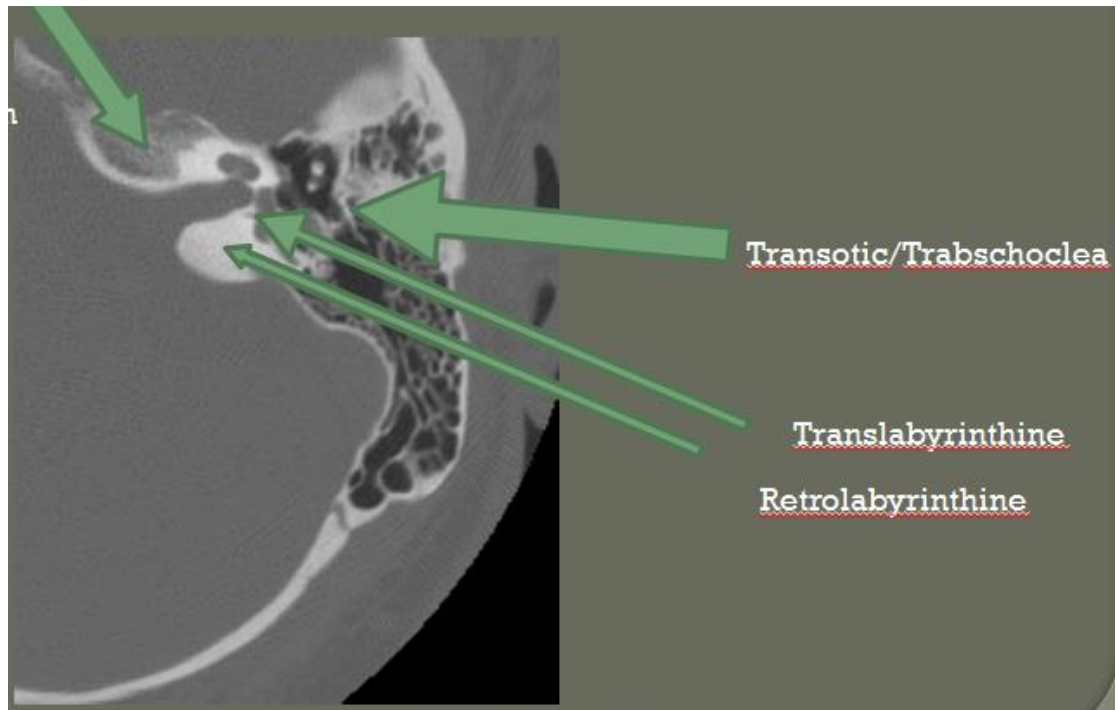
✓ IV antibiotics are often the **first-line treatment of petrous apicitis**

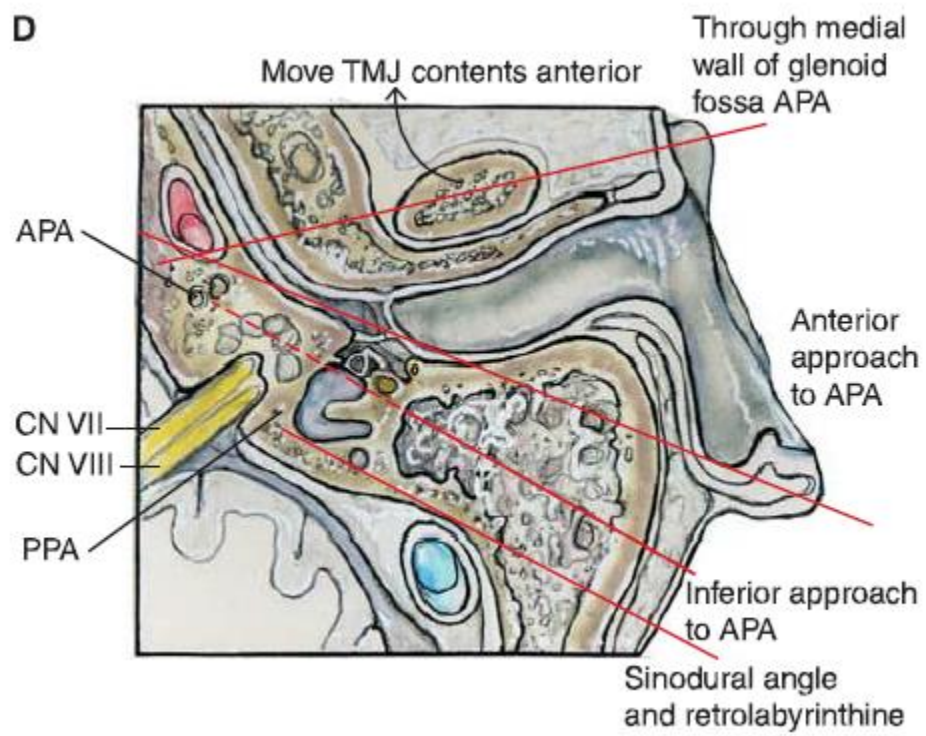
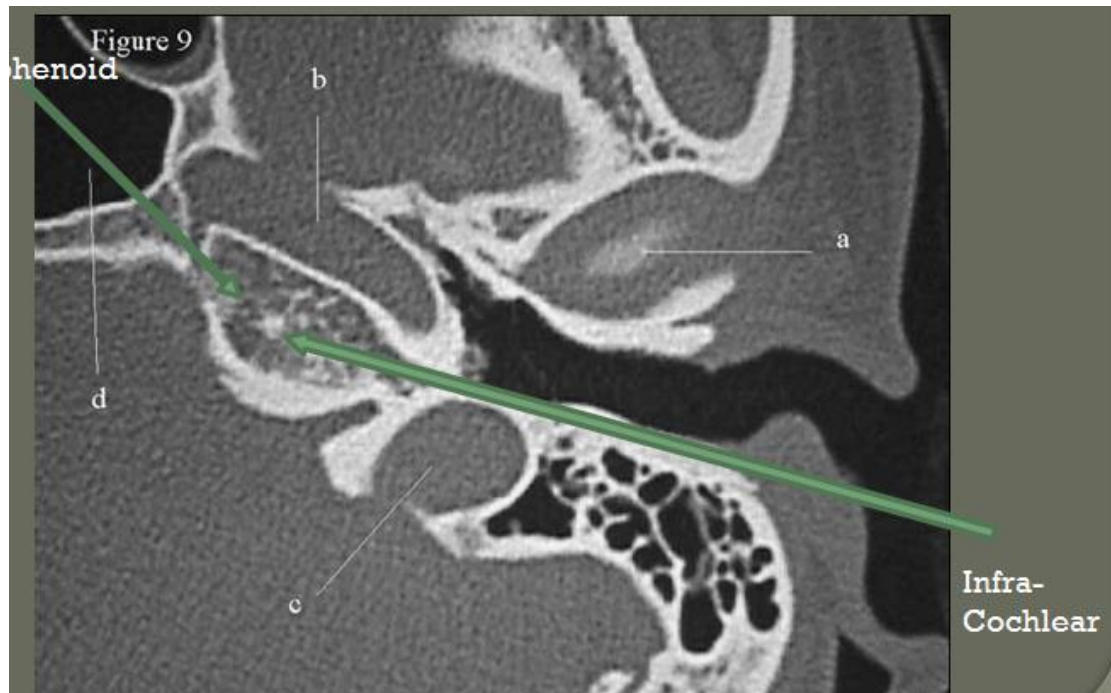
- Most cases of acute petrositis can now be cured with antibacterial therapy alone.
- Long duration of treatment. 3-6 week
- Serial CRP & ESR rates to monitor for response of bony infections
- Surgical drainage is required when presence abscess, necrotic bone, or persistent infection despite medical therapy
- Should precede and follow surgical intervention

✓ **PETROUS APICECTOMY**

- Surgical access to the petrous apex. becomes necessary for:-
 - Drainage of infected air cells "Petrositis (petrous apicitis) "Petrous Apex Syndrome"
 - Cholesteatomas (congenital)
 - Cholesterol granulomas and mucosal cysts
 - Biopsy of various mass lesions
 - **Benign tumors**
 - Meningioma
 - Schwannoma
 - **Malignant tumors**
 - Chondrosarcoma / Chondroma
 - Chordoma
 - Plasmacytoma
 - Metastatic lesion
 - Langerhans cell histiocytosis
- Access to the petrous apex Depending on the specific anatomy
 - May be obtained via the retrolabyrinthine infracochlear, or subarcuate **air cell tracts**.
 - Most patients, the petrous apex can be drained via a CWU approach to the temporal bone.
 - Some may be necessary to perform a canal wall down mastoidectomy or possibly a middle cranial fossa approach to access this area
 - Typically, the margins of the exposure include the cochlea superiorly, the carotid artery anteriorly, and the internal jugular vein and bulb posteriorly.

- **Surgical approach to the petrous apex** depends on:
 1. Available air-cell pathways "below, above, posterior, and anterior to the labyrinth"
 2. The portion of the apex involved "posterior, and anterior"
 3. Hearing , non-hearing ear "Sparing the otic capsule surgically is preferred in patients with serviceable hearing "
- The classic procedures designed to access the **posterior petrous apex** include:
 1. Infralabyrinthine approach
 2. Translabyrinthine approach "Sinodural"
 3. Retrolabyrinthine approach
 4. Subarcuate "provide drainage via cells that extend over the superior semicircular canal and through the "hole in the doughnut," working through the center of the superior semicircular canal"
- Procedures used to access the **anterior petrous apex** include:
 1. Infracochlear approach.
 2. Transotic approach " FN not mobilized"
 3. Transcochlear approach " transposition of FN"
 4. Middle fossa approach " used when the disease involves the anteriorpettous apex but spares the middle ear and mastoid"
 5. Anterior approach through the glenoid fossa "described by Ramadier and was popularized by Lempert. The exposure involves removal of the anterior canal wall and condyle of the mandible; exposure of the epitympanum; avulsion of the tensor tympani; opening of the tensor semicanal; and then dissection in the triangle between the carotid artery (posterior), the cochlea (superior), and the middle fossa dura (anterior). This approach can be modified by preserving the anterior canal wall and condyle.))"
 6. Endoscopic Trans-sphenoidal"the presence of venous sinuses between the petrous apex and sphenoid, such as the cavernous sinus, can make this approach challenging. It can be considered for lesions located in the medial section of the petrous apex abutting and/or prolapsing into the posterior wall of the sphenoid sinus"
- In a non-hearing ear (Passing through Otic capsule):
 - Translabyrinthine
 - Transcochlear (transotic) approach
 - Provides **superior** access for complete removal of mass lesions
 - One disadvantage of these two approaches is that they could potentially expose the cerebrospinal fluid (CSF) to the infectious process.





3. Facial Paralysis

- ✓ It can occur as a complication of both **acute** and **chronic** otitis media.
- ✓ 2nd most common extracranial complication from AOM facial palsy
- ✓ Less commonly presents as a complication of COM
- ✓ Facial paralysis can also be associated with
 - Coalescent mastoiditis
 - Masked mastoiditis
 - Petrous apicitis.

❖ Acute Otitis Media

- ✓ Facial nerve is normally well protected in its **bony canal**.
- ✓ Sometimes, the bony canal is **dehiscent** "Tympanic segment most common", and the nerve lies just under the middle ear mucosa.
- ✓ It is in these cases that inflammation of middle ear spreads to epi- and perineurium, causing facial paralysis.
- ✓ Often presents in children with incomplete paresis
- ✓ Comes on **abruptly** is usually short-lived with appropriate treatment , good prognosis "rarely lasts longer than 3 weeks"
- ✓ Facial nerve function fully recovers if acute otitis media is controlled with **systemic antibiotics**.
- ✓ **Myringotomy** or **cortical mastoidectomy** may sometimes be required.

❖ Chronic Otitis Media

- ✓ Either results from **cholesteatoma** or from **penetrating granulation tissue**.
- ✓ **Cholesteatoma** destroys bony canal and then causes pressure on the nerve, further aided by oedema of associated inflammatory process.
- ✓ Site of lesion depends on the anatomy of the cholesteatoma.
 - Most commonly, the nerve is compressed in the **tympanic segment**.
 - Other sites of injury include :-
 - **Geniculate ganglion** region (anterior epitympanic cholesteatoma),
 - **Mastoid segment** (facial recess/sinus tympani/ retrofacial cholesteatoma)
- ✓ Facial paralysis is **insidious** but **slowly** progressive , full paralysis, **worse** prognosis
- ✓ Treatment is **urgent** **exploration of the middle ear and mastoid**.
 - Facial canal is inspected from the **geniculate ganglion** to the **stylomastoid foramen**.
 - Diamond burs should be used to carefully remove the bone of the fallopian canal
 - If granulation tissue or cholesteatoma has entered the bony canal, **uncapped** in the area of involvement.
 - Granulation tissue **surrounding** the nerve is **removed**
 - If it actually **invades** the nerve sheath, it is **left in place**.

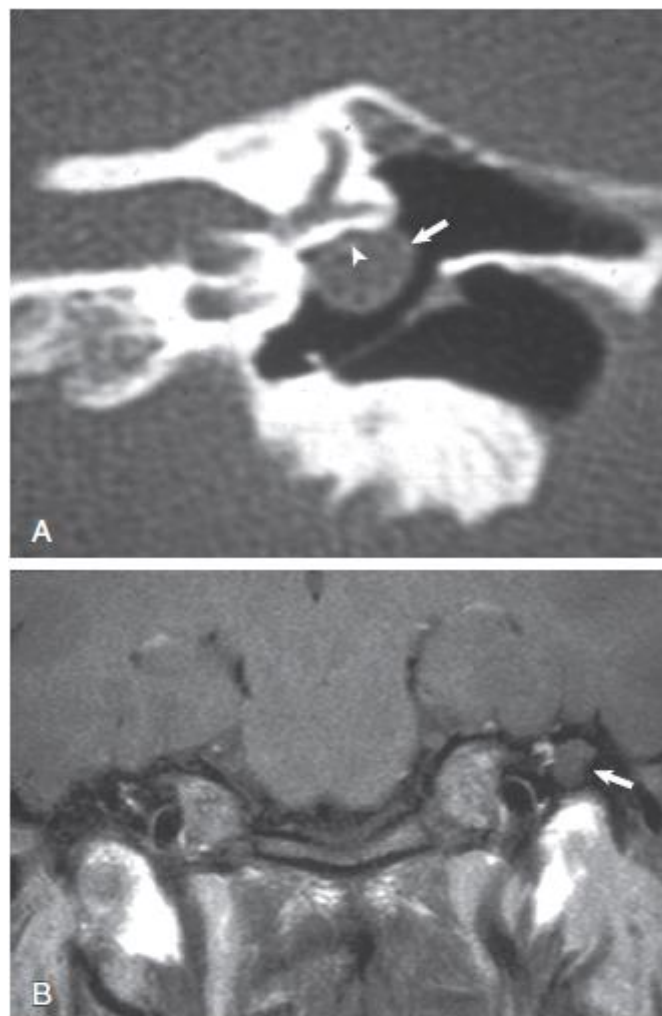
- If a segment of the nerve has been **destroyed** by the granulation tissue, **resection of nerve and grafting** are better left to a second stage when infection has been controlled and fibrosis has matured.
- Significantly improved outcomes have been seen when surgical decompression was performed within 1 week of the onset of facial paresis
- Remember—a facial palsy occurring in the presence of chronic ear disease is **not Bell's palsy** and active treatment is needed if the palsy is not to become permanent.
- Systemic steroid therapy is probably helpful to reduce inflammation and edema in the **acute phases** of facial paralysis for any type of OM

✓ **Four Indications for Mastoidectomy in Facial nerve Palsy secondary to Otitis Media complication**

1. History of COM
2. Onset of paralysis >2 weeks after onset of AOM
3. ENOG showing >90% Degeneration after 6 days (poor prognosis if left)
4. Failure of paralysis to resolve after appropriate Medical management

✓ **Diagnosis**

- ✓ Clinical by examination alone : AOM in very straightforward cases
- ✓ CT scan:- very useful and will likely demonstrate the pathology and site of lesion COM
- ✓ MR imaging is helpful if lesions other than cholesteatoma are considered
- ✓ ENOG (electroneuronography) , Electromyography (EMG) be helpful in predicting prognosis for recovery in some cases (see facial palsy topic)



Coronal temporal bone computed tomography scanning (A) and T1-weighted magnetic resonance imaging (B) show cholesteatoma (arrow) eroding the facial nerve canal (arrowhead).

5. Fistula of Labyrinth

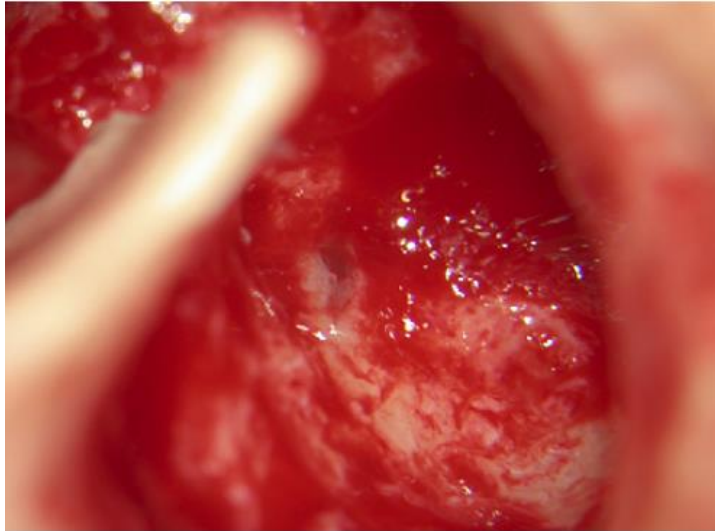
- ✓ There is thinning or erosion of bony capsule of labyrinth, usually of the **horizontal semicircular canal** , rarely cochlea superior canal and posterior canal ,vestibule
- ✓
- **Aetiology**
- ✓ The causes are:
 - A. Chronic suppurative otitis media with cholesteatoma "osteolysis and uncovering of the labyrinth " **occurred in 7% of the cholesteatomas in the large series by Gersdorff and colleagues "**
 - Can occur from resorption of the otic capsule due to inflammatory mediators in the absence of cholesteatoma, which typically occurs in COM with granulation.
 - B. Neoplasms of middle ear, e.g. carcinoma or glomus tumour.
 - C. Surgical or accidental trauma to labyrinth.
- **Clinical features**
- ✓ Frequently asymptomatic and only discovered on CT imaging or at surgery
- ✓ Because part of membranous labyrinth is exposed and becomes **sensitive to pressure changes.**
- ✓ Patient complains of transient vertigo often induced by
 - Pressure on tragus
 - Cleaning the ear
 - Valsalva manoeuvre
 - Tullio phenomenon (vertigo secondary to auditory stimuli)
- ✓ Vary degree sensorineural hearing loss is found in most of these patients (68%)

- **Dx:-**
 - ✓ The definitive diagnosis for a fistula is only made intraoperatively
 - ✓ CT may reveal bony erosion of the labyrinth 60 % of cases
 - ✓ Diagnosed by "**fistula test**" which can be performed in two ways.

- **(a) Pressure on tragus**
 - ✓ Sudden inward pressure is applied on the tragus, increases air pressure in the ear canal and stimulates the labyrinth.
 - ✓ Patient will complain of **vertigo**.
 - ✓ **Nystagmus** may also be induced with **quick component towards the ear under test " stimulated ear "**

- **(b) Siegle's speculum** (pneumatoscapy)
 - ✓ When positive pressure is applied to ear canal
 - ✓ Patient complains of **vertigo usually with nystagmus**.
 - ✓ The **quick component of nystagmus would be towards the affected ear** (ampullopetal displacement of cupula).
 - ✓ High false negative rate , this classic picture is **not sensitive** in the preoperative identification pt of fistula
 - ✓ Periodic vertigo or significant disequilibrium is found in 62% to 64% of patients who have fistulae preoperatively
 - ✓ The fistula test is positive in 32% to 50% of patients who are found ultimately to have fistulae during surgical exploration.
 - ✓ Sensorineural hearing loss is found in most of these patients (68%), **it is not a sensitive indicator of fistula**
 - ✓ **Presence** of "**sensorineural hearing loss vertigo, or a positive fistula test**" in a patient who has a **cholesteatoma** should raise the suspicion for a fistula
 - BUT**
 - ✓ **Absence** of "**sensorineural hearing loss vertigo, or a positive fistula test**" in a patient who has a **cholesteatoma** does not guarantee an intact bony labyrinth.

- **CT**
 - ✓ Preoperative CT imaging detection of an exposed labyrinth, facial nerve, or dura, to aid in surgical planning
 - ✓ Detect fistulae accurately on preoperative CT has been reported as 60%
 - ✓ CT scans are no more sensitive than history and physical
 - ✓ examination in detecting labyrinthine fistulae
- The definitive diagnosis for a fistula is only made intraoperatively "observed at surgery as a "blue line" parallel to the underlying semicircular canal lumen
- Surgical approach is to assume the presence of a fistula in every cholesteatoma case, to prevent unexpected complications.

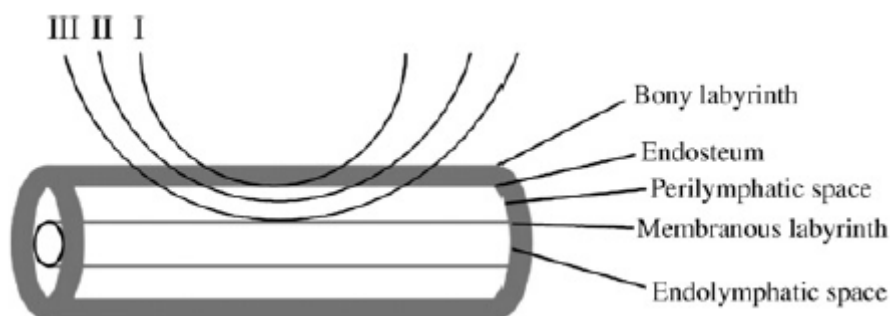


Intraoperative picture of a horizontal canal fistula.



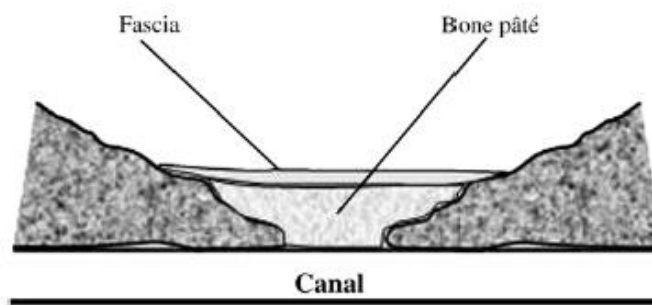
coronal CT scan of a postoperative right ear, revealing a horizontal canal fistula.

- ✓ Staging system introduced by **Dornhoffer and Milewski** is the classification used in the authors' department, and is used in this article to discuss fistulae and their management
- ✓ **Type I Fistula**: Bony erosion and intact endosteum.
- ✓ **Type IIa Fistula**: Endosteum is violated, but the perilymphatic space is preserved.
- ✓ **Type IIb Fistula**: Perilymph is violated by disease or inadvertently suctioned.
- ✓ **Type III Fistula**: Membranous labyrinth and endolymph have been disrupted by disease or surgical intervention



- **Treatment**

- In **chronic suppurative otitis media or cholesteatoma**:- **tympanomastoidectomy " mastoid exploration"** is often required to eliminate the cause cholesteatoma
- Most appropriate management of the fistula remains an ongoing debate.
- Most appropriate approach to the fistula is to perform a **canal wall down mastoidectomy**, remove the bulk of the cholesteatoma, and leave the fistula covered with the **matrix**
- Removal of the matrix increases the risk of sensorineural hearing loss, and that by removing the sac itself
- Risk of significant sensorineural hearing loss as a result of surgical manipulation highly **controversial** topic
- Pressure from the cholesteatoma is relieved and further bony erosion or infectious complications are unlikely
- Graft site (fascia) OR shaped cap of bone secured with fibrin glue if fistula is exposed



The repair of a labyrinthine fistula after removal of cholesteatoma.

- Some investigators advocate the complete removal of the cholesteatoma over the fistula, with repair of the bony defect in all circumstances
- The **size, extent, and location** of the fistula should be considered when determining whether complete cholesteatoma removal should be attempted
- Contraindication of removal of cholestatoma matrix : **Fistu-La**
 - F: Firmly adherent
 - I: Infection
 - S: Size > 2 mm " if smaller can remove safely "
 - T : Tympanoium (promontory/cochlea) high risk of SNHL
 - U : Only hearing ear
 - LAAAA
- No hearing loss resulted from the removal of **type I , IIa fistulae**
- 50% significant hearing loss postoperatively from the removal **type IIb and type III fistulae**
- **Systemic antibiotic** therapy should be instituted before and after operation to prevent spread of infection into the labyrinth.
- **Intraoperative steroids** experienced stable or improved hearing 90% of the time

5. Labyrinthitis

✓ There are 2 types of labyrinthitis:

- (a) Diffuse **serous "toxic"** labyrinthitis
- (b) Diffuse **suppurative** labyrinthitis

❖ Diffuse Serous Labyrinthitis

- ✓ It is **diffuse intralabyrinthine inflammation without pus formation** and is a reversible condition if treated early.
- ✓ Due to alteration of the inner ear tissue fluid environment due to bacterial toxins enter the inner ear via the round or oval window, or via a labyrinthine fistula.

• Aetiology

- A. Most often it arises from **pre-existing Fistula of Labyrinth "circumscribed labyrinthitis"** associated with chronic middle ear suppuration or cholesteatoma.
- B. In acute infections of middle ear, cleft inflammation spreads through annular ligament or the round window.
- C. It can follow stapedectomy or fenestration operation.
- D. Congenital labyrinthine deformities, such as Mondini deformity and enlarged vestibular aqueducts

• Clinical features

- **Mild cases** complain of **vertigo and nausea**
- **Severe cases**, **vertigo is worse with marked nausea, vomiting and even spontaneous nystagmus**.
- Quick component of nystagmus is **towards the affected ear**. "stimulate"
- As the inflammation is **diffuse**, cochlea is also affected with some degree of sensorineural hearing loss.
- **Serous labyrinthitis**, if not checked, may pass onto **suppurative labyrinthitis** with total loss of **vestibular and cochlear** function.
- Endolymphatic hydrops can be seen pathologically associated with serous labyrinthitis.

- **Treatment**

Medical	Surgical
a. Bed rest, his head immobilised with affected ear above. b. Antibacterial therapy is given in full doses to control infection. c. Steroid d. Labyrinthine sedatives, e.g. prochlorperazine (Stemetil) or dimenhydrinate (Dramamine), are given for symptomatic relief of vertigo ,nausia. e. Myringotomy is done if labyrinthitis has followed acute otitis media and the drum is bulging. Pus is cultured for specific antibacterial therapy	○ To treat the source of infection. ○ Cortical mastoidectomy (in acute mastoiditis) ○ Modified radical mastoidectomy (in chronic middle ear infection or cholesteatoma) ○ Medical treatment should always precede surgical intervention

❖ **Diffuse Suppurative Labyrinthitis**

- ✓ This is diffuse bacterial pyogenic infection invasion the labyrinth with.
- ✓ Results in rapid destruction of inner ear contents, with **permanent** loss of vestibular and cochlear functions

• **Aetiology**

- ✓ It usually follows serous labyrinthitis, pyogenic organisms entering through a pathological or surgical fistula.

• **Clinical features**

- ✓ There is severe vertigo with nausea and vomiting due to acute vestibular failure. Spontaneous nystagmus will be observed with its quick component towards the healthy side. "paralytic"
- ✓ Patient is markedly toxic.
- ✓ There is total loss of hearing (permanent)
- ✓ Relief from vertigo is seen after 3-6 weeks due to adaptation.
- ✓ Suppurative process within the labyrinth can gain access to the subarachnoid space via the fundus of the cochlea or the cochlear aqueduct.
- ✓ Otogenic sources are thought to be a common etiology of bacterial meningitis in childhood.
- ✓ **Conversely**, suppurative meningitis may extend into the labyrinth and result in a **secondary suppurative (meningogenic) labyrinthitis**.
- ✓ This phenomenon underlies the frequent association between meningitis and hearing loss.
- ✓ Ossification of the labyrinth (**labyrinthitis ossificans**) is frequently seen following suppurative labyrinthitis, particularly due to **S. pneumoniae**, and can complicate cochlear implantation
- ✓ Early evaluation of hearing following meningitis and early cochlear implantation when appropriate can result in substantially better outcomes for patients with postmeningitic deafness.

• **Treatment**

- ✓ It is same as for serous labyrinthitis.
- ✓ Rarely, drainage of the labyrinth is required, if intralabyrinthine suppuration is acting as a source of intracranial complications, e.g. meningitis or brain abscess

B. INTRACRANIAL COMPLICATIONS OF OTITIS MEDIA

- 1. Extradural Abscess or granulation tissue**
- 2. Subdural Abscess**
- 3. Meningitis**
- 4. Brain Abscess**
- 5. Lateral" sigmoid" Sinus Thrombophlebitis (Syn. Sigmoid Sinus Thrombosis) (Occluding ,Non occluding)**
- 6. Otitic Hydrocephalus**

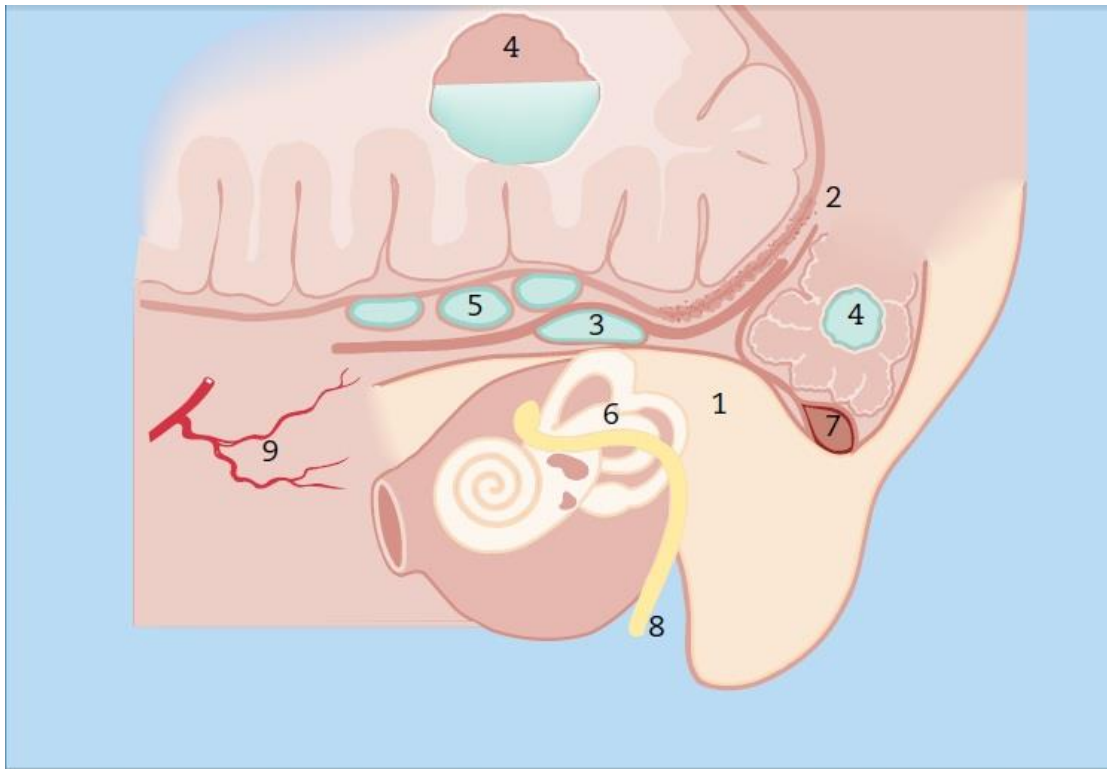
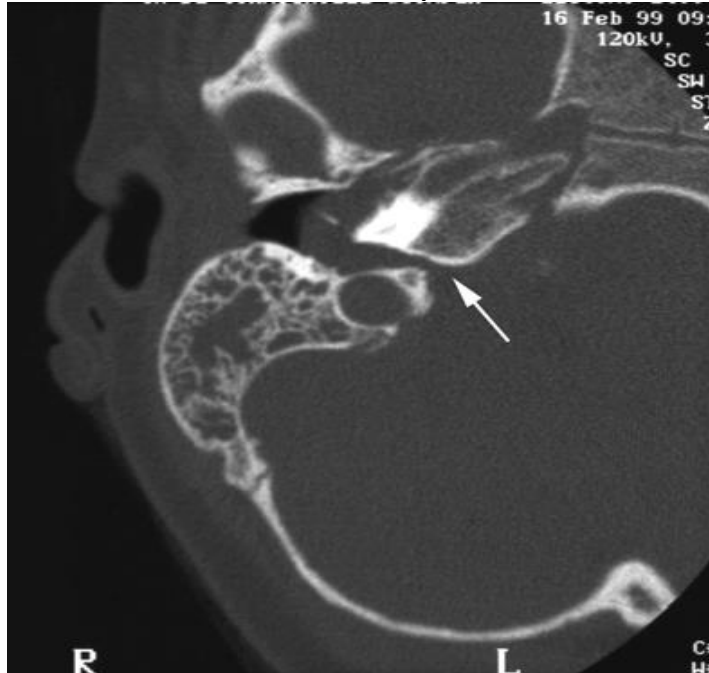


Fig. 10.1 Complications of chronic otitis media. 1, Acute mastoiditis; 2, Meningitis; 3, Extradural abscess; 4, Brain abscess (temporal lobe and cerebellum); 5, Subdural abscess; 6, Labyrinthitis; 7, Lateral sinus thrombosis; 8, Facial nerve paralysis; 9, Petrositis.

❖ **Routes of Spread into the Intracranium:**

1. Direct extension from bone erosion
2. Lymphatic or hematogenous spread
3. Invasion through normal anatomic structures (labyrinth)
4. Spread through iatrogenic or traumatic defects
5. Extension through **Hyrtle's Fissure** (embryologic remnant that connects hypotympanum to the subarachnoid space , **The cleft extends between the bony labyrinth and the jugular bulb. , associate with CSF otorrhea**)



1. Extradural Abscess (Epidural Abscess) Or Granulation Tissue

- ✓ It is collection of pus between the **bone and dura**.
- ✓ It may occur both in **acute and chronic infections of middle ear**.

❖ Pathology

- ✓ In **acute otitis media**, bone over the dura is destroyed by **hyperaemic decalcification**,
- ✓ In **chronic otitis media** it is destroyed by cholesteatoma OR coalescent mastoiditis
- ✓ **Direct extension** via bone erosion pus comes to lie directly in contact with dura. (most common)
- ✓ Also occur by **venous thrombophlebitis** (in this case, bone over the dura remains intact.)
- ✓ Extradural abscess may lie in :-
 - Dura of middle or posterior cranial fossa
 - Dura of lateral venous sinus (perisinus abscess).
- ✓ The affected dura may be covered with **granulations or appear unhealthy and discoloured**. (pachymeningitis).
- ✓ Frequently associated with lateral sinus thrombophlebitis, meningitis, and cerebritis or brain abscess.

❖ Clinical Features

- ✓ **Most of the time are asymptomatic and silent**, "not differ from those found in COM" **and are discovered accidentally during cholesteatoma surgery or CT scan for other purposes**
- ✓ No sensitive or specific symptoms suggestive of this disease process.
- ✓ However, Extradural Abscess is suspected when there is:
 - **(i)** Persistent headache on the side of otitis media.
 - **(ii)** Severe pain in the ear.
 - **(iii)** General malaise with low-grade fever.
 - **(iv)** Pulsatile purulent ear discharge.
 - **(v)** Disappearance of headache with free flow of pus from the ear (spontaneous abscess drainage).

❖ **Diagnosis is made on**

✓ **Contrast-enhanced CT :-**

- May reveal erosion of the sigmoid plate or tegmen
- Rim enhancing lentiform epidural fluid collection



✓ **MRI**

- Superior to CT in demonstrating small intracranial suppurative lesions.
- Hyperintense relative to CSF on T1
- Isointense to CSF on T2

❖ **Treatment**

(a) **Mastoidectomy**

- ✓ Required to deal with the **causative disease process**.
- ✓ Extradural abscess and granulation tissue is evacuated by removing overlying bone till the limits of healthy dura without granulation tissue is evident on all margins of the abscess are reached.
- ✓ Cases where **bony plate of tegmen tympani or sigmoid sinus plate is intact** but there is suspicion of an abscess, the intact bony plate is removed to evacuate any collection of pus.

(b) **Antibiotic**

- ✓ High dose for a minimum of 5 days
- ✓ Closely observed for any further complications, such as sinus thrombosis, meningitis or brain abscess.
- ✓ Postoperative antibiotics are continued at least until the symptoms of the abscess and otitis have resolved

2. Subdural Abscess

- ✓ This is collection of pus **between dura and pia-arachnoid membranes**
- ✓ Subdural abscess more commonly occurs in the frontal region from sinusitis, but may result from ear disease
- ✓ Subdural empyema **rarely** occurs due to COM
- ✓ More typical complication of meningitis in infants (H. influenza , bilateral)
- ❖ **Pathology**
 - ✓ Infection spreads from the ear by **direct extension** via erosion of bone and dura or by **thrombophlebitic process** in which case intervening **bone remains intact**.
 - ✓ Pus rapidly spreads in subdural space and comes to lie against the convex surface of cerebral hemisphere causing **pressure symptoms**.
 - ✓ With time, the pus may get loculated at various places in subdural space.
- ❖ **Clinical Features**
 - ✓ Signs and symptoms of subdural abscess are due to
 - a. Meningeal irritation
 - b. Thrombophlebitis of cortical veins of cerebru
 - c. Raised intracranial tension.
 - ✓ **(a) Meningeal irritation**
 - Headache "earlies and most persistent symptom", fever , malaise, increasing drowsiness, neck rigidity and positive Kernig's sign.
 - ✓ **(b) Cortical venous thrombophlebitis**
 - Veins over the cerebral hemisphere undergo thrombophlebitis leading to **aphasia, hemiplegia, hemianopia , coma**.
 - Jacksonian seizures(partial seizure) OR status epilepticus.([from cortical damage](#))
 - ✓ **(c) Raised intracranial tension**
 - **papilloedema, ptosis and dilated pupil (IIIrd nerve involvement),** and involvement of other cranial nerves.

- ❖ **CT scan or MRI are required for diagnosis** (reveals crescent shaped enhancement that does not cross midline)



- ❖ **Treatment**

- ✓ It is a neurological emergency :- **series of burr holes or a craniotomy** is done to drain subdural empyema "**neurosurgical emergency**"
- ✓ **IV antibiotics** are administered to control infection.
- ✓ Once infection is under control (**stable**) , attention is paid to causative ear disease which may require mastoidectomy.
- ✓ Lumbar puncture should not be done as it can cause herniation of the cerebellar tonsils.
- ✓ The prognosis is poor.

3. Meningitis

- ✓ It is inflammation of leptomeninges " thin meninges" (pia and arachnoid)
- ✓ Usually with bacterial invasion of CSF in subarachnoid space.
- ✓ It is the **most common** intracranial complication of otitis media (especially in children < 5 years old)
- ✓ It can occur in both **acute (infants and children)** and **chronic otitis media (adults)**
- ✓ AOM is the most common secondary cause of meningitis
- ✓ 10–20% risk of postmeningitic partial or total, unilateral or bilateral SNHL
- ✓ May cause ossification of the labyrinth or cochlea
- ✓ All patients with meningeal signs should undergo otoscopic examination to rule out AOM and CSOM

❖ Mode of infection

- ✓ In **infants** and children **Blood-borne infection** is common;
- ✓ In **adults**, it follows **chronic ear disease**, which spreads directly by bone erosion or retrograde thrombophlebitis or channels (Hyrts' fissures);
- ✓ In **one-third** of the patients with meningitis, another intracranial complication may coexist.
- ✓ The rapid onset of meningitis with AOM in a child with sensorineural hearing loss may indicate the presence of an inner ear malformation that allows communication through the oval or round windows to the vestibule, cochlea. and internal auditory canal, traumatic stapes dislocation or perilymphatic fistula
- ✓ Increased risk with Mondini's aplasia (from dilated vestibular aqueduct)
- ✓ **Pathogens:**
 - S. pneumoniae (most common)
 - H. influenzae (non-typable),
 - Nisseria meningitides

❖ Clinical Features

- ✓ Symptoms and signs of meningitis are due to
 1. Presence of infection
 2. Raised intracranial tension
 3. Meningeal and cerebral irritation.
- **(i)** Fever often with chills and rigors.
- **(ii)** Headache.
- **(iii)** Neck rigidity.
- **(iv)** Photophobia and mental irritability , lethargy
- **(v)** Nausea and vomiting (sometimes projectile).
- **(vi)** Drowsiness which may progress to delirium or coma.
- **(vii)** new onset seizures
- **(viii)** Cranial nerve palsies and hemiplegia.

- ✓ **Examination** will show:
 - (i) **Neck rigidity**
 - (ii) **Positive Kernig's sign** (extension of leg with thigh flexed on abdomen causing back pain)
 - (iii) **Positive Brudzinski's sign** (flexion of neck causes flexion of hip and knee)
 - (iv) Tendon reflexes are exaggerated initially but later become sluggish or absent,
 - (v) Papilloedema (usually seen in late stages).
- ❖ **Diagnosis**
 - ✓ CT or MRI with contrast will help to make the diagnosis.
 - ✓ It may also reveal another associated intracranial lesion.
 - ✓ show characteristic meningeal enhancement and rule out additional intracranial complications
 - ✓ Lumbar puncture and CSF examination **confirm** the diagnosis (**CSF is turbid, cell count is raised and may even reach 1000/ml with predominance of polymorphs; protein level is raised, sugar is reduced and chlorides are diminished.**)
 - ✓ CSF is always cultured to find the causative organisms and their antibiotic sensitivity.

Table 4. Normal CSF characteristics and main pathological alterations

Characteristics	Normal	Meningitis		
		Acute bacterial	Viral	Chronic
Pressure (mm/H ₂ O)	100-200	N or ↑	N or ↑	N or ↑
Aspect	Clear	Turbid/purulent	clear/cloudy	Clear/cloudy
Color	Clear	white	Clear	Clear/white
Cytology (mm ³)	Until 4	> 1,000	500-1,000	<500
Cell type	Lymphocytes	Neutrophils	Lymphocytes	Lymphocytes
Protein (mg/dL)	V 5-10 SO 10-25 L 15-45	↑↑	Normal or ↑	Normal or ↑
Glucose (mg/dL)	2/3 from serum	↓	Normal	Normal or ↓
Lactic acid (mmol/L)	<3.5	>3.5	<3.5	<3.5

❖ **Treatment**

✓ **Medical**

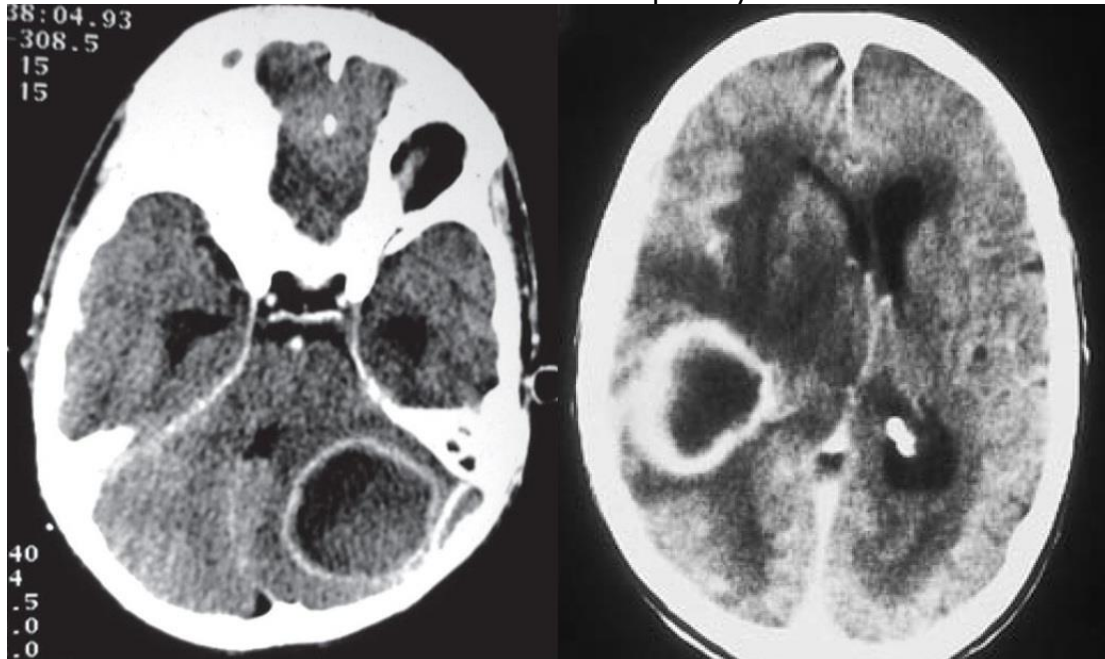
- **Antimicrobial therapy**:- third-generation cephalosporins and vancomycin, should be administered while diagnostic tests are ordered and arranged "gram-negative and anaerobe coverage"
- **Culture and sensitivity of CSF** will further aid in the choice of antibiotics.
- **Corticosteroids** combined with antibiotic therapy significantly reduce the rates of death , neurological & audiological "SNHL" complications., should start early

✓ **Surgical**

- ✓ Meningitis following **acute otitis media** may require **myringotomy or cortical mastoidectomy**.
- ✓ Indications for **mastoidectomy** in otogenic Meningitis include:
 - Presence of cholesteatoma
 - Coalescent mastoiditis
 - Bony erosion with direct extension of disease
 - Persistence of symptoms despite maximal medical therapy
- ✓ Surgery is undertaken as soon as general condition of patient permits.
- ✓ It may be done urgently, if there has been no satisfactory response to medical treatment.

4. Otogenic Brain Abscess

- ✓ 50% of brain abscesses in adults and 25% in children are otogenic in origin.
- ✓ In adults, usually follows chronic suppurative otitis media with cholesteatoma,
- ✓ In children, usually follows acute otitis media "rare"
- ✓ Most common sites are the temporal lobe or cerebellum
- ✓ Cerebral abscess is seen twice as frequently as cerebellar abscess.

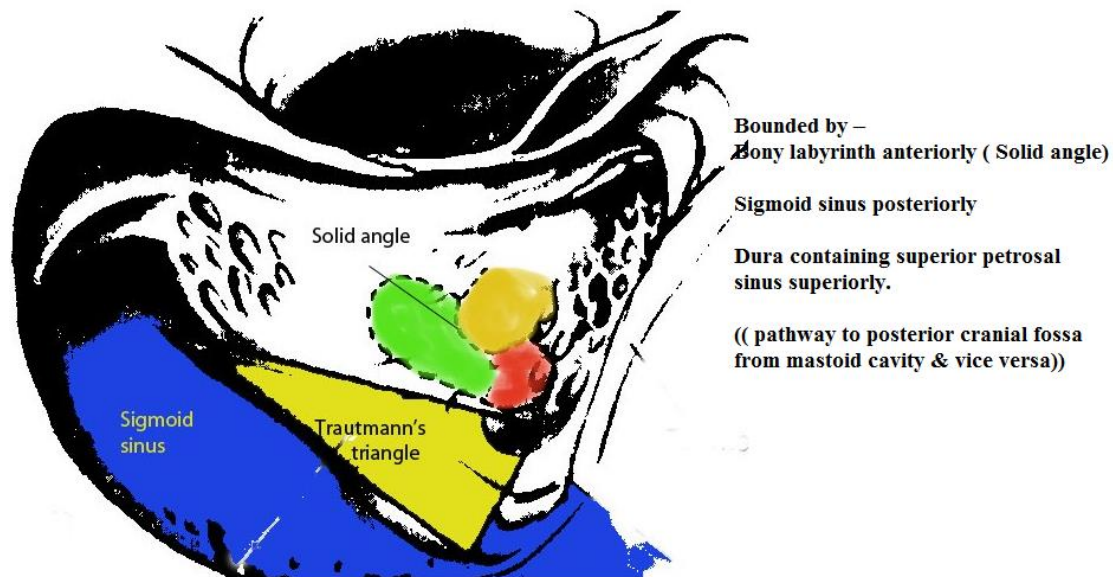


left-sided cerebellar abscess

temporal lobe abscess

❖ Route of Infection

- ✓ Direct extension of middle ear infection through the tegmen (cerebral) , Trautmann's triangle (Cerebellar)
- ✓ Retrograde thrombophlebitis, in which case the tegmen will be intact.
- ✓ Often it is associated with extradural abscess, sigmoid sinus thrombophlebitis or labyrinthitis.



❖ **Bacteriology**

✓ Both aerobic and anaerobic organisms are seen.

✓ **Aerobic** include

- Streptococci pyogenic
- Strep. pneumoniae,
- Staphylococcus sp
- Proteus sp

✓ **Anaerobic** "rarely seen"

- Peptostreptococcus
- Bacteroides fragilis.
- H. influenzae

❖ **Pathology** (Brain abscess develops through four stages):

✓ (a) Stage of invasion (initial encephalitis)

- It often passes unnoticed as symptoms are slight.
- Patient may have headache, low-grade fever, malaise and drowsiness.

✓ (b) Stage of localisation (latent abscess)

- There are no symptoms during this stage.
- Localize the pus by formation of a capsule.
- The stage may last for several weeks.

✓ (c) Stage of enlargement (manifest abscess)

- Abscess begins to enlarge.
- A zone of oedema appears round the abscess and is responsible for aggravation of symptoms.
- seizures, LOC seen
- Clinical features at this stage are due to:

1. Raised intracranial tension.
2. Disturbance of function in the cerebrum or cerebellum, causing focal symptoms and signs.

✓ (d) Stage of termination (rupture of abscess)

- An expanding abscess in the white matter of brain ruptures into the ventricle or subarachnoid space resulting in fatal meningitis.

❖ **Clinical Features**

✓ Brain abscess is often associated with other complications, such as extradural abscess, perisinus abscess, meningitis, sinus thrombosis and labyrinthitis, and **thus the clinical picture may be overlapping.**

✓ **Clinical features can be divided into:**

- **(i)** Those due to raised intracranial tension,
- **(ii)** Those due to **area of brain affected**. They are the localizing features.

✓ **(a) Symptoms and signs of raised intracranial tension**

- **(i) Headache.** severe and generalised, worse in the morning.
- **(ii) Nausea and vomiting.** usually projectile.
- **(iii) Level of consciousness.** Lethargy, which progresses to drowsiness, confusion, stupor and finally coma.
- **(iv) Papilloedema** is absent in early cases. Appears late when raised intracranial tension has persisted for 2-3 weeks.
- **(v) Slow pulse and subnormal temperature.**

✓ **(b) Localizing features**

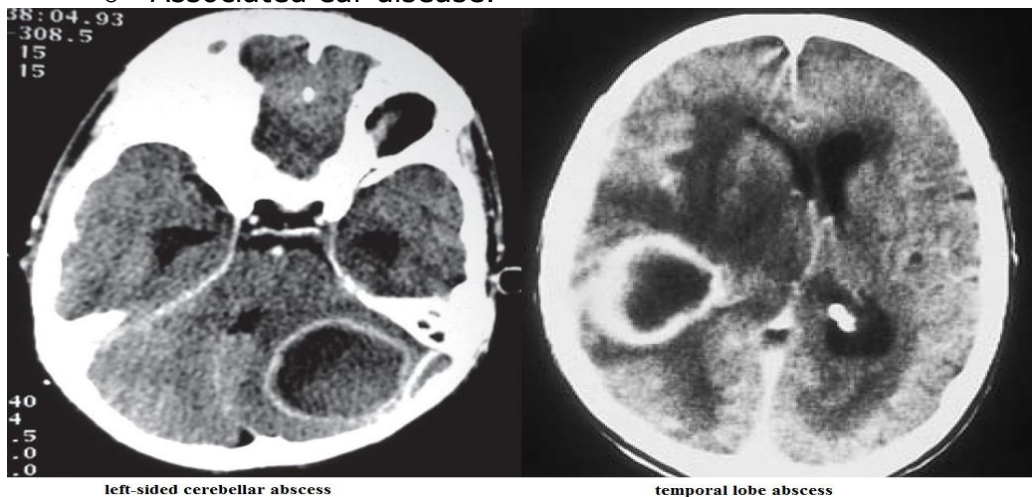
Localizing features	
Temporal lobe abscess	Cerebellar abscess
• (i) Nominal aphasia. If abscess involves dominant hemisphere, patient fails to tell the names of common objects such as key, pen, etc. but can demonstrate their use. • (ii) Contralateral upper quadrantic Homonymous hemianopia. This is due to pressure on the optic radiations. • (iii) Contralateral motor paralysis. • (iv) Epileptic fits. • (v) Pupillary changes and oculomotor palsy. It indicates transtentorial herniation.	• (i) Headache & Neck stiffness • (ii) Spontaneous nystagmus is common and slow irregular and generally to the side of lesion. • (iii) Ipsilateral hypotonia and weakness. • (iv) Ipsilateral ataxia. Patient staggers to the side of lesion. • (v) Past-pointing and intention tremor can be elicited by finger nose test. • (vi) Dysdiadokokinesia. Rapid pronation and supination of the forearm shows slow and irregular movements on the affected side. • (vii) Vertigo

❖ **Diagnosis**

- ✓ Any patient with chronic ear disease who develops pain or headache should be suspected of having **intracranial extension**.
- ✓ Any patient who has otogenic meningitis, labyrinthitis or lateral sinus thrombosis **may** also have a brain abscess.
- ✓ Confirmation and localization of the abscess will require further investigation.

1. **CT**

- Helps to find the site and size of an abscess., bony erosion of the mastoid
- It also reveals associated complications such as:
 - Extradural abscess
 - Sigmoid sinus thrombosis
 - Associated ear disease.



2. **MRI** shows soft-tissue lesions with more detail than CT but gives no bone detail.

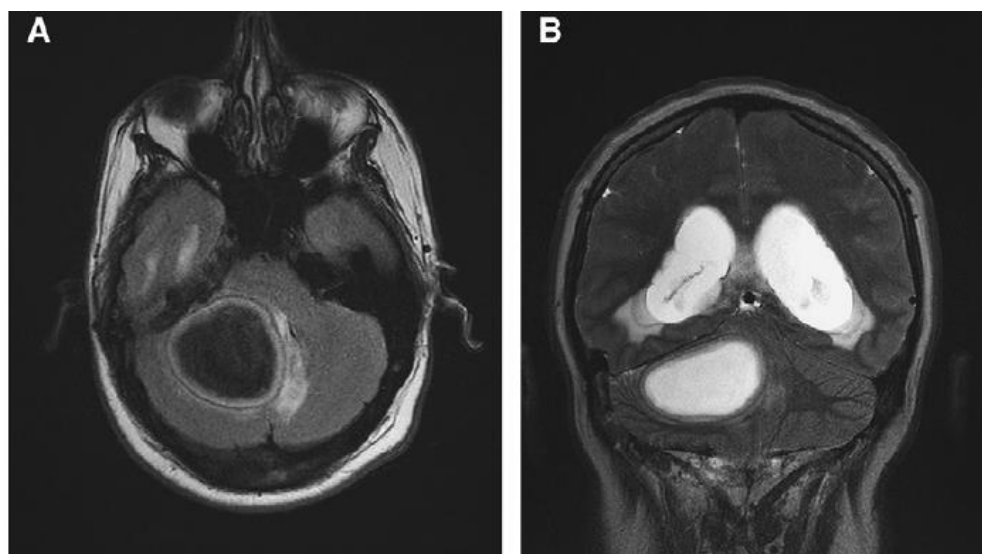


Fig. 7. An axial T1-weighted MRI (A) and a coronal T2-weighted MRI (B) demonstrating a large right cerebellar brain abscess with resulting hydrocephalus.

✓ **Lumbar puncture**

- CSF will show
 - Some rise in pressure
 - Increase in protein
 - Normal glucose level.
 - WBC of raised but is much less than seen in cases of meningitis.

✓ **CT findings in Temporal Bone Infection Contraindicating Lumbar Puncture**

- Lateral Shift of midline structures
- 4th Ventricle Obstruction
- Loss of Cisterns (Basal, Suprachiasmal, Superior Cerebellar, or Quadrigeminal Plate Cisterns)

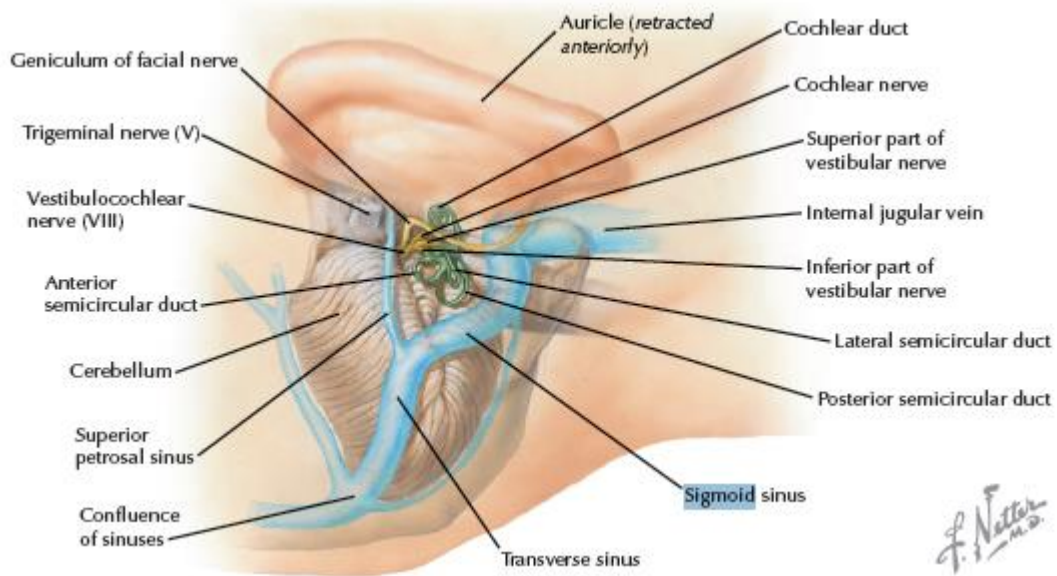
Treatment		
Medical	Neurosurgical	Otologic
○ High doses IV antibiotics gram positives, gram negatives, and anaerobe ○ Raised intracranial tension can be lowered by dexamethasone, 4 mg i.v. 6 hourly or mannitol 20% in doses of 0.5 g/kg body weight. ○ Discharge from the ear should be treated by suction clearance and use of topical ear drops.	○ The choice of surgical procedure is left to the judgement of the neurosurgeon. ○ <u>Options include:</u> i. Aspiration through a burr hole ii. Excision of abscess, iii. Open incision of the abscess and evacuation of pus.	○ Associated ear disease which caused the brain abscess needs attention. ○ AOM :myringotomy with evacuation of the purulent effusion is sufficient ○ COM would require mastoidectomy to remove the irreversible disease and to exteriorise the infected area ○ Surgery of the ear is undertaken only after the abscess has been controlled by antibiotics and neurosurgical treatment.

❖ **PROGNOSIS**

- ✓ The prognosis of brain abscess has improved with the use of antibiotics and modern diagnostic methods but still carries a high mortality rate may be 70%.
- ✓ Left untreated, death from brain abscess occurs from pressure coning, rupture into a ventricle or spreading encephalitis.

5. Lateral Sinus Thrombophlebitis (Sigmoid Sinus Thrombosis)

- ✓ It is an inflammation of inner wall of lateral venous sinus (sigmoid and/or transverse sinus) with formation of a **thrombus**.
- ✓ 17% to 19% of intracranial complications

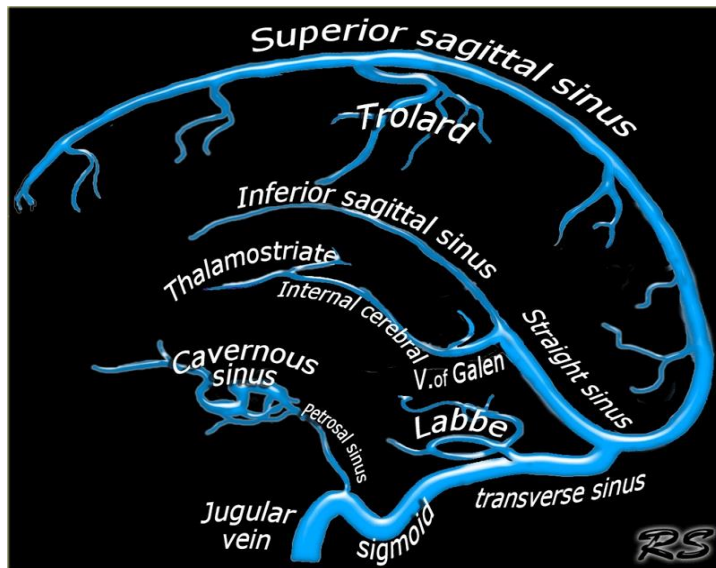


❖ Aetiology

- ✓ It occurs as a complication of:
 - Chronic suppuration of middle ear and cholesteatoma
 - Acute coalescent mastoiditis
 - Granulation tissue
 - AOM "common factor in children"

❖ **Pathology**

Pathological process can be divided into the following stages:	
(a) Formation of perisinus abscess Or granulation	○ Abscess or granulation forms in relation to outer dural wall of the sinus. ○ Overlying bony dural plate may have been destroyed by coalescent bone erosion or cholesteatoma. ○ Sometimes, it remains intact when route of infection was by thrombophlebitis of mastoid emissary veins
(b) Endophlebitis and mural thrombus formation	Inflammation spreads to inner wall of the venous sinus with deposition of fibrin, platelets, and blood cells leading to thrombus formation within the lumen of sinus.
(c) Obliteration of sinus lumen and intrasinus abscess	○ Mural thrombus enlarges to occlude the sinus lumen completely. ○ Organisms may invade the thrombus causing intrasinus abscess which may release infected emboli into the blood stream causing septicaemia.
(d) Extension of thrombus	○ Thrombus breaks down due to intrasinus abscess, spread & begins to seed to: ✓ Superior sagittal sinus ✓ Cavernous sinus ✓ Mastoid emissary vein ✓ Jugular bulb or jugular vein "risk of septic pulmonary emboli"



❖ Bacteriology

- Beta-hemolytic Streptococcus
- S. pneumoniae
- Pseudomonas
- Staphylococcus (including MRSA)
- H. influenza
- Klebsiella, Enterococcus, Proteus
- Anaerobes

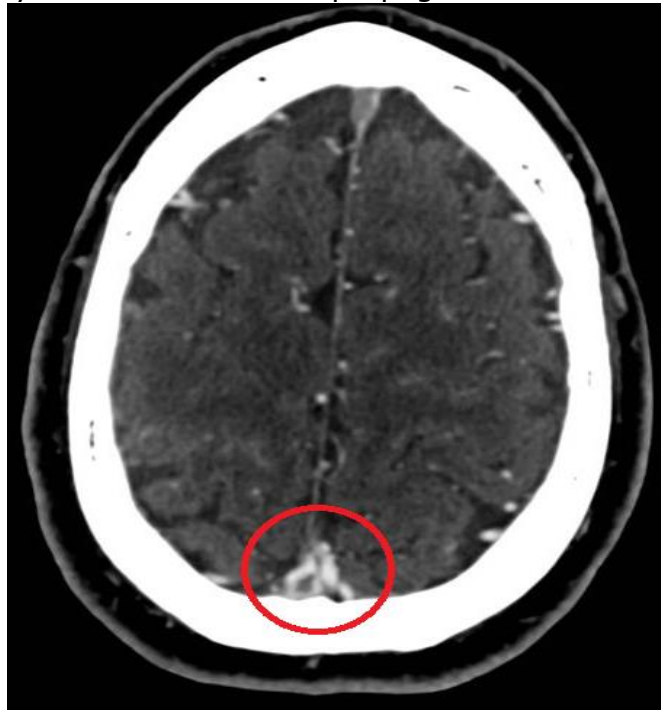
❖ Clinical Features

- ✓ More recent articles have pointed out that this classic pattern clinical features is not seen as frequently, current antibiotic therapy
1. **Swinging pyrexia**— Up to 40°C. "picket fence" (due to septicaemia)
"single high-fever reading should alert the clinician to the possibility of sigmoid sinus thrombophlebitis "
 2. **Rigors.**
 3. **Polymorph leucocytosis.**
 4. **Positive Tobey–Ayer test (Quesckenstedt's test):**
 - Compression of **contralateral** internal jugular vein rise in CSF pressure.
 - Compression of **ipsilateral** internal jugular vein no rise (secondary to obstruction).
 5. **Griesinger's sign** (Pain over mastoid from occlusion of the mastoid emissary vein)
 6. **Neck pain and tenderness** along the anterior border of the sternocleidomastoid muscle
 7. **Meningeal signs**— Sometimes.
 8. **Positive blood cultures**, especially if taken during a rigor.
 9. **Papilloedema**—Sometimes.
 10. **Metastatic abscesses**—Prognosis poor.
 11. **Cortical signs**—Facial weakness, hemiparesis.

❖ **Dx :-**

✓ Typically determined with imaging study:

- **CT with contrast** may reveal enhancement within the sinus , characteristic Empty Delta sign "enhancement of the triangular sinus wall around non-enhancing intraluminal thrombus produces"
- **MRI** more sensitive, reveals increased signal intensity in both T1 and T2 weighted images
- **MRV/MRA** may reveal total/partial occlusion, can be used serially to evaluate for clot propagation or resolution.



empty delta sign

❖ **Complications**

- **1.** Septicaemia and pyaemic abscesses in lung, bone, joints or subcutaneous tissue.
- **2.** Meningitis and subdural abscess.
- **3.** Cerebellar abscess.
- **4.** Thrombosis of jugular bulb and jugular vein with involvement of IXth, Xth and XIth cranial nerves.
- **5.** Cavernous sinus thrombosis. There would be chemosis, proptosis, fixation of eyeball and papilloedema.
- **6.** Otitic hydrocephalus, when thrombus extends to sagittal sinus via confluens of sinuses decreased venous drainage from. intracranial hypertension ,sudden worsening of a severe headache , high mortality rate

❖ **Management :-**

1. Surgery

- ✓ Presence of COM with or without cholesteatoma treatment is a **Surgical**
 - **Minimum** mastoidectomy with removal of chronic infection, granulation, and cholesteatoma is required.
 - Sigmoid sinus is exposed and the surrounding epidural abscess or granulation is removed.
- ✓ The best way to manage the sinus itself is a point of **controversy** in the otology literature. **Most texts recommend** a diagnostic **needle aspiration** of the affected sinus, once it is exposed surgically.
 - If the aspiration reveals normal blood return, then the sinus is left intact;
 - If the aspiration is negative or reveals frank pus, the sinus is opened (Venotomy) and at least a portion of the infected clot is evacuated.
- ✓ Recent reports demonstrated that if the surrounding granulation tissue and inflammation are removed through a mastoidectomy, the sinus will recanalize without clot evacuation
- ✓ Report demonstrated that with sinus thrombosis in the presence of **AOM**, myringotomy and IV antibiotics resolved the infection, and the sinus was shown to recannulate in three subjects without mastoidectomy
- ✓ **Ligation of the internal jugular vein** is not necessary unless there is evidence of continued septic embolization after surgical intervention and IV antibiotics. considered in the presence of **septic emboli**.
- ✓ Post op repeat MRI and MRV should be performed to rule out the development of a secondary intracranial complication such as brain abscess, or propagation of the thrombus into the superior sagittal sinus.

2. Antibiotics

- ✓ After surgical intervention, the patient should remain on IV antibiotics for at least 2 weeks

3. Anticoagulants

- ✓ When the thrombus is isolated to the sigmoid sinus , not been shown to improve recanalization
- ✓ Not necessary unless the clot is shown to involve the sagittal sinus transverse sinus or cavernous sinus, or signs of increased intracranial pressure persist despite medical management.

6. Otitic Hydrocephalus

- ✓ Increased ICP **secondary** to Acute or Chronic Middle Ear disease
- ✓ **With or without** Lateral/Sigmoid Sinus Thrombosis
- ✓ **WITHOUT** evidence of Meningitis or Abscess "normal CSF findings"

❖ Mechanism

- ✓ Pathophysiology is not understood completely
- ✓ Lateral sinus thrombosis accompanying middle ear infection causes obstruction to venous return.
- ✓ If thrombosis extends to superior sagittal sinus, it will also impede the function of arachnoid villi to absorb CSF.
- ✓ Both these factors result in raised intracranial tension.
- ✓ Multiple cases have been described in the absence of otitis or otologic surgery

❖ Clinical Features

✓ Symptoms

- **(a)** Severe headache, may be accompanied by nausea and vomiting.
- **(b)** Diplopia due to paralysis of VIth cranial nerve.
- **(c)** Blurring of vision due to papilloedema or optic atrophy.

✓ Signs

- **(a)** Papilloedema
- **(b)** Nystagmus due to raised intracranial tension.
- **(c)** Lumbar puncture. CSF pressure exceeds 300 mm of water (normal 70-120 mm H₂O). It is otherwise **normal** in cell, protein and sugar content and is bacteriologically sterile.

❖ Diagnosis of otitic hydrocephalus is one of exclusion

❖ Treatment

- ✓ The aim is to reduce CSF pressure to prevent **optic atrophy and blindness**.

This is achieved **medically** :-

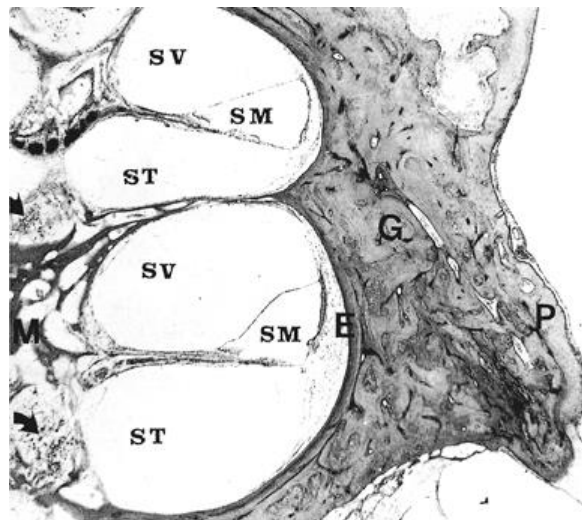
- Initially Acetazolamide "Diamox " (500 mg bid) & corticosteroids
- Mannitol and Furosemide (0.5 g/Kg IV)
- Serial lumbar puncture or placement of a lumbar drain.
- Draining CSF into the peritoneal cavity VP shunt

✓ **Surgical** :-

- Middle ear infection may require antibiotic therapy and mastoid exploration to deal with sinus thrombosis ,Debride Granulations

Otosclerosis (OS):

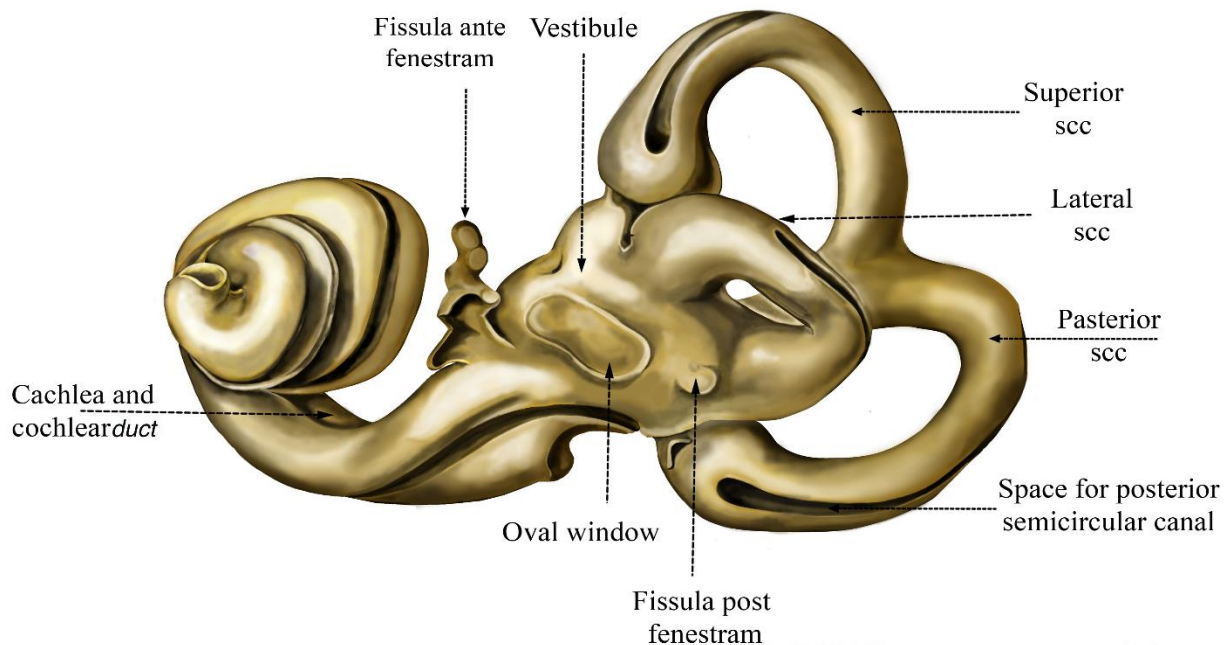
- Fibrous osteodystrophy of human otic capsule.
- Primary disease of the bony labyrinth causing abnormal resorption and deposition of bone.
- Incidence of 1% among Caucasian population.
- Age of onset between 20-40 years.
- Females are affected twice as males.
- Most common cause of progressive conductive hearing loss in adults.
- Definitive diagnosis can be made only during exploratory tympanotomy.
- **Otic Capsule:**
 - o It is the bony labyrinth.
 - o Has three layers:
 - 1. Endosteal layer:**
 - Innermost layer.
 - Lines the bony labyrinth.
 - 2. Endochondral layer:**
 - Develops from the cartilage and later ossifies into bone.
 - It is in this layer that some islands of cartilage are left unossified that later give rise to otosclerosis.
 - 3. Periosteal layer:**
 - Outermost layer.
 - Covers the bony labyrinth.



E: Endosteal layer
G: Endochondral layer
P: Periosteal layer

- **Embryology**

- Maturation of bony labyrinth plays a role in pathogenesis of OS.
- Otic capsule arises from mesenchyme surrounding the otic vesicle at 4 weeks of embryologic development.
- At 8 weeks, cartilaginous framework is initiated.
- At 16 weeks, endochondral osseous replacement of cartilaginous framework begins in 14 centers.
- In some people, complete bony replacement does not occur and leaves cartilage in certain locations.
- **Fissula ante fenestram:**
 - Located anterior to oval window (OW).
 - Last area of endochondral bone formation in the labyrinth.
 - Most common site of OS (80-90%).
- Other areas of otosclerotic lesions:
 - Border of round window (RW)
 - 2nd most common site of OS (30%).
 - Apical medial wall of cochlea
 - Area posterior to cochlear aqueduct
 - Region adjacent to the semicircular canals
 - Stapes footplate



- **Histopathology:**

- Three forms of otosclerotic lesions:

1. Otospongiosis (Early active phase):

- Early lesions appear adjacent to fissula ante fenestram as sheets of connective tissue that replace bone.
- Osteocytes resorb bone around preexisting blood vessels, which causes widening of the vascular channels and dilation of microcirculation.
- Otoscopic exam can reveal the reddish hue caused by these lesions (**Schwartz sign**).
- Formation of new spongy bone occurs which has enlarged marrow spaces and rich with blood vessels and connective tissue.
- Healthy surrounding bone has few viable osteocytes and chondrocytes and is relatively avascular.
- With (H&E) staining, this new spongy bone appears densely blue (**The blue mantles of Manasse**) which can be found also in 20% of normal temporal bones.

2. Transitional phase

3. Otosclerosis (Late phase):

- Predominant finding is formation of sclerotic, dense bone in areas of previous osseous resorption.
- The vascular spaces that were once dilated are narrowed due to bony deposition.
- Although OS begins in endochondral bone, as the spongiosis and sclerosis continue, the endosteal and periosteal layers also become involved.
- Advanced lesions spread across stapedial annular ligament and cause stapedial fixation.
- If the lesion progresses medially to involve endosteum of the cochlea, SNHL results.
- It may spread in both directions, resulting in a mixed sensorineural-conductive hearing loss.

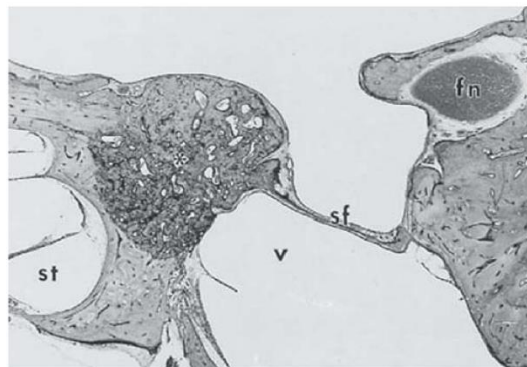


Figure 154.1 Photomicrograph of temporal bone section in a 79-year-old Caucasian male demonstrating a mature otosclerotic focus (*) involving the promontory and anterior stapes footplate (sf). st, scala tympani; v, vestibule; fn, facial nerve. (x20) (Courtesy of M.M. Paparella, director, and S. Lamey, coordinator, Otopathology Laboratory, University of Minnesota).

- **Etiology of OS:**

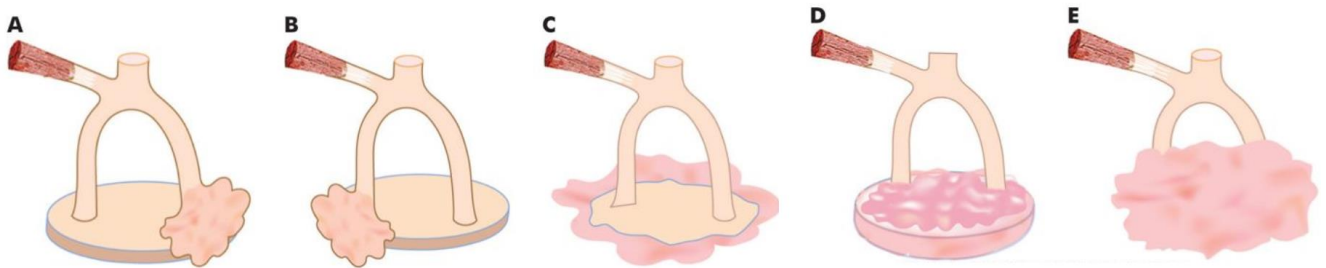
- Exact cause of otosclerosis is not known.
- The following facts have been documented in OS:
 - **Heredity:**
 - Autosomal dominant transmission with incomplete penetrance (25-40%).
 - 30% of cases are sporadic.
 - 50-60% of otosclerotics have positive family history.
 - **Race:**
 - White races are affected more than Negros.
 - **Sex:**
 - Females are affected twice as often as males.
 - **Age of onset:**
 - Hearing loss starts between 20-40 years of age.
 - Rare before 10 years.
 - Juvenile OS may progress more rapidly.
 - **Viral infection:**
 - Electron microscopic and immunohistochemical studies of OS lesions have shown RNA related to measles virus.
 - It is likely that otosclerosis is a viral disease as has been suggested for Paget's disease.
 - **Autoimmune:**
 - No evidence suggesting this etiology.

- **Types of Otosclerosis:**

- Areas of otosclerosis involvement dictate the clinical presentation.

1. Stapedial OS:

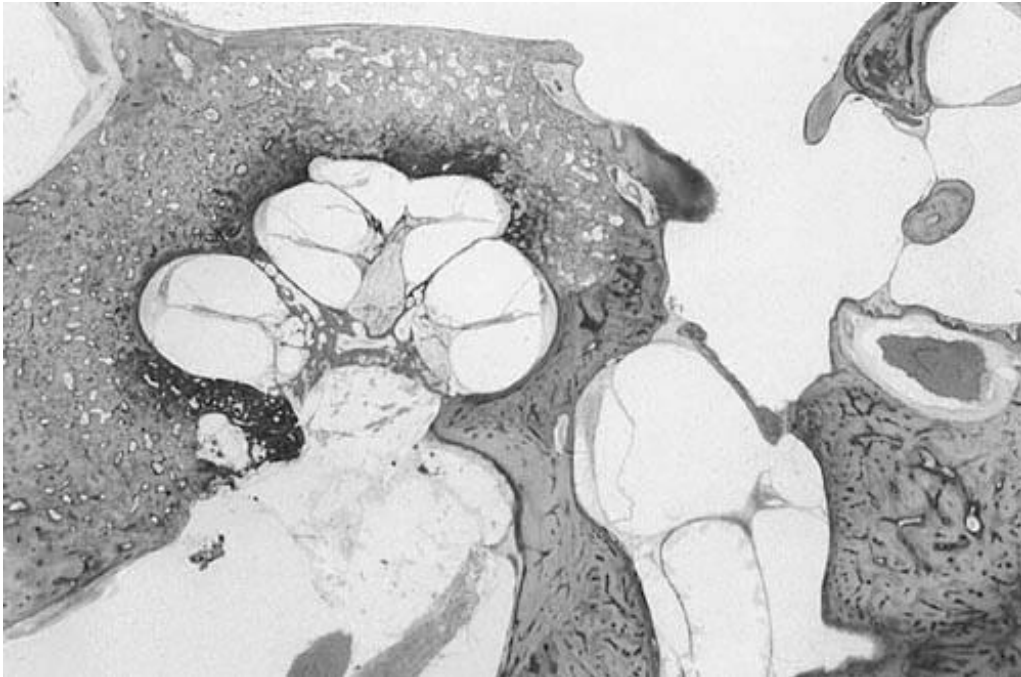
- Most common type of OS involves the stapes.
- Cause fixation of the stapes footplate.
- CHL is the presenting symptom.
- Types:
 - **Anterior focus:**
 - Most common type.
 - Located at Fissula ante fenestram
 - Anterior to oval window.
 - **Posterior focus:**
 - Located at Fissula post fenestram
 - Posterior to oval window.
 - **Circumferential:**
 - Located around the margin of the stapes footplate.
 - **Biscuit:**
 - Located in the footplate without involvement of annular ligament.
 - Minimal fixation may occur.
 - Stapes footplate can become mobilized inadvertently during stapes procedure, placing the patient at higher risk of post-op SNHL.
 - **Obliterative:**
 - Completely obliterate the oval window niche.



2. Cochlear OS:

- Involves region of round window or other areas in the otic capsule.
- Cause SNHL due to liberation of toxic materials into the inner ear fluid or direct extension of lesions into the cochlea.
- SNHL is usually associated with significant stapedial OS.
- Isolated pure SNHL can be seen without associated CHL.
- Rarely associated with vertigo.

- Shambaugh criteria to identify patients presenting with isolated SNHL due to OS (Cochlear OS):
 1. Schwartze sign in either ear.
 2. Family history of OS.
 3. Unilateral CHL consistent with OS and bilateral, symmetric SNHL.
 4. Audiogram with flat or cookie-bite curve with excellent discrimination.
 5. Progressive pure cochlear loss beginning at the usual age of onset for OS.
 6. CT showing demineralization of cochlea typical for OS
 7. Stapedial reflex showing diphasic "on-off effect" seen before stapedial fixation.



- **Clinical presentation:**

○ History:

- Slowly progressive hearing loss over period of years.
- 70% of cases presented with bilateral involvement.
- More apparent to the patient at age 30-40 years.
- More common in females (2:1).
- Hearing improves in noisy situations (Paracusis of Willis) because people speak louder in noisy surroundings.
- Tinnitus present in 75% of cases.
- Rarely associated with vestibular symptoms
 - Mild but persistent.
 - SSCCD and Meniere disease should be ruled out.
- No history of infections or trauma.
- Positive family history of hearing loss in 60% of cases.
- Pregnancy associated with acceleration of otosclerosis.

○ Physical Examination:

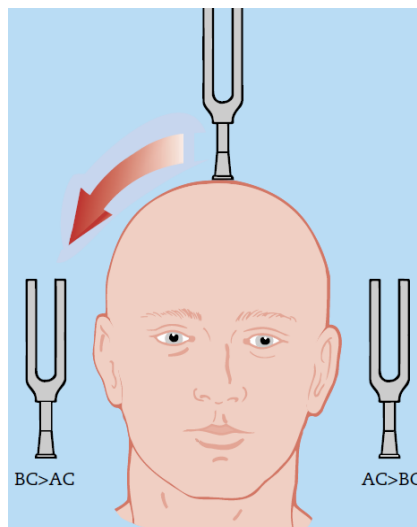
▪ **Oto-microscopic examination:**

- Intact and mobile TM.
- **Schwartz sign:**
 - Red appearance seen through TM due to hyperemia of the promontory mucosa.
 - Seen in 10% of active OS due to increased vascularity.



▪ **Tuning fork:**

- Shows picture of CHL.
- Weber will lateralizes to the ear with greater CHL.
- Rinne will be negative (BC > AC) in the affected ears.



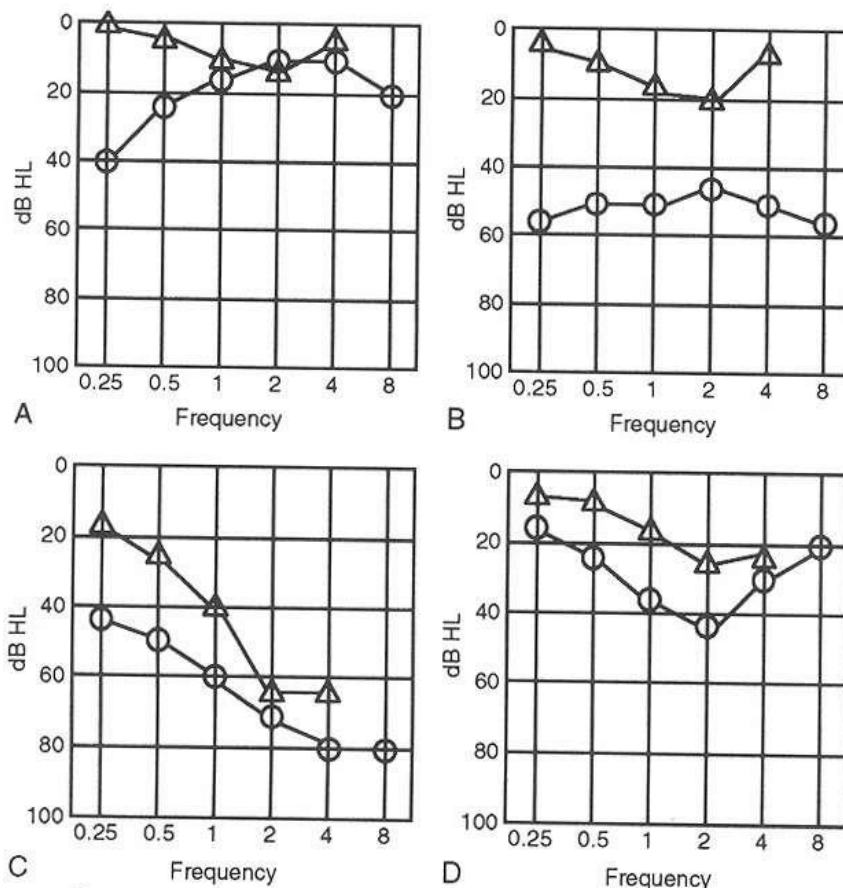
○ Audiometry:

▪ **Pure tone audiometry:**

- Shows mainly CHL with ABG begins in low frequencies.
- In absence of cochlear involvement, maximum CHL produced by complete stapes fixation is 60-65dB.
- In cochlear otosclerosis, AC thresholds continue to worsen and start to have MHL or SNHL, with high frequencies becoming severely affected.

• Carhart notch:

- Depression of BC occurs at different frequencies but maximum at 2000 Hz.
 - 5 dB at 500 Hz
 - 10 dB at 1000 Hz
 - 20 dB at 2000 Hz
 - 5 dB at 4000 Hz
- It is a mechanical artifact secondary to stapes fixation and the change in the normal ossicular resonance, which is around 2000 Hz in human.
- Disappears after successful stapedectomy.
- Occurs in any condition which reduces the inertial vibration of the stapes footplate (tympanosclerosis, ossicular fixation)

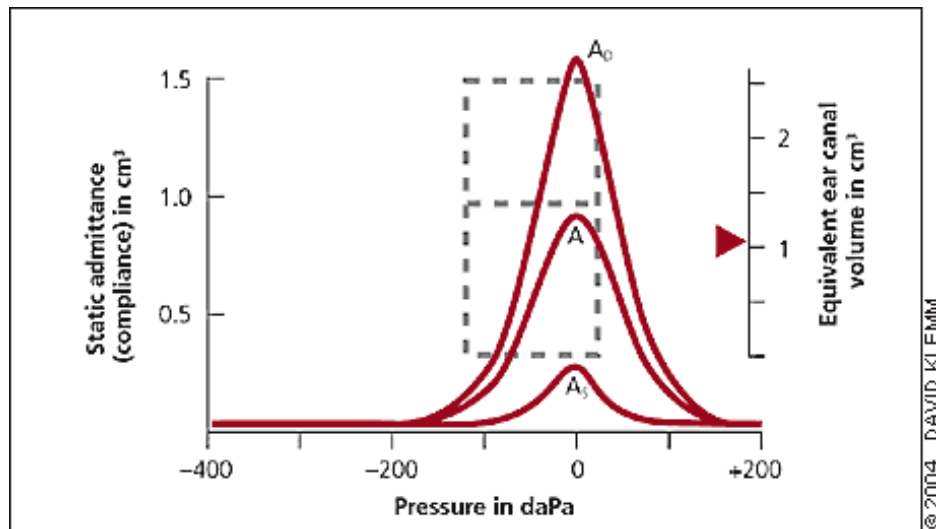


▪ **Speech audiometry:**

- Word recognition scores are usually excellent in patients with OS even in the later stages of the disease process.
- Discrimination will be affected with cochlear involvement.

▪ **Tympanometry:**

- Type A in 95% of patients with OS.
- 5% will show curve of ossicular stiffness. (Type As).
- Multi-frequency tympanometry:
 - Based on the analysis of tympanograms at a wide range of frequencies between 226 and 2,000 Hz.
 - Changes in the transmission characteristics of middle ear system can be easily determined from the changes in the resonant frequency.
 - Otosclerosis increase the stiffness of middle ear system due to the fixation of the stapes and therefore increases resonant frequency.



▪ **Stapedial reflex:**

- Very sensitive indicator of otosclerosis.
- Earliest evidence of OS is Diphasic pattern/on-off effect:
 - Brief increases in compliance occur at onset and end of the stimulus when the probe is in the affected ear.
 - Occurs as a result of the movement of posterior portion of footplate to move independent of the fixed anterior portion of the footplate due to elasticity of footplate.
 - Pathognomonic of early stapedial fixation.
 - Seen prior to development of a detectable ABG in PTA.
- As the disease progresses, stapes gets fixed and both ipsilateral and contralateral stapedial reflexes become absent.
- Presence of stapedial reflexes with significant CHL warrants evaluation for SSCD.

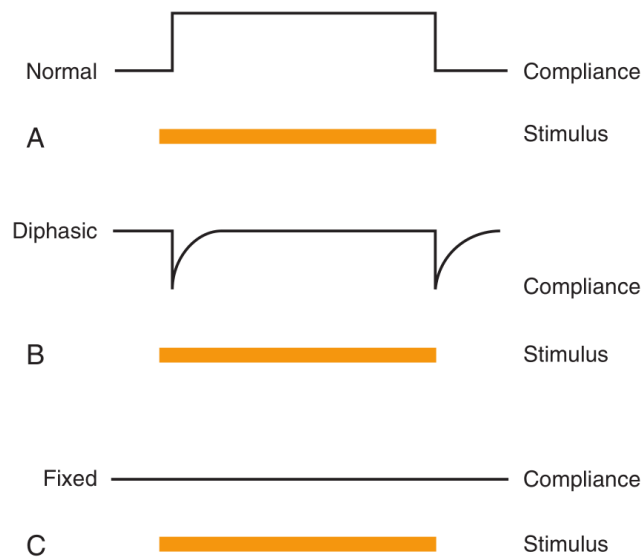
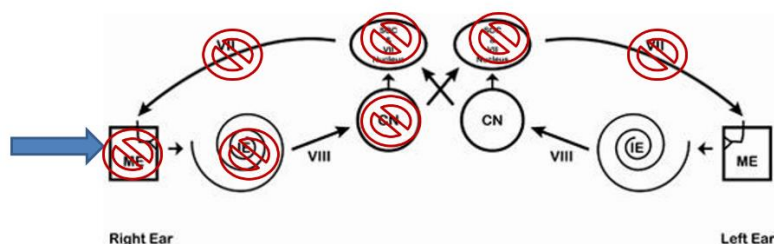


FIGURE 144-5. Progressive changes in the configuration of the acoustic reflex with stapedial fixation. **A**, Healthy reflex with a sustained change in compliance as long as the stimulus is on. **B**, Diphasic reflex with an on-off pattern. This is seen in cases of early otosclerotic stapedial fixation. **C**, As the stapes becomes fixed, the diphasic reflex is replaced by an absent acoustic reflex.



○ Vestibular Tests:

- Indicated in patients with vestibular symptoms to rule out SSCCD and Meniere disease.
- **Vestibular Evoked Myogenic Potential (VEMP):**
 - cVEMP have become a standard test in the workup of a patient with SSCCD or Meniere's disease.
 - Patients with SSCCD will have decreased thresholds (increased sensitivity) to sounds.
 - Patients with Meniere's disease will have increase threshold or absent response to sounds.

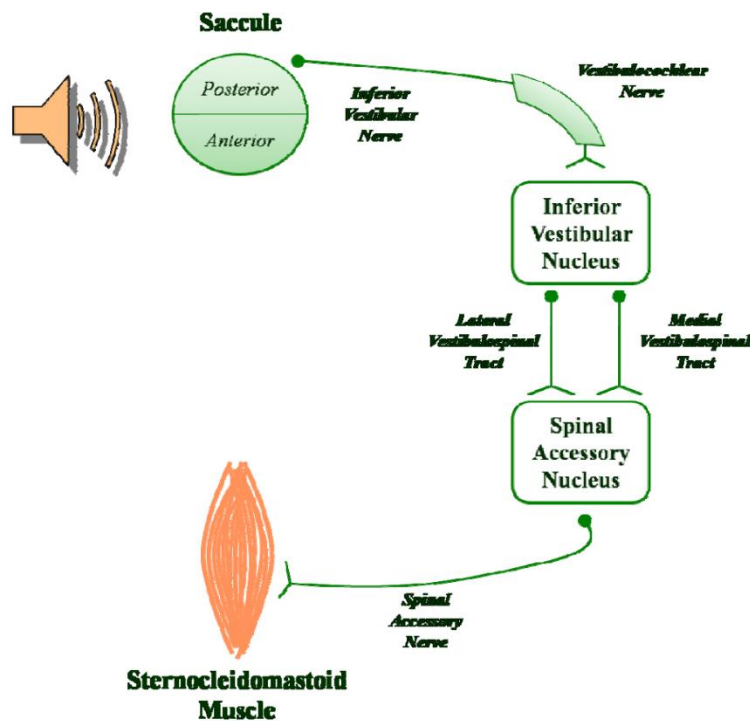


Figure 3. Diagram of the cVEMP neural pathway evoked by an air-conducted stimulus.

	cVEMP	oVEMP
Muscle	SCM	Orbital (inferior oblique)
Response stimulation	- ve	+ ve
End-organ excitation	Saccule	Utricle
Nerve	Inf. Vest. N.	Sup. Vest. N.

- Imaging Studies:
 - **HRCT CT temporal bone:**
 - Indicated in all patients with OS.
 - Objectives:
 - Confirm the diagnosis.
 - Rule out other differential diagnosis.
 - Pre-op planning.
 - Stapedial OS:
 - Hypodense demineralised plaque can be seen in the region of fissula ante fenestram.
 - Cochlear OS:
 - Per-cochlear hypodense “double ring” sign represents demineralized foci in the otic capsule.
 - Basal turn ossification in severe cases.
 - **Normal CT does not rule out otosclerosis.**

Fig. 1 Axial (a) and coronal (b) HRCT images of the right temporal bone in an adult patient with right-sided CHL. A hypodense demineralised plaque (arrow) is noted in the region of the fissula ante fenestram in keeping with fenestral otosclerosis



Fig. 12 Axial HRCT images of the right (a) and left (b) temporal bone in an adult patient with severe bilateral SNHL. The bilateral pericochlear hypodense ‘double ring’ (arrow) is in keeping with bilateral cochlear otosclerosis

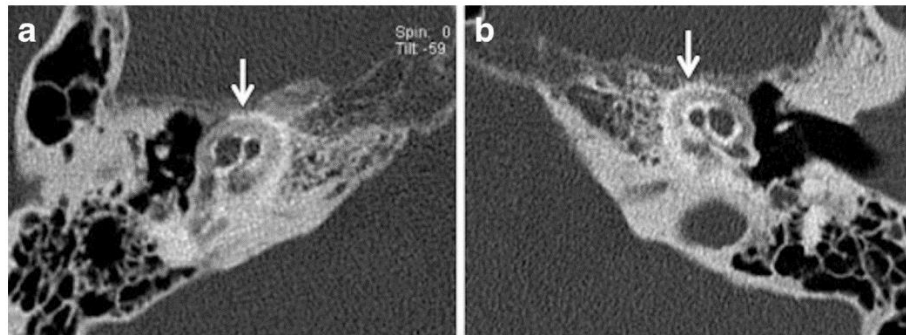


Fig. 4. Axial computed tomography scan of the petrous bone in patients with otosclerosis. (A) Grade 1: solely fenestral involvement, otospongiotic lesion on the anterior border of the vestibulum (white arrow). (B) Grade 2A: double ring effect (black arrow). (C) Grade 3: diffuse confluent cochlear involvement with an unrecognizable cochlea.

Table 1 Reporting checklist for preoperative HRCT of the temporal bone in otosclerosis

	Reporting points	Clinical and surgical relevance
1.	Size and location of plaques	Size and location of plaques may correlate with severity of CHL/air-bone gap
2.	Status of oval window	Complete obliteration may require surgical drilling prior to prosthesis insertion
3.	Status of round window	Obliteration may result in a poor result after stapedectomy
4.	Facial nerve canal	Floppy facial nerve may complicate oval window surgery or render this impossible
5.	Concurrent middle ear pathology	Inflammatory disease must be treated prior to surgery
6.	Ossicular chain integrity	Ossicular fixation, fusion and fracture may compound CHL
7.	Sinus plate and jugular bulb	Dehiscent jugular bulb may complicate surgery
8.	Inner ear pathology	Congenital cochlear and inner ear anomalies may preclude surgery
9.	Opposite ear	Disease is bilateral in 80-85 % cases, even in absence of symptoms

Table 2 CT grading of otosclerosis (Symons/Fanning 2005)

CT grading of otosclerosis	Location of plaques
Grade 1	Solely fenestral
Grade 2	Patchy localised cochlear disease (+/- fenestral involvement) • To basal turn (grade 2A) • To middle turn (grade 2B) • Around lateral aspect of basal, middle, apical turns (grade 2C)
Grade 3	Diffuse confluent cochlear involvement (+/- fenestral involvement)

▪ **MRI:**

- Indicated in patients with cochlear OS planned for CI to evaluate the patency of cochlea.
- Cochlear OS:
 - Ring of peri-cochlear and peri-labyrinthine intermediate signal on T1.
 - Mild-moderate enhancement in T1 post contrast.
 - Obliteration of fluid space in basal turn of cochlea in severe cases.

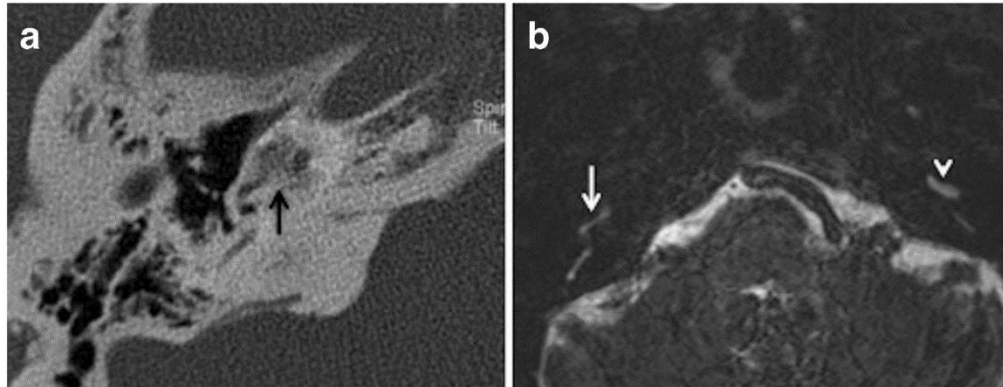


Fig. 17 **a** Axial HRCT of the right temporal bone in a patient with known bilateral cochlear otosclerosis. There is almost complete obliteration of the basal turn of the right cochlea (*arrow*) by the otosclerotic process. **b** Axial MRI of the temporal bones in the same patient as (17a) at the level

of the cochlea. There is significant obliteration of the fluid space in the basal turn of the right cochlea (*arrow*). The normal fluid space in the basal turn of the left cochlea (*arrowhead*) is shown for comparison

**TABLE
154.1**



DIAGNOSIS OTOSCLEROSIS

History

Progressive hearing loss
Family history of OS
Tinnitus
Possible vestibular symptoms (rule out Ménière or SSCD)
Otitis media/otorrhea (absent)
Head trauma (absent)

Physical examination

Tympanic membrane (normal)
Schwartz sign
Rinne test (negative at 256 and 512 Hz)
Weber test (lateralizes to side with greater CHL)

Ancillary studies

Audiogram (assess for CHL, mixed HL, SNHL, Carhart notch)
Tympanometry (type A or As; absent or on-off stapedial reflex)
Imaging (CT scan showing radiolucent areas around bony labyrinth)

CHL, conductive hearing loss; CT, computed tomography; HL, hearing loss; SNHL, sensorineural hearing loss; SSCD, superior semicircular canal dehiscence.

- **Differential diagnosis of OS:**

- **Tympanosclerosis:**
 - Mimic OS but with history of recurrent AOM or VT insertion,
 - TM is thickened with associated myringosclerosis.
- **Incus/malleus fixation:**
 - Lateral ossicular chain fixation, malleus and/or incus become fixed in epitympanum (at superior malleolar ligament).
 - Results in immobility of all the ossicles.
 - Occurs congenitally or may be acquired through tympanosclerosis.
 - Entire ossicular chain must be examined with every exploratory tympanotomy to avoid overlooking this lesion.
 - Almost always associated with type As tympanogram.
- **Congenital footplate fixation:**
 - Presents at earlier age juvenile OS.
 - Can be detected at age of 3 years, whereas juvenile OS was not detectable until age of 10 years.
 - Less likely to have positive family history (10%) compared to juvenile OS (50%).
 - Incidence of congenital malleus fixation is higher in children with congenital footplate fixation (25%) than in children with juvenile OS (3%).
 - 80% of juvenile OS patients had post-op closure of ABG to within 10dB compared to 44% only in patients with congenital footplate fixation.
- **Ossicular discontinuity:**
 - Flaccid TM on pneumatic otoscopy.
 - Type Ad tympanogram.
 - History of recurrent COM suggests incus necrosis.
 - TM may be normal or thickened or atrophic.
 - Fibrous union of incudostapedial joint can produce ABG wider in high frequencies than in the lower frequencies.
 - History of temporal bone trauma:
 - Fracture of stapes super-structure or incus long process have a similar audiometric configuration as OS.
 - Fibrous union forms with resultant resolution of CHL.
 - Distorted TM surface landmarks occasionally provide evidence of prior temporal bone trauma.
- **OME and ME masses:**
 - Audiometry and physical examination will help to make the diagnosis apparent.

- **Superior Semicircular Canal Dehiscence:**
 - Patients will present with low frequency CHL that is not due to middle ear pathology.
 - Autophony and pulsatile tinnitus.
 - Vertigo induced by loud noises.
 - Normal stapedial reflex.
 - CT scan will show SSCD.
- **Paget Disease (Osteitis Deformans):**
 - Disease with diffuse bony involvement that is histologically similar to OS.
 - In contrast to OS, Paget disease begins in the periosteal layer and involves the endochondral bone last.
 - Temporal bone involvement can produce SNHL, but stapes involvement or fixation rarely occurs.
 - Elevation of the serum alkaline phosphatase level and involvement of other skeletal bones are often seen.

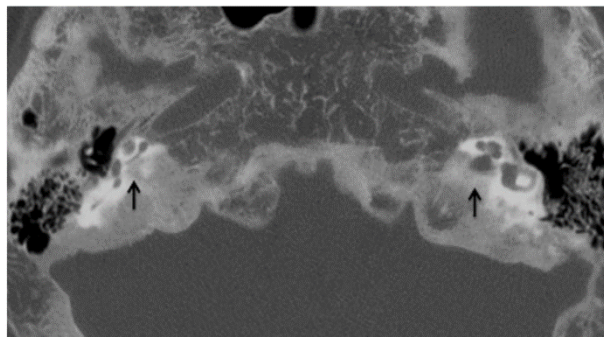


Fig. 16 Axial HRCT of the skull base in an adult patient with known Paget's disease. The hypodense appearance of bilateral otic capsules (*arrows*) may mimic otosclerosis; however the diffuse skull base involvement indicates the true pathology

- **Osteogenesis Imperfecta (Van der Hoeve syndrome):**
 - Autosomal dominant defect of osteoblast activity.
 - Should be suspected in presence of:
 - Blue sclera
 - Progressive CHL
 - Multiple fractures.
 - CHL is secondary to stapes fixation and these patients can have stapes surgery with results similar to those with otosclerosis.



- **Treatment of OS:**

○ **Observation:**

- Indications of observation:
 - Unilateral OS.
 - Mild CHL.
 - Young age.
- Follow-up for progression with audiograms every 6-12 months.
- Patients with special needs should be encouraged to consider surgery or amplification even if they have unilateral loss.

○ **Conventional Hearing Aids:**

- All patients with OS and CHL should be offered HA during counseling.
- Indications of HA:
 - Patients not fit medically for surgery.
 - Patients refusing surgery.
 - Only hearing ear.
- Patients with stapedial OS have excellent discrimination and HA may provide effective treatment.
- Patients with advanced OS and severe MHL can obtain serviceable hearing with HA after stapedectomy.
- HA avoids risk of SNHL that could occur from stapes surgery.
- However, HA is not capable of providing a good satisfaction compared to a successful stapes surgery.

○ **Medical Management:**

- Indications of Medical management:
 - Cochlear otosclerosis.
 - Vestibular symptoms
 - Pre-op stabilization
- Stabilize active OS to prevent progression of CHL, SNHL and dizziness.

1. Sodium Fluoride:

- Fluoride ions replace hydroxyl ions forming a more stable fluorapatite complex (instead of hydroxyapatite crystal) that resists osteoclastic degradation.
- 60mg daily to obtain the maximum bone calcifying effect.
- Evaluation of efficacy is based on:
 - Disappearance of Schwartz's sign
 - Stabilization or improvement in hearing
 - Improvement in CT appearance of otic capsule.
- Contraindication:
 - Chronic nephritis
 - Rheumatoid arthritis
 - Pregnancy, lactation and children

2. Vitamin D.

3. Calcium supplement.

○ **Stapes Surgery:**

- Objectives of stapes surgery:
 1. Open oval window for sound transmission to labyrinth.
 2. Reconstruct the conductive bridge between incus and the labyrinth.
 3. No complications.
- Indications of stapes surgery:
 - **Socially unacceptable CHL or MHL:**
 - >30 dB ABG
 - Negative Rinne with 512 Hz tuning fork.
 - Good speech discrimination (>60%).
- Poor candidates of stapes surgery:
 - **Young patients:**
 - High risk of OW re-closure after successful initial procedure.
 - High risk of AOM and eustachian tube dysfunction.
 - **Biscuit and obliterative OS:**
 - 62% success rate with closure of ABG to 10 dB.
 - Higher risk of SNHL from drilling (4%).
 - **Revision surgery:**
 - Lower success rate 65%.
 - Higher risk of post-op SNHL (7%).
 - HA is good alternative.
- Important pre-op considerations:
 - **Unilateral OS:**
 - Optimal results can be obtained only if:
 - Hearing in the two ears becomes nearly equal after surgery.
 - **Bilateral OS:**
 - Initial stapes surgery should be done for the poorer hearing ear.
 - Better ear can be operated after 6–12 months of uncomplicated first ear surgery due to risk of delayed post-op SNHL.
 - **Children:**
 - Children < 5 years with CHL >30db should be fitted with HA and surgery postponed in unilateral cases until the child is old enough to participate in the decision process.
 - Children > 5 years with CHL >30db and bilateral congenital stapes fixation, and SRT >35 dB may be considered surgical candidates if the child or parents do not accept HA.

- Results of stapes surgery:
 - **Hearing:**
 - Post-op hearing results reveal closure of ABG within 10dB of the pre-op BC level in 90% of patients.
 - Immediate hearing improvement is noted in some patients intra-op or in the recovery area.
 - Other patients report gradual hearing improvement with occasional associated vestibular symptoms due to serous labyrinthitis.
 - Balance disturbances usually resolve within 1-2 days up to few weeks.
 - Post-op audiogram done 2-3 months to allow enough time for middle ear fluid and blood to resorb.
 - 10% of patients experience either worsening hearing or no improvement
 - 2% of patients suffer persistent SNHL.
 - 1% of patients will have profound SNHL.
 - **Tinnitus:**
 - 85% of patients with tinnitus improved after stapes surgery.

- Relative contraindications of stapes surgery:
 - 1. Certain Occupations:**
 - **Occupation requiring repeated exposure to barometric pressure changes (Scuba divers):**
 - Greater risk for post-op fistula and prosthesis dislocation.
 - **Occupation requiring excellent balance:**
 - Risk of post-op vertigo and dizziness.
 - **Occupation requiring taste function (Chefs):**
 - Risk of post-op dysgeusia secondary to stretching or cutting the chorda tympani nerve.
 - 2. TM perforation:**
 - Greater risk of post-op SNHL.
 - TM should be fully repaired before attempted stapes surgery (i.e., staging of the ear).
 - 3. Active Meniere:**
 - Greater risk of post-op SNHL.
 - When endolymphatic space is dilated (endolymphatic hydrops), saccule may be enlarged to and adheres to undersurface of stapes footplate and stapes procedure can injure the saccule resulting in profound SNHL.
 - Patients should be free of symptoms for ≥ 6 months before undergoing stapes surgery.
 - 4. Active OS:**
 - Positive Schwartze sign
 - Pregnancy
 - TM atelectasis
 - 5. Severe Eustachian tube dysfunction**
 - 6. History of cholesteatoma**
 - 7. Active otitis externa.**
 - 8. Active otitis media.**
 - 9. OME.**
- Absolute contraindications of stapes surgery:
 - 1. Only hearing ear.**
 - 2. Better hearing ear.**

- Types of stapes surgery:
 - Aims to perform a fenestration over the footplate into the vestibule, which varies in size depends on the technique.
 - With larger fenestrations, there is a larger conductive advantage gained in hearing but the vestibule is more prone to trauma which leads to severe post-op vertigo or disequilibrium which usually resolves shortly.
 - Either technique can be used in the hands of an experienced surgeon to obtain satisfactory and stable long-term hearing results.
- **Total Stapedectomy:**
 - First stapes surgery described for OS.
 - Steps:
 - Trans-canal approach.
 - Tympanomeatal flap is elevated from 6-12 o'clock, 6mm lateral to the annulus.
 - TM is elevated with the annulus.
 - Chorda tympani nerve is identified and preserved.
 - Scutum (Medial most posterior-superior EAC wall) is removed to visualize the OW, pyramidal process, tympanic segment of facial nerve.
 - IS joint is separated.
 - Lateral ossicular chain movement is assessed to rule out lateral chain fixation.
 - Stapes superstructure and footplate are palpated.
 - RW reflex is assessed.
 - Stapedial tendon is divided.
 - Placing control holes in footplate.
 - Anterior and posterior crus are fractured.
 - Stapes suprastructure is fractured and extracted.
 - Entire footplate is removed and replaced with graft (temporalis fascia, vein graft, tragal perichondrium, gelfoam) and wire or piston prosthesis.
 - Advantages:
 - Better post-op hearing gain at lower frequencies (250 and 500Hz) compared to partial stapedectomy or stapedotomy.
 - Disadvantages:
 - Higher risk of damage to the inner ear and resultant vertigo or SNHL.

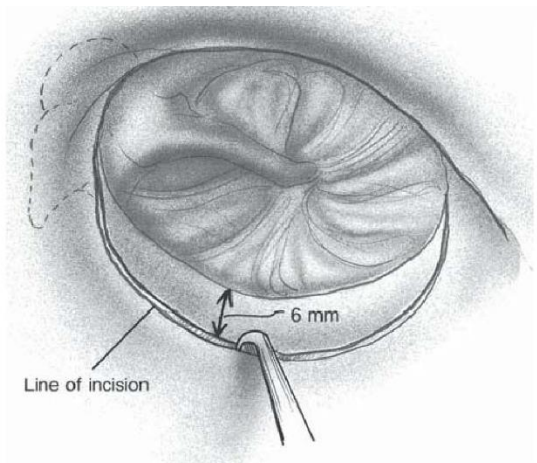


Figure 154.4 Typical design for a tympanomeatal flap.

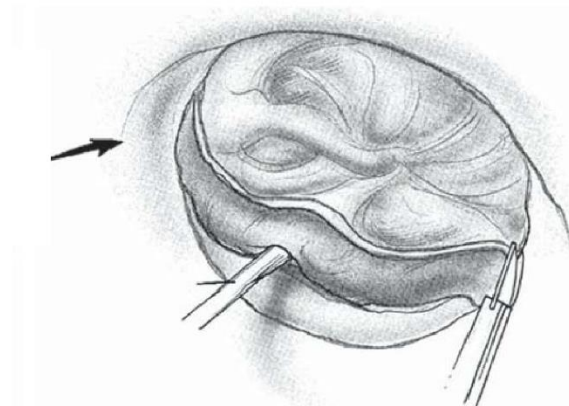


Figure 154.5 Technique for tympanomeatal flap elevation. The skin is elevated to the tympanic sulcus, and then the annulus is elevated from the sulcus.

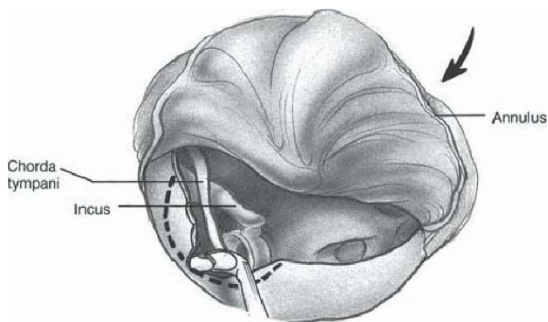


Figure 154.6 The tympanomeatal flap is now elevated. The dotted line indicates the area of bone removal for exposure to the OW.

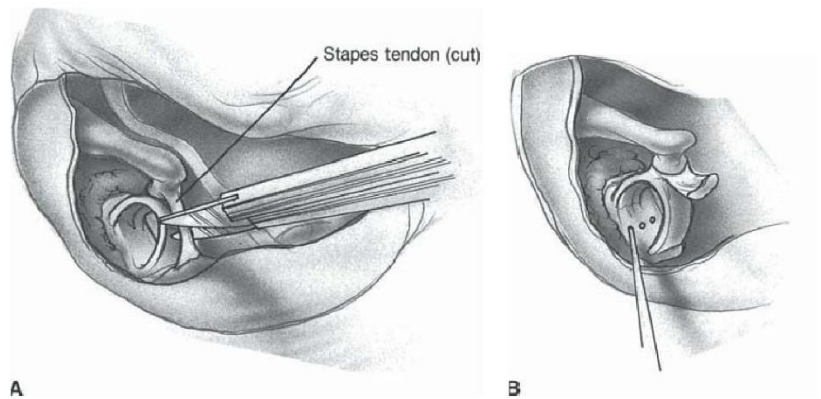


Figure 154.7 A: The tympanomeatal flap is now elevated, and bone has been removed. The stapedial tendon is divided by a pair of microscissors. **B:** Control holes are placed in the stapedial footplate.

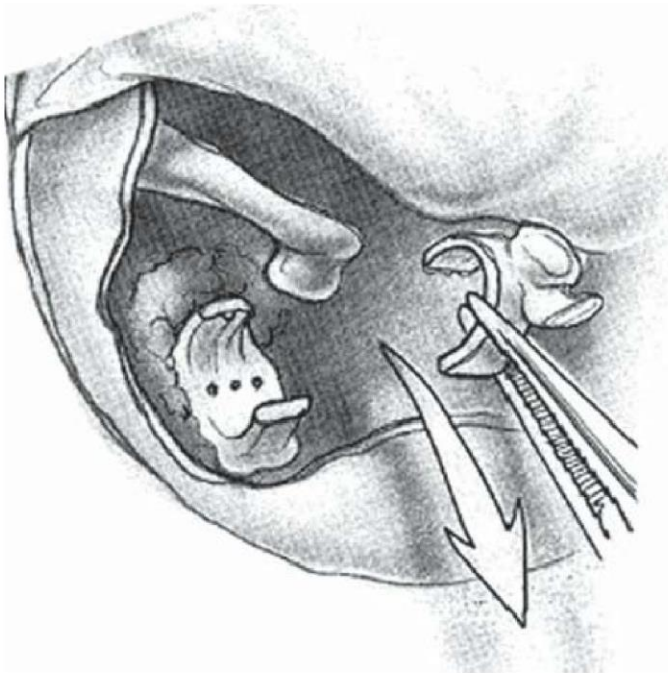


Figure 154.8 The stapedial superstructure is removed after fracturing of the anterior and posterior crus.

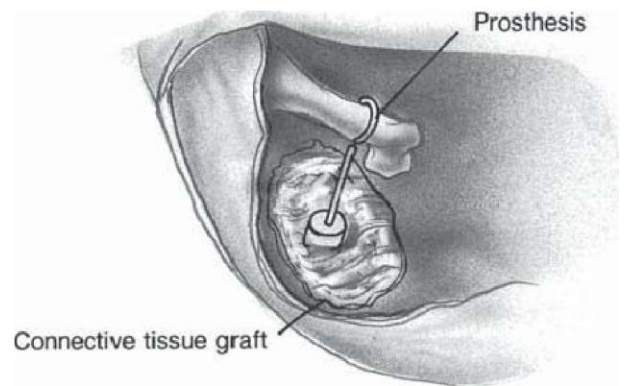


Figure 154.10 Prosthesis in place.

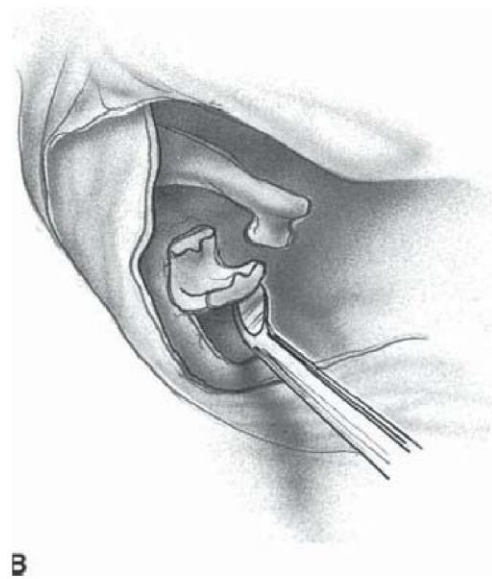
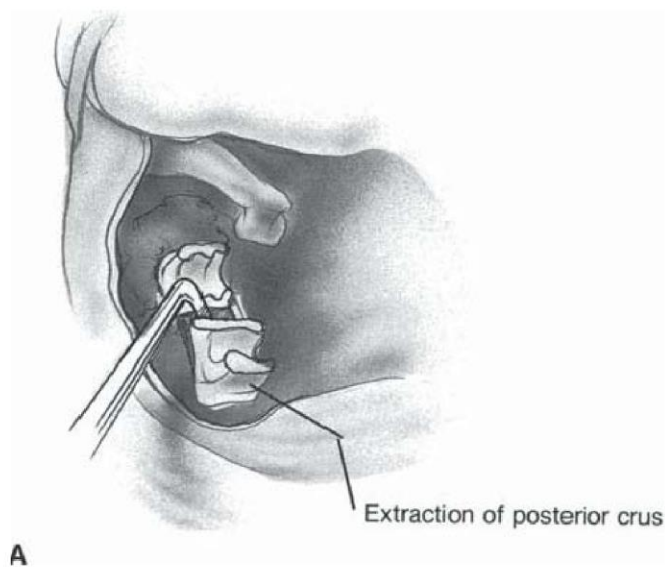


Figure 154.9 **A:** The posterior half of the stapedial footplate is removed with a sharp pick. **B:** The anterior half of the stapedial footplate is removed by a joint knife or Hough hoe.

- **Partial Stapedectomy:**

- Modification of total stapedectomy.
- Considered for patients with isolated anterior fixation at the fissula ante fenestram.
- Steps:
 - Similar initial steps of Total Stapedectomy.
 - IS joint is not separated.
 - Stapedial tendon is not divided.
 - Removal of only the anterior footplate and anterior crus.
 - Connective tissue graft is placed over the exposed area.
- Advantages:
 - Better post-op hearing gain at higher frequencies (4,000 Hz) compared to total stapedectomy.
 - Benefit for patients working in noisy environment due to preservation of stapedial tendon.

- **Stapedotomy:**

- New trend in stapes surgery.
- Longstanding results of stapedotomy procedure have been comparable to total stapedectomy.
- Steps:
 - Similar initial steps of Total Stapedectomy.
 - IS joint is separated.
 - Stapedial tendon is divided.
 - Anterior and posterior crus are fractured.
 - Stapes suprastructure is fractured and extracted.
 - Small circular fenestra is created (with sikeeter or laser) in the center of footplate.
 - A prosthesis is inserted into stapedotomy opening and attached to lenticular process of incus.
 - Clotted blood or soft tissue graft can be used to seal the oval window.
 - Incus is gently palpated to observe the motion of the prosthesis.
- Advantages:
 - Better post-op hearing gain at higher frequencies (4,000 Hz) compared to total stapedectomy.
 - Lower risk of damage to the inner ear and resultant vertigo or SNHL compared to total or partial stapedectomy.



Figure 154.12 A Skeeter drill is used to complete the stapedotomy.

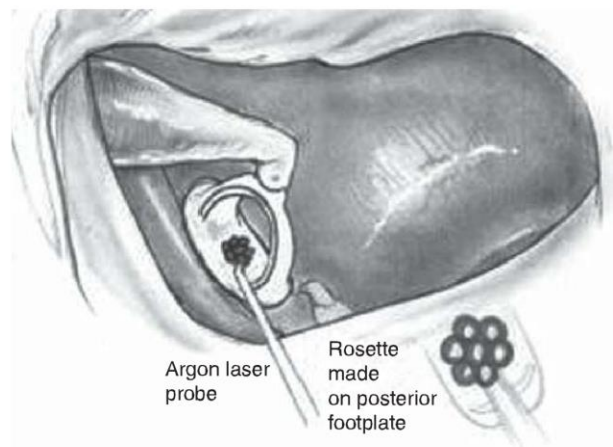


Figure 154.11 A rosette pattern is created with a laser in the midportion of the footplate. This step can be completed prior to down fracturing the stapes or after this step.

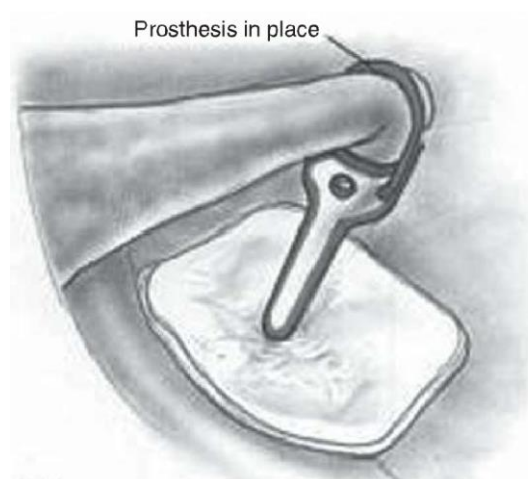


Figure 154.13 A tissue graft can be used to cover the stapedotomy prior to placement of the prosthesis to decrease the risk of a postoperative perilymph fistula. The prosthesis is then engaged between the incus lenticular process and the stapedotomy.

- **Stapes Mobilization:**
 - Performed only in selected group of patients in whom a small point of fixation from OS can be seen and where improved stapes mobility can be clearly demonstrated.
 - High risk of re-fixation.
- Laser vs Microdrill during stapedotomy:
 - The surgeon's operative experience and skill are considered the most important factors in determining the success and incidence of complications.
 - No significant differences in post-op PTA results.
 - No significant difference in post-op SNHL rates.
 - **Microdrill (Skeeter):**
 - More useful for thick footplate.
 - **Laser:**
 - May reduce the mechanical trauma to the stapes, causing decreased labyrinthine irritation and possibly better results.
 - Most common types (Argon, KTP, CO2)
- Types of stapes prostheses:
 1. **Wire teflon piston:**
 - Technically difficult and can result in delayed failure.
 - Wire loop must be manually crimped to incus.
 - If crimped too tightly, incus necrosis may result because of attenuation of the fragile blood supply.
 - If crimped too loosely, vibration of the wire loop may create a gradual erosion through the incus.
 2. **Heat-activated self-crimping prosthesis:**
 - Provides a reliable, consistent snug fit of the wire loop on the incus.
 - Once positioned on the incus, a laser, bipolar, or heating filament can be used to self-crimp the prosthesis.
 3. **Bucket-handle prosthesis**



- Intra-op consideration and complications:

- 1. Bleeding:**

- Intra-op bleeding can be troublesome during the delicate parts of the stapedectomy.
 - Most common cause of bleeding is mucosal trauma.
 - Intra-op bleeding can significantly hamper surgery during the hyperemic or active phase of OS.
 - Some surgeons recommend pre-op sodium fluoride in these patients to stabilize the lesion.

- 2. TM perforation (2%):**

- May occur intra-op during elevation of TM from the sulcus in the posteroinferior area.
 - Avoided by careful identification of annular ligament.
 - Perforation does not prevent completion of the operation.
 - Management:
 - Small perforations usually heal with placement of a small piece of Gelfoam or paper patch over the perforation
 - Larger tears may require an underlay myringoplasty using tissue or sometimes require deferring stapes surgery.

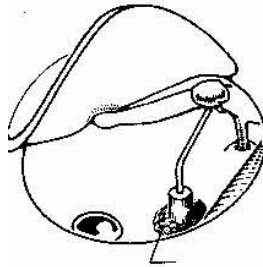
Persistent post-op perforations should be repaired within 4-6 weeks after surgery to prevent any problems with transcanal contamination of the middle ear and a subsequent otitis media and SNHL.

- 3. Lateral Ossicular Chain Fixation (1-10%):**

- Lateral chain movement should be assessed after separation of incudostapedial joint.
 - Fixed malleus head is rare and usually associated with stapes fixation.
 - Management:
 - If no stapes fixation:
 - Incus replacement prosthesis, or
 - TORP with cartilage graft.
 - If there is stapes fixation:
 - Removal of malleus head and incus and the use of malleus to OW prosthesis.

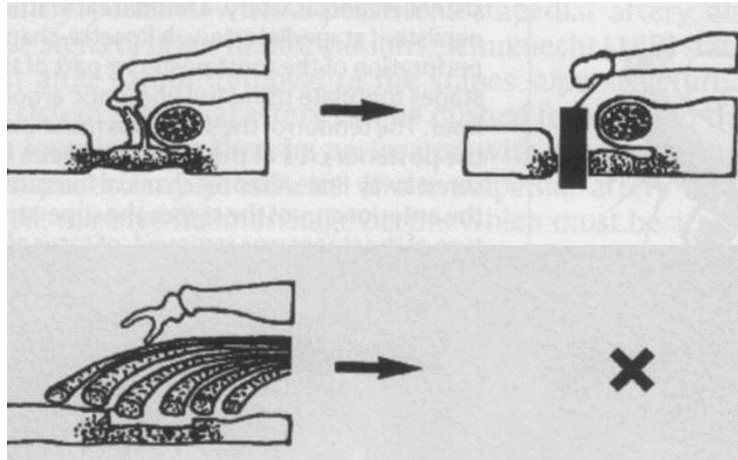
4. Incus subluxation/dislocation or fracture of long process of incus:

- Occurs during:
 - Curettage of scutum.
 - Separation of IS joint
 - Placement of prosthesis
- Management:
 - Placing the prosthesis on the remaining incus (Notched bucket-handle prosthesis) with excellent hearing results.
 - Staging the procedure and coming back at later time may allow incus to heal and be utilized in future.
 - For dislocated incus, remove the incus and use malleus to OW prosthesis.
 - Malleus to OW prosthesis is more difficult to place has poorer post-op hearing results.



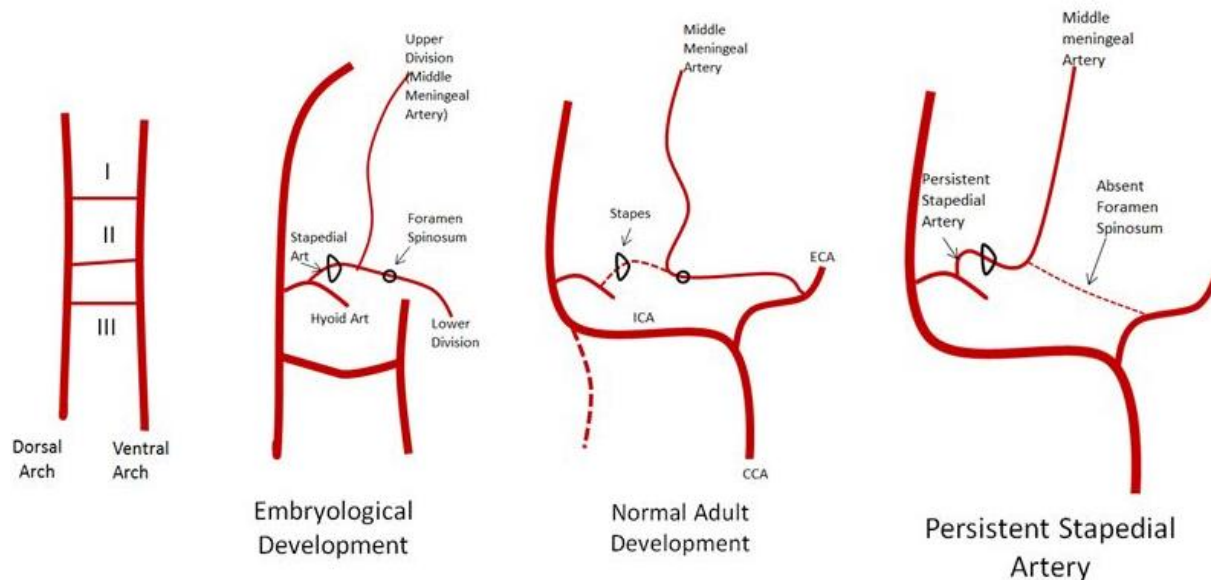
5. Exposed overhanging facial nerve:

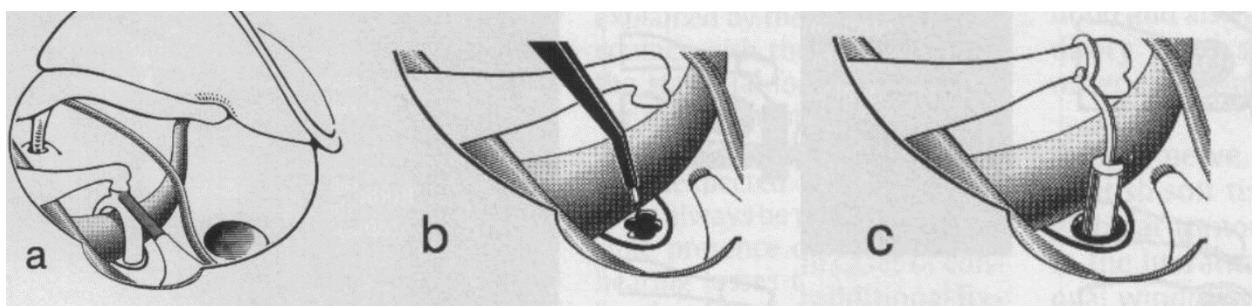
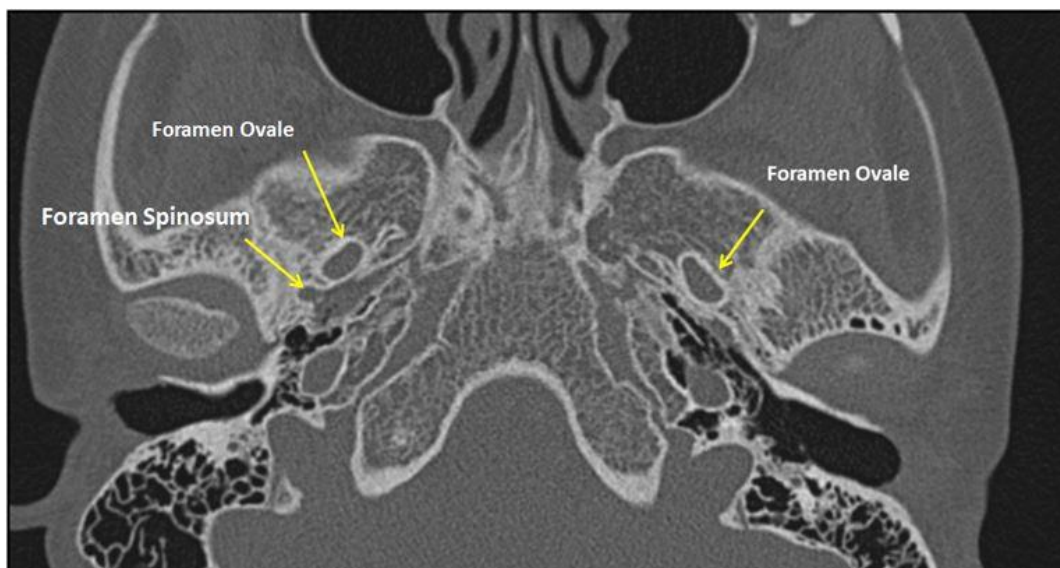
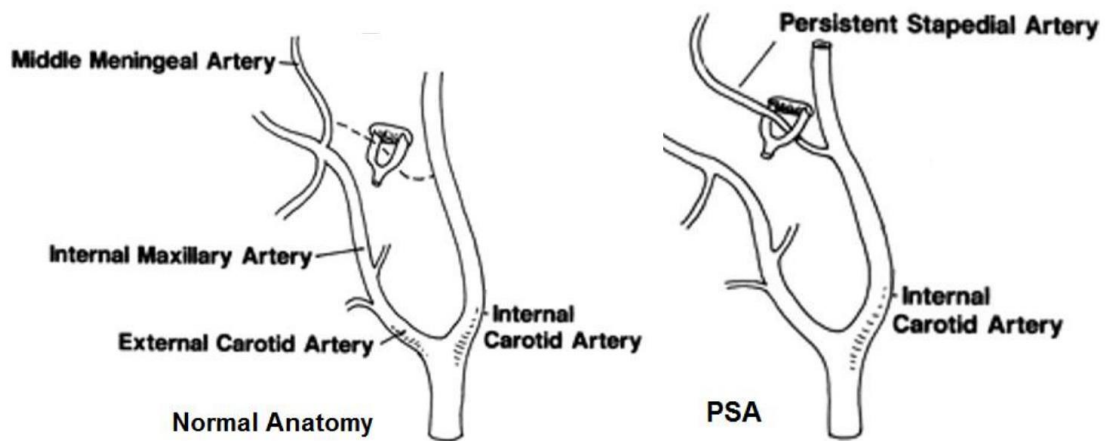
- Exposed facial nerve occurs in 9% of stapes procedures.
- Usually, it does not present a problem intra-op unless it blocks access to the footplate and makes completion of the procedure impossible.
- Management:
 - Retracting the nerve gently superiorly with a small suction catheter, while the drill or laser is used to create the fenestra.
 - A prosthesis touching the facial nerve does not create a problem for post-op hearing or facial function.
 - If the facial nerve is causing a significant block to the access then the procedure should be aborted.



6. Persistent Stapedial Artery:

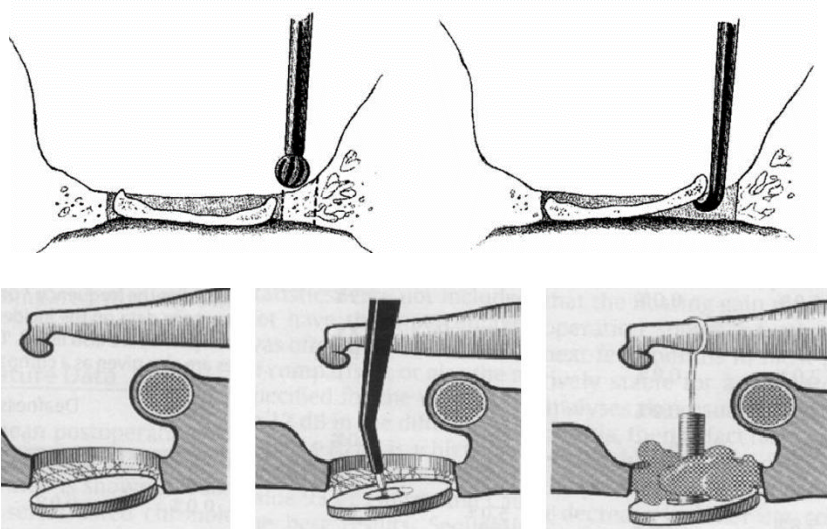
- Embryologic remnant of second branchial arch.
- Originate from internal carotid artery and pass in obturator foramen between the crus of stapes footplate.
- Can be predicted pre-op by CT that shows absent foramen spinosum which contains middle meningeal artery.
- Management:
 - The vessel must be divided or displaced to gain access to the footplate with high risk of bleeding.
 - Persistent stapedial artery can be difficult to coagulate with bipolar cautery or laser and surgery should be aborted.





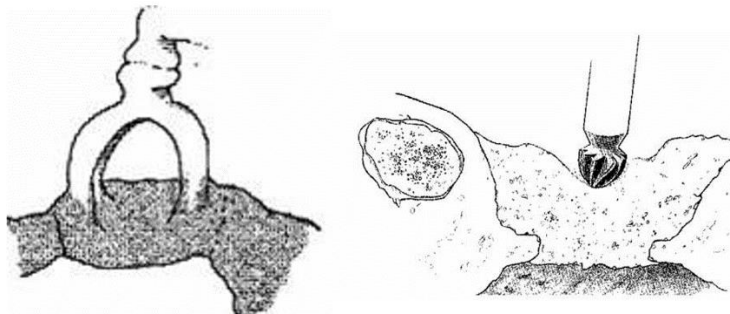
7. Floating footplate:

- Fixed footplate suddenly becomes mobile after stapes superstructure is removed with risk of depressing footplate into the vestibule.
- Rare with the use of laser or microdrill, especially when the crura are left in place until after the prosthesis is placed.
- Attempts to remove the depressed footplate can result in significant SNHL and vertigo.
- Management:
 - If control holes were made:
 - Small right angle hooks or needles placed through the control holes made previously to lift the footplate out of the vestibule.
 - If no control holes made:
 - Drilling a small hole in the promontory at the inferior edge of the footplate then a small hook can be used to elevate footplate gently out of the oval window.
 - Connective tissue graft and prosthesis can be placed lateral to depressed footplate after making the fenestra with microdrill or laser with unpredictable results.
 - May abort the procedure and wait for the footplate to refix and re-operate later (laser minimize floating footplate).



8. Solid or obliterated footplate:

- When present one ear, obliterative OS is present in contralateral ear in 50% of cases.
- Higher risk of SNHL from drilling (4%).
- Management:
 - Microdrill to create a fenestra.
- Partial or complete re-closure of oval window following primary stapedectomy may occur which is considered to be a cause of early failure (within one year) and some authors advocate the use of sodium fluoride post-op in these patients.



9. Perilymph gusher:

- Profuse perilymph gusher (CSF leak) immediately on opening the vestibule.
- Perilymph volume is on the scale of microliters so this abundant flow represents CSF.
- Rare with incidence of 0.03%.
- Associated with congenital footplate fixation in pediatric patients.
- Causes:
 - Modiolar defect with communication to fundus of IAM.
 - Patent cochlear aqueduct. or more commonly a modiolar
- Total stapedectomy in presence of gusher increases the risk of post-op SNHL.
- The control holes placed before footplate removal allows for early identification of this problem.
- Management:
 - Elevation of head of the bed.
 - Small fenestra stapedotomy.
 - Placement of a tissue seal over footplate defect.
 - Lumbar drain may be needed to decompress the subarachnoid space to allow for adherence of the graft to the defect in footplate.

▪ Post-op care:

- Bed rest and head elevation 30 degrees:
 - Reduce the perilymph pressure in the vestibule.
- If no vertigo or dizziness is present, the patient can be discharged home after 2-4 hours of surgery if under LA.
- Instructions:
 - Dry ear precautions until TM is healed.
 - Avoid flying for 5 days after surgery.
 - Avoid nose blowing, straining, diving and lifting heavy objects for 4 weeks after surgery.
 - Cough and sneeze with the mouth open.

▪ Post-op complications:

1. SNHL:

- Most devastating surgical complication.
- SNHL may be mild or isolated to high frequencies.
- 2% of patients will have persistent SNHL.
- 1% of patients will have profound SNHL
- Surgical trauma is the most common cause of permanent SNHL following stapes surgery.
- Laser is associated with a lower incidence of SNHL.
- Risk factors:
 - Extensive drilling (obliterative type).
 - Prior traumatic mobilization.
 - Bleeding (Blood is ototoxic).
 - Surgical instrument trauma.
- Management:
 - Prednisone is started immediately and tapered over 10 days.
 - Course begins with 60 mg daily for 5 days and on the sixth day, the dose is reduced to 40 mg for 1 day, followed by a 10-mg/day reduction until the tenth day.

2. Serous labyrinthitis:

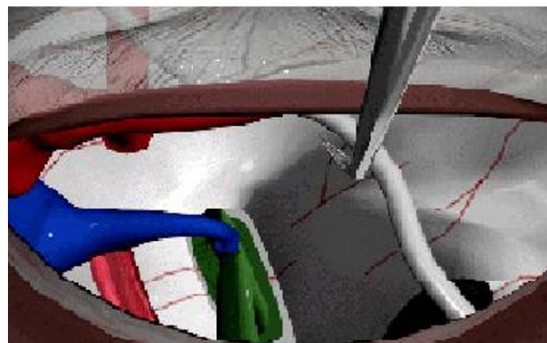
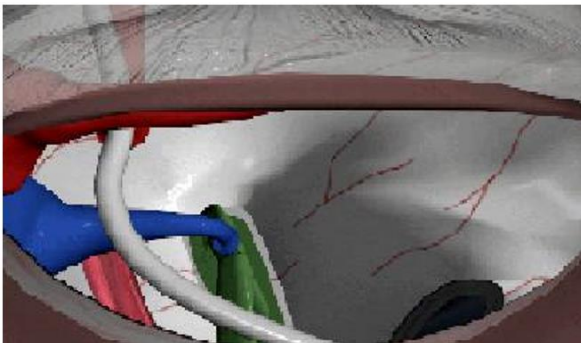
- Common after stapes surgery because of a certain amount of inflammation within the inner ear.
- Patients may exhibit mild unsteadiness, positional vertigo, or high-frequency SNHL.
- Irritative nystagmus (to operated ear) occur usually with serous labyrinthitis.
- Paralytic nystagmus (to non-operated ear) occurs if there is inner ear injury.
- Management:
 - Spontaneous recovery occurs within several days to weeks.

3. Acute otitis media:

- Rare post-op complication
- High risk of SNHL in the operated ear.
- The newly created OW partition allows a middle ear infection to quickly involve the labyrinth (suppurative labyrinthitis) and potentially meningitis.
- Management:
 - Intensive systemic antibiotic therapy for otitis media should initiated immediately to avoid any significant sequelae.

4. Chorda tympani injury:

- Occurs in up to 30% of cases.
- Less severe symptoms are reported with complete sectioning of the nerve than with stretching or partial tearing.
- If the nerve has been stretched or otherwise injured, it is preferable to section it.
- Symptoms include:
 - Dry mouth
 - Tongue soreness,
 - Metallic taste
- Management:
 - Spontaneous recovery occurs within 3-4 months.



5. Partial of total facial weakness:

- Facial nerve paralysis occurs less than 0.5%
- Caused by:
 - Delayed bell's palsy from re-activation of herpes virus without injury to facial nerve (after 5 days post-op).
 - Injury or stretching to dehiscent facial nerve (immediate post-op).
 - Local anesthetic effect (immediate post-op).
- Management:
 - Usually partial weakness and complete recovery occurs within 6 weeks with prednisone treatment in case of delayed bell's palsy.

6. Reparative granuloma:

- Granulation tissue around OW.
- Rarely seen today (0.1%).
- Risk factors for reparative granuloma:
 - Total stapedectomy
 - Use of gelfoam or fat as OW sealant
 - Presence of powder on operating gloves.
- Hallmark of post-op granulation is progressive or sudden SNHL after earlier post-op hearing improvement.
- Occurs within 1-6 weeks post-op.
- Suspected when the commonly seen symptoms of serous labyrinthitis (SNHL and vertigo) persist beyond several days after operation or worsen with time.
- Examination shows a reddish discoloration in posterosuperior quadrant of TM.
- CT scan can be helpful in the diagnosis and to assess status of the prosthesis/vestibule.
- The overall outcome of this potentially devastating process is related to early diagnosis and treatment.
- Management:
 - Prompt recognition and removal of the granuloma from around the oval window, removal of OW seal and prosthesis with replacement using a different material.

7. Perilymph fistula:

- Rare with incidence of 3-10%.
- May occur in the early post-op period or many years later.
- The most common cause of fistula in fistula exploration surgery is total stapedectomy.
- Incidence is much less after stapedotomy with small fenestra technique.
- Presenting symptoms is fluctuating or progressive MHL, tinnitus and vertigo.
- May preceded by history of sudden barometric pressure change or trauma.
- Can be avoided by instructing the patient post-op to avoid nose blowing, flying, diving, & lifting heavy objects.
- Management:
 - Revision surgery with careful removal of the prosthesis.
 - Tissue seal is placed over the open oval window and the prosthesis is replaced.
 - Laser is helpful to remove granulation and scar tissue.

8. Persistent or progressive CHL:

- Most common causes for failure of primary stapes surgery:
 1. Displaced prosthesis (50%).
 2. Erosion of incus long process (25%)
 3. OS bony regrowth (15%)
 4. Adhesions around prosthesis
 5. Missed pathology:
 - Malleus fixation
 - Round window OS
- Management:
 - Revision surgery and removal of prosthesis is associated with a higher incidence of SNHL and a reduced closure of ABG.
 - Hearing aids.

**TABLE
154.2**



**COMPLICATIONS
OTOSCLEROSIS SURGERY**

Intraoperative

Bleeding (high jugular bulb, tympanomeatal flap, persistent stapedial artery, mucoperiosteum)

Facial nerve injury (<1%)

Perilymph gusher

Fracture/dislocation of incus

Floating footplate

Postoperative

Acute otitis media

TM perforation

CHL (middle ear effusion, displaced prosthesis, incus erosion)

SNHL, vertigo, tinnitus (due to intraoperative trauma, labyrinthitis, reparative granuloma, perilymph fistula)

Facial nerve palsy (local anesthetic, intraoperative trauma, delayed Bell palsy)

CHL, conductive hearing loss; OS, otosclerosis; SNHL, sensorineural hearing loss; TM, tympanic membrane.

**TABLE
154.3**

**CLINICAL INDICATORS FOR
STAPEDECTOMY**

Strategy

Indicators (one of the following)

CHL without other source of conductive abnormality
(e.g., perforated TM, serous otitis media)

Laboratory tests (required)

Audiogram with air–bone gap, speech reception threshold,
and discrimination scores

Other tests (as indicated)

Impedance audiometry

Type of anesthesia (as indicated)

Location of service (as indicated)

Process

Criteria for discharge

Recovery from anesthesia

Absence of significant vertigo

Ability to ambulate without assistance

Absence of signs of infection

Absence of significant ear drainage

Outcome

Results

Follow-up

Improvement in hearing

Health of TM

The American Academy of Otolaryngology-Head and Neck Surgery and the American Society for Head and Neck Surgery have published clinical indicators for surgical procedures. These clinical indicators are educational statements that have been drafted to assist surgeons in their practice and to promote discussion. These indicators are not practice guidelines nor do they represent standards of practice with which individuals must conform.

TM, tympanic membrane.

- **Bone Anchored Hearing Aid (BAHA):**
 - Option for patients with CHL or MHL who cannot wear HA.
 - BAHA bypasses the ossicular chain and amplifies sound that stimulates the cochlea directly through bone conduction.
- **Cochlear Implant (CI):**
 - Indications:
 - **Advanced OS with SD scores <30%.**
 - **Advanced OS with CT scan showing grade 2C or 3 regardless the SD score.**
 - Complications are higher compared patients without OS and includes:
 - Failure of electrode insertion due to basal turn obliteration.
 - Partial electrode insertion due to basal turn obliteration.
 - Misplacement of electrode due to basal turn obliteration.
 - Facial nerve stimulation in 20% of patients due to increased conductivity of otospongiotic bone which required reduction in stimulus levels of the electrodes or totally deactivating the causative electrodes.
 - Reduced performance of the implant due to progressive otosclerotic changes in the cochlea.

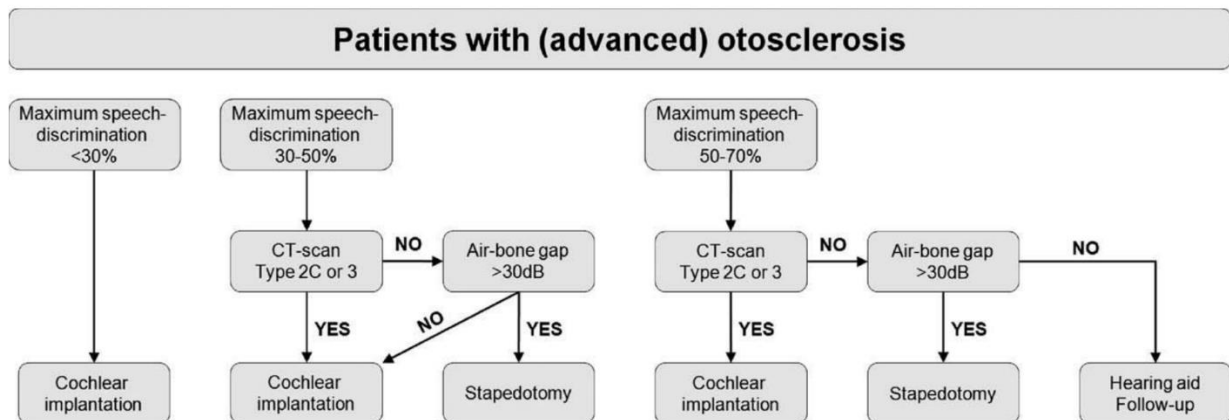


Fig. 2. Algorithm guideline to counsel patients with (advanced) otosclerosis. Algorithm is based on the speech discrimination score, computed tomography (CT) classification, and the air-bone gap, and will guide the surgeon to either cochlear implantation, stapedotomy, or a hearing aid and follow-up.

CPA and Petrous Apex Lesions

- **Cerebellopontine Angle (CPA):**
- Subarachnoid space (cistern).
- Inverted triangular-shaped.
- Located within lateral posterior fossa.
- Extends into the medial aspect of IAC (Porus Acusticus).
- **Contains:**
 1. CSF
 2. Arachnoid tissue
 3. CN-VII
 4. CN-VIII
 5. Anterior inferior cerebellar artery (AICA)
 6. Flocculus of the cerebellum
 7. Foramen Lushka of 4th ventricle
- **Borders:**
 - **Superiorly:** Cerebellar tentorium, **CN-V**
 - **Anteriorly:** Lateral aspect of clivus
 - **Medially:** Brainstem
 - **Laterally:** Petrous bone
 - **Posteriorly:** Cerebellum
 - **Inferiorly:** Cerebellar tonsil, **CN IX, X, XI**

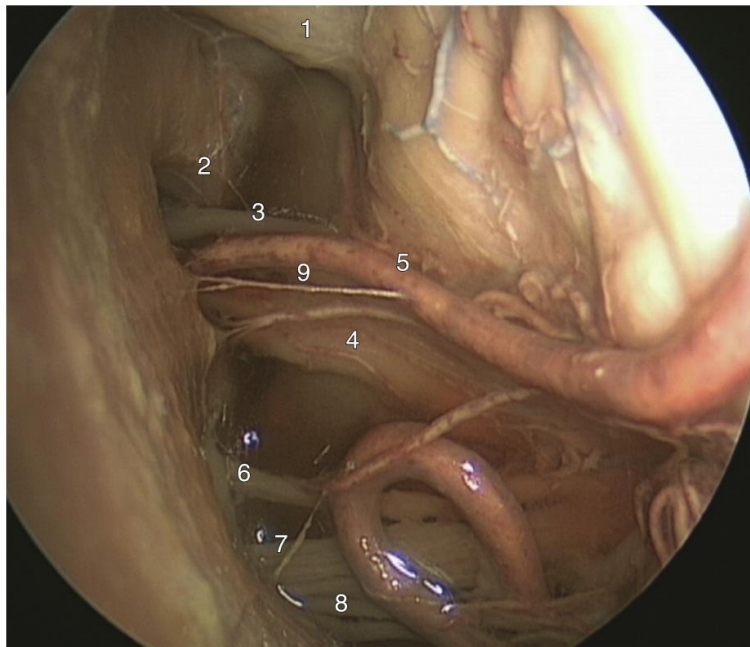


FIGURE 173-17. Anterior view of the cerebellopontine angle and medial wall of petrous bone as in an endoscopic transclival approach (right side). 1, cranial nerve V (trigeminal); 2, interior auditory canal porus; 3, cranial nerve VII (facial nerve); 4, cranial nerve VIII (vestibulo-cochlear nerve); 5, anterior inferior cerebellar artery; 6, cranial nerve IX; 7, cranial nerve X; 8, cranial nerve XI; 9, internal auditory artery. (Courtesy Oswaldo Laércio M. Cruz, Helder Tedeschi, and Albert Rhoton.)

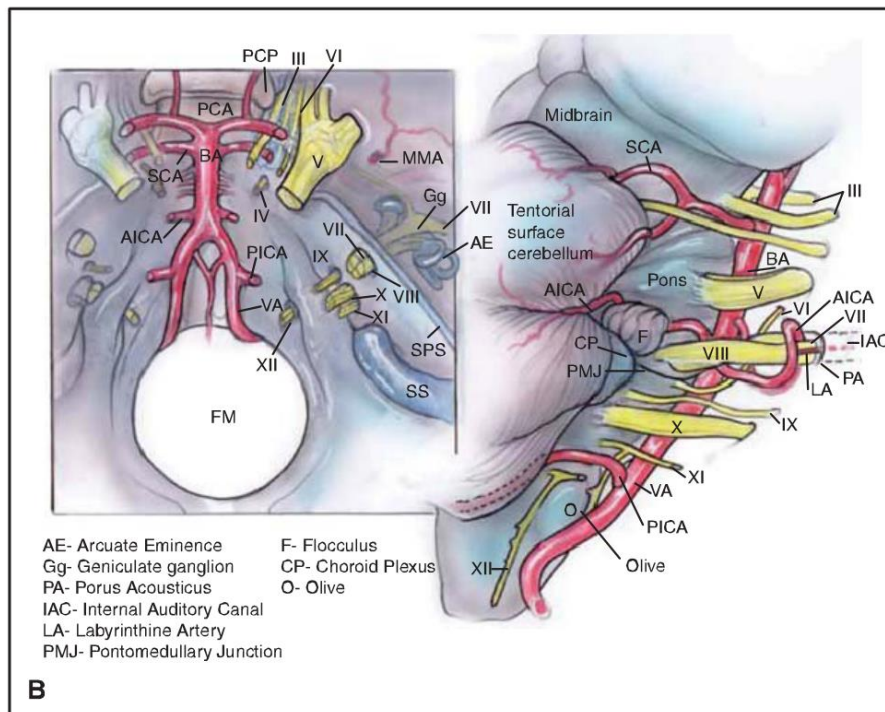
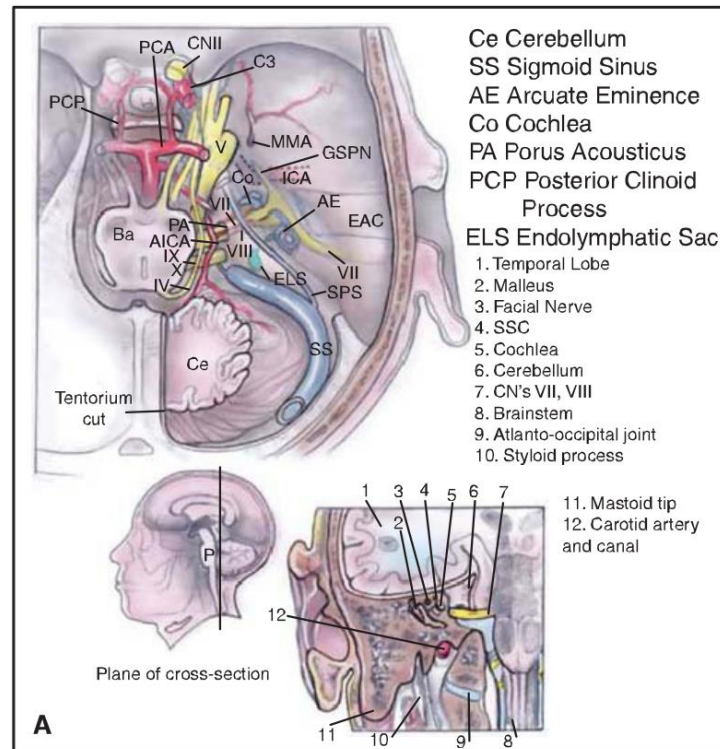
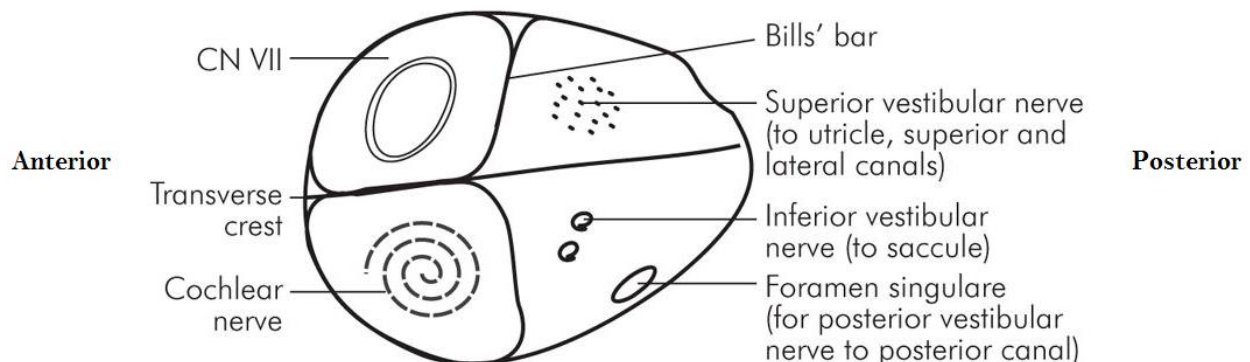
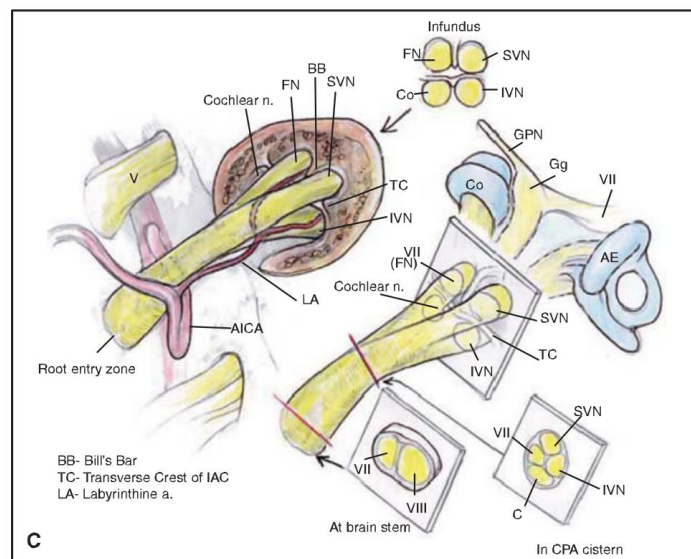
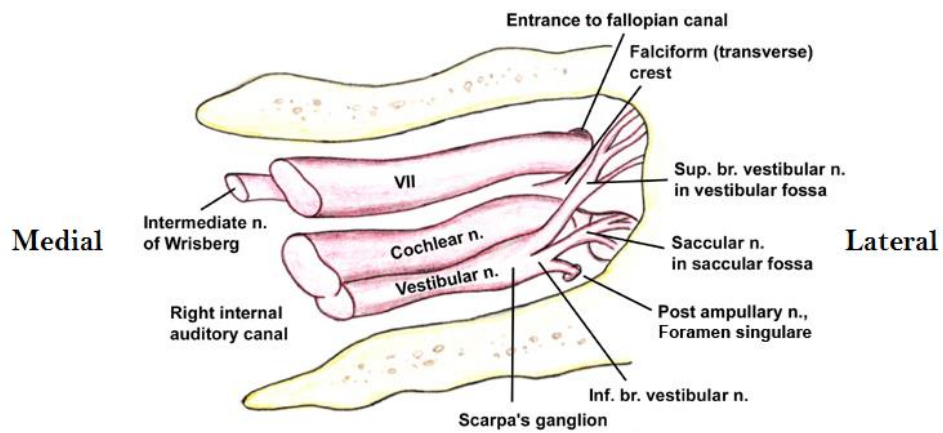
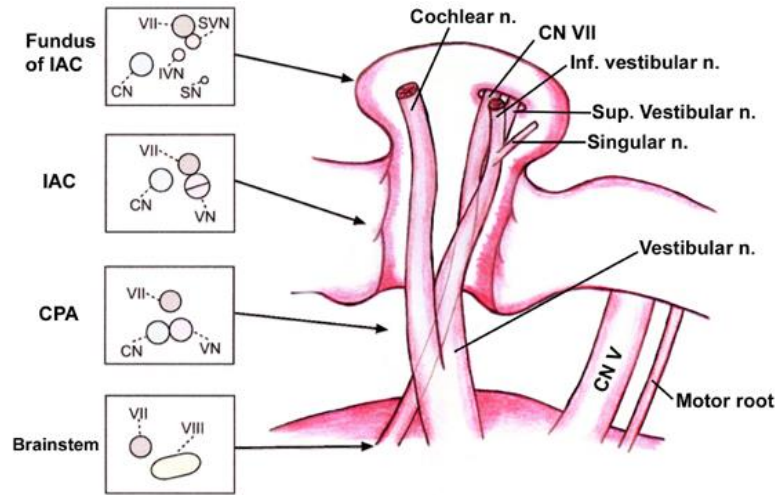


Figure 159.1 Axial and coronal (A) and sagittal views (B) through the skull at the level of the IACs and CPAs. V, trigeminal nerve; VII, facial nerve; VIII, cochleovestibular nerve; IAC, internal auditory canal.

- Course of neurovascular structures within CPA:
 - Trigeminal nerve (**CN-V**) travels within the superior portion of CPA cistern from the lateral pons to the Meckel cave.
 - Facial (**CN-VII**) and Vestibulocochlear (**CN-VIII**) nerves exit the brainstem at the lateral ponto-medullary junction adjacent to foramen of Luschka and travel within the CPA to enter Porus Acusticus.
 - **CN VII and VIII are encased in glial tissue in CPA.**
 - Glossopharyngeal (**CN-IX**), Vagus (**CN-X**) and Accessory (**CN-XI**) nerves extend from the medulla across the inferior aspect of CPA and enter the pars nervosa of jugular foramen.
 - Anterior inferior cerebellar artery (**AICA**), branch of basilar artery, takes a variable course within CPA and loops to enter Porus Acusticus.
 - Gives a branch, Labyrinthine artery, which travels laterally in IAC to supply the cochlea and labyrinth.
- **Internal Auditory Canal (IAC):**
- Directed antero-laterally from CPA cistern.
- Length of 1.2-1.4 cm from Porus Acusticus (medial aspect of IAC) to the Fundus (lateral aspect of IAC).
- Falciform (transverse) crest:
 - Divides the IAC into superior and inferior compartments.
- Bill bar (vertical crest):
 - Divides superior compartment into anterior and posterior portions.
 - Inferior compartment does not have a bony vertical crest.
- **CN VII and VIII are encased in Schwann cells in IAC.**
- **Obersteiner-Redlich zone** is the Glial/Schwann junction.
- Contents (Seven-up Coke-down):
 - Anterior-Superior compartment: **CN-VII**
 - Anterior-Inferior compartment: **CN-VIII**
 - Posterior-Superior compartment: **Superior vestibular nerve**
 - Posterior -Inferior compartment: **Inferior vestibular nerve**





- **Petrous Apex:**
- Pyramid-shaped structure formed by medial portions of temporal bone.
- Its apex is pointing anteromedially and its base located posterolaterally.
- **Boundaries:**
 - Laterally: Inner ear
 - Medially: Petro-occipital fissure
 - Anteriorly: Petro-sphenoidal fissure and ICA
 - Posteriorly: Posterior cranial fossa
- **Divided by IAC into:**
 - **Anterior portion:**
 - Large
 - Types of Pneumatization:
 - **Diploic (filled with bone marrow) in 60%.**
 - **Pneumatized (filled with air cells) in 30%.**
 - From extension of air cells from mastoid along:
 - Infra-labyrinthine.
 - Anterior tract
 - Superior tract
 - Posteromedial tract
 - Sub-arcuate tract
 - Pneumatization is asymmetric in 10% of individuals.
 - **Sclerotic in 10%.**
 - **Posterior portion:**
 - Small
 - Dense otic capsule

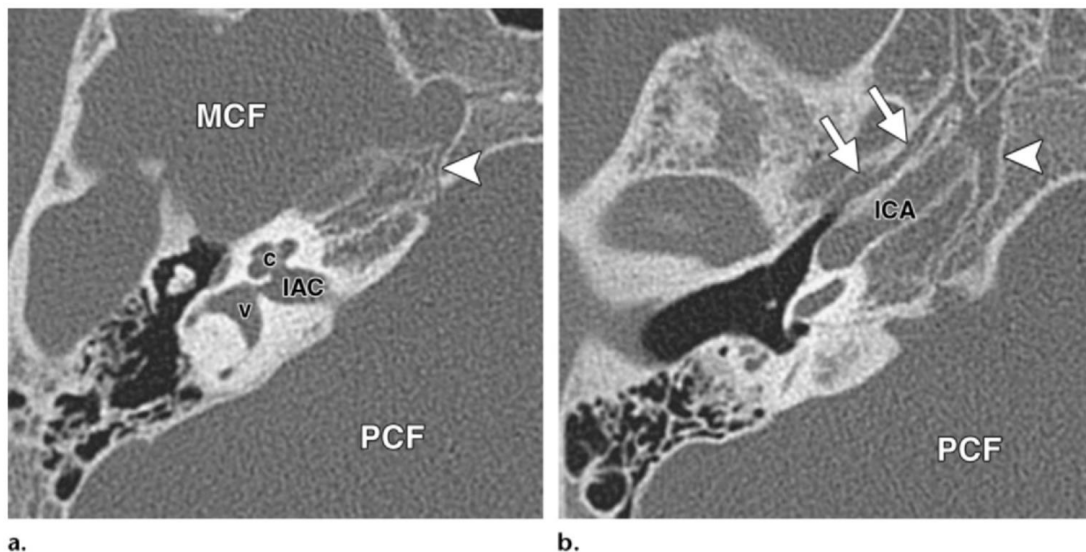


Figure 1. Normal petrous apex at CT. Axial CT images of the right temporal bone, obtained at the level of the IAC (**a**) and slightly more inferiorly at the level of the horizontal petrous carotid canal (**b**), show the normal petrous apex, which is bounded anteriorly by the petrosphenoidal fissure (arrows in **b**) and middle cranial fossa (*MCF* in **a**), posteriorly by the posterior cranial fossa (*PCF*), and medially by the petroclival fissure (arrowhead). The IAC divides the petrous apex into an anterior portion that usually contains marrow and a denser posterior portion that is derived from the otic capsule. *C* in **a** = cochlea, *ICA* in **b** = ICA canal, *V* in **a** = vestibule.

Lateral Skull Base Lesions:

- Classification:
 - **Intra-Dural:**
 - Extra-Axial:
 - **CPA Lesions:**
 - Vestibular Schwannoma **(85%)**
 - Meningioma **(10%)**
 - Epidermoid **(5%)**
 - Arachnoid Cyst **(1%)**
 - Non-Vestibular schwannomas **(1%)**
 - Others:
 - Lipoma
 - Aneurysm AICA
 - Metastasis
 - Intra-Axial:
 - **Brain tissue lesions:**
 - Brainstem Glioma (Most common pediatric CPA lesion).
 - Hemangio-blastoma
 - Medullo-blastoma
 - **Extra-Dural:**
 - Petrous Apex lesions:
 - Cholesterol Granuloma
 - Epidermoid
 - Mucocele
 - Petrous apicitis
 - Jugular foramen lesions:
 - Paragangliomas (Glomus Jugular Tumors)
 - Lower Cranial nerves schwannomas.
 - Clival lesions:
 - Chordoma
 - Chondrosarcoma
 - Plasmacytoma
 - **Trans-Dural:**
 - Clival and temporal bone lesions.

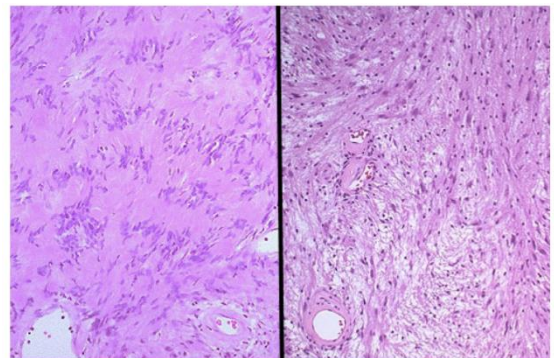
Because lateral skull base lesions cannot be directly visualized, imaging plays a crucial role in the diagnosis and for patient care, as treatment approaches depend on the specific disease process and the nearby structures involved.

CPA Lesions:

- CPA lesions are the most common neoplasms in the posterior fossa.
- Accounts for 10% of all intracranial tumors.
- Intra-dural extra-axial lesions.
- Most CPA lesions are benign:
 - o Vestibular Schwannoma **(85%)**
 - o Meningioma **(10%)**
 - o Epidermoid **(5%)**
 - o Arachnoid Cyst **(1%)**
 - o Non-vestibular schwannomas **(1%)**
- Primary malignancies or metastatic lesions account for <2% of CPA lesions.

Vestibular Schwannoma (VS):

- Most common CPA tumor (85%).
- Acoustic neuroma is a misnomer:
 - o Not arise from the cochlear nerve.
 - o Not a neuroma.
- Epidemiology
 - o Peak presentation is in the 5th-6th decades.
 - o When they occur in patients with neurofibromatosis type 2 (NF2), usually present by the 3rd decade.
 - Unilateral VS appearing < age of 30 years mandate close evaluation of contralateral ear.
 - o No sex predilection.
- Histopathology:
 - o Circumscribed encapsulated benign tumor (unlike neuromas).
 - o Arises from Schwann cells of vestibular portion of CN-VIII.
 - Inferior division is more commonly affected by VS (90%).
 - Equally affected in Cumming.
 - Originated most commonly laterally within IAC in the intra-canalicular segment of vestibular nerve near **Scarpa ganglion**.
 - Less commonly originated at glial-Schwann cell junction (Obersteiner-Redlich zone).
 - o Histologic Types:
 - **Antoni Type A:**
 - More cellular.
 - Uniform spindle cells.
 - Parallel nuclei.
 - Arranged in fascicles.
 - Verocay bodies.
 - **Antoni Type B:**
 - Less cellular.
 - Less uniform.
 - Looser stroma.
 - Cystic degeneration.



These are the classic microscopic appearances of a schwannoma, which is benign. Note the more cellular "Antoni A" pattern on the left with palisading nuclei surrounding pink areas (Verocay bodies). On the right is the "Antoni B" pattern with a looser stroma, fewer cells, and myxoid change.

- Etiology:
 - Sporadic in **95%** of cases.
 - Associated with Neurofibromatosis type 2 (NF2) in **5%** of cases.
- **Neurofibromatosis:**
- Autosomal dominant inheritance.
- Types:
 - **NF-1 (von Recklinghausen Disease)**
 - Mutation in neurofibromin 1 on chromosome 17.
 - Most commonly present with cutaneous manifestation.
 - **5% risk of unilateral vestibular schwannoma.**
 - Diagnostic criteria (at least 2 of the following characteristics):
 1. ≥ 6 Café-au-lait spots:
 - Size > 5 mm in pre-pubertal patients.
 - Size > 1.5 cm in post-pubertal patients.
 2. ≥ 2 Neurofibromas of any type or ≥ 1 Plexiform neurofibroma.
 3. Axillary or groin freckling
 4. Optic nerve glioma
 5. ≥ 2 Lisch nodules (iris hamartomas)
 6. Distinct bony lesions
 7. First-degree relative with NF-1



- **NF-2 (Central Neurofibromatosis):**
 - Caused by mutation of NF2 gene (tumor suppressor gene) located on chromosome 22q12.
 - Responsible to encode protein "Merlin/ schwannomin".
 - Absence of Merlin results in promotion of cellular proliferation and tumorigenesis.
 - Early onset < 21 years old.
 - **95% risk of bilateral vestibular schwannomas.**
 - Subtypes of NF2:
 1. **Wishart:** Severe type.
 2. **Gardner:** Mild type.
 - Diagnostic criteria:
 - **Bilateral VS.**
 - OR
 - **First-degree relative with NF-2.**
 - **+ One of the following:**
 - Unilateral vestibular schwannoma
 - Any two of meningioma, glioma, schwannoma, or juvenile cataract.

- Pathophysiology of VS:

- Slow-growing tumor within the IAC.
 - **Growth rate of 1-2 mm/year.**
- Typically encroaching and displacing neural structures without direct invasion.
- Expands medially into the medial aspect of IAC (Porus Acusticus).
- Signs and symptoms of VS depends on its size (Jackler staging):
 - Small size VS (≤ 1 cm).
 - Medium size VS (1-2.5 cm).
 - Large size VS (2.5-4 cm).
 - Giant size VS (> 4 cm).
- Invade the vestibular nerves and compress the cochlear and facial nerves and the labyrinthine artery:
 - Leads to vestibular and cochlear dysfunction.
 - However, facial nerve is resilient to dysfunction despite the compression and thinning of the nerve.
 - Facial weakness can be present in large tumors.
- Extension into the CPA leads to compression of cerebellum and brainstem:
 - Causes cranial neuropathy, hydrocephalus and death.
- Very rare malignant potential ($< 1\%$).

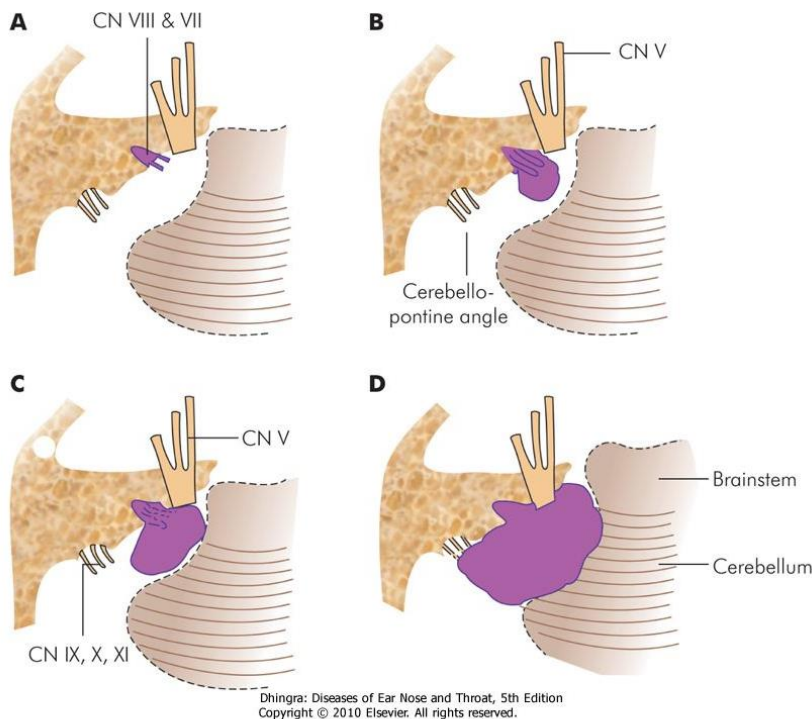


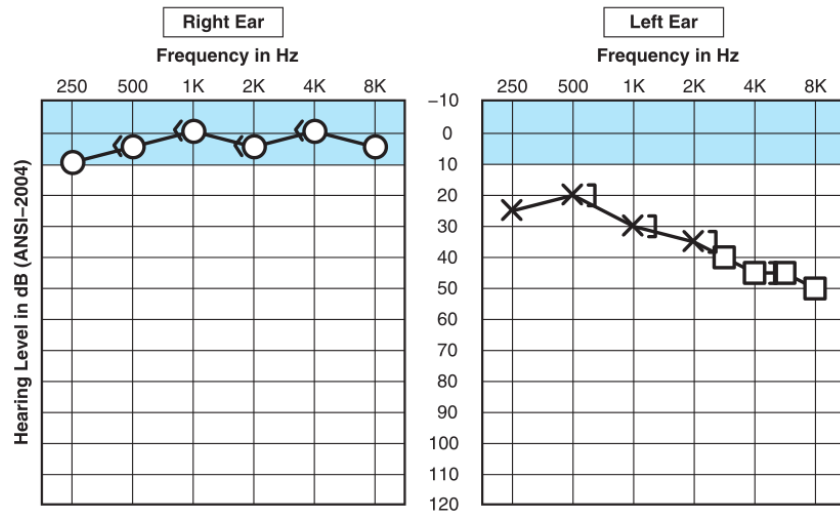
Figure 18.2 Acoustic neuroma and its expansion: (A) Intracanalicular. (B) Tumour extending into cerebellopontine angle. (C) Tumour pressing on CN V. (D) Very large tumour pressing on CN V, IX, X, XI, and brainstem and cerebellum.

- Clinical picture of VS:
 - **Asymptomatic:**
 - Diagnosed incidentally on imaging studies for other indications.
 - Occurs with:
 - Small VS within IAC.
 - VS arising in CPA rather than IAC.
 - **Unilateral symptoms:**
 - **Unilateral (Asymmetrical) SNHL (95%):**
 - Resulted from VS growth in IAC.
 - Progressive high frequency SNHL.
 - Up to 20% of patients with VS present with sudden SNHL.
 - Only 1-5% of patients with asymmetrical SNHL or sudden SNHL are having VS.
 - Even a sudden SNHL with complete recovery can be caused by VS.
 - **Unilateral tinnitus (80%):**
 - Resulted from VS growth in IAC.
 - Only 1-5% of patients with unilateral tinnitus are having VS.
 - **Disequilibrium (10%):**
 - Resulted from VS growth in IAC.
 - Mild balance disturbance.
 - Tumor extension into the labyrinth may result in episodic vertigo, similar to Meniere disease.
 - **Unilateral Facial nerve involvement:**
 - Resulted from VS growth in IAC.
 - Hitselberger's sign (30-50%):
 - Reduced sensation of posterior EAC and concha.
 - Compression of Sensory branches of FN (early).
 - Facial paresis (Suspect other invasive tumors).
 - Motor fibers are more resistant and are affected late.
 - **Unilateral Trigeminal nerve involvement:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Unilateral facial numbness and reduced corneal reflex.
 - **Unilateral Abducens CNs involvement:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Diplopia.
 - **Unilateral Lower CNs involvement:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Unilateral Glossopharyngeal involvement:
 - Dysphagia.
 - Unilateral Vagus involvement:
 - Deviated uvula away from involved side.
 - Unilateral VF paralysis.
 - Unilateral Spinal Accessory involvement:
 - Unilateral shoulder drop.
 - Unilateral Hypoglossal involvement:
 - Deviated tongue toward the involved side.

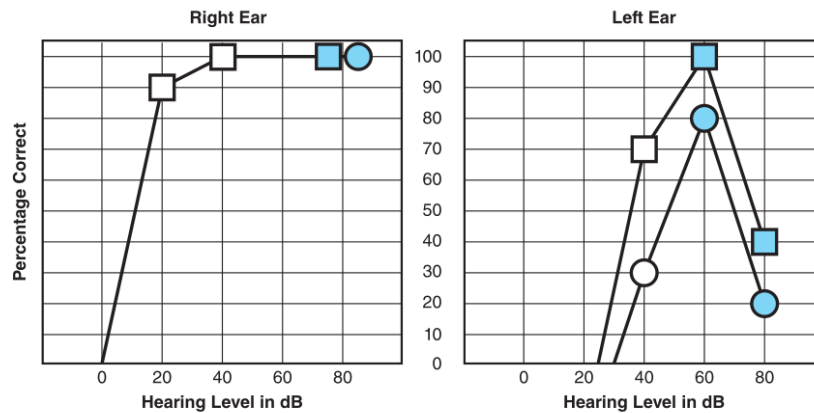
- **Brun Nystagmus:**
 - Resulted from large VS growth in CPA (>3 cm in size) compressing ipsilateral cerebellar flocculus.
 - Combination of central and peripheral Nystagmus.
 - If right CPA tumor:
 - Right-beating central nystagmus with right gaze (toward the lesion).
 - Left-beating vestibular nystagmus with left gaze (away from the lesion).
- **Hydrocephalus:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Compression and obstruction of 4th ventricle.
- **Ataxia:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Compression of cerebellum.
- **Death:**
 - Resulted from large VS growth in CPA (>3 cm in size).
 - Compression of brainstem.
- Diagnosis of VS:
 - **Audiology Tests (Initial screening):**
 - **Pure Tone Audiometry (PTA):**
 - Asymmetric HF-SNHL (65%):
 - Average difference of ≥ 15 dB in AC thresholds between ears at 500, 1000, 2000 and 3000 Hz.
 - Sudden SNHL (20%).
 - Normal PTA (5%).
 - **Speech Audiometry:**
 - Impaired speech discrimination score (SDS) out of proportion to PTA (even with mild hearing loss).
 - Positive for Roll over:
 - Paradoxical decrease in SDS with increasing speech intensity (dB).
 - **Stapedial (Acoustic) Reflex:**
 - Absent Reflex or Positive Reflex Decay (90%):
 - Inability to maintain the stapedial reflex for a sustained signal at 10 dB SL for 10 seconds
 - **Auditory Brainstem Response (ABR):**
 - Most sensitive and specific audiological test.
 - Less sensitive than MRI for small tumors.
 - Not considered initial diagnostic tool for VS except in patients with contraindication for MRI (pacemaker).
 - Sensitivity of ABR depends on the size of VS:
 - 60-80% sensitive for small VS < 1cm.
 - 90% sensitive for medium and large VS.
 - Findings indicate Retro-cochlear pathology:
 - Abnormal Morphology.
 - Delayed inter-aural wave V latency (>0.2 ms).
 - Delayed wave I-III latency (>2.0 ms)

Table 15-1. Results of various tests to differentiate a cochlear from a retrocochlear lesion

	Normal	Cochlear lesion	Retrocochlear lesion
• Pure tone audiogram	Normal	Sensorineural hearing loss	Sensorineural hearing loss
• Speech discrimination score	90-100%	Below 90%	Very poor
• Roll over phenomenon	Absent	Absent	Present
• Recruitment	Absent	Present	Absent
• SISI score	0-15%	Over 70%	0-20%
• Threshold tone decay test	0-15 dB	Less than 25 dB	Above 25 dB
• Stapedial reflex	Present	Present	Absent
• Stapedial reflex decay (page 109)	Normal	Normal	Abnormal
• E.R.A	Normal interval between wave I & V	Normal interval between wave I & V	Wave V delayed or absent

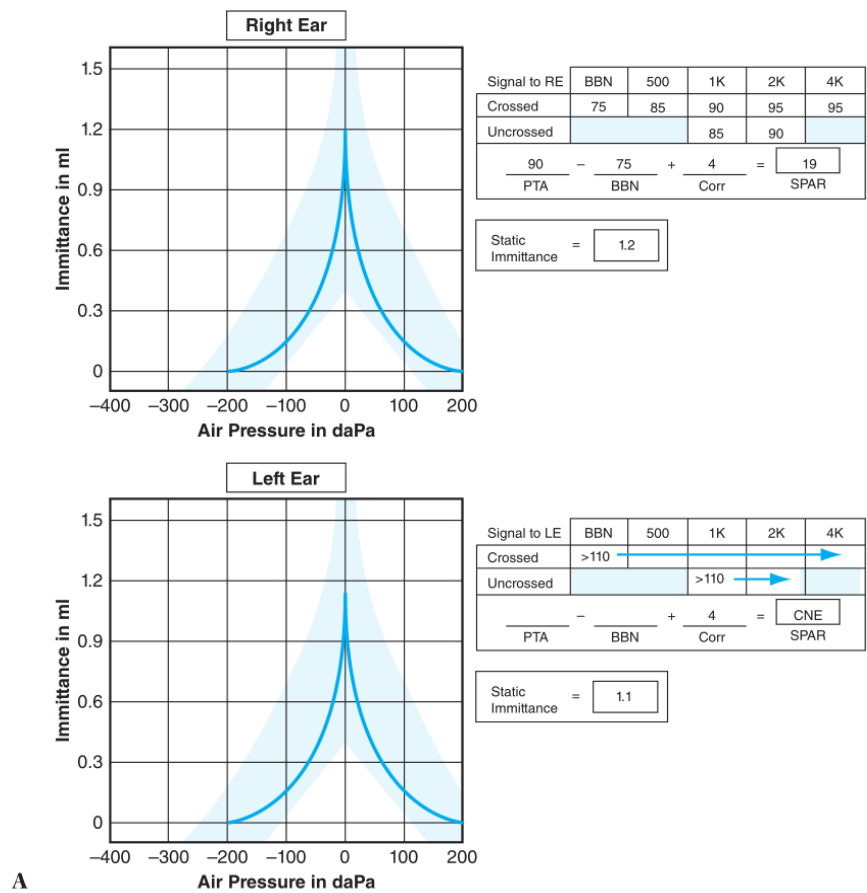


Pure-tone audiometric results show normal hearing sensitivity on the right and a mild, relatively flat sensorineural hearing loss on the left.

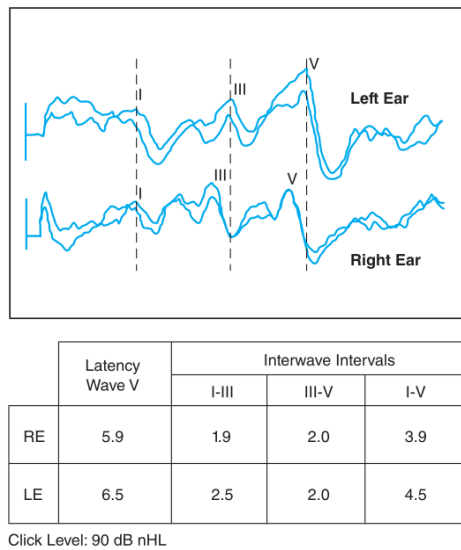


Key to Symbols		Summary	
Unmasked	Masked	Right Ear	Left Ear
WRS	 ○	5 dB	ST 30 dB
SSI	 □	dB	SAT dB
		100 %	WRS _M 80 %
		100 %	SSI _M 100 %
		100 %	DSI 100 %

Speech audiometric results show rollover of the performance-intensity functions on the left ear.



Immittance measures are consistent with normal middle-ear function.
Left crossed and left uncrossed refl exes are absent, consistent with left afferent disorder.



Auditory brainstem response results show delayed latencies and prolonged interpeak intervals on the left ear, consistent with retrocochlear site of disorder.

○ **Vestibular Tests (Not screening tests):**

▪ **Videonystagmography (VNG):**

- Caloric testing is the most useful component of VNG testing in VS patients.
 - Identify a unilateral vestibular weakness in tumors that have affected or originate from superior vestibular nerve.
 - Small VS of inferior vestibular nerve may have a normal caloric response and VNG entirely.
- VNG may add prognostic information by identifying the nerve of origin when a hearing preservation operation is considered.
 - Theoretically better results may be obtained with resection of tumors of superior vestibular nerve.

▪ **Vestibular Evoked Myogenic Potential (VEMP):**

- Assist in determining the nerve of origin.
- Absent or reduced cVEMP can be demonstrated if VS originated from inferior vestibular nerve.
- However, normal cVEMP does not rule out the presence of inferior vestibular nerve tumor.
- Presence of abnormal VEMP response is related more to the size and ensuing neural compression of tumor within IAC than by the nerve of origin.

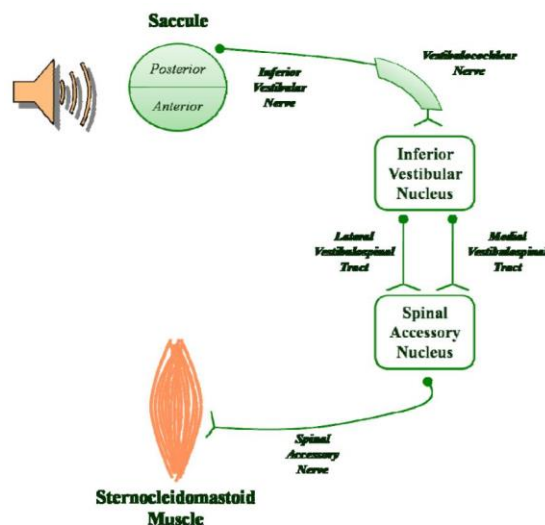


Figure 3. Diagram of the cVEMP neural pathway evoked by an air-conducted stimulus.

	cVEMP	oVEMP
Muscle	SCM	Orbital (inferior oblique)
Response stimulation	- ve	+ ve
End-organ excitation	Saccule	Utricle
Nerve	Inf. Vest. N.	Sup. Vest. N.

- **Imaging studies:**

- **MRI with Gad:**

- Gold standard imaging study in evaluation of patients with suspected VS:
 - Asymmetrical SNHL or speech discrimination score.
 - Unilateral tinnitus.
- Sensitive to small intra-canalicular VS < 1cm.
- General features:
 - Widening of IAC.
 - Ice cream cone/Mushroom appearance:
 - Tumor filling IAC and extends into CPA.
 - Fundal cap:
 - Presence of CSF within IAC, lateral to tumor.
 - Positively affect hearing preservation outcomes in middle cranial fossa approach for tumor resection.
 - Form acute angle with the underlying bone.
- MRI Sequences:
 - **T1:**
 - HYPO-intense to brain.
 - **T1 C+ (Most sensitive):**
 - Marked heterogeneous tumor enhancement.
 - Can detect as small VS as 1-2 mm.
 - **T2:**
 - HYPER-intense to brain.
 - HYPO-intense to CSF.
 - **Heavily T2 Fast-Spin Echo (FSE):**
 - Achieve the accuracy of contrasted T1 without the need for contrast.
- Tumor size measurement:
 - Standard documentation of tumor size is recording the linear measurements in 3 dimensions.
 - Tumor volume measurement is more sensitive in monitoring small changes of tumor growth overtime.

- **CT with Contrast:**

- Provide adjunctive information regarding bony structures surrounding CPA lesions.
- Not ideal as screening test for evaluation of CPA lesions:
 - Can miss lesions < 1cm.
- Indicated in suspected VS patients who can't undergo MRI or if MRI is unavailable.
- Findings:
 - Heterogeneous contrast-enhanced mass in IAC.
 - Expansion of IAC (Average IAC width is 5mm).
 - Acute angles at the bone-tumor interface.

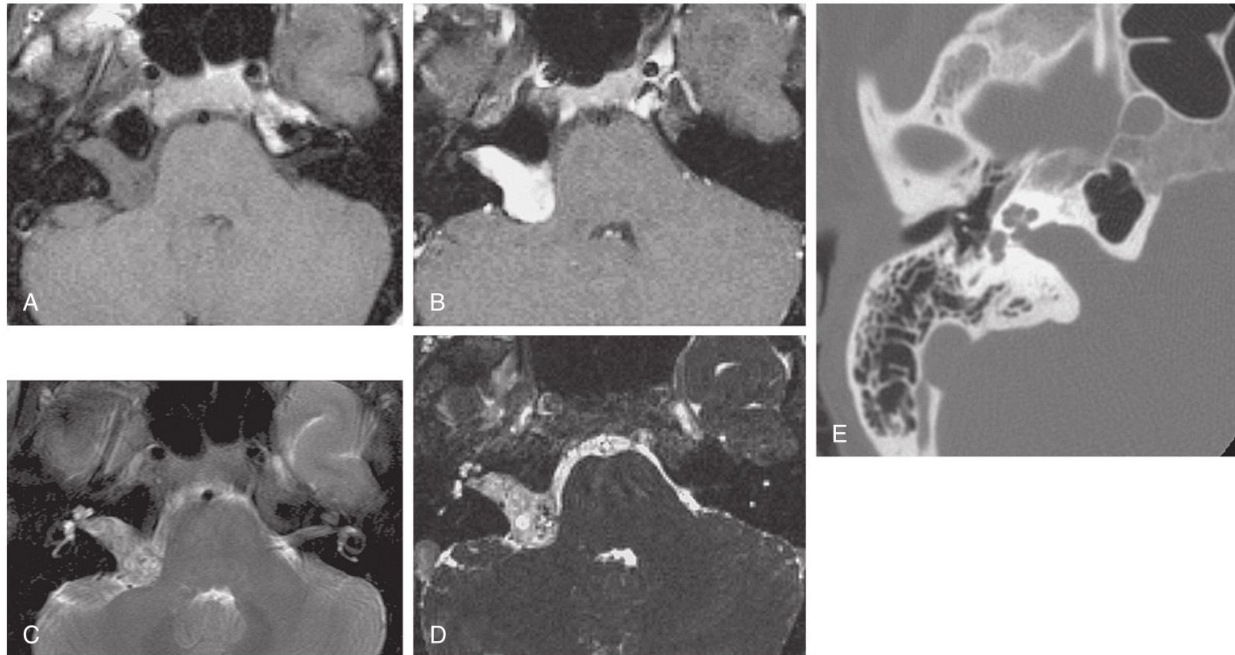


FIGURE 177-2. A medium-sized vestibular schwannoma, demonstrated by magnetic resonance imaging (A to D) and computed tomography (CT) (E). A and B, T1-weighted images before and after intravenous administration of gadolinium contrast. The tumor in the right internal auditory canal (IAC), which is mushrooming into the cerebellopontine angle and enlarging the canal, is isointense to mildly hypointense to the cerebellum and enhances markedly with contrast. C and D, T2-weighted and constructive interference in steady-state (CISS) images. The tumor is moderately hyperintense on the T2-weighted images. CISS images precisely demonstrate the cochlea and cystic and calcified components of the tumor and surrounding vascular structures. E, CT bone windows. Note the enlargement of the IAC. (Modified from Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, p 245.)

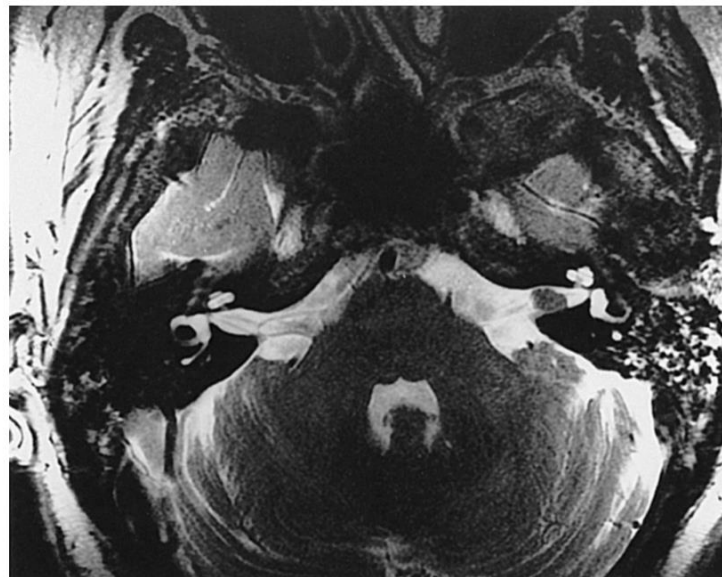


FIGURE 177-1. On T2-weighted fast spin echo magnetic resonance imaging, this axial reconstruction shows a 5-mm intracanalicular tumor (at left).

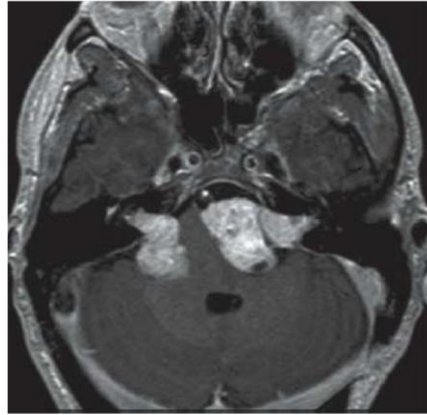


Figure 159.3 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating bilateral enhancing vestibular schwannomas with brainstem compression.

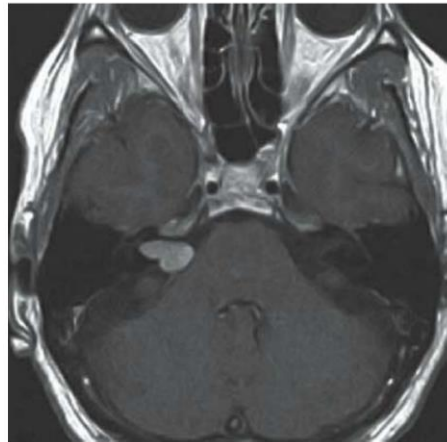


Figure 159.2 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating an enhancing right vestibular schwannoma with an "ice cream cone" or "mushroom" appearance.



Figure 159.4 T2 fast spin-echo magnetic resonance image of a left intracanalicular vestibular schwannoma. Note the presence of CSF in the fundus lateral to the tumor.

- Treatment of VS:

○ **Observation with serial imaging**

▪ Indications:

1. **Asymptomatic patients.**
2. **Small tumors without brainstem compression.**
3. **Only hearing ear.**
4. **Elderly (>65 years).**
5. **Non-operative patients.**

▪ Goal:

- At the time of diagnosis, rate of tumor growth is unknown and unpredictable.
- Observation allows enough time to determine the tumor growth rate, and if no growth is demonstrated, treatment is not necessary.
- Hearing aids are used when appropriate.

▪ Follow-up plan:

- Initially with MRI after 6 months then annually if no growth was detected with volumetric measurements.

• Indication of intervention:

- Rapid tumor growth (>2.5 mm/year) regardless of tumor size.

▪ Advantages:

- Afford years of residual hearing.
- Avoid the complications of the intervention.

▪ Disadvantages:

- Deterioration of hearing loss during the observation period and the opportunity for hearing-preservation approach may be lost.

○ **Stereotactic Radiation (Radiosurgery):**

- Administration of radiation (13-14 Gy) to a precise location.
- Induce radiation damage to the target area while minimizing the effect to the adjacent structures.

▪ Modalities:

- Gamma Knife
- Cyber Knife

▪ Goals:

- Provide long-term tumor growth control **(90-100%)**.
 - No radiographic enlargement over 2 mm in tumor diameter or 10% change in tumor volume.
- Avoid the risk of surgical intervention.
- Prevent acute loss of neurologic function.

▪ Indications:

1. **Tumors < 2-3 cm.**
2. **Recurrence after surgery.**
3. **Only hearing ear.**
4. **Elderly (>65 years).**
5. **Non-operative patients.**

- Advantages:
 - Avoid morbidity of surgical intervention.
 - Rapid return to normal activity.
- Disadvantages:
 - **Stop the growth without removing the tumor.**
 - No pathologic diagnosis.
 - Post-radiation scarring makes salvage surgery difficult.
 - Risk of post-radiation tumor expansion:
 - Occurs in 25% of patients.
 - Transient tumor swelling (2-4 mm).
 - May take 6 months to 5 years to resolve.
 - Increase the risk of complications.
 - Complications:
 - **Hearing loss:**
 - Occurs gradually months to years later.
 - Due to radiation of the cochlea.
 - Maintaining a cochlear dose < 6.9 Gy is important in preserving residual hearing.
 - Hearing preservation rate 50-86%.
 - **Chronic vestibular dysfunction:**
 - Present in 15-25% of patients.
 - **Facial and trigeminal neuropathies:**
 - 0-5% of patients.
 - Risk increase with doses > 13 Gy.
 - **Hydrocephalus:**
 - Without evidence of tumor growth.
 - Occurs in up to 5% of patients.
 - More common with large tumors due proteinaceous debris obstruction of CSF flow.
 - **Radiation-induced Malignancy:**
 - In young patients.
 - 1:1,000 risk over 5-30 years period.
- Follow up plan:
 - Post-irradiation MRI scans are obtained initially after 6 months and then annually to observe the effect of the stereotactic radiation.
 - After radiation, tumors show signs of treatment effect by loss of central enhancement within the first 6-12 months which correlate with tumor necrosis.



FIGURE 179-1. The Gamma Knife helmet. (Courtesy of Elekta.)

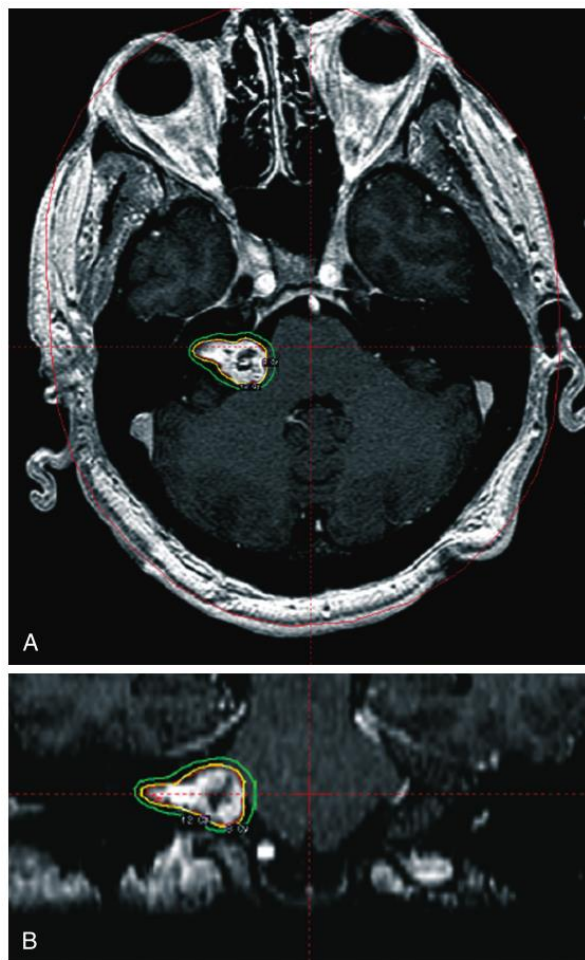
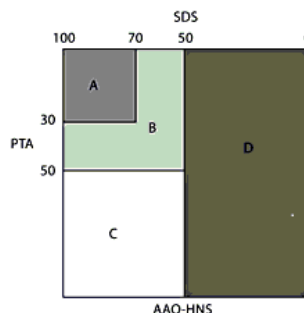
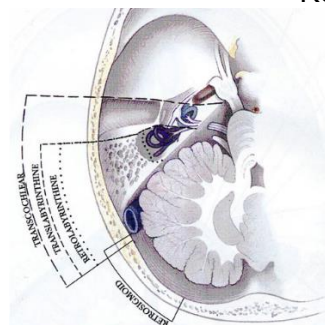


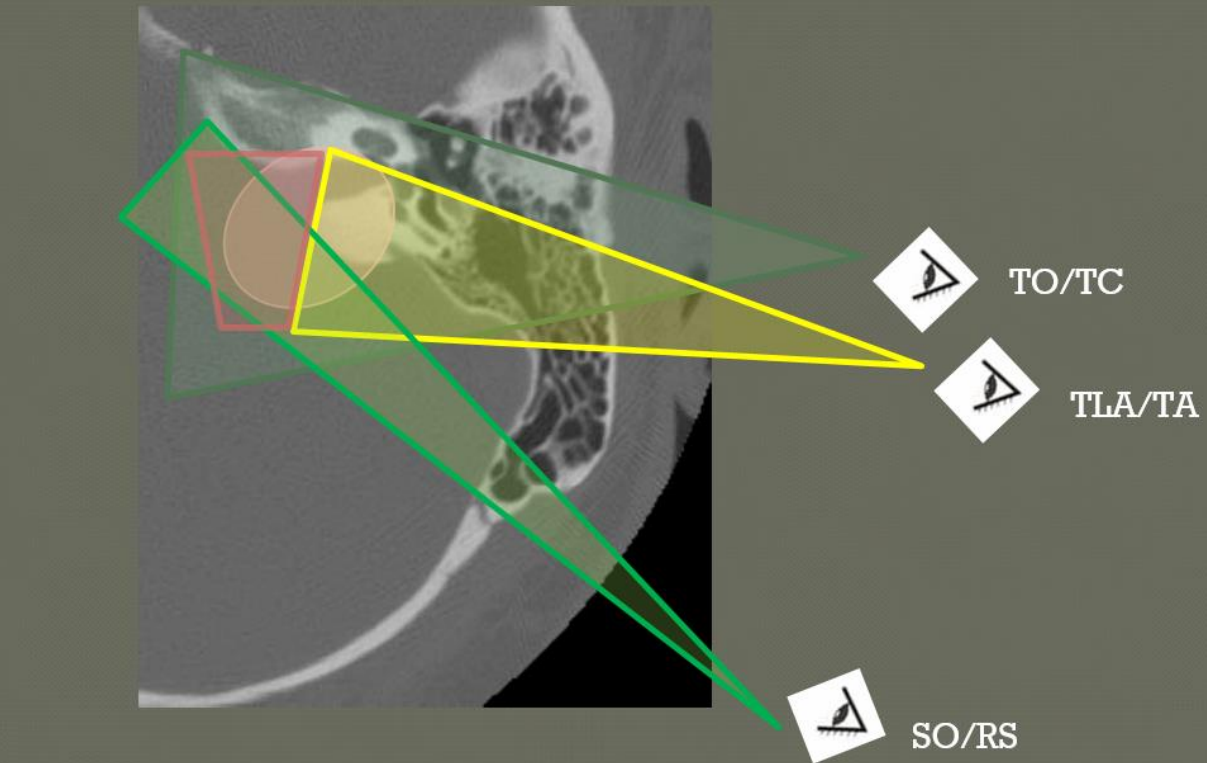
FIGURE 179-2. Axial (A) and coronal (B) images of a radiation dose plan for a vestibular schwannoma. Multiple shots can be arranged within the tumor at various radiation doses to give homogeneous exposure to the tumor while avoiding unnecessarily high levels of radiation to adjacent structures.

○ **Microsurgery:**

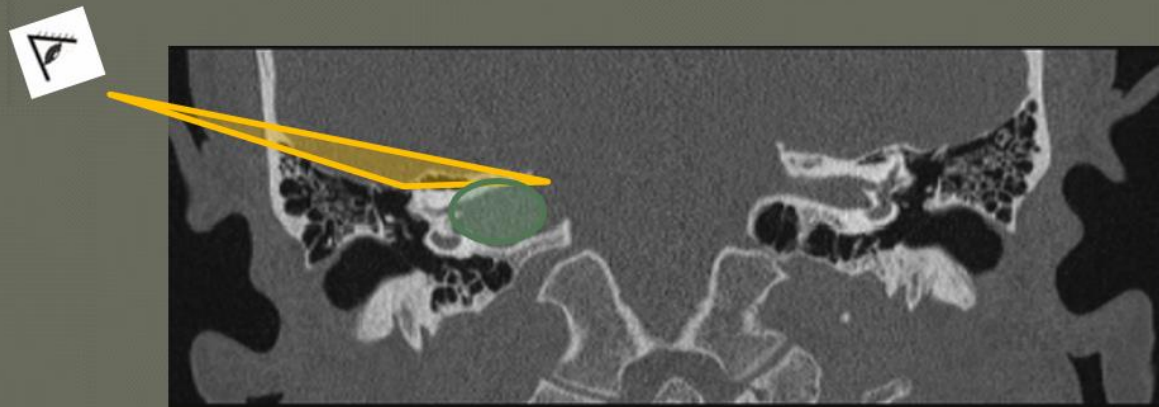
- Indications:
 - 1. Tumors > 2-3 cm.**
 - 2. Recurrence after stereotactic radiation.**
- Goals:
 - Complete tumor resection, tumor de-bulking or IAC decompression.
 - Preserving facial nerve and auditory function if present preoperatively.
- Surgical approaches for CPA lesions depends on:
 - Hearing status.
 - Tumor size.
 - Tumor location.
 - Facial nerve status.
- Types of Surgical approaches for CPA lesions:
 - **Hearing preservation approaches:**
 - Candidate:
 - 1. Patients with:**
 - **Serviceable hearing:**
 - Grade A ASHA:
 - $PTA \leq 30$ dB and $SDS \geq 70\%$
 - Grade B ASHA:
 - $PTA \leq 50$ dB and $SDS \geq 50\%$
 - **Tumor size < 2 cm.**
 - 2. NF2**
 - 3. Only hearing ear.**
 - Examples:
 - 1. Middle Cranial Fossa**
 - 2. Retrosigmoid/Suboccipital**
- **Non-Hearing preservation approaches:**
 - Candidate:
 - 1. Non-serviceable hearing (ASHA grade C or worse).**
 - 2. Tumors > 2 cm.**
 - Examples:
 - 1. Trans-labyrinthine.**
 - 2. Trans-otic/Trans-cochlear.**
 - 3. Combined approach (Trans-labyrinthine and Retro-sigmoid/Sub-occipital).**



Approaches to CPA



MCF



○ **Trans-Labyrinthine Approach:**

- Non-hearing preservation approach.
- Most common approach for resection VS.
- Indications:
 1. **Non-serviceable hearing (Grade C or worse) with any tumor size.**
 2. **Large Tumor size > 2 cm:**
 - Hearing in large tumors is unlikely to be preserved by any approach.
 - Trans-labyrinthine approach carries the highest rate of preservation of facial nerve function.
- Contraindications (By Dr.Shami):
 1. Only hearing ear.
 2. Ipsilateral cholesteatoma.
- Steps:
 - Trans-mastoid labyrinthectomy and skeletonization of sigmoid sinus and posterior fossa dura.
 - Permit identification of facial nerve with wide exposure of IAC and CPA.
 - De-bulking and resection of the tumor.
 - Eustachian tube closure.
 - Subtotal petrosectomy, cavity obliteration, and blind sac closure of the external auditory canal.
- Advantages:
 - Most direct route.
 - Excellent exposure.
 - Not limited by tumor size.
 - Minimal cerebellar retraction.
 - **Highest rate of facial nerve preservation (98%).**
 - **Superior exposure of entire course of facial nerve.**
 - **Preservation rate is lower with tumor size >1.5 cm.**
 - **Immediate repair of facial nerve is possible.**
- Disadvantages:
 - Sacrifices the hearing.
 - Extent of mastoid pneumatization may constrict the surgical field.
 - Risk of CSF leak from the wound.

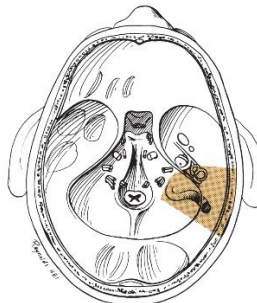


FIGURE 177-27. Surgical exposure in the translabyrinthine approach to the cerebellopontine angle (CPA). This approach permits direct access to the CPA without retraction of the cerebellum.

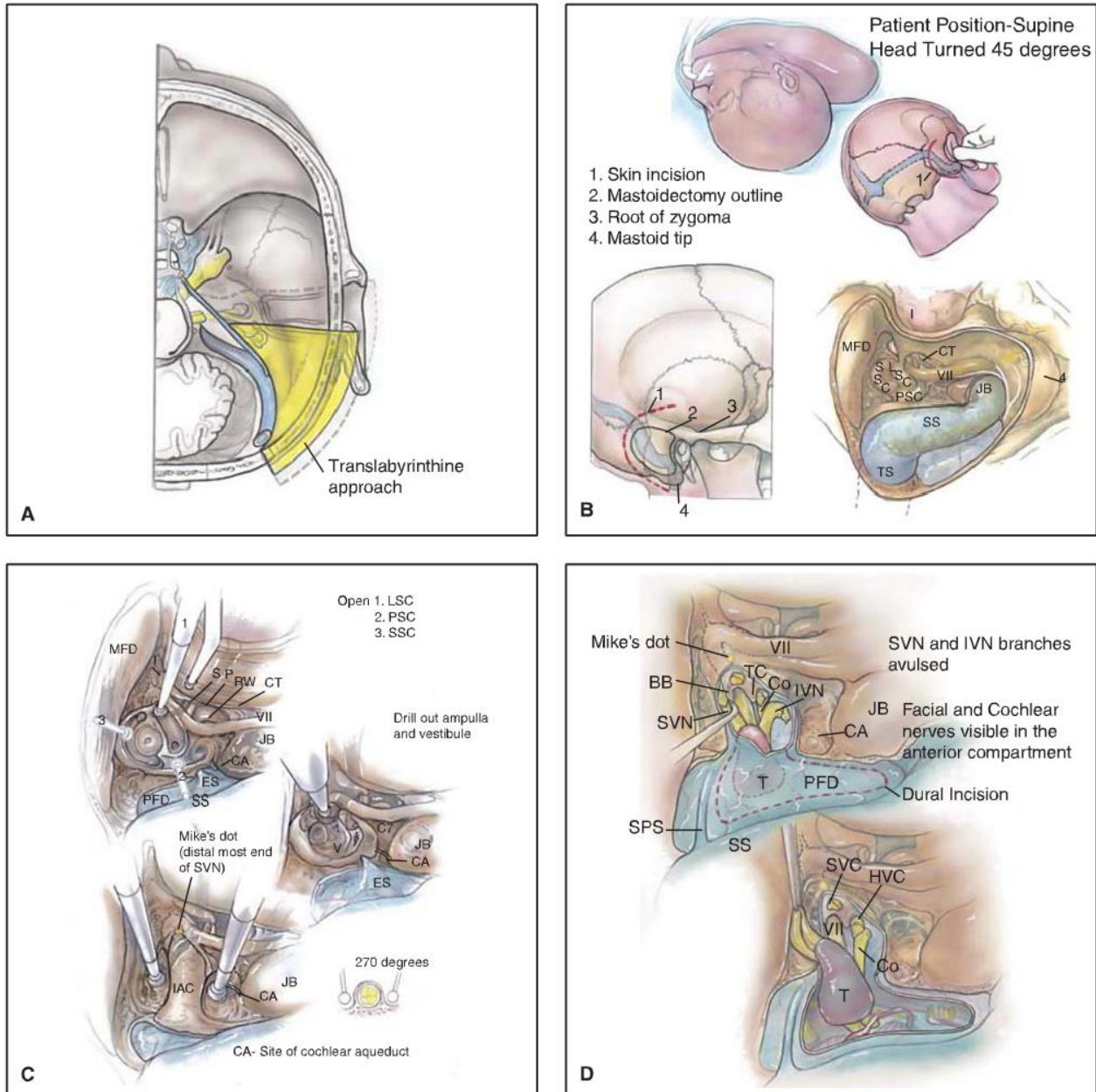


Figure 159.12 Translabyrinthine tumor removal. **A:** The tumor removal is accomplished between the posterior external auditory canal and internal auditory canal anteriorly and the sigmoid sinus posteriorly avoiding prolonged cerebellar retraction. **B:** Patient positioning and skin incision is shown with the complete mastoidectomy then carried out. **C:** Labyrinthectomy with identification of the internal auditory canal and skeletonization. **D:** Dural incision of the internal auditory canal and posterior fossa. CT, chorda tympani; MFD, middle fossa dura; SSC, superior semicircular canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal; VII, facial nerve; TS, transverse sinus; SS, sigmoid sinus; JB, jugular bulb; PFD, posterior fossa dura; S, stapes; P, Promontory; RW, round window; ES, endolymphatic sac; CA, cochlear aqueduct; BB, Bill's bar; TC, transverse crest; Co, cochlear nerve; IVN, inferior vestibular nerve; T, tumor; SPS, superior petrosal sinus; SVN, superior vestibular nerve; SVC, cut superior vestibular nerve; IVC, cut inferior vestibular nerve. (Continued on next page)

○ **Trans-Cochlear Approach:**

- Non-hearing preservation approach.
- Extension of trans-labyrinthine approach obtained by displacement of facial nerve posteriorly and removal of the cochlea to provide surgical access anteriorly.
- Indications:
 - 1. Extensive lesions of petrous apex and clivus.**
- Advantages:
 - Wide exposure of skull base with access to petrous apex and clivus.
- Disadvantages:
 - Sacrifices the hearing.
 - Risk of temporary facial nerve paralysis.

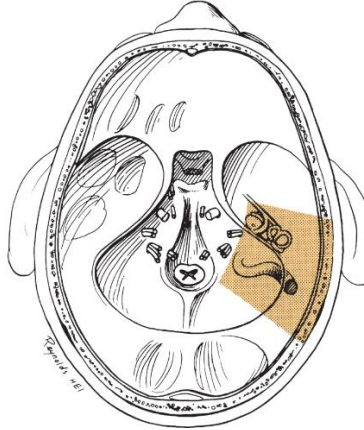


FIGURE 177-35. The transcochlear approach to the cerebellopontine angle petrous tip and clivus. Removal of the cochlea and anterior portion of the petrous bone permits wide visualization of the clivus and skull base.

○ **Trans-Otic Approach:**

- Non-hearing preservation approach.
- Similar to Trans-Cochlear approach but not involving transposition of facial nerve.
- Advantages:
 - Less risk of facial nerve weakness compared to trans-cochlear approach.
- Disadvantages:
 - Sacrifices the hearing.
 - Limited exposure compared to trans-cochlear approach.

○ **Combined Approach:**

- Non-hearing preservation approach.
- Indications:
 - 1. Large CPA tumor size > 4 cm**
- Steps:
 - Trans-labyrinthine with Retro-sigmoid/Sub-occipital facilitate exposure for resection large CPA tumors.
 - Extension of tumor anteriorly to petroclival region may requires addition of Trans-cochlear approach.

○ **Middle Cranial Fossa Approach:**

- Hearing preservation approach.
- Indications:
 1. **Serviceable hearing and lateral intra-canalicular tumor with minimal CPA involvement (<2 cm).**
- Steps:
 - Temporal craniotomy permits exposure of IAC after identification of superior SCC and geniculate ganglion.
- Advantages:
 - Highest rate of hearing preservation (80%).
 - Lowest rate of CSF leak.
 - No intra-dural drilling:
 - Low rate of post-operative headache compared to Retro-sigmoid/Sub-occipital Approach.
- Disadvantages:
 - Limited to small lateral intra-canalicular lesions.
 - More difficult technically.
 - Poor exposure to posterior fossa.
 - Requires temporal lobe retraction:
 - Risk of aphasia or seizure.

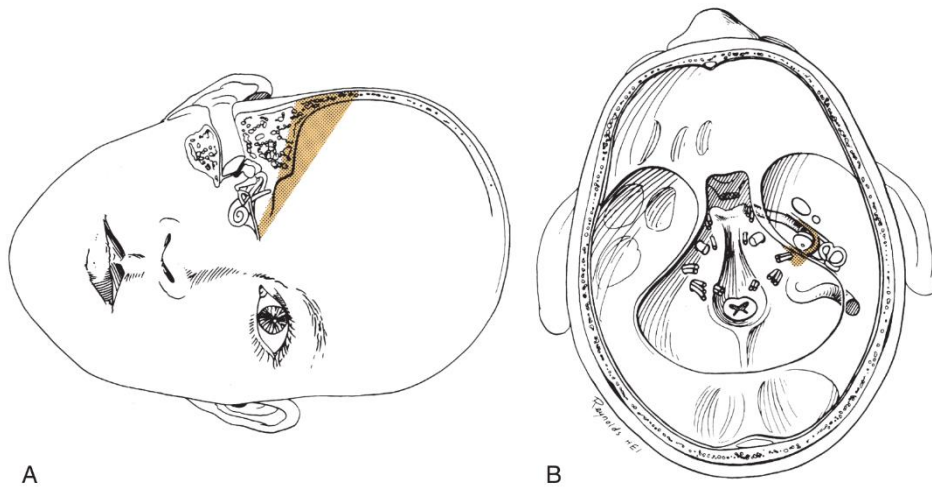


FIGURE 177-31. A, Coronal view: surgical exposure in the middle fossa approach to the cerebellopontine angle (CPA). A temporal craniotomy permits retraction of the temporal lobe and access to the floor of the middle fossa and internal auditory canal (IAC). **B,** Axial view: surgical exposure in the middle fossa approach to the CPA. The shaded area indicates the area of bone removal on the floor of the middle fossa. Note that although wider exposure is possible at the porus, limited space is available between the cochlea and the superior semicircular canal as the surgeon approaches the fundus of the IAC.

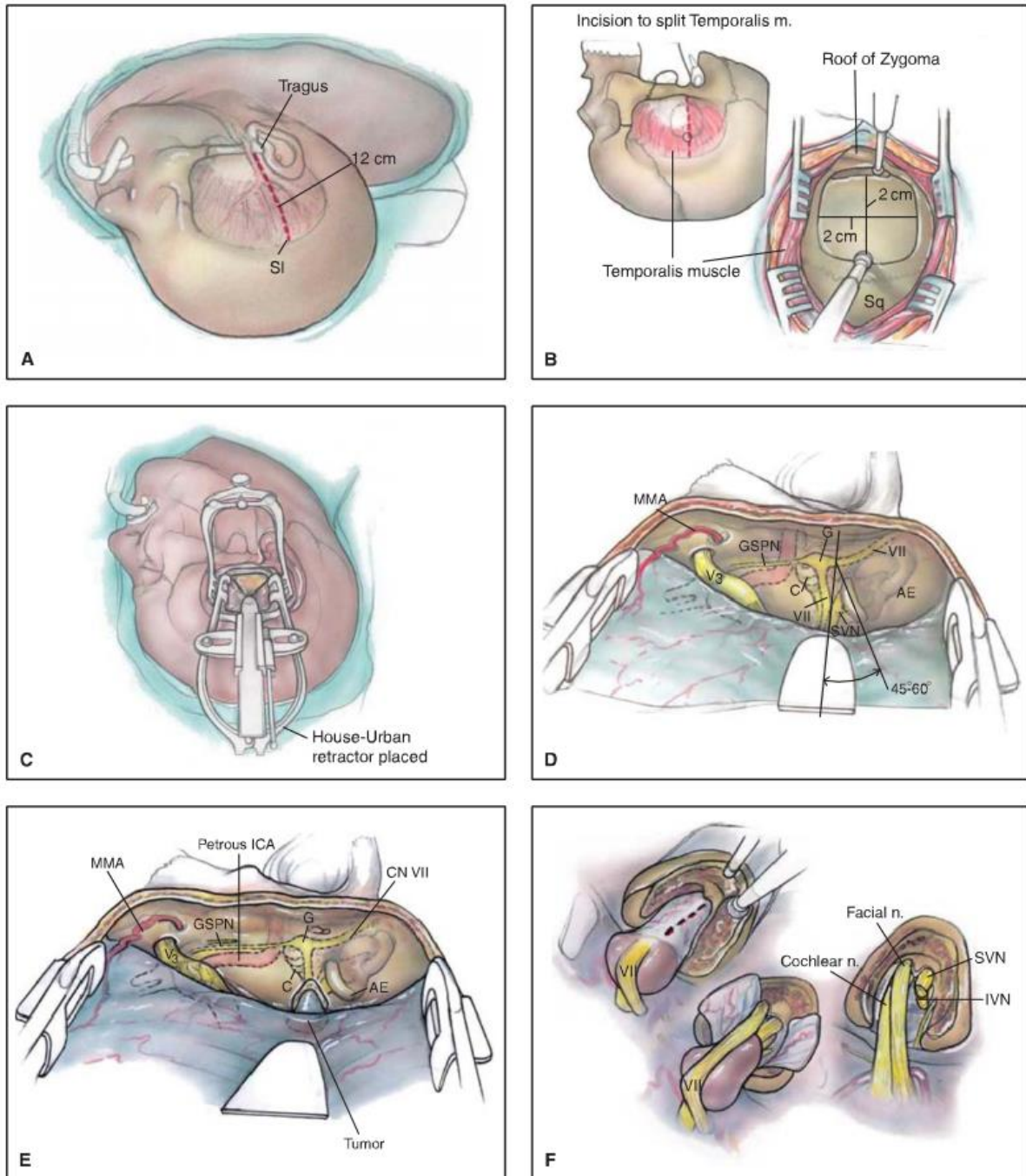


Figure 159.10 Middle fossa approach. **A** and **B**: A 12 cm incision is followed by a 4 x 4 cm craniotomy. **C**: The retractor is positioned so that the structures of the temporal bone can be identified. With the greater superficial petrosal nerve and the arcuate eminence as guides, bone removal is begun and the IAC is identified. **D**: The dura then can be incised away from the facial nerve under high magnification. **E**: The internal auditory canal is identified. **F**: The dura is opened sharply avoiding the facial nerve. Fine hooks are used to separate the superior vestibular nerve from the facial nerve. The IAC is filled with fat. TL, temporal lobe; GSPN, greater superficial petrosal nerve; ICA, internal carotid artery; BB, the Bill bar; VII, facial nerve; SV, superior vestibular nerve; T, tumor; M, malleus; I, incus; SSC, superior semicircular canal; AE, arcuate eminence; G, geniculate ganglion; C, cochlea; MMA, middle meningeal artery. (Continued on next page)

○ **Retro-sigmoid/Sub-occipital Approach:**

- Hearing preservation approach.
- Indications:
 - 1. **Serviceable hearing and CPA or medial intra-canalicular tumors of any size.**
- Steps:
 - Trans-mastoid decompression of sigmoid sinus and retro-sigmoid craniotomy permit access to CPA tumors without disturbing the labyrinth.
 - Permits direct access to the medial two thirds of IAC while preserving the inner ear.
- Advantages:
 - Hearing preservation (30-60%).
 - Provide the most panoramic view of posterior fossa.
 - Deals with tumors of any size in CPA or medial IAC.
- Disadvantages:
 - Limited exposure to lateral IAC.
 - **Highest rate of CSF leak.**
 - **Highest rate of facial nerve injury.**
 - **Highest rate of post-operative headache (10-30%).**
 - Intra-dural spread of bone dust from intra-dural drilling.
 - **Highest rate of air embolism:**
 - Through diploic veins in skull into jugular system and sigmoid sinus.
 - **Requires cerebellar retraction:**
 - **Risk of postoperative imbalance.**

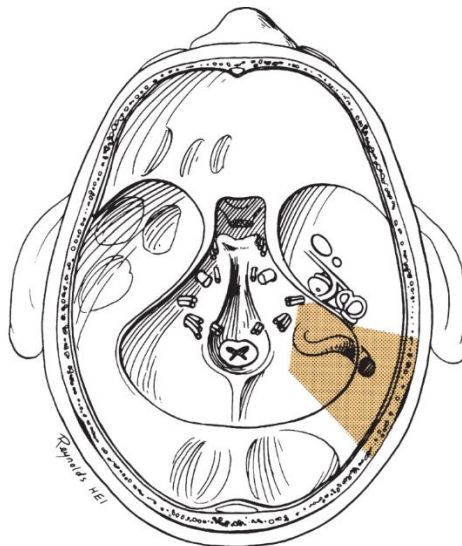


FIGURE 177-29. Surgical exposure in the retrosigmoid approach to the cerebellopontine angle (CPA). This approach permits wide access to the CPA; however, the cerebellum is between the surgeon and the CPA, thus cerebellar support (retraction) is required to permit complete visualization of the CPA.

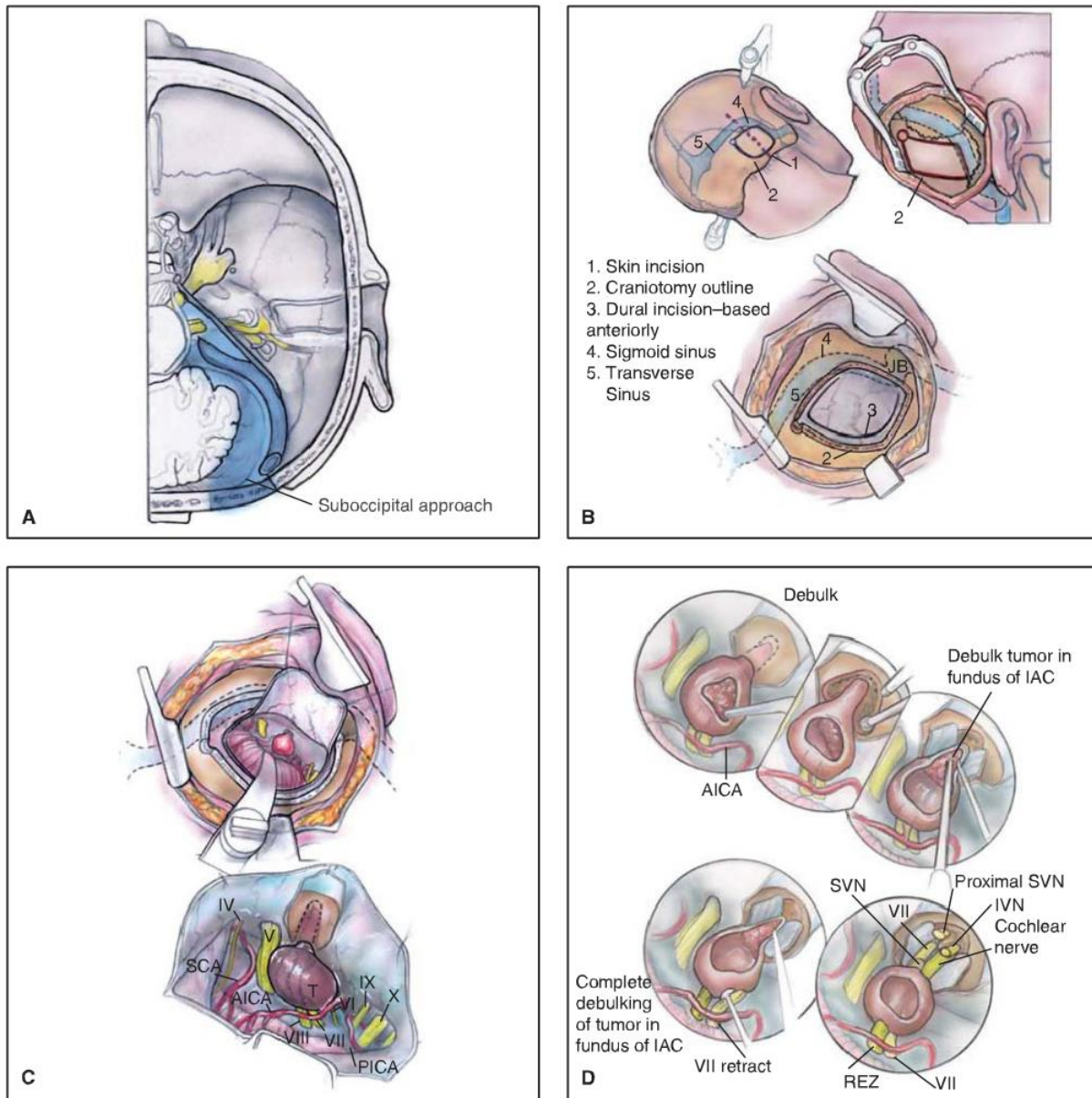


Figure 159.11 Retrosigmoid-suboccipital approach. **A:** The approach between the posterior temporal bone and cerebellum is outlined in blue. **B:** A 3 × 3-cm craniotomy is made with the cutting bur, the sigmoid sinus serving as the anterior limit and the transverse sinus as the superior limit. **C:** The cerebellum is retracted exposing the tumor and the internal auditory canal is drilled away after reflecting dura off the posterior temporal bone. **D:** The tumor is sequentially debulked and removed. SS, sigmoid sinus; TC, transverse crest; C, cochlear nerve; LA, labyrinthine artery; AICA, anterior inferior cerebellar artery; VII, facial nerve; IX, glossopharyngeal nerve; X, vagal nerve; XI, spinal accessory nerve; JB, jugular bulb; SCA, superior cerebellar artery; PICA, posterior inferior cerebellar artery; SVN, superior vestibular nerve; REZ, root entry zone. (Continued on next page)

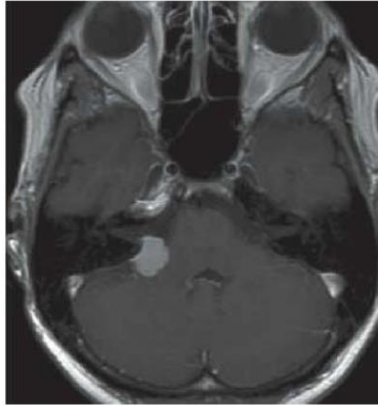


Figure 159.7 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating an enhancing lesion at the right CPA suitable for a suboccipital microsurgical resection.

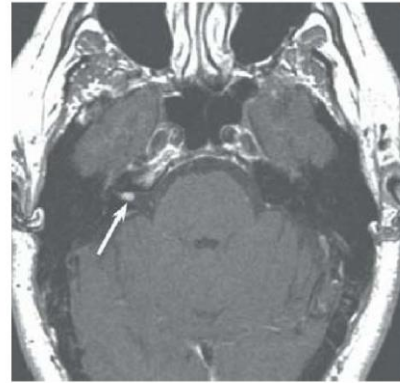


Figure 159.6 Axial T1-weighted contrast enhanced MRI image at the level of the IAC demonstrating a small enhancing intracanalicular right vestibular schwannoma (arrow) suitable for a middle cranial fossa microsurgical resection.

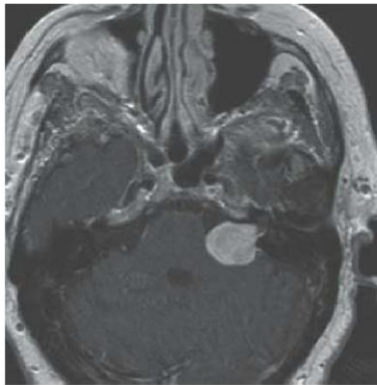


Figure 159.8 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating an enhancing left vestibular schwannoma suitable for either translabyrinthine or suboccipital microsurgical resection.

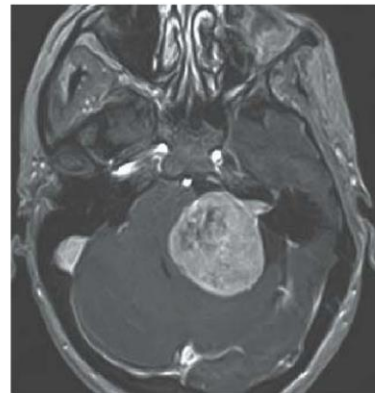


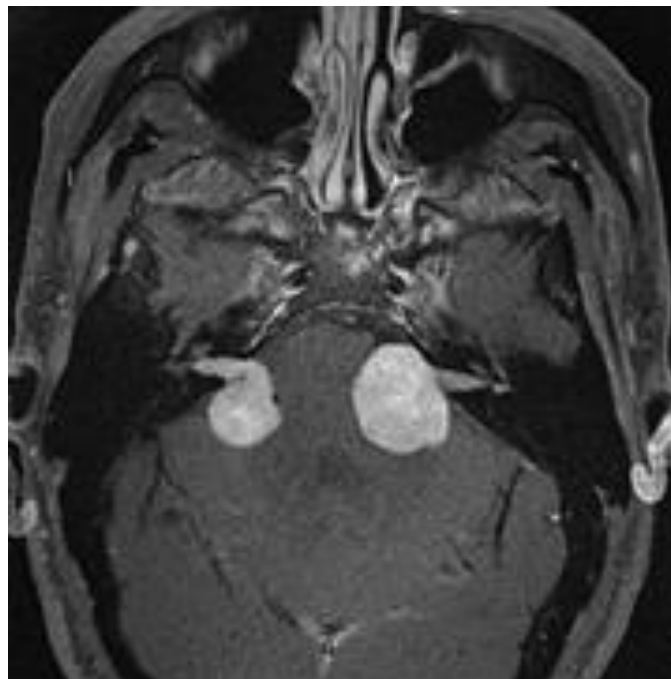
Figure 159.9 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating a large enhancing left vestibular schwannoma suitable for a combined microsurgical resection.

- **Hearing Rehabilitation:**

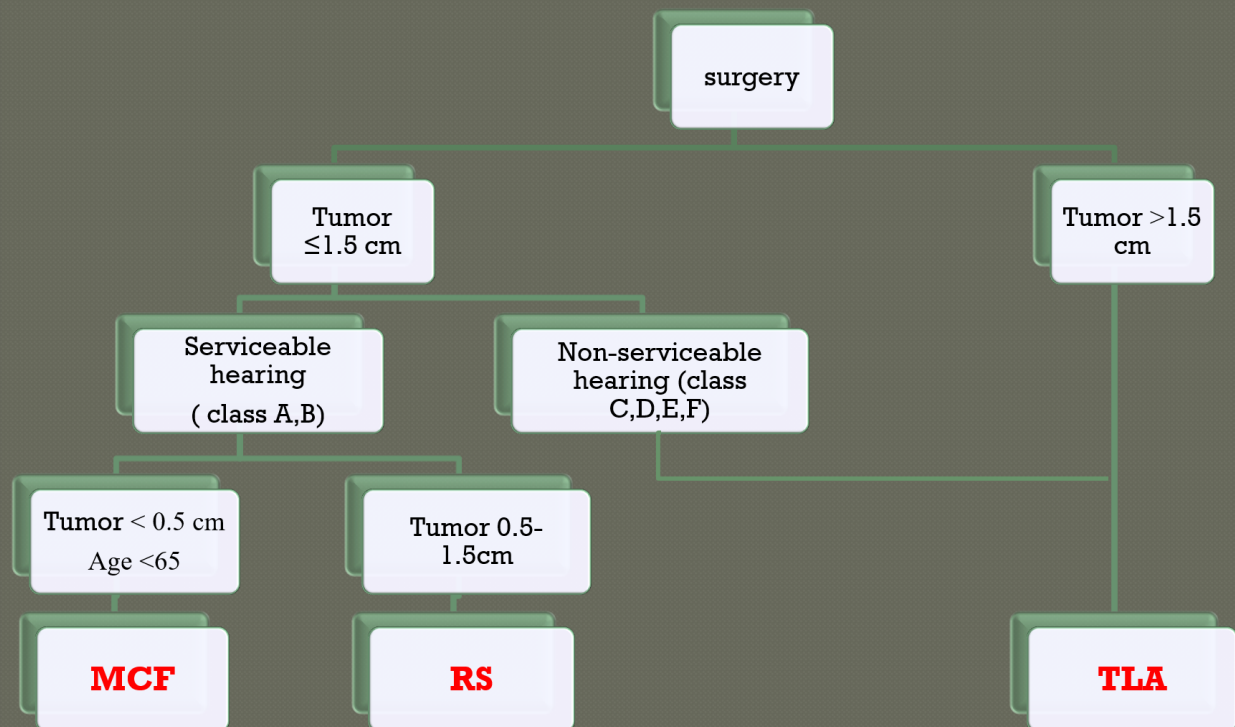
- Options of hearing rehabilitation after non-hearing preservation surgery of unilateral VS:
 - Contralateral routing of signals (CROS) hearing aid.
 - Bone-anchored hearing aid (BAHA).

- **Treatment plan for NF2:**

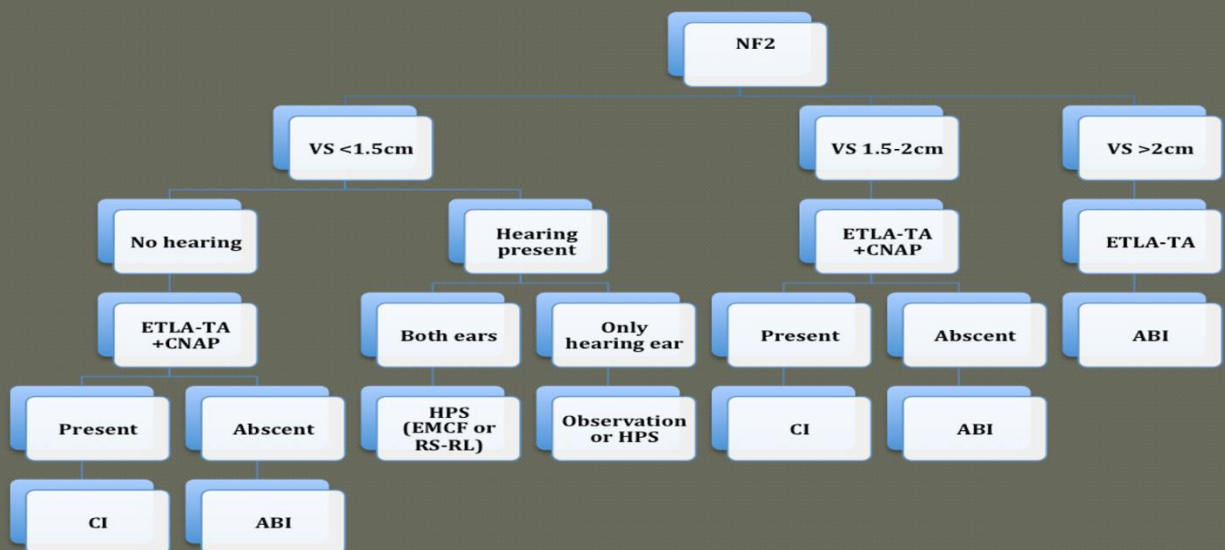
- Preservation of hearing is vital.
- Early detection and intervention provide long-term functional hearing.
- Consider screening the family with audiometry and MRI.
- Approach:
 - **Surgical resection of the larger VS first:**
 - Consider hearing preservation approach:
 - Retro-sigmoid/sub-occipital or Middle cranial fossa approaches.
 - If cochlear nerve was preserved intra-op:
 - Consider cochlear implantation.
 - If cochlear nerve was sacrificed intra-op:
 - Consider Auditory brainstem implant (ABI).
 - **If hearing was preserved in the previously operated side:**
 - Consider surgical removal of the second tumor.
 - **If hearing couldn't be preserved in the previously operated side:**
 - Consider observation for the second tumor with serial MRI strategy.
 - Consider stereotactic radiation for the second tumor.



VS surgery



NF2 & hearing rehabilitation



- Post-operative complication:
 - **Hearing loss:**
 - Hearing preservation is low with any approach if tumor size is more than 2 cm.
 - Highest rate of hearing preservation is with middle cranial fossa approach (80%).
 - **Facial weakness:**
 - Post-op facial nerve function is dependent on:
 - Size of tumor
 - Adherence of tumor to facial nerve
 - Experience of surgical team.
 - Highest rate of facial nerve preservation is with trans-labyrinthine approach.
 - Highest rate of facial nerve injury is with retro-sigmoid/sub-occipital approach.
 - Facial nerve transection ideally is managed by immediate repair or with an interposition graft.
 - **CSF leak:**
 - Reported from 1-10% of all cases.
 - Highest rate of CSF leak is with retro-sigmoid/sub-occipital approach.
 - **Headache:**
 - Highest rate of postoperative headache is with retro-sigmoid/sub-occipital approach.
 - Secondary to intra-dural spread of dust as a result of intra-dural drilling.
 - **Meningitis:**
 - Serious concern in postoperative period.
 - Mean time of onset is 8 days postoperatively.
 - Rate of 1-10% of all cases.
 - **Recurrence of tumor:**
 - Rate of <1% of all cases.
 - **Mortality:**
 - Rate of <1% of all cases.

**TABLE
159.1**



**EMERGENCIES
AFTER TUMOR REMOVAL**

Emergency	Management
Intracranial hemorrhage	Remove dressing and open wound, to allow rapid decompression of brainstem; move to operating room for control of hemorrhage.
Pneumocephalus	If unstable, emergency craniotomy to remove air; otherwise, discontinue lumbar drain and observe.
Meningitis	Lumbar puncture for culture and sensitivity; appropriate antibiotics intravenously; surgical closure of any CSF leak.
CSF leak	Pressure dressing; if leak persists despite lumbar drain, surgical closure.

Meningioma:

- 2nd most common CPA tumor (10%).
- Most common intracranial extraaxial tumor
 - o Constitutes up to 20% of all primary intracranial tumors.
- Epidemiology
 - o Most common in middle-age women.
 - o More common in female (2:1).
 - o Peak presentation is in the 5th-6th decades.
- Histopathology:
 - o Arises from the arachnoid villi cap (endothelial) cells.
 - o Grossly, appears as globular mass firmly adherent to the dura mater.
 - o Has characteristic speckles scattered throughout the tumor, corresponding to the microscopic psammoma bodies.
 - o WHO histopathologic classifications:
 - 1. Grade I (Benign):**
 - Most common form (90%).
 - Display whorls of spindle cells with presence of calcified psammoma bodies.
 - Variants: Meningothelial, fibrous, transitional psammomatous and angiolastic.
 - 2. Grade II (Atypical):**
 - Forms 7% of all meningiomas.
 - Display increased cellularity, atypical nuclei and areas of necrosis.
 - Variants: Choroid, clear cell.
 - 3. Grade III (Anaplastic/Malignant):**
 - Forms 2% of all meningiomas.
 - Display high proliferation indices with brain invasion.
 - Variants: Papillary, rhabdoid, anaplastic.
- Etiology:
 - o Sporadic in most cases.
 - o Associated with Neurofibromatosis type 2 (NF2) in some cases.
- Pathophysiology:
 - o Benign but locally aggressive encapsulated tumors.
 - Displaces but does not invade adjacent neural tissue.
 - Invades the adjacent bone by extension along haversian canals (without destruction).
 - Adjacent bone is hyperostotic in 25% of cases.
 - o Occurs at different anatomic sites:
 - **Para-sagittal region (Most common location).**
 - Falx
 - Convexity
 - Tuberculum sellae
 - Sphenoid ridge
 - **Petrous face (CPA)**
 - Tentorium
 - Clivus

- Posterior fossa meningiomas arise outside the IAC on either:
 - Posterior surface of petrous bone.
 - Along the sigmoid sinus.
- Because they arise outside the IAC, become large before they produce signs and symptoms of CN-VIII compression (late presentation).
- Clinical picture:
 - **Similar to VS:**
 - Small tumors:
 - Hearing loss, tinnitus and imbalance.
 - Larger tumors
 - Involvement of other cranial nerves and hydrocephalus.
- Diagnosis:
 - **Audio-vestibular Tests:**
 - Similar findings of VS once CN-III is compressed.
 - Cannot distinguish between VS and meningiomas.
 - Low sensitivity of detecting meningiomas compared to VS because meningiomas do not originate within IAC.
 - **Imaging:**
 - Most important tool in the diagnosis.
 - **CT scan with contrast:**
 - Homogeneous enhanced tumors (compared to VS).
 - Areas of calcifications (25%).
 - Bony hyperostotic changes.
 - **MRI with Gad:**
 - General features:
 - No widening of IAC.
 - Broad dural base enhancement.
 - Dural tail:
 - Extension of enhancement along underlying dural surface (50-75%).
 - Form obtuse angle with the underlying bone.
 - Tumor progression may result in erosion of cranial base and invasion into neural foramina or the middle ear.
 - **T1:**
 - HYPO-intense to brain.
 - **T1 C+:**
 - Heterogeneous enhancement
 - More heterogeneous than VS.
 - Less marked enhancement than VS.
 - **T2:**
 - Variable intensity.

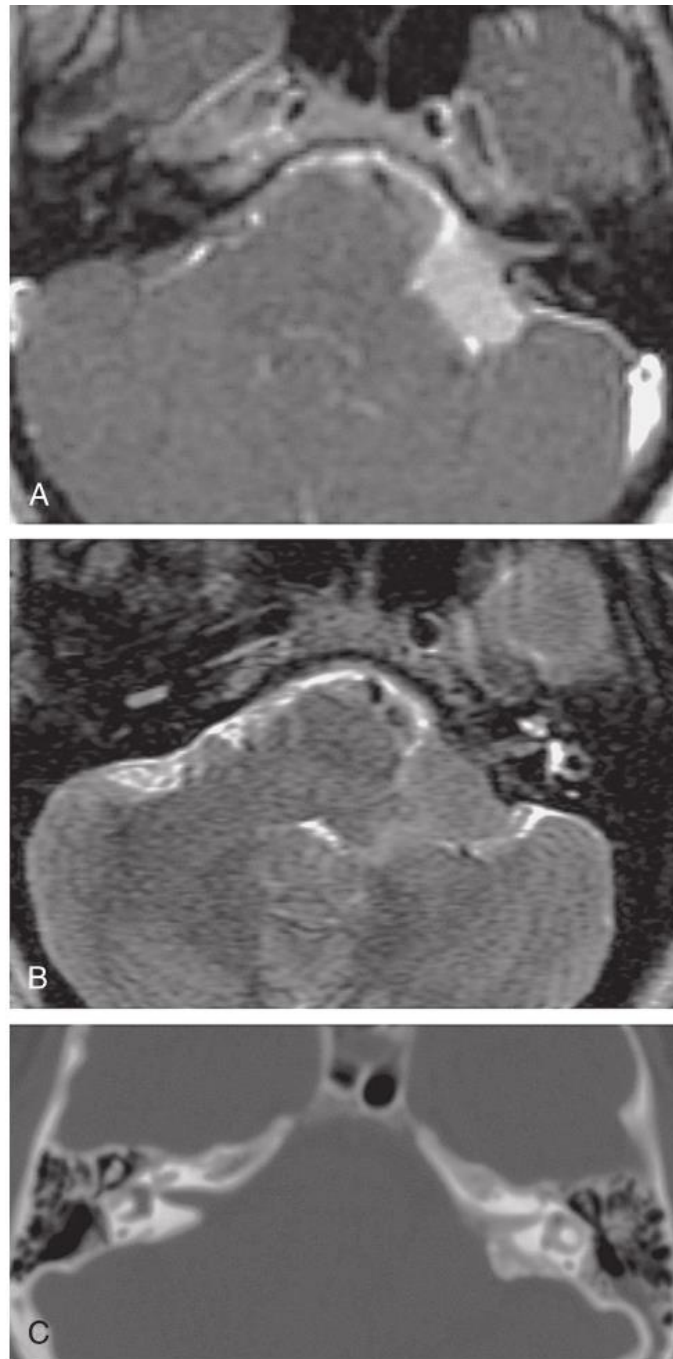


FIGURE 177-4. Cerebellopontine angle meningioma with classic features on magnetic resonance imaging (MRI) and computed tomography (CT). **A**, Post-gadolinium T1-weighted MRI: The tumor is eccentric to the internal auditory canal (IAC) and broadly dura based with obtuse angles at the bone-tumor interface. This hemispherical tumor shows uniform enhancement. Note the lack of IAC involvement. **B**, T2-weighted MRI. The tumor is homogeneously isointense to gray matter. **C**, CT bone windows. The tumor shows extensive hyperostosis underlying the dural base. (From Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, pp 245-250.)

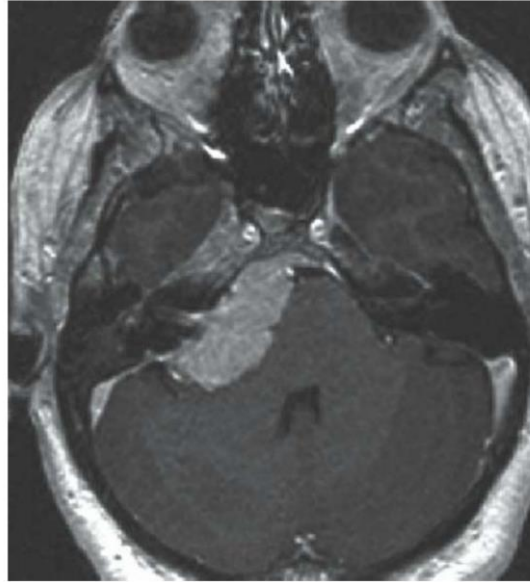


Figure 159.15 Axial T1-weighted contrast-enhanced MRI image at the level of the IAC demonstrating a right CPA meningioma with broad dural attachment and enhancement.

- Treatment (Similar to VS):
 - **Observation with serial imaging**
 - Indications:
 1. **Small tumors in asymptomatic patients.**
 2. **Only hearing ear (?)**
 3. **Elderly (>65 years).**
 4. **Non-operative patients.**
 - **Stereotactic Radiation (Radiosurgery):**
 - Indications:
 1. **Recurrence after surgery.**
 2. **Only hearing ear (?)**
 3. **Elderly (>65 years).**
 4. **Non-operative patients.**
 - **Microsurgery:**
 - Gold standard treatment for meningioma with progressive symptoms.
 - Similar approaches as for VS.
 - Complete resection is not always feasible.
 - Decompression of surrounding neurovascular structures provides:
 - Symptomatic relief
 - Tissue for histopathologic diagnosis.

Primary Cholesteatoma (Epidermoid):

- 3rd most common CPA tumor (5%).
- Histopathology:
 - o Stratified squamous epithelial lining surrounds a desquamated keratin.
- Etiology:
 - o Congenital, originates from epithelial rests within the CPA or the temporal bone (Most common).
 - o Acquired, from extension from the mastoid, middle ear, or petrous.
- Pathophysiology:
 - o Benign slow-growing lesions, expanding into areas of least resistance.
 - Tend to erode surrounding bone and encase neurovascular structures.
 - Produce variable shapes with irregular surfaces.
 - May extend in dumbbell pattern into middle cranial fossa.
 - o As the lesions expand, compression and irritation of surrounding structures produce the signs and symptoms, which will be apparent at the second to fourth decades.
- Clinical picture:
 - o **Similar to VS:**
 - Distinguishing clinical manifestations:
 - Facial twitching.
 - Progressive facial paralysis (earlier presentation).
- Diagnosis:
 - o **Audio-vestibular Tests:**
 - Similar findings of VS once CN-III is compressed.
 - Cannot distinguish between VS and epidermoid.
 - o **Imaging:**
 - Most important tool in the diagnosis.
 - **CT scan with contrast:**
 - Hypodense irregular lesions.
 - No enhancement with intravenous contrast.
 - Remodeling of surrounding bone.
 - **MRI with Gad:**
 - **T1:**
 - o HYPO-intense to brain.
 - **T1 C+:**
 - o **NO enhancement**
 - o Unlike VS, meningiomas and chondromas.
 - **T2:**
 - o HYPER-intense to brain.
 - **Fluid-Attenuated Inversion Recovery (FLAIR):**
 - o **HYPER-intense to CSF.**
 - o Unlike arachnoid cysts.
 - **Diffusion-weighted Imaging (DWI):**
 - o **HYPER-intense to brain (Restricted diffusion).**
 - o Unlike arachnoid cysts.

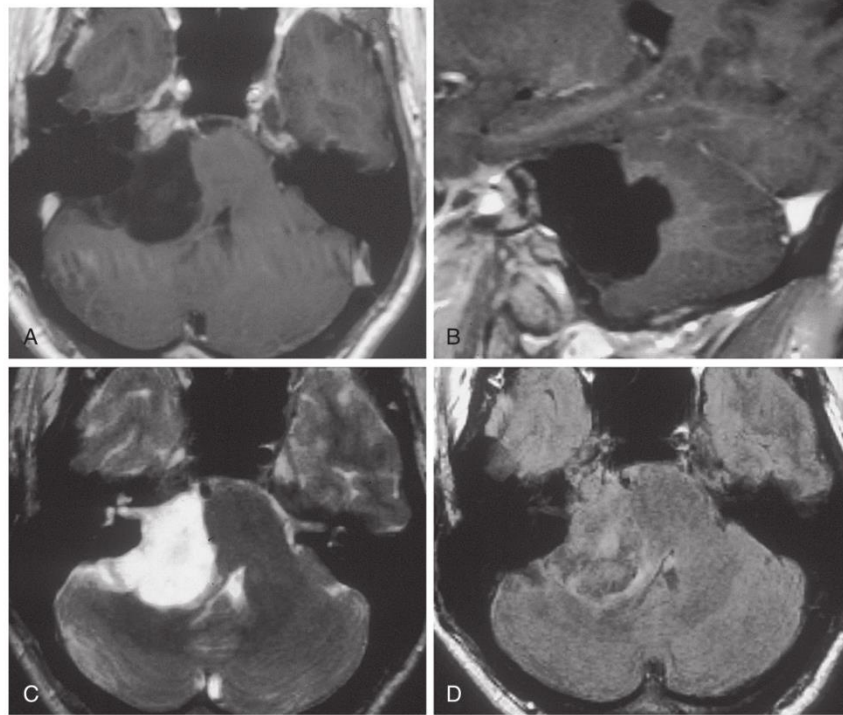


FIGURE 177-6. Cerebellopontine angle (CPA) epidermoid, demonstrated by magnetic resonance imaging. **A** and **B**, T1-weighted axial and sagittal images after gadolinium administration. The CPA mass that extends to the prepontine cistern is isointense to cerebrospinal fluid (CSF) and shows no enhancement. **C**, T2-weighted image. The tumor, with its irregular cauliflower-like margins, is isointense to CSF, as in T1-weighted images; it is compressing the fourth ventricle, but the internal auditory canal is normal. **D**, On this fluid-attenuated inversion recovery sequence image, the epidermoid has heterogeneous hyperintense foci, which enables differentiation of an epidermoid from an arachnoid cyst in this case. (From Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, pp 245-250.)

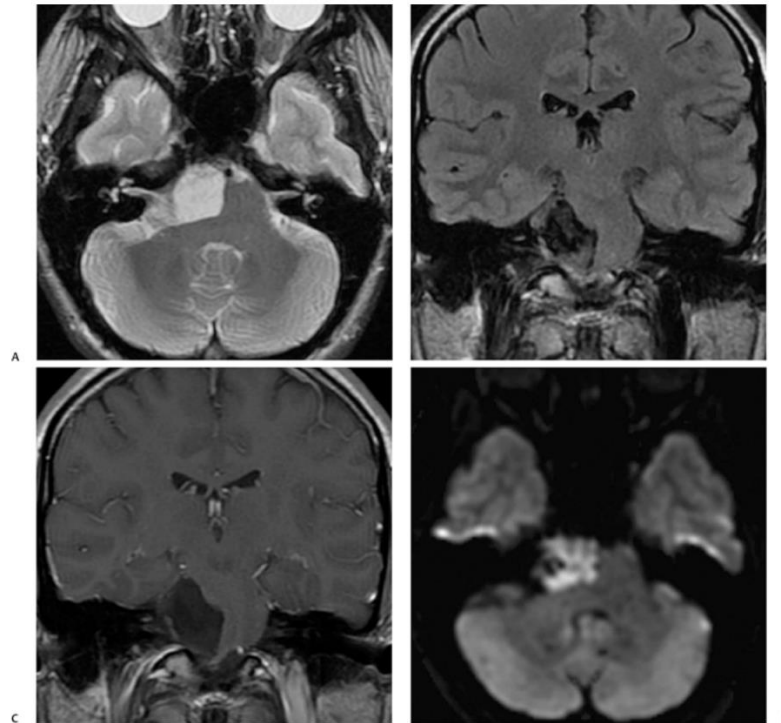


Fig. 8.16 Cerebellopontine angle (CPA) epidermoid in a 19-year-old with an 8-month history of headaches but no hearing loss or facial nerve dysfunction. **(A)** Axial T2-weighted magnetic resonance image shows the extraaxial hyperintense right CPA mass with no adjacent parenchymal edema. **(B)** Coronal fluid attenuated inversion recovery (FLAIR) image shows the mass to be hyperintense relative to cerebrospinal fluid. There is no hydrocephalus. **(C)** Contrast-enhanced T1-weighted magnetic resonance image shows no enhancement. **(D)** Diffusion weighted image shows restricted diffusion and confirms that there is no extension in the internal auditory canal.

- Treatment:

○ **Microsurgery :**

- Gold standard treatment for epidermoids.
- Similar approaches as for VS.
- Complete resection is not always feasible.
- Lesions that envelop vital neurovascular structures can be managed with subtotal resection and monitored with imaging since these are slowly growing lesions.
- Recurrence rates of:
 - 23% after total resection.
 - 27% after subtotal resection.
- Majority of recurrences required surgical intervention.

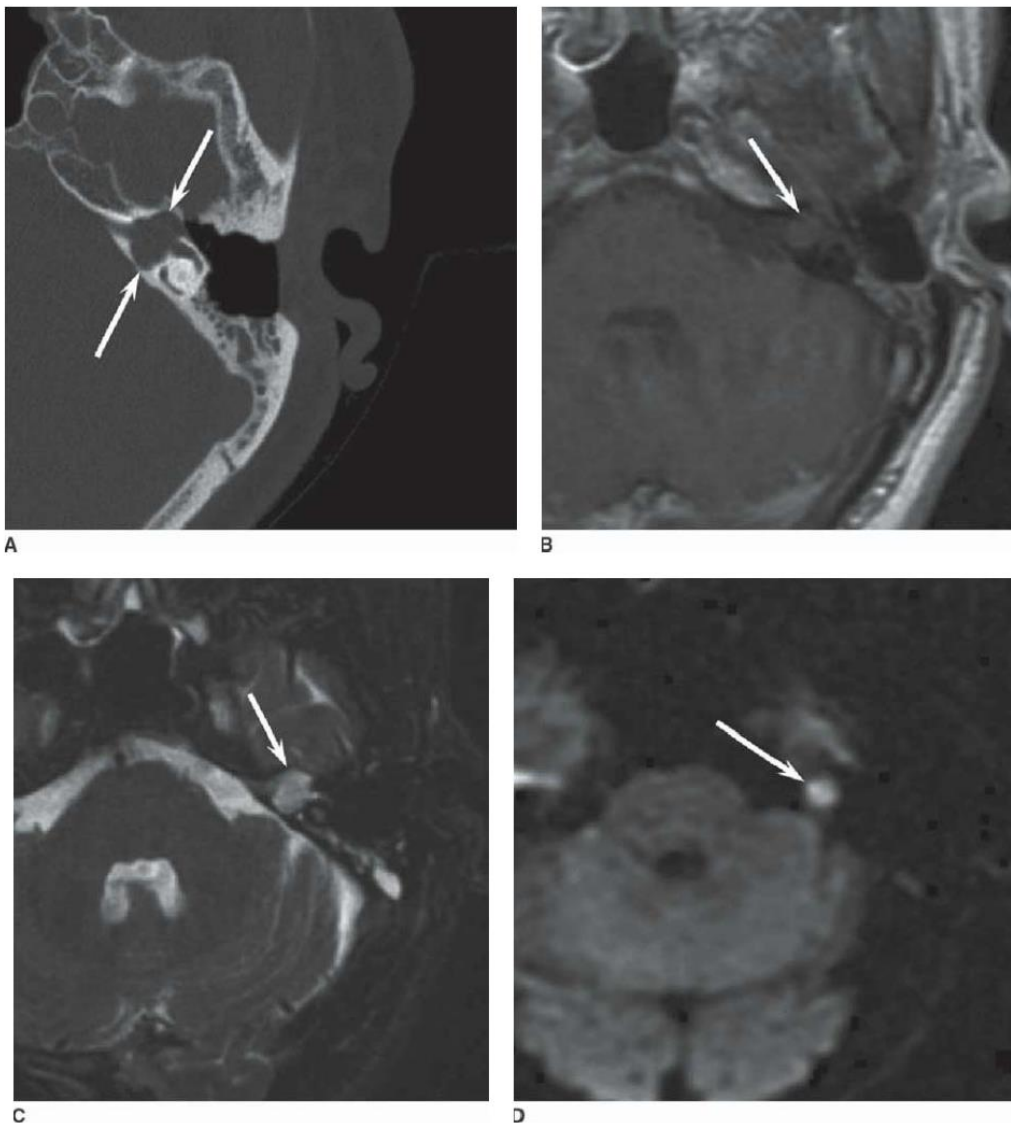


Figure 159.16 Radiographic appearance of recurrent acquired cholesteatoma. Axial CT of the temporal bone demonstrating a mastoidectomy defect with a smooth expansile mass (arrows) eroding the inner ear (A). This mass is isointense to brain and nonenhancing on axial T1-weighted contrast-enhanced MRI image (B), but hyperintense on T2-weighted images (C) and diffusion-weighted images (D). The restricted diffusion distinguishes recurrent cholesteatoma from scar.

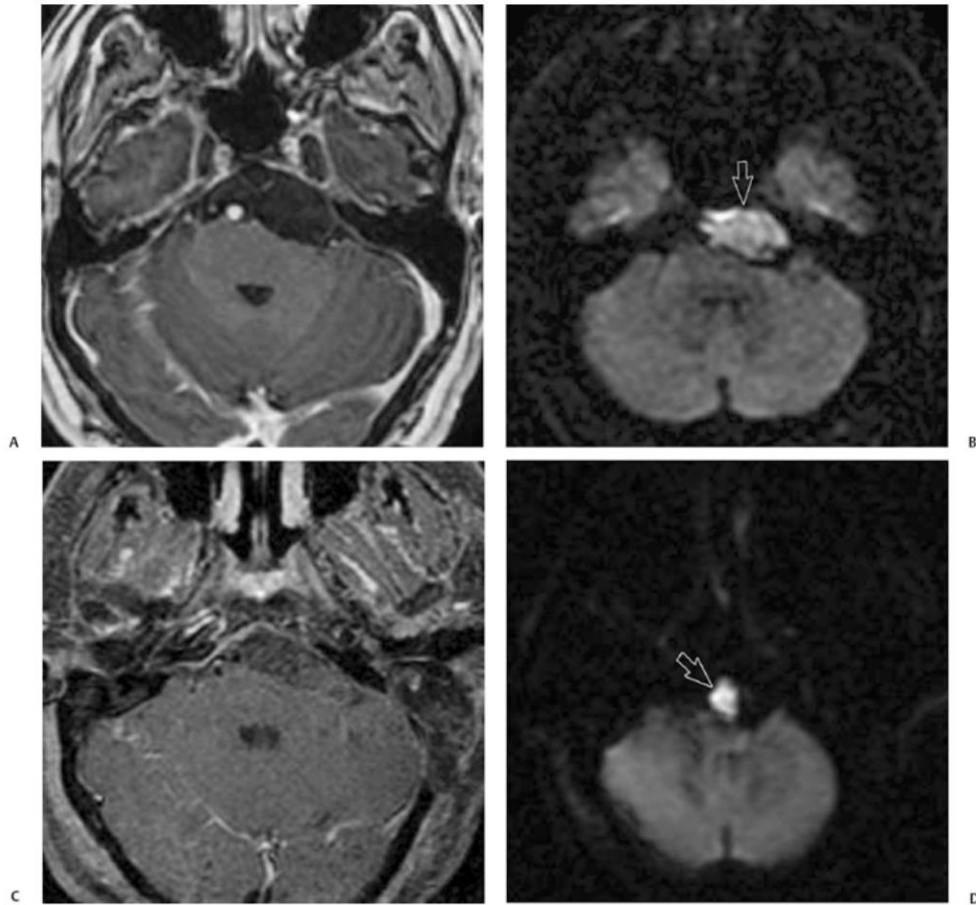


Fig. 8.17 Epidermoid. (A) Preoperative gadolinium-enhanced T1-weighted magnetic resonance image (T1WI). (B) Diffusion weighted image shows a preponine epidermoid tumor with characteristic restricted diffusion. (C) Gadolinium-enhanced T1WI performed following subtotal surgical resection shows heterogeneous signal, and residual tumor cannot be distinguished from postoperative changes. (D) Diffusion weighted imaging confirms residual epidermoid, seen as focal high signal intensity.

Arachnoid Cysts:

- Account for 1% of tumors within CPA.
- Congenital malformation of arachnoid result in sac filled with normal CSF.
- Typically asymptomatic but can cause compression of CN-VII and VIII.
- Smooth lesions displacing neurovascular structures without invasion.
- Imaging (Iso-intense to CSF):
 - o T1: HYPO-intense.
 - o T1 C+: Non-enhancing.
 - o T2: HYPER-intense.
 - o **FLAIR: HYPO-intense.**
 - o **Diffusion weighted: HYPO-intense (No restricted diffusion).**
- Treatment:
 - o Observation if asymptomatic.
 - o Decompression via suboccipital approach for symptomatic cysts.

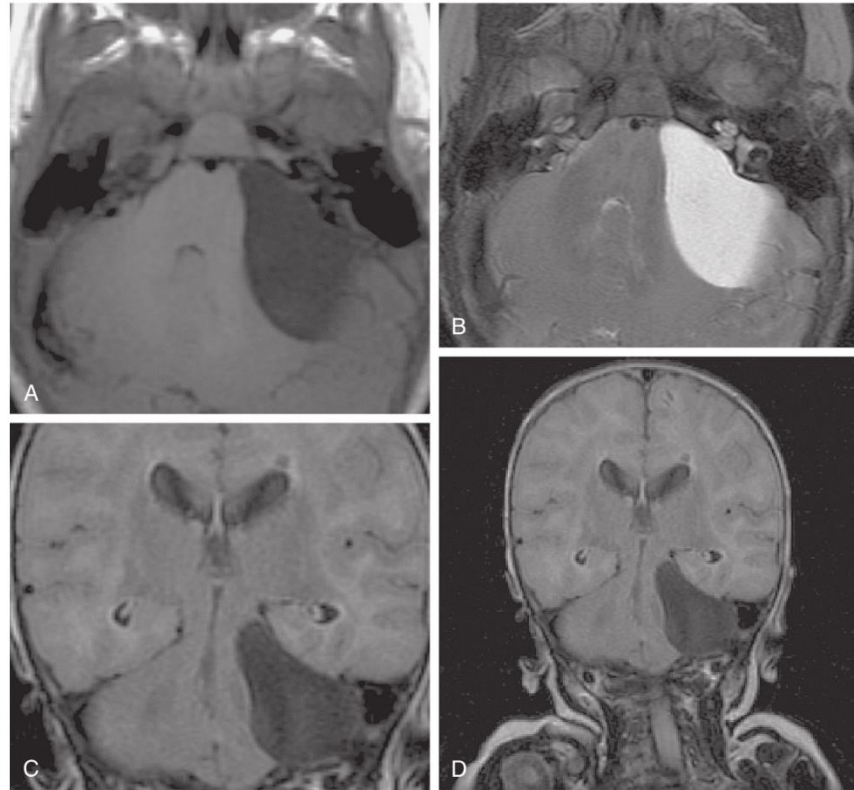


FIGURE 177-7. Cerebellopontine angle arachnoid cyst, demonstrated by magnetic resonance imaging. **A**, T1-weighted image. **B**, T2-weighted image. **C** and **D**, T1-weighted coronal and axial images, respectively. The cyst is isointense to cerebrospinal fluid on all sequences. It is homogeneous and has a smooth surface, which can be helpful in differentiating it from an epidermoid. (From Maya MM, Lo WWM. Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, pp 245-250.)

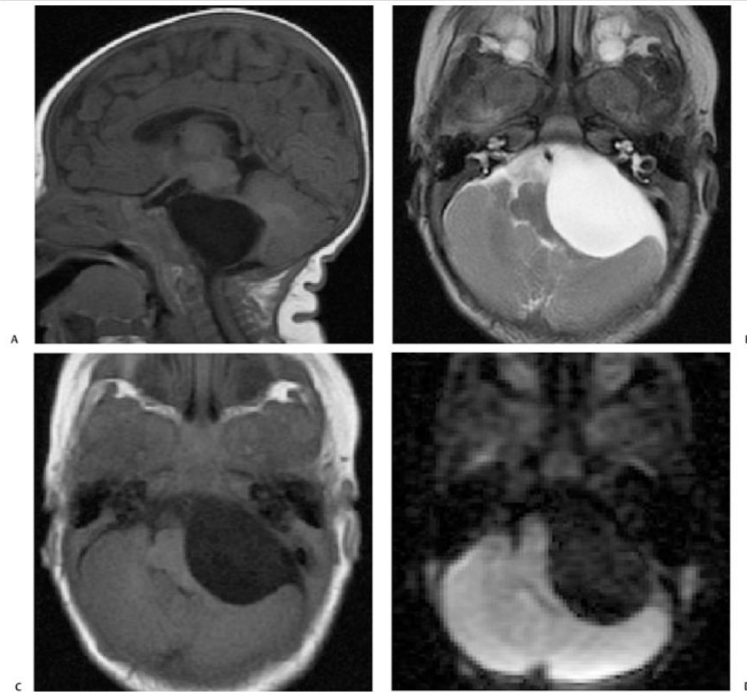


Fig. 8.18 Cerebellopontine angle (CPA) arachnoid cyst. **(A)** Sagittal T1-weighted and **(B)** axial T2-weighted magnetic resonance images show a smoothly contoured fluid-intensity left CPA mass, which suppresses completely on axial fluid attenuated inversion recovery (FLAIR) imaging **(C)**. No abnormal signal is seen in the adjacent medulla or left cerebellar hemisphere. **(D)** Axial diffusion-weighted image shows low signal intensity (no restricted diffusion), confirming the cerebrospinal fluid/fluid nature of this mass. Despite significant local mass effect, there is no obstructive hydrocephalus in this child who presented with macrocrania and head tilt. (Courtesy of Gary Hedlund, MD.)

Non-Vestibular Schwannomas:

- Account for 1% of tumors within CPA.
- More common in NF2 patients.
- Schwannomas may arise on any other cranial nerve in the posterior fossa:
 - **Trigeminal (2nd most common after CN-VIII).**
 - Facial
 - Glossopharyngeal
 - Vagal
 - Accessory
 - Hypoglossal
- On imaging studies, Non-VS have the same characteristics as those of VS except for their location.
- Distinguished by their different location and by symptoms of dysfunction of cranial nerve of origin.
- Specific features on Non-VS:
 - **Trigeminal Schwannomas:**
 - Most common non-vestibular schwannomas.
 - Arise either:
 - **Intra-durally:**
 - From the nerve root in CPA and Meckel cave.
 - **Extra-durally:**
 - From gasserian ganglion in middle cranial fossa.
 - Symptoms:
 - Ipsilateral facial hypesthesia in CN-V distribution.
 - CT imaging:
 - Enhancing expansion of Meckel cave or foramen lacerum.
 - MRI imaging
 - Enhancing lesion in anterior-posterior orientation adjacent to Meckel cave.
 - May be difficult to distinguish from a meningioma.
 - Treatment:
 - Lesions extending into Meckel cave can be treated with radiation or can be resected through the anterolateral intradural approach (Dolenec's) and anterior petrosal approach.

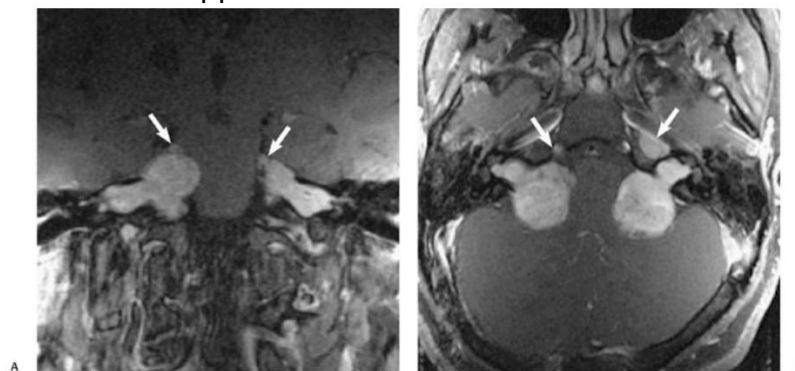


Fig. 8.22 Neurofibromatosis type II. (A) Coronal and (B) axial gadolinium enhanced T1-weighted magnetic resonance images show bilateral lobulated, enhancing cerebellopontine angle–internal auditory canal (IAC) vestibular schwannomas extending to the fundus of the IAC bilaterally, and compressing the pons. Note also the presence of bilateral enhancing masses involving the fifth cranial nerves and Meckel's cave (arrows) consistent with schwannomas.

○ **Facial Schwannomas:**

- May arise anywhere along the course of the facial nerve.
 - CPA is involved in more than 50% of tumors.
 - Difficult to distinguish from VS if limited to IAC.
- Symptoms:
 - Facial weakness if tumor is very large (unlike FN hemangioma).
 - **Facial twitching:**
 - **Distinguish it from VS but NOT from Epidermoid.**
- Electroneuronography (ENoG):
 - Reduced in facial nerve schwannomas even when no facial weakness or tic is present.
 - Normal in VS until the tumor becomes very large.
- CT imaging:
 - Erosion of IAC or labyrinthine facial nerve canal.
 - Expansion of geniculate ganglion and fallopian canal.
- MRI imaging
 - Enhancing dumbbell-shaped expansion of intra-temporal facial nerve.
 - Enhancing expansion of geniculate ganglion region.
- Treatment:
 - **Observation** for Grade I-III HB.
 - **Surgical decompression** for declining facial nerve function via middle fossa or trans-mastoid approaches.
 - **Resection and cable grafting** for nonfunctional facial nerve (Grade V-VI HB).

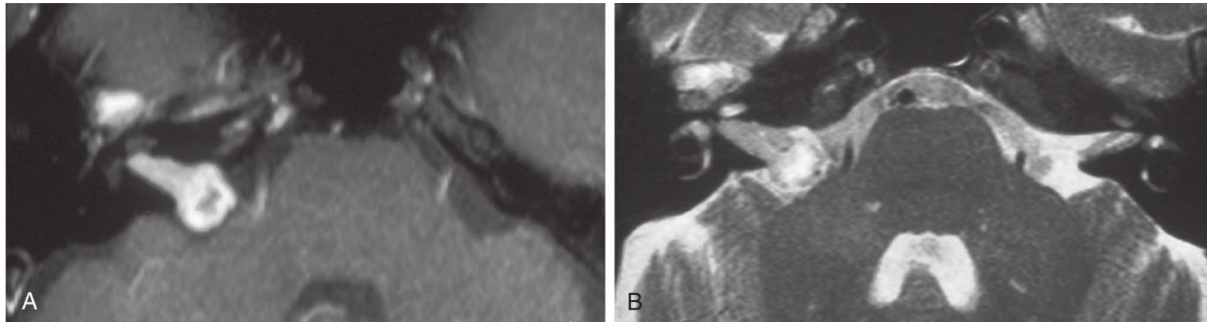


FIGURE 177-8. Facial schwannoma of internal auditory canal, demonstrated by magnetic resonance imaging. A and B, Postgadolinium T1- and T2-weighted images. The patient presented with progressive sensorineural hearing loss with later development of facial palsy. The enhancing intracanalicular tumor is indistinguishable from an acoustic schwannoma. Note also the extension of the tumor to the geniculate ganglion. (From Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, pp 245-250.)

○ **Lower CNs Schwannomas:**

- Typically involve the inferior aspect of posterior fossa.
- Symptoms:
 - Related to the nerve of origin but expansion of tumor may affect all of lower cranial nerves.
 - Residual hearing is more likely to be preserved with lower cranial nerve neuromas compared to VS.
- Imaging:
 - Enhancing lesion of the CPA.
 - Expansion of jugular foramen (CN-IX-X-XI).
 - Expansion of hypoglossal canal (CN-XII).
- Treatment:
 - Similar to VS.

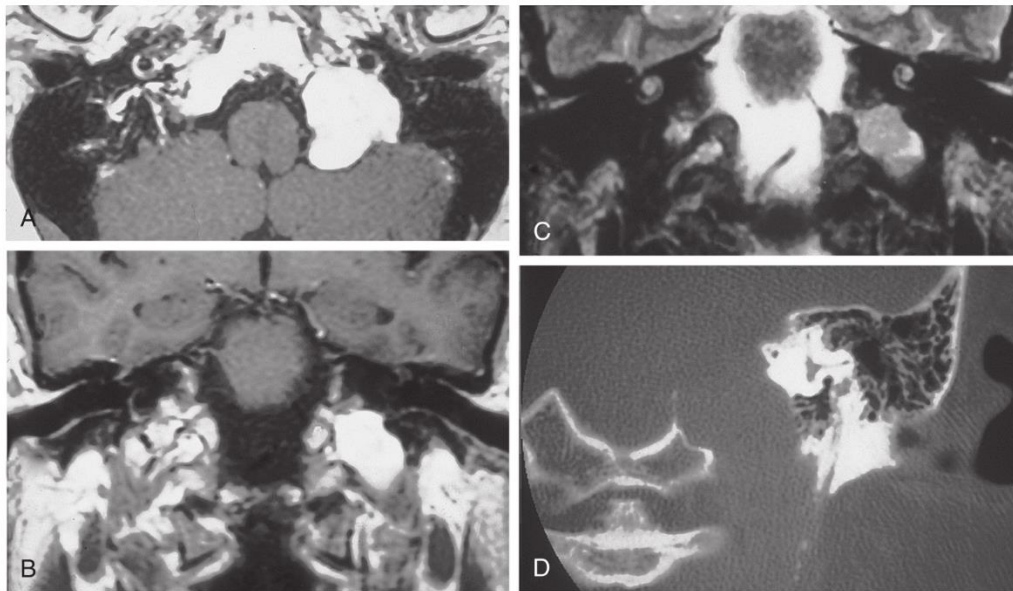


FIGURE 177-10. Jugular foramen schwannoma, demonstrated by magnetic resonance imaging (MRI) and computed tomography (CT). **A** and **B**, Axial and coronal T1-weighted postgadolinium images. The large tumor in the left jugular fossa extends inferiorly and shows avid enhancement. **C**, Coronal T2-weighted image depicts the extracranial portion of the tumor, which is hyperintense to gray matter. **D**, CT bone windows, coronal image. Expansion of the jugular foramen is symmetric, and the margins are well defined. (From Maya MM, Lo WW, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby, pp 245-250.)

Lipomas:

- Account for 1% of tumors within CPA.
- Congenital malformation resulting in hamartomatous collections of mature adipose tissue.
- Grow very slowly and induce similar symptoms to other CPA lesions.
- Smooth lesions displacing neurovascular structures without invasion.
- Imaging:
 - o T1: HYPER-intense.
 - o T1 C+: Non-enhancing.
 - o T2: HYPER-intense.
 - o **Post-contrast Fat-suppressed T1: HYPO-intense.**
- Treatment:
 - o Observation if asymptomatic.
 - o Decompression via suboccipital approach for symptomatic lesions.

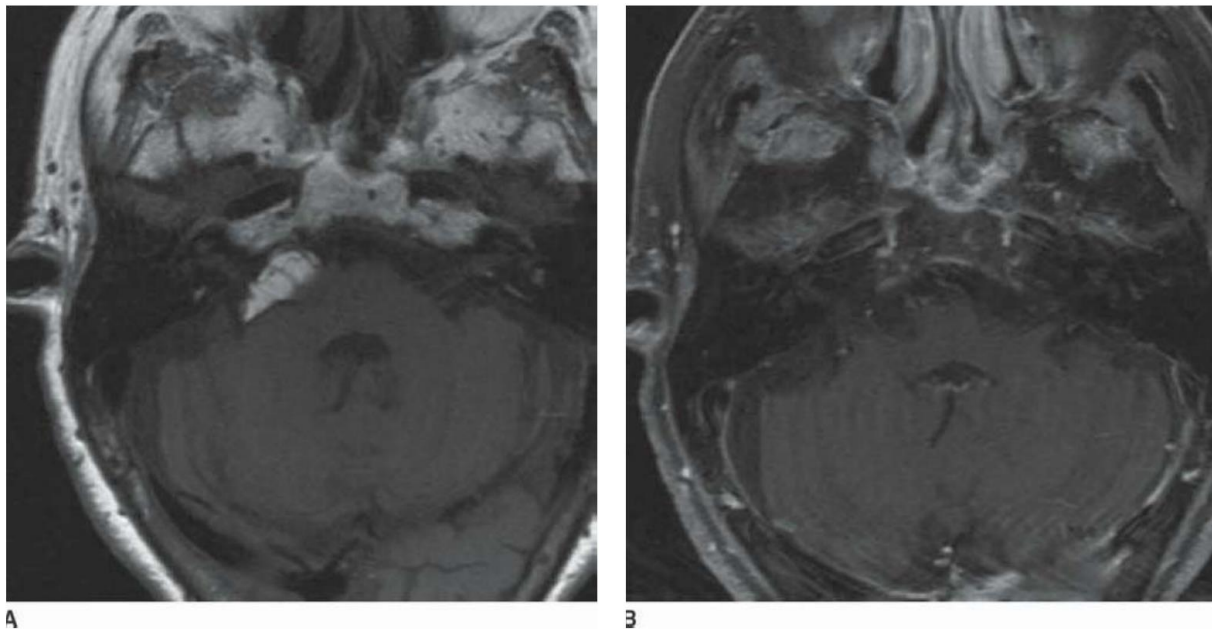


Figure 159.17 Axial T1-weighted unenhanced MRI image at the level of the IAC demonstrating a right CPA lipoma (A). The diagnosis is confirmed with a postcontrast fat-suppressed T1 MRI image as this lesion appears dark (B).

TABLE 177-2. Imaging Features of the Three Most Common Cerebellopontine Angle Lesions

Feature	Acoustic Neuromas	Meningiomas	Epidermoids
Location	Centered on the internal auditory canal	Eccentric to the internal auditory canal	Anterolateral or posterolateral to brainstem
Bone changes	Most enlarge the internal auditory canal	Occasional hyperostosis	Occasional erosion
Shape	Spherical or ovoid, acute bone-tumor angle	Hemispherical, rarely plaque-like; may herniate through tentorium; obtuse bone-tumor angle	Variable; tends to dumbbell into middle fossa or contralateral cerebellopontine angle
CT density	Mostly isodense	Slightly hypodense; some calcified	Mostly hypodense; occasional peripheral calcium
CT enhancement	Moderate to marked; often inhomogeneous	Marked and homogeneous	Nonenhancing
T1-weighted MRI	Isointense or hypointense	Isointense or hypointense	Hypointense
Gadolinium enhancement	Marked	Moderate	Nonenhancing
T2-weighted MRI	Isointense or hypointense	Variable	Hyperintense

Modified from Lo WM: Tumors of the temporal bone and cerebellopontine angle. In Som PM, Bergeron RT, editors: *Head and neck imaging*, St Louis, 1991, Mosby, pp 420-445.

CT, computed tomography; MRI, magnetic resonance imaging.

Table 135-2**Imaging Characteristics of Cerebellopontine Angle Lesions**

Lesion	T1	T2	T1 + Contrast	Notes
Vestibular schwannoma	Isointense to brain	Slightly hyperintense	Enhances	
Meningioma	Isointense to brain	Hypointense/hyperintense	Enhances	T2 signal depends on calcium content
Arachnoid cyst	Hypointense	Hyperintense	No enhancement	Signal characteristics mirror CSF
Epidermoid	Hypointense	Hyperintense	No enhancement	T2 signal slightly more dense than CSF, DWI does not restrict
Lipoma	Hyperintense	Hypointense	No enhancement	T1 signal disappears with fat suppression

CSF, cerebrospinal fluid; DWI, diffusion-weighted imaging.

Radiological DDx of extra-axial CPA lesions

lesion	T1	T2	Gad	content	Special features
VS	↓ ↔	↓ ↔	Yes, marked	May contain micro/macrocysts	• IAC involved • Acute bone tumor angle
meningioma	↓ ↔	Variable ↓ ↑	Yes, moderate		• Dural tail • Obtuse bone tumor angle • Hyperostosis • Calcification
Epidermoid cyst	↓	↑	No	Heterogeneous	• Encases neurovascular structures • Irregular surface
Arachnoid cyst	↓	↑	No	Homogeneous	• isointense to CSF on all sequences • Displaces neurovascular structures • Smooth surface
Lipoma	↑	↑	No	Homogeneous	Decrease intensity with fat suppression
Cranial nerves schwannoma	↓ ↔	↑ ↔	Yes		Follow anatomical course of corresponding nerve
Metastasis	↓	↑	Possible		Brain edema
aneurysm	↓	↓	Possible (in thrombosed aneurysm)		Well circumscribed

Petrous Apex Lesions:

- Extra-dural lesions.
- Most common petrous apex lesions:
 - o Cholesterol Granuloma
 - o Epidermoid
 - o Mucocele
 - o Petrous apicitis
- Normal variants of petrous apex (**Leave Me Alone**):
 - o Asymmetric Petrous Apex
 - o Petrous Apex Effusion
- **Imaging of Normal Petrous Apex:**
- **T1:**
 - o Diploic: HYPER-intense to Brain.
 - o Pneumatized or sclerotic: HYPO-intense to Brain.
- **T1 C+:**
 - o Diploic: No enhancement.
 - o Pneumatized or sclerotic: No enhancement.
- **T2:**
 - o Diploic: HYPO-intense to Brain.
 - o Pneumatized or sclerotic: HYPO-intense to Brain.
- **Fat-Suppression:**
 - o **Diploic: HYPO-intense to Brain.**
 - o Pneumatized or sclerotic: HYPO-intense to Brain.

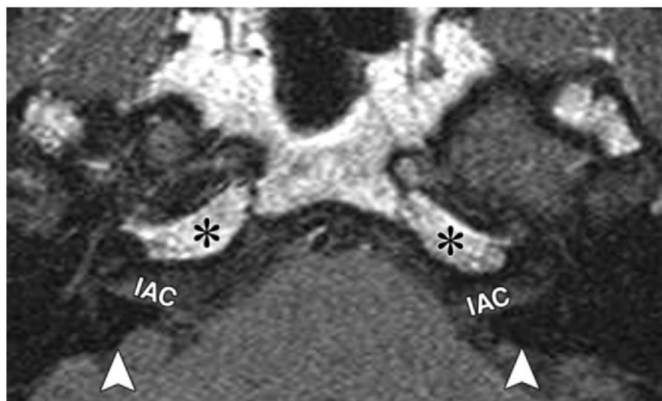


Figure 2. Normal petrous apex at MR imaging. Axial unenhanced T1-weighted MR image at the level of the IACs shows the normal petrous apex. Note the high-signal-intensity fatty marrow in the anterior petrous apex (*) and the low signal intensity of the denser posterior petrous apex (arrowheads). In pneumatized or sclerotic apices, the anterior portion may normally demonstrate low signal intensity due to the absence of fatty marrow.

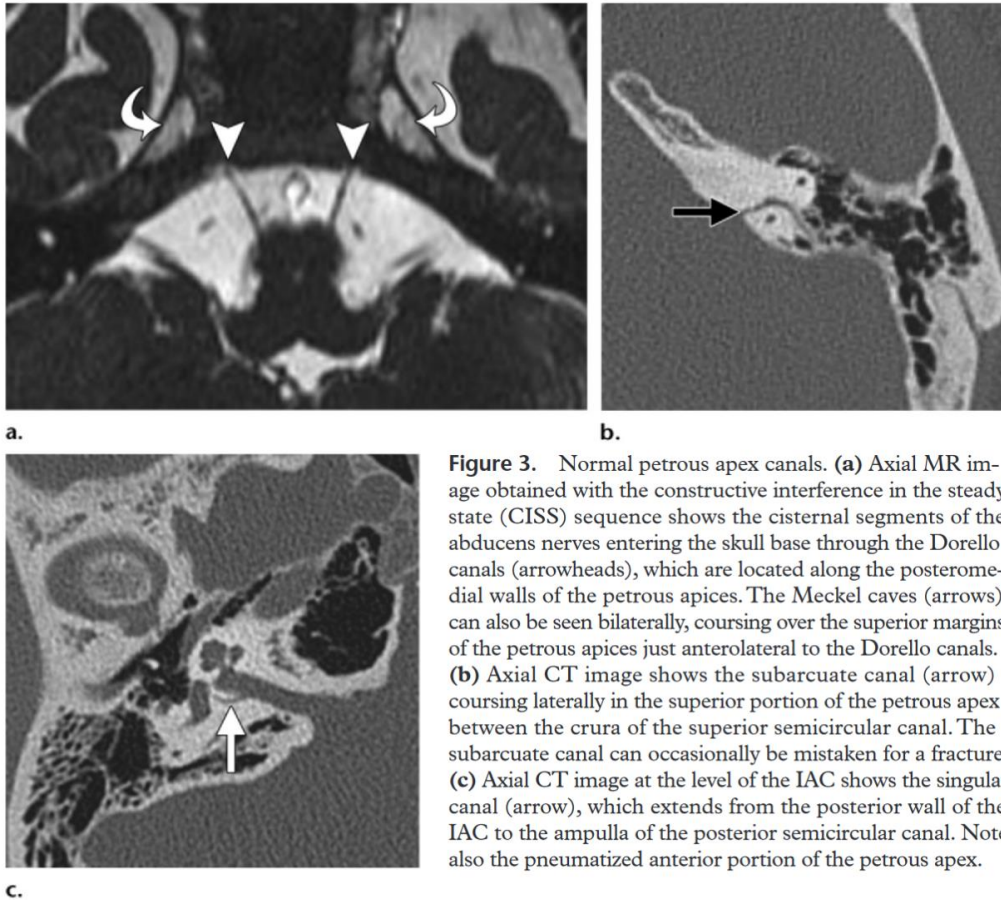
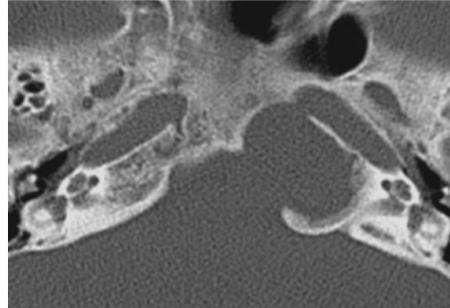


Figure 3. Normal petrous apex canals. **(a)** Axial MR image obtained with the constructive interference in the steady state (CISS) sequence shows the cisternal segments of the abducens nerves entering the skull base through the Dorello canals (arrowheads), which are located along the posteromedial walls of the petrous apices. The Meckel caves (arrows) can also be seen bilaterally, coursing over the superior margins of the petrous apices just anterolateral to the Dorello canals. **(b)** Axial CT image shows the subarcuate canal (arrow) coursing laterally in the superior portion of the petrous apex between the crura of the superior semicircular canal. The subarcuate canal can occasionally be mistaken for a fracture. **(c)** Axial CT image at the level of the IAC shows the singular canal (arrow), which extends from the posterior wall of the IAC to the ampulla of the posterior semicircular canal. Note also the pneumatized anterior portion of the petrous apex.

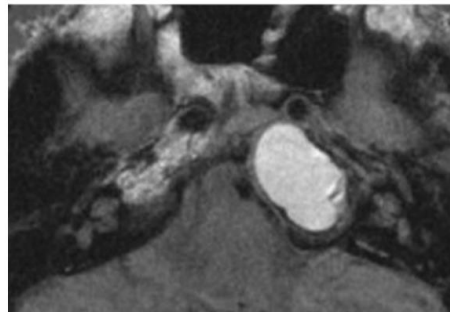
Cholesterol Granuloma:

- Most common petrous apex lesions.
- Slow-growing expansile cystic lesion.
- Occur 20 times more frequently than epidermoids.
- Uncommon compared to VS.
- Etiology:
 - o Long-standing history of otitis media in a patient with well-pneumatized petrous apex.
- Pathophysiology:
 - o Occlusion of air cell system results in accumulation of secretions and hemorrhage into the air cells followed by foreign body reaction and progressive granuloma formation.
 - o Contains viscous brown fluid, granulation tissue, and cholesterol crystals, which are often contained within a thick fibrous capsule that lacks a true epithelial lining.
- Clinical picture:
 - o Similar to VS with the extension of the mass into the CPA.
 - o Gradenigo's syndrome:
 - Otorrhea.
 - Lateral rectus palsy.
 - Trigeminal/retroorbital pain.

- Imaging:
 - **CT Scan:**
 - General features:
 - Smooth, round and expansive isodense lesion.
 - NO enhancement with contrast (may have rim-enhancement).
 - Highly pneumatized contralateral petrous apex.
 - **MRI:**
 - **T1:**
 - HYPER-intense to brain.
 - Unlike Epidermoids.
 - **T1 C+:**
 - No Enhancement.
 - **T2:**
 - HYPER-intense to brain.
 - **Fat-suppression:**
 - HYPER-intense to brain.
 - Unlike asymmetric pneumatization.
- Treatment:
 - **Observation:**
 - For asymptomatic lesions.
 - **Drainage:**
 - For symptomatic lesions.



a.



b.

Figure 4. Cholesterol granuloma. (a) Axial CT image shows a smoothly expansile lesion in the left petrous apex that causes bone dehiscence along its posteromedial margin. (b) On a corresponding axial nonenhanced T1-weighted MR image, the lesion is well circumscribed and has homogeneous high signal intensity, a finding characteristic of a cholesterol granuloma.

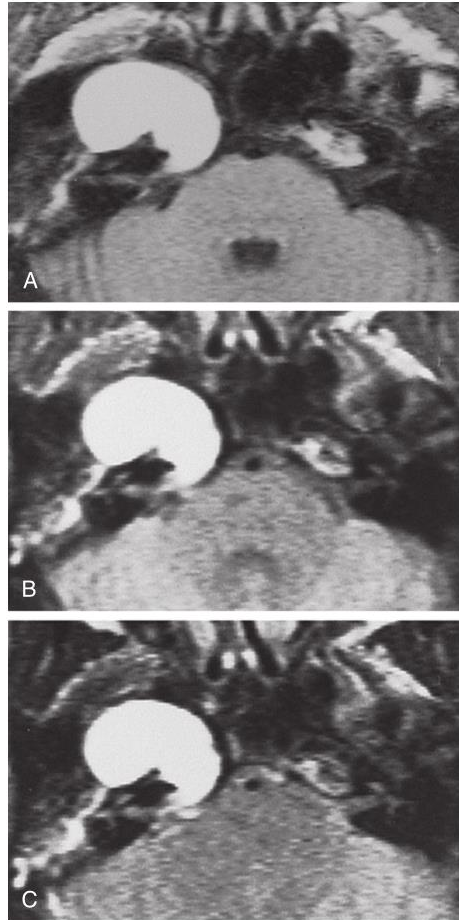


FIGURE 177-14. Petrous apex cholesterol granuloma demonstrated by magnetic resonance imaging. **A to C**, T1-weighted, proton density, and T2-weighted images, respectively. The lesion is hyperintense on all three images. Note the hypointense rim medial to the lesion accentuated on **B** and **C**, attributable to a hemosiderin deposit or chemical shift artifact. (From Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby.)

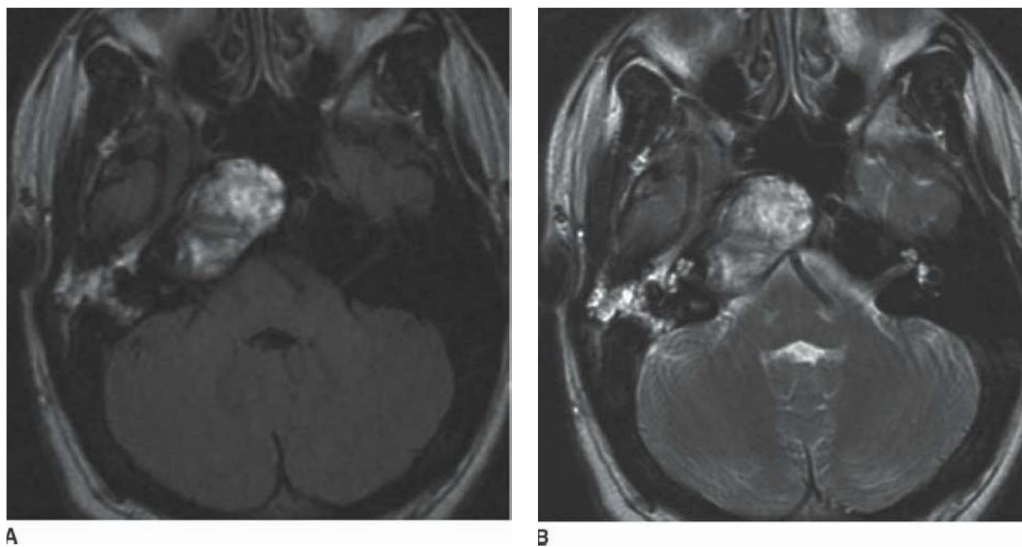
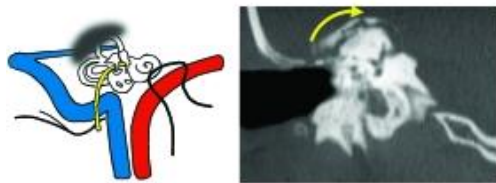


Figure 159.22 Cholesterol granuloma of the right petrous apex is hyperintense on both T1-weighted MRI images (**A**) and T2-weighted images (**B**).

Epidermoid (Petrous Bone Cholesteatoma):

- Accounts for 10% of all petrous apex lesions.
- Most commonly are congenital.
- Similar to CPA Epidermoid.
- Moffat-Smith classification of Petrous Bone Cholesteatoma:
 1. Supra-labyrinthine
 2. Supra-labyrinthine-apical
 3. Infra-labyrinthine
 4. Infra-labyrinthine-apical
 5. Massive-labyrinthine
 6. Massive-labyrinthine-apical
 7. Apical

Supralabyrinthine



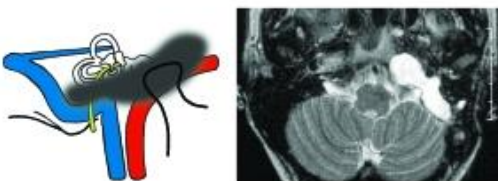
Supralabyrinthine-apical



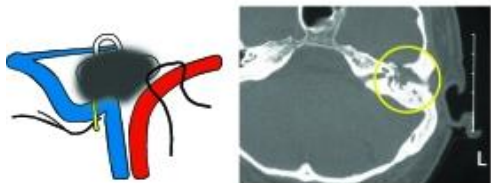
Infralabyrinthine



Infralabyrinthine-apical



Massive labyrinthine



Massive labyrinthine-apical



Apical



- Imaging:

○ **CT scan with contrast:**

- Hypodense smooth expansile lesions.
- No enhancement with intravenous contrast.
- Remodeling of surrounding bone.

○ **MRI with Gad:**

▪ **T1:**

- HYPO-intense to brain.
- Unlike cholesterol granulomas.

▪ **T1 C+:**

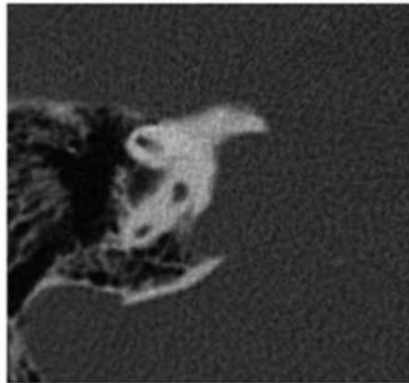
- NO enhancement

▪ **T2:**

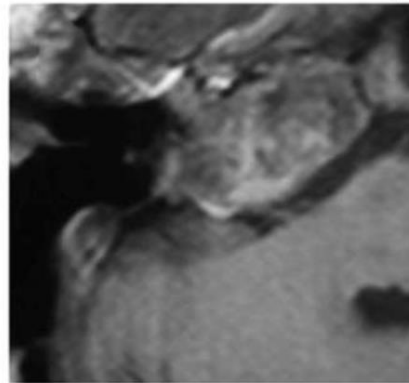
- HYPER-intense to brain.

▪ **Diffusion-weighted Imaging (DWI):**

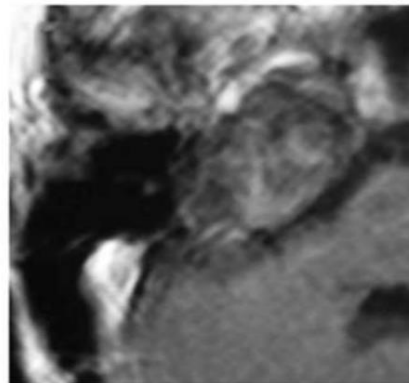
- HYPER-intense to brain (Restricted diffusion).
- Unlike cholesterol granulomas.



(c)



(d)



- Treatment:

○ **Surgical excision**

Petrous Apex Mucocele:

- Uncommon.
- Resulted from post-inflammatory obstruction of a pneumatized petrous apex air cells.

- Imaging:

- **CT scan with contrast (Similar to Cholesterol granuloma):**

- Smooth, round and expansive hypodense lesion.
 - NO enhancement with contrast.
 - Highly pneumatized contralateral petrous apex.

- **MRI with Gad:**

- **T1:**

- HYPO-intense to brain.
 - Unlike cholesterol granulomas.

- **T1 C+:**

- NO enhancement

- **T2:**

- HYPER-intense to brain.

- **Diffusion-weighted Imaging (DWI):**

- HYPO-intense to brain (NO restricted diffusion).
 - Unlike epidermoids.

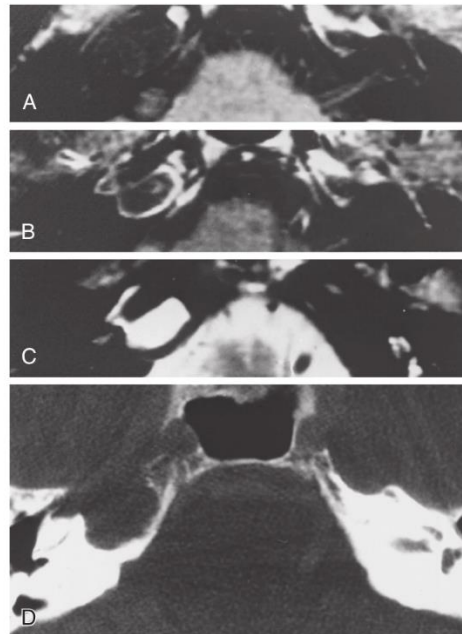


FIGURE 177-17. A to C, Axial magnetic resonance images of a petrous apex mucocele: T1-weighted without gadolinium (**A**), T1-weighted with gadolinium (**B**), and T2-weighted (**C**). The lesion is hypointense on the T1-weighted image and shows only rim enhancement with the addition of gadolinium. Note the hyperintensity of the mucocele on the T2-weighted image. **D,** Axial computed tomography scan of the same lesion. Note that the expansile lesion has eroded the petrous apex.

- Treatment:

- **Observation:**

- For asymptomatic lesions.

- **Drainage:**

- For symptomatic lesions.

Petrous Apicitis:

- Uncommon.
- Infectious process caused by medial extension of acute otitis media into a pneumatized petrous apex.
- Imaging:
 - **CT scan with contrast:**
 - Irregular destruction of petrous apex.
 - No expansion.
 - **MRI with Gad:**
 - **T1:**
 - HYPO-intense to brain.
 - **T1 C+:**
 - **Enhancement**
 - **T2:**
 - HYPER-intense to brain.

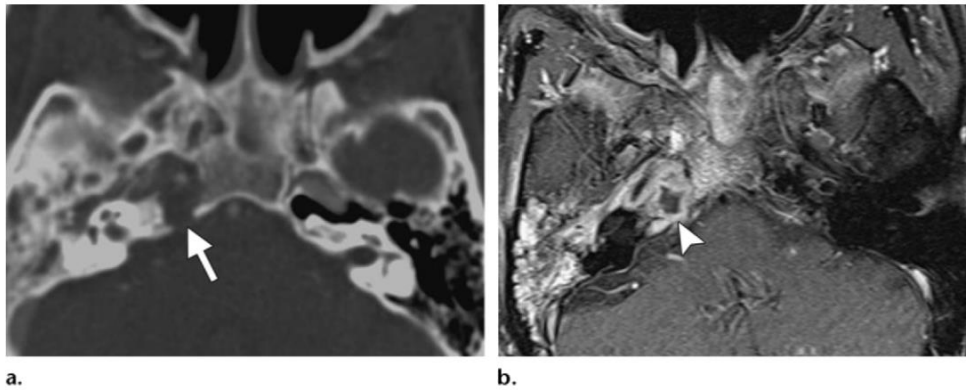


Figure 7. Petrous apicitis. **(a)** Axial contrast-enhanced CT image shows opacification of the right mastoid, middle ear, and anterior petrous air cells. There is cortical erosion along the posterior margin of the petrous apex (arrow). Note the pneumatization of the left petrous apex. **(b)** Gadolinium-enhanced fat-suppressed T1-weighted MR image shows diffuse enhancement throughout the right mastoid and middle ear, anterior petrous apex, and clivus. There is also a focal rim-enhancing fluid collection in the anterior petrous apex (arrowhead), a finding consistent with an abscess.

- Treatment:
 - **Medical management:**
 - **Surgical drainage:**
 - If not responding to medical management.

Asymmetric Petrous Apex:

- Normal variant.
- Petrous apex pneumatization is asymmetric in up to 10% of patients.
- HYPER-intense fatty marrow may be mistaken for a **cholesterol granuloma** on T1-weighted images.
- Differentiated from cholesterol granuloma by:
 - o Normal trabeculated petrous apex on CT.
 - o Lack of mass effect.
 - o HYPO-intense on fat suppression MRI.

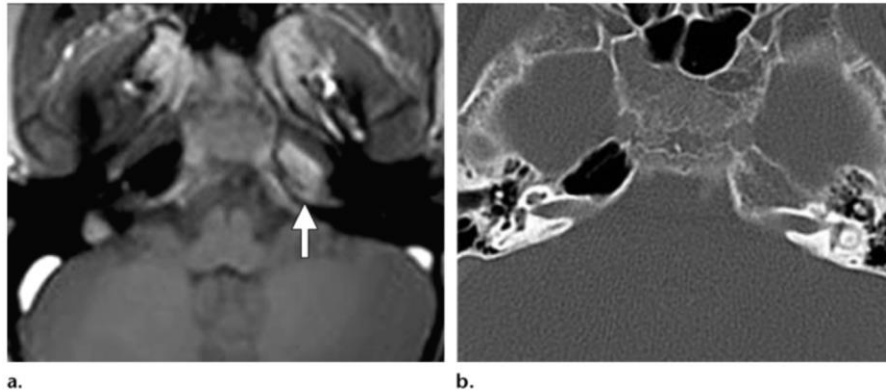


Figure 23. Asymmetric petrous apex pneumatization mimicking a lesion. (a) Axial unenhanced T1-weighted MR image shows asymmetric high signal intensity in the left anterior petrous apex (arrow), a finding that might be mistaken for a cholesterol granuloma or soft-tissue mass. (b) Corresponding CT image shows that the right petrous apex is pneumatized. Therefore, it is evident that the high signal intensity in the left petrous apex in a simply represents normal fatty marrow in nonpneumatized bone. Note the normal bone trabeculae in the left anterior petrous apex.

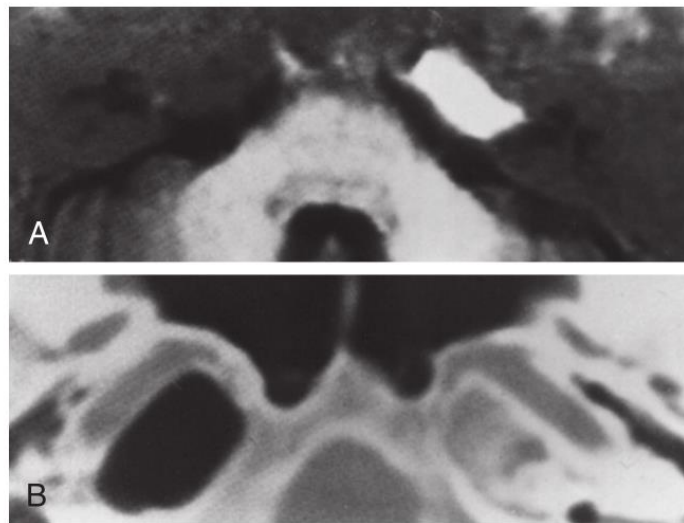


FIGURE 177-15. Asymmetric pneumatization of the petrous apex demonstrated on an axial T1-weighted image (A) and an axial computed tomography scan (B). Note that the area of hyperintensity on the T1-weighted image could be confused with a neoplasm. The computed tomography scan reveals extensive pneumatization on the contralateral side and limited pneumatization on the side with hyperintense signal on magnetic resonance imaging.

Petrous Apex Effusion:

- As in the mastoids, effusions can develop within pneumatized petrous apex cells and can occasionally mimic masses.
- Differentiated from masses by:
 - Normal trabeculated petrous apex on CT.
 - Lack of mass effect.
 - T1: HYPO-intense (unlike cholesterol granulomas).
 - T1 C+: No enhancement.
 - T2: HYPER-intense (similar to CSF intensity).

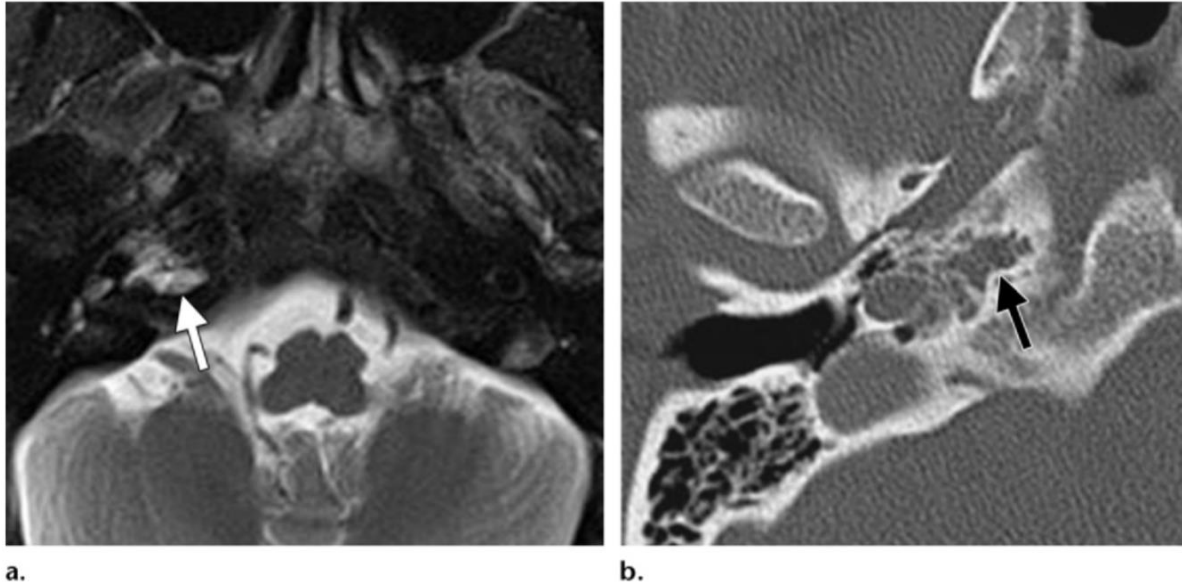


Figure 24. Petrous apex effusion. **(a)** Axial T2-weighted MR image shows asymmetric high signal intensity in the right anterior petrous apex (arrow). **(b)** Axial CT image shows that the abnormality represents opacified air cells in the anterior petrous apex (arrow). The lack of osseous destruction or bone remodeling helps distinguish trapped fluid from a tumor or mucocele.

TABLE 8–5. Cystic Lesions of the Petrous Apex

Lesion	CT	Expansile	MRI T1	MRI T2	Enhancing
Mucocele	Smooth bony remodeling	Yes	Low signal	Bright	No
Petrous apicitis	Irregular bony remodeling	No	Low	Bright	Yes
Cholesteatoma	Smooth bony remodeling	Yes	Low	Bright	No
Cholesterol Granuloma	Smooth bony remodeling	Yes	Bright	Bright	No

Lesion	T1	T2	Gad	Special features
Cholesterol granuloma	↑	↑	No	Smooth expansile lesion
PBC	↓	↑	No	Smooth bone erosion
Chordoma	↓	↑	Yes	Heterogeneous
Effusion	↑	↑	Mucosal enhancement	•No bone erosion •Preserve partitions between cells •Pneumatized apex
Petrous apicitis	↓	↑	Rim enhancement	•Irregular bone erosion •Hx/O OM
Carotid aneurysm	↓	↓	Rim enhancement	Smooth bone erosion

TABLE 177-3. Imaging Features of Petrous Apex Cystic Lesions

Features	Cholesterol Granuloma	Epidermoid	Mucocele
Computed tomography density	Isodense	Hypodense	Hypodense
T1-weighted MRI appearance	Hyperintense	Hypointense	Hypointense
Gadolinium contrast MRI appearance	Nonenhancing	Nonenhancing	Rim enhances; mass is nonenhancing
T2-weighted MRI appearance	Hyperintense	Hyperintense	Hyperintense
Borders of lesion	Smooth	Scalloped	Smooth
Diffusion-weighted imaging	No restricted diffusion (hypointense)	Restricted diffusion (hyperintense)	No restricted diffusion (hypointense)

Modified from Lo WM: Tumors of the temporal bone and cerebellopontine angle. In Som PM, Bergeron RT, editors: *Head and neck imaging*, St Louis, 1991, Mosby, pp 420-445.
MRI, magnetic resonance imaging.

Table 135-1

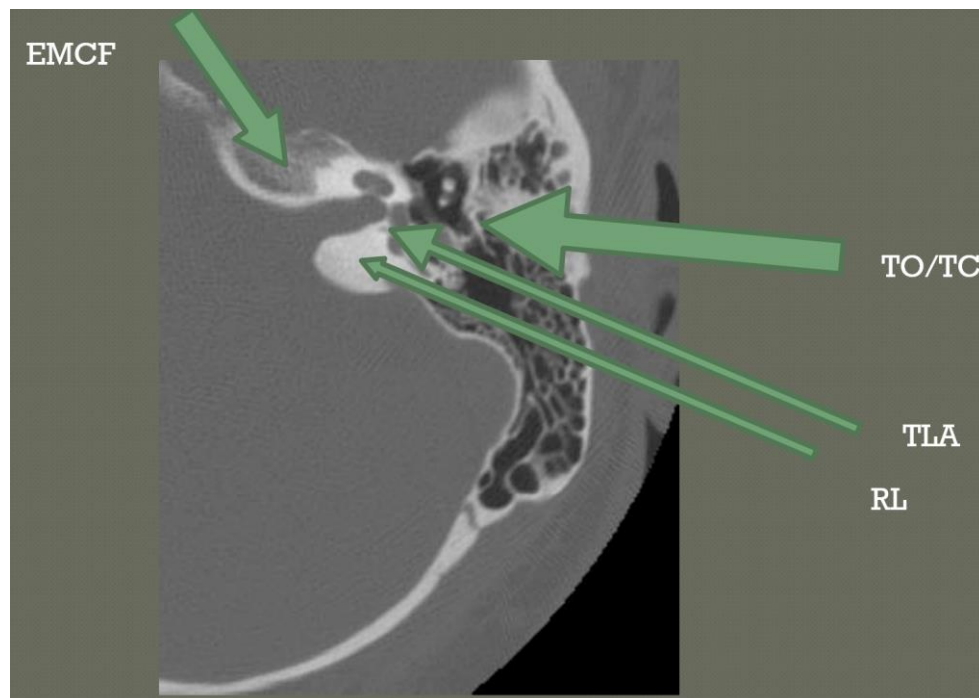
Imaging Characteristics of Petrous Apex Lesions

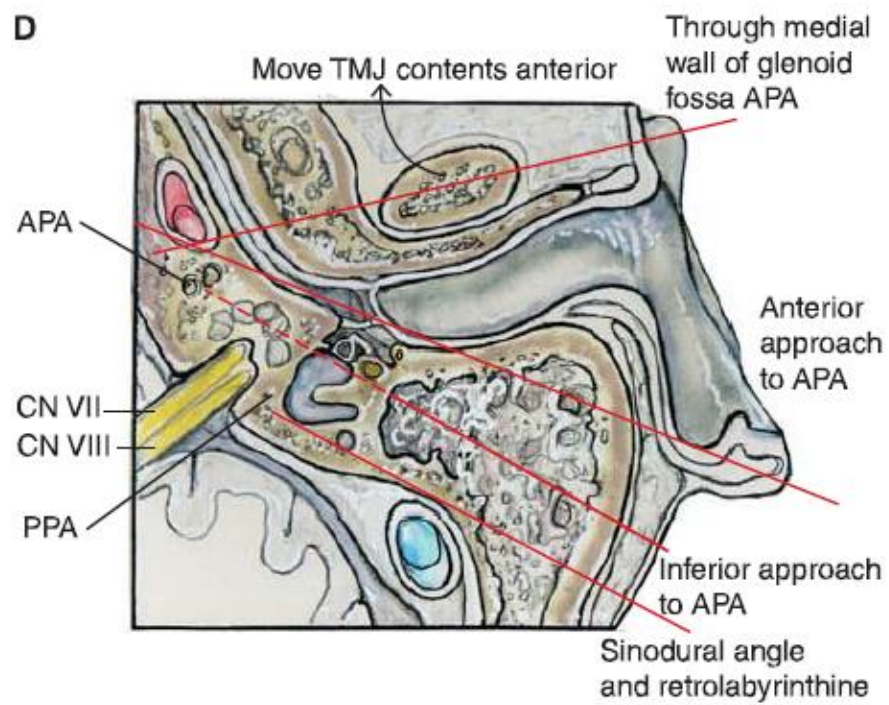
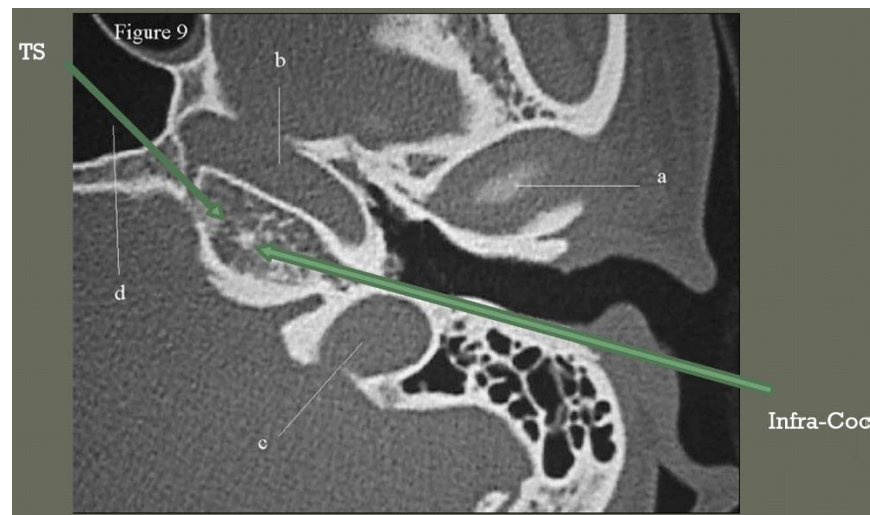
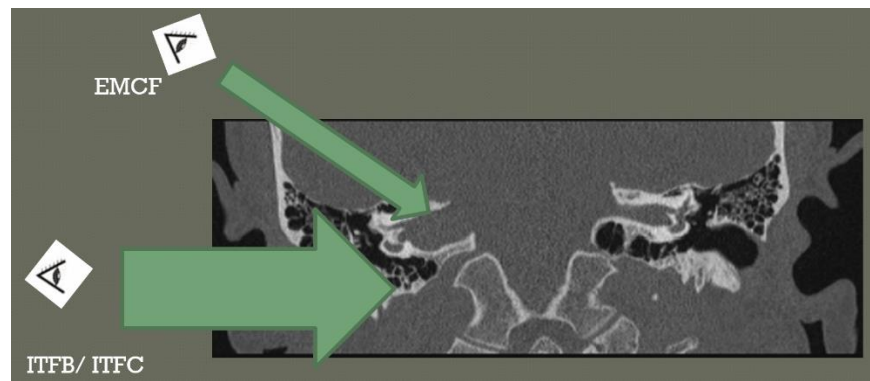
Lesion	CT	T1	T2	T1 + Contrast	Notes
Asymmetric pneumatization	Bone marrow filled, no air cells	Hyperintense to intermediate intensity	Hypointense	No enhancement	T1 signal disappears with fat suppression
Retained secretions	Air cell trabecular preservation, nonexpansile	Hypointense	Hyperintense	No enhancement	
Cholesterol granuloma	Air cell trabecular breakdown, expansile	Hyperintense	Hyperintense	No enhancement	
Mucocele	Air cell trabecular breakdown, expansile	Hypointense	Hyperintense	Rim enhancement	
Cholesteatoma	Air cell trabecular breakdown, expansile	Hypointense	Hyperintense	May have rim enhancement if granulation tissue present	DWI does not restrict (bright signal)

DWI, diffusion-weighted imaging.

Surgical Approach to Petrous Apex lesions:

- Depends on:
 - **Available air-cell pathways:**
 - Below, above, posterior and anterior to the labyrinth.
 - **Portion of petrous apex involved:**
 - Anterior or Posterior.
 - **Hearing status:**
 - Sparing the otic capsule surgically is preferred in patients with serviceable hearing.
- Surgical approaches for Posterior Petrous Apex lesions:
 - **Hearing preservation approaches:**
 - Infra-labyrinthine approach
 - Retro-labyrinthine approach
 - Subarcuate approach
 - **Non-hearing preservation approaches:**
 - Trans-labyrinthine approach.
- Surgical approaches for Anterior Petrous Apex lesions:
 - **Hearing preservation approaches:**
 - Extended middle cranial fossa approach
 - Glenoid fossa approach
 - Infra-cochlear approach
 - Endoscopic trans-sphenoid approach (for Drainage)
 - Infra-temporal approach (Fisch type A-B)
 - **Non-hearing preservation approaches:**
 - Trans-otic/Trans-cochlear approaches.





Jugular Foramen Lesions:

- Extra-dural lesions.
- Most common JF lesions:
 - o Paraganglioma (Glomus Tumor)
 - o Lower Cranial Nerves Schwannoma

Paraganglioma (Glomus Tumor):

- 2nd most common temporal bone tumor (after VS).
- Most common vascular tumor of temporal bone.
- Most common Jugular Foramen Lesion.
- Epidemiology:
 - o Peak presentation is in the 4th-6th decades.
 - o Familiar paragangliomas present earlier.
 - o More common in females (3:1).
- Histopathology:
 - o Benign tumor of neuroendocrine paraganglial chemoreceptor cells originating from neural crest cells adjacent to CN-IX and X.
 - o Contain nests of non-chromaffin-staining cells (Zellballen) septated by a prominent fibrovascular stroma.
 - o Typically non-functional tumors but capable of clinically significant catecholamine secretion (norepinephrine) in **(5%) of patients:**
 - Causing paroxysmal hypertension, headache and palpitations.

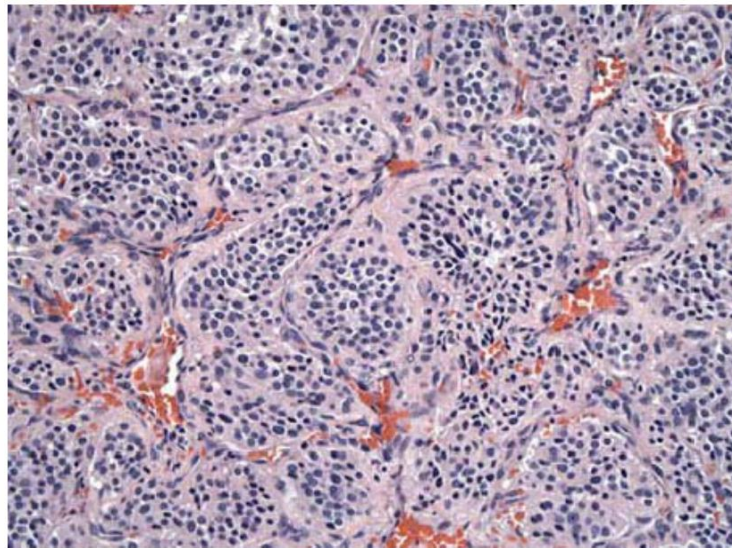


Figure 147.2 Paraganglioma: Histologic section showing tumor cells organized into nests, termed *zellballen*, separated by fibro-vascular stroma.

- Pathophysiology:
 - Slow-growing locally destructive tumors.
 - Invade neurovascular structures of skull base.
 - Follow the path of least resistance.
 - Distributed along parasympathetic nerves in skull base and neck.
 - Occur more frequently on the left side.
 - Commonly supplied by the ascending pharyngeal artery.
- **Rule of 10:**
 - **10% are Familiar:**
 - Autosomal dominant.
 - Present earlier.
 - More aggressive.
 - More commonly multifocal (50%).
 - More commonly secrete vasoactive substances.
 - **10% are Multiple.**
 - **10% in Pediatric.**
- Classification of Glomus tumors:
 - 1. Carotid Body Tumor (Glomus Caroticum):**
 - Most common type in the head and neck **(70%)**.
 - Arises from carotid body.
 - Typically does not involve the temporal bone.
 - 2. Glomus Jugulare:**
 - Most common type in the temporal bone.
 - Arises from adventitia of jugular bulb in the jugular foramen.
 - 3. Glomus Tympanicum:**
 - Arises from the promontory along the course of Jacobson's nerve (tympanic branch of CN-IX).
 - Confined to middle ear space.
 - Present early with pulsatile tinnitus and CHL.
 - 4. Glomus Vagale:**
 - Arises from paraganglia around the Vagus nerve at skull base.

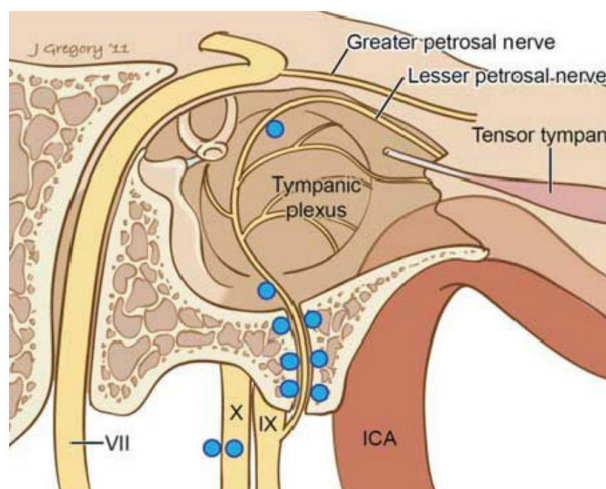


Figure 127.3 Distribution of frequent locations for glomus bodies in the temporal bone.

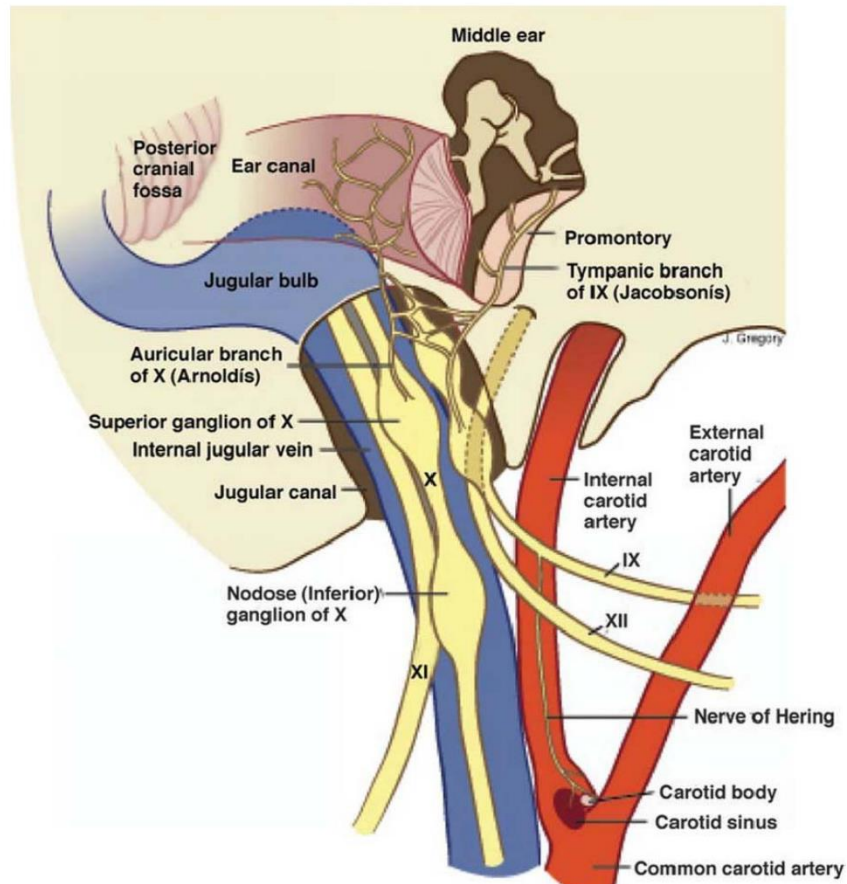


Figure 127.2 Schematic representation of lower cranial nerves at the level of jugular foramen.

- Clinical picture (Jugulotympanic Paragangliomas):
 - **Symptoms of the mass:**
 - Pulsatile tinnitus (Most common symptom).
 - Conductive hearing loss.
 - Aural fullness.
 - Dizziness.
 - Bloody otorrhea.
 - Facial weakness.
 - Multiple cranial nerve palsies (CN IV–XII):
 - Jugular foramen syndrome/Vernet syndrome: (CN IX–XI).
 - Villaret's syndrome (CN IX–XII + sympathetic chain).
 - Collet Sicard syndrome (CN IX–XII).
 - **Symptoms of secreting tumors:**
 - Labile hypertension
 - Palpitations
 - Flushing
 - Diaphoresis
 - Diarrhea

- **Signs:**
 - Reddish-blue pulsatile mass medial to the inferior TM.
 - Brown's sign:
 - Blanching of the mass with positive pressure on pneumatic otoscopy.
 - Aquino's sign:
 - Decreasing of the pulsation of the mass with ipsilateral carotid artery compression.
 - Hemorrhagic aural polyp extending in EAC.
 - Auscultation over the mastoid or infra-auricular area reveals an audible bruit.

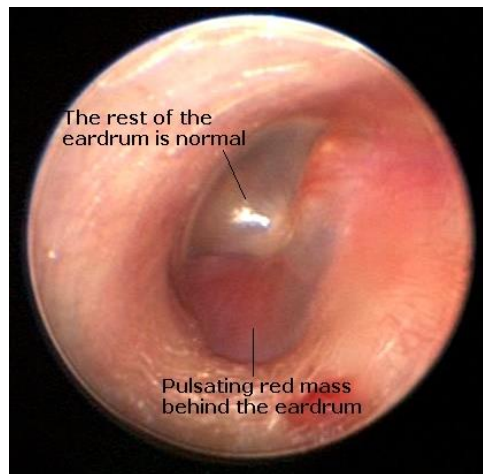


Figure 127.6 Modes of spread for jugular paragangliomas.

- Diagnosis (Jugulotympanic paragangliomas):
 - **24-hours urine catecholamine screen:**
 - For vanillylmandelic acid and metanephrine.
 - Used as screening test for actively secreting paragangliomas.
 - **CT with contrast:**
 - Marked contrast enhancement.
 - Characteristic bone destruction (Moth-eaten pattern):
 - Erosion of jugulo-tympanic spine.
 - Enlargement of jugular foramen with irregular bony destruction.
 - Involvement of middle ear and mastoid.
 - **MRI with Gad:**
 - Goals:
 - Confirm the diagnosis.
 - Identify intracranial extension and differentiate between intra-dural and extra-dural extension.
 - Detection of concurrent paragangliomas.
 - **T1:**
 - HYPO-intense to brain.
 - **T1 C+:**
 - Marked enhancement.
 - **T2:**
 - Salt and pepper appearance:
 - Pepper: High-flow signal voids of feeding arteries.
 - Salt: Subacute hemorrhage within the tumor.
 - **Angiography:**
 - Assist in the diagnosis and for pre-operative embolization.
 - **Radionuclide Imaging:**
 - Indium octreotide scanning (111 In-octreoscan):
 - 90% specificity and sensitivity in detecting head and neck paragangliomas.
 - Very effective to screen for secondary tumors.
 - Effective postoperatively to screen for recurrent disease.
 - 123 I-metaiodobenzylguanidine (MIBG):
 - Shows similar sensitivities and specificities to octreoscan.
 - 18 F-PDG-PET:
 - Best for evaluation of malignant and metastatic paragangliomas.
 - Sensitivities of 74-88%.

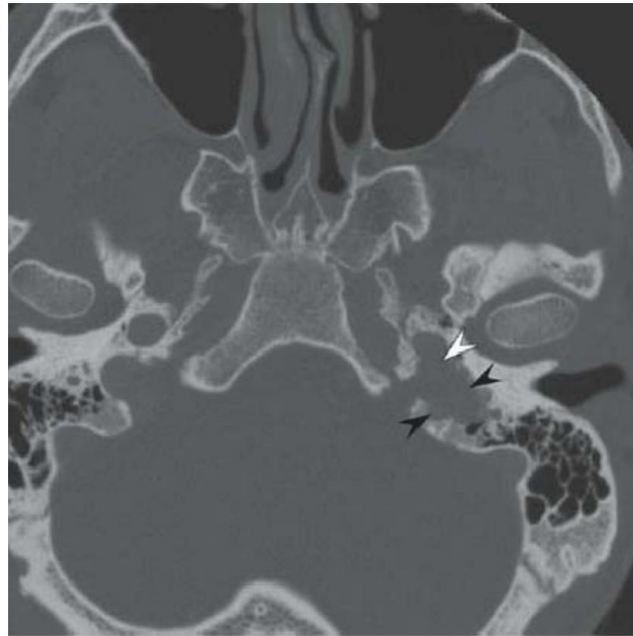


Figure 147.3 Glomus jugulare. Axial CT showing classic bony erosion of the left jugular fossa and caroticojugular spine (*black arrowheads*). The tumor abuts the posterior aspect of the vertical petrous carotid artery (*white arrowhead*).



Figure 159.19 Axial temporal bone CT image at the level of the jugular foramen demonstrating permeative bony erosion (*arrowheads*) and loss of the caroticojugular spine caused by a glomus jugulare tumor.



FIGURE 177-11. Glomus jugulare tumor demonstrated by computed tomography. **A**, Axial image shows demineralization of the bony margins of the jugular foramen on the right side (*black arrows*). Demineralization of the fine cortical line is usually demonstrated along the lateral aspect of the vascular portion of the jugular foramen. Compare with intact cortical plate on the opposite side (*white arrow*). Note the demineralization of the bony wall that separates the carotid canal from the jugular foramen. **B**, Enlarged axial image shows the demineralization of the lateral cortical plate (*arrowhead*) and slight demineralization of the bony plate between the carotid canal and the jugular foramen (*white arrow*). Note the proximity of the demineralized bone to the vertical section of the facial nerve canal (*black arrow*). **C**, Axial image enlarged through the normal side shows the intact white cortical line (*arrow*) along the lateral aspect of the jugular foramen. The integrity of this line essentially excludes a glomus jugulotympanic tumor. **D**, Coronal image shows demineralization of the margins (*arrowheads*) as well as the soft tissue of the intratympanic portion of the tumor (*arrow*). (From Maya MM, Lo WWM, Kovanlikaya I: Temporal bone tumors and cerebellopontine angle lesions. In Som PM, Curtin HD, editors: *Head and neck imaging*, ed 4, Philadelphia, 2003, Mosby.)

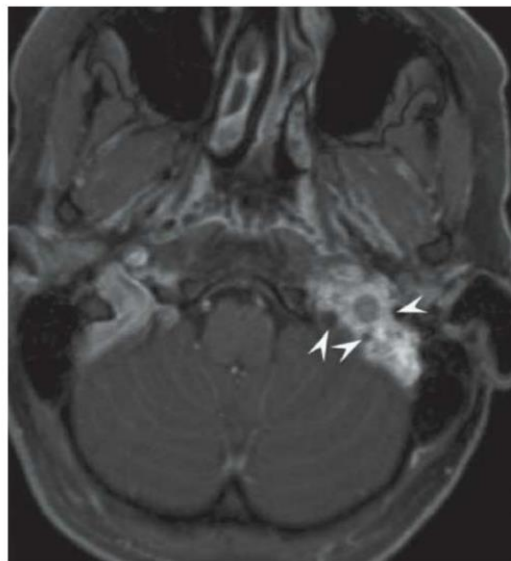


Figure 159.20 Axial T1-weighted contrast-enhanced MRI image at the level of the left jugular foramen demonstrating an enhancing glomus jugulare tumor with dark intratumoral flow voids (*arrowheads*).

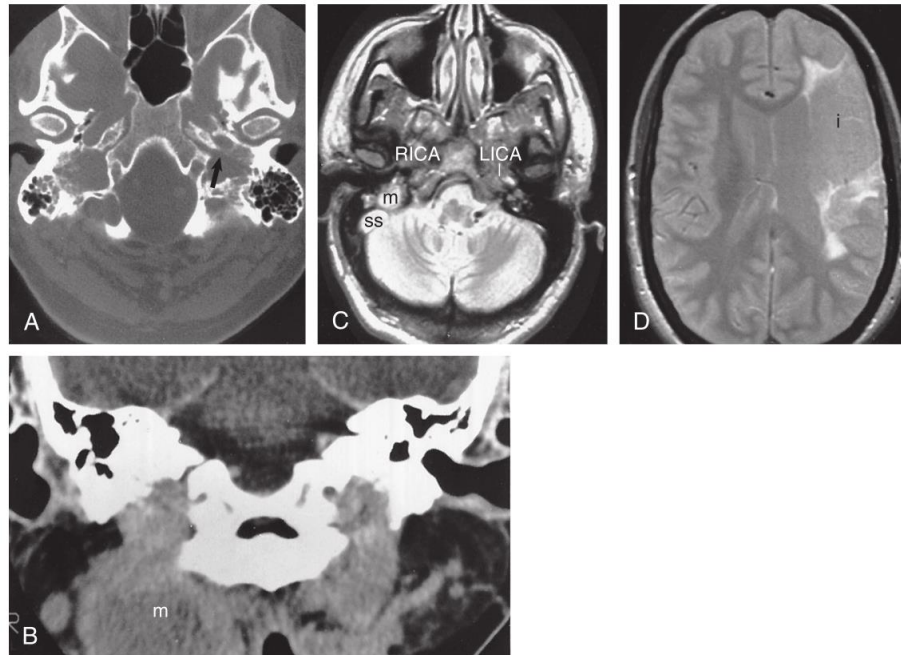


FIGURE 176-29. Familial paragangliomas in a 53-year-old woman. The tumors included bilateral glomus jugulare paragangliomas, a right-sided glomus vagale paraganglioma, and bilateral carotid body paragangliomas. **A**, Axial view, computed tomography (CT), bone algorithm. This CT study is performed with a small field of view and a bone algorithm to increase the resolution of bone edges. The bilateral glomus jugulare paragangliomas are destroying bone from the pars vasculosa laterally. On the left side, the lesion is destroying the septum between the internal carotid artery and the internal jugular vein—common findings in a glomus jugulare paraganglioma (*arrow*). **B**, Coronal view, CT with intravenous contrast. Both glomus jugulare paragangliomas extend into the neck. However, another large mass can be seen on the right, a glomus vagale paraganglioma (*m*). **C**, Axial view, T2-weighted magnetic resonance imaging (MRI). The high-signal-intensity lesion, a glomus jugulare paraganglioma (*m*), has occluded the sigmoid sinus, which shows high-intensity signal (*ss*). The left internal carotid artery (*LICA*) shows signal of pathologic high intensity. This vessel has been occluded by a large paraganglioma in the neck (*not shown*). Compare the signal intensity of this vessel with the normal dark black appearance of internal carotid artery flow on the right (*RICA*). Although MRI cannot provide the fine bony resolution of CT (see **B**), it gives more information about vascular patency. **D**, The *LICA* occlusion has resulted in a middle cerebral arterial distribution infarct (*i*).

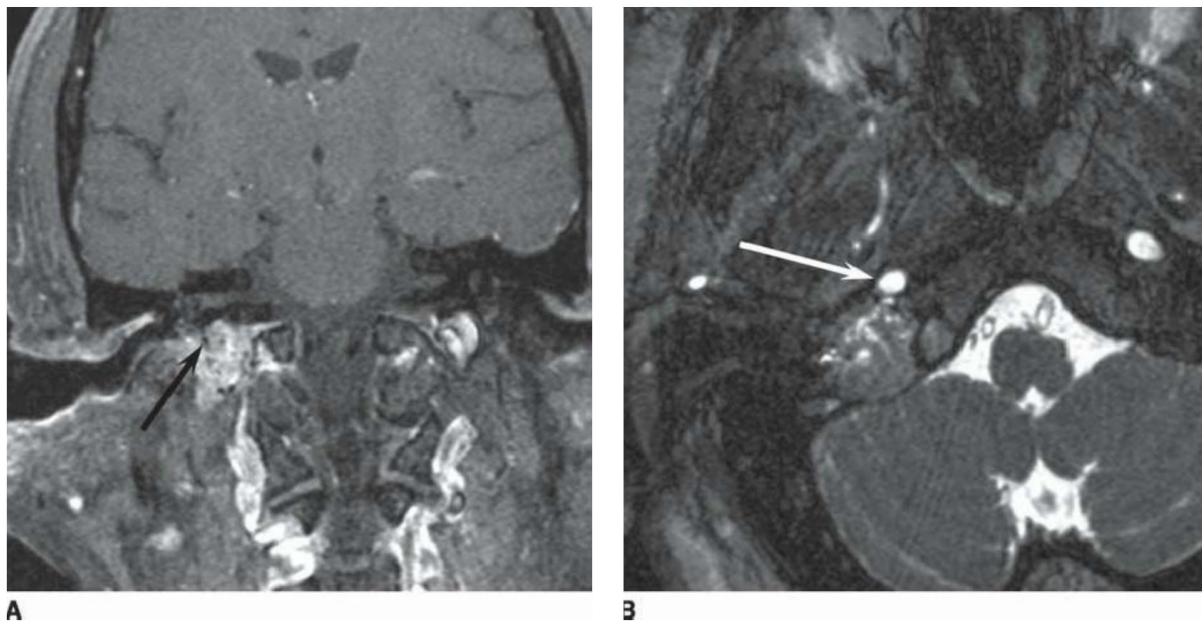


Figure 127.9 Jugular paraganglioma. **A**: Coronal enhanced fat-suppressed T1-weighted image shows avid enhancement with focal round flow voids (*arrow*) indicating large feeding vessels. **B**: Axial fat-suppressed T2-weighted image demonstrates the proximity of the tumor to the vertical portion of the petrous internal carotid artery (*arrow*) as well as the intracranial but extradural component of the tumor in the jugular foramen posteriorly.

- Staging (Jugulotympanic paragangliomas):
 - Two surgical classification systems are used pre-operatively based on the radiographic findings:
 - **Fisch classification system:**
 - Doesn't differentiate between tympanic and jugular paragangliomas.

Table 1. Classification of glomus jugularis tumors of the temporal bone as proposed by Fisch (4).

Type	Description
A	Tumors restricted to middle ear (glomus tympanicum tumors)
B	Tumors restricted to tympanomastoid site
C	Tumors involving the infra-labyrinth portion towards the petrous apex
D1	Tumor with intracranial invasion (<2 cm)
D2	Tumor with intracranial invasion (>2 cm)

**TABLE
127.3**

**FISCH CLASSIFICATION
OF JUGULOTYMPANIC
PARAGANGLIOMAS**

Class A	Tumors arising on the tympanic plexus confined to the middle ear
Class B	Tumors arising from the inferior tympanic canal in the hypotympanum with middle ear and/or mastoid invasion, but jugular bulb and carotid canal intact
Class C	Tumors arising in the dome of the jugular bulb and involving the overlying cortical bone
C1	Tumors eroding the carotid canal but not involving the carotid artery
C2	Tumors involving the vertical petrous carotid artery
C3	Tumors involving the horizontal carotid canal but not foramen lacerum
C4	Tumors involving the foramen lacerum and cavernous sinus
Class D	Tumors with intracranial extension
De1	Extradural extension of <2 cm medial dural displacement
De2	Extradural extension of >2 cm medial dural displacement
Di1	Intradural extension of <2 cm
Di2	Intradural extension of >2 cm
Di3	Neurosurgically unresectable tumors

- **Glasscock-Jackson classification system,**
 - Differentiate between tympanic and jugular paragangliomas.

TABLE 127.4	GLASSCOCK-JACKSON CLASSIFICATION OF JUGULOTYMPANIC PARAGANGLIOMAS
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Glomus tympanicum

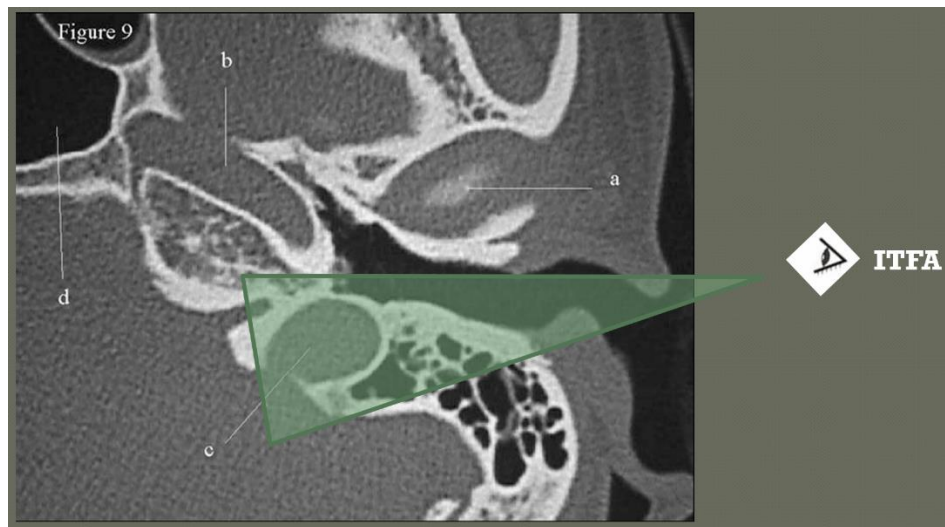
Type 1	Small mass limited to the promontory
Type 2	Tumor completely filling the middle ear
Type 3	Tumor filling the middle ear and mastoid
Type 4	Tumor completely filling the middle ear, extending into the mastoid or through the external auditory canal. May also extend anteriorly to involve the carotid artery

Glomus jugulare

Type 1	Small tumor involving the jugular bulb, middle ear, and mastoid
Type 2	Tumor extending under the internal auditory canal. May have intracranial extension
Type 3	Tumor extending to the petrous apex. May have intracranial extension
Type 4	Tumor extending beyond the petrous apex into the clivus or infratemporal fossa. May have intracranial extension.

- Treatment (Jugulotympanic paragangliomas):
 - **Microsurgery:**
 - Traditional treatment for head and neck paragangliomas.
 - Fisch infratemporal fossa approaches were developed to specifically resect temporal bone paragangliomas.
 - Pre-operative embolization is useful adjunct in large tumors.
 - Leads to tumor shrinkage and decreased intraoperative bleeding which helps in doing complete surgery.
 - Surgery should be done within 48 hours to avoid recruitment of collateral circulation.
 - Anesthesia requirements should take into consideration actively secreting tumors, which require alpha and beta adrenergic blockade.

- **Glomus Tympanicum (Fisch class A):**
 - Small tumors confined to the middle ear cavity.
 - Approached by trans-canal/inferiorly based tympanomeatal flap:
 - Embolization is not required.
 - Bipolar electrocautery microforceps can be used to shrink the tumor and resect it.
- **Jugulotympanic paragangliomas (Fisch class B):**
 - Large tumors without involvement of jugular bulb or jugulocarotid spine.
 - Approached by combined post-auricular/end-aural approach:
 - CWU mastoidectomy and extended facial recess.
 - Sacrifices chorda tympani and remove the vaginal process of tympanic bone to expose the hypotympanum.
- **Jugulotympanic paragangliomas (Fisch class C and D):**
 - Large tumors with involvement of jugular bulb or intracranial extension.
 - Approached by Fisch Type A infratemporal fossa approach:
 - Extended mastoidectomy with removal of EAC, TM, malleus and incus.
 - Carotid artery is skeletonized.
 - Tumor is removed from carotid, middle ear and sigmoid/jugular bulb/jugular vein.
 - ET tube should be permanently occluded to avoid postoperative CSF rhinorrhea.
 - Intracranial extension is addressed by:
 - Opening the dura in pre-sigmoid area down to tumor extension at the level of jugular bulb.
 - Petrous apex extension is addressed by:
 - Trans-labyrinthine/Trans-cochlear approach.



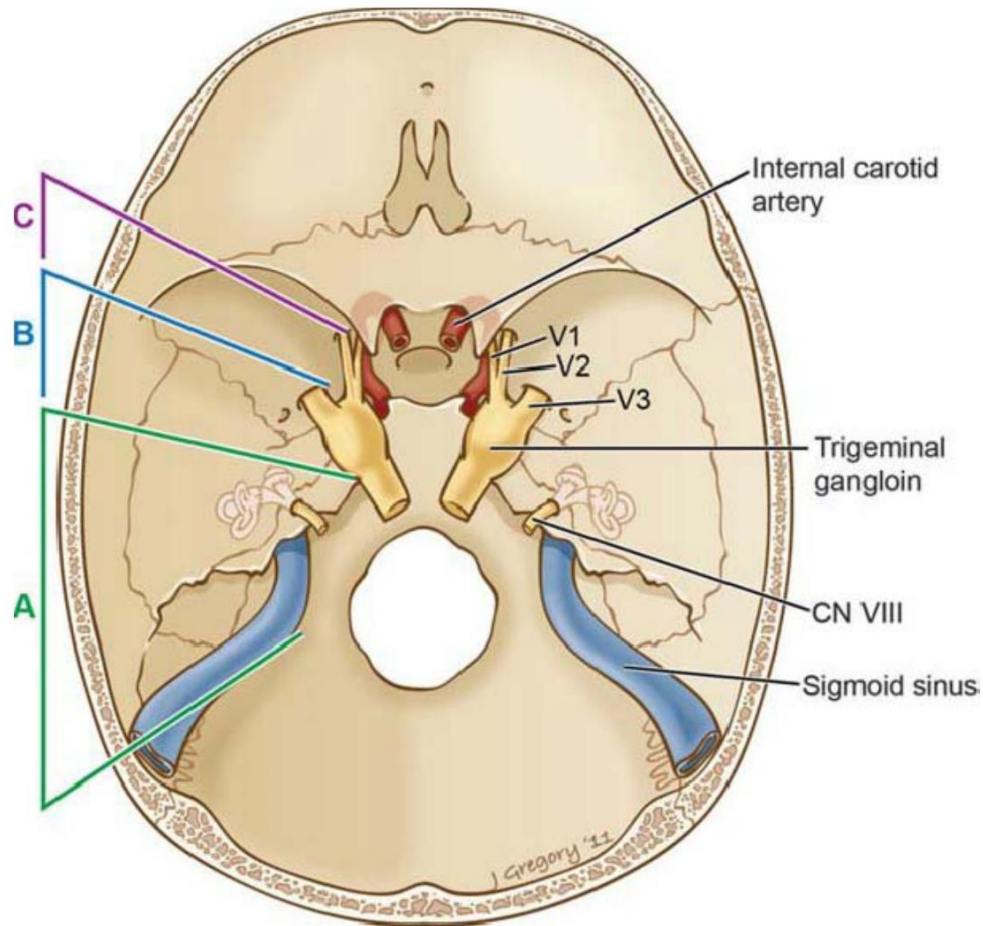


Figure 127.12 Fisch classification of infratemporal fossa approaches. Type A, exposure to the level of the anterior eustachian tube, middle meningeal artery/foramen spinosum. Type B, exposure to the foramen ovale, V3. Type C, exposure to the foramen rotundum, V2.

- **Fisch Type A:**
 - Allows access to lesion of jugular foramen and anterior petrous apex.
 - Indications:
 - Types C and D glomus jugulare tumors.
 - Petrous apex epidermoids.
- **Fisch Type B:**
 - Allows access for lesions of anterior petrous apex and mid-clivus.
- **Fisch Type C:**
 - Allows access for extensive lesions involving eustachian tube, clivus, and parasellar region.
 - Anterior extension of type B, in which the pterygoid process is drilled.

- **Radiotherapy:**
 - Not modality of choice for treatment of surgically resectable glomus tumors of temporal bone.
 - Indicated mainly for:
 - Recurrences.
 - Un-operable patients.
 - Un-resectable lesions.

**TABLE
159.2**
CPA LESION RADIOLOGY SUMMARY

Lesion	T1 (Relative to Brain)	T2 (Relative to Brain)	Contrast Enhancement	CT	Other Findings
Cranial nerve schwannoma	Isointense	Isointense, with foci of high intensity	Strong, with cystic areas	• Smooth expansion of bone (IAC, jugular foramen, fallopian canal)	Fundal cap—VS Ice cream cone—VS
Meningioma	Isointense	Isointense	Strong	• Calcifications • Hyperostosis	
Paraganglioma	Isointense	Isointense	Strong	• Permeative erosion of bone • Erosion of caroticojugular spine	Salt and pepper appearance
Endolymphatic sac tumors	Hyperintense	Hyperintense	Strong	• Extensive bony destruction	Characteristic location
Metastases	Variable	Hyperintense	Strong, but heterogeneous	• Irregular bony destruction	Melanoma—T1 hyperintensity
Chordoma	Hypointense	Hyperintense	Moderate	• Central skull base destruction	Originate at the clivus
Chondrosarcoma	Hypointense	Hyperintense	Weak	• Irregular bony destruction • Calcified central matrix	Occur along petroclival suture
Epidermoid cyst (cholesteatoma)	Hypointense	Hyperintense	None	• Erosion of bone	Bright on T2 FLAIR
Dermoid cyst	Hyperintense	Hyperintense	None	• Erosion of Bone • Calcification	Bright on DWI Bright on T2 FLAIR
Arachnoid cyst	Hypointense	Hyperintense	None	• Sculpted bone	Dark on T2 FLAIR
Lipoma	Hyperintense	Hypointense	None	• Lower density than fluid	Suppresses with fat sat
Cholesterol granuloma	Hyperintense	Hyperintense	None	• Smooth expansion of bone	

Temporal Bone Trauma:

- Epidemiology:

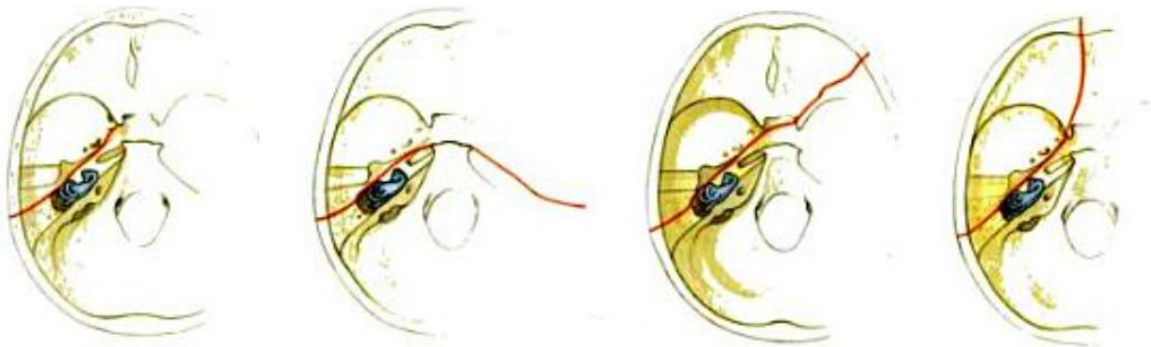
- Associated with severe traumatic forces.
- Most commonly due to motor vehicle accidents.
- Occurs mainly in men (3:1).
- Mainly during 2nd to 4th decades of life.
- Most temporal bone fractures are Unilateral.
- Bilateral TB fractures in 20% of cases.

- Pathophysiology:

- Result from either posterior or lateral blows to the head.
- Takes the path of least resistance along the structurally weakened points.

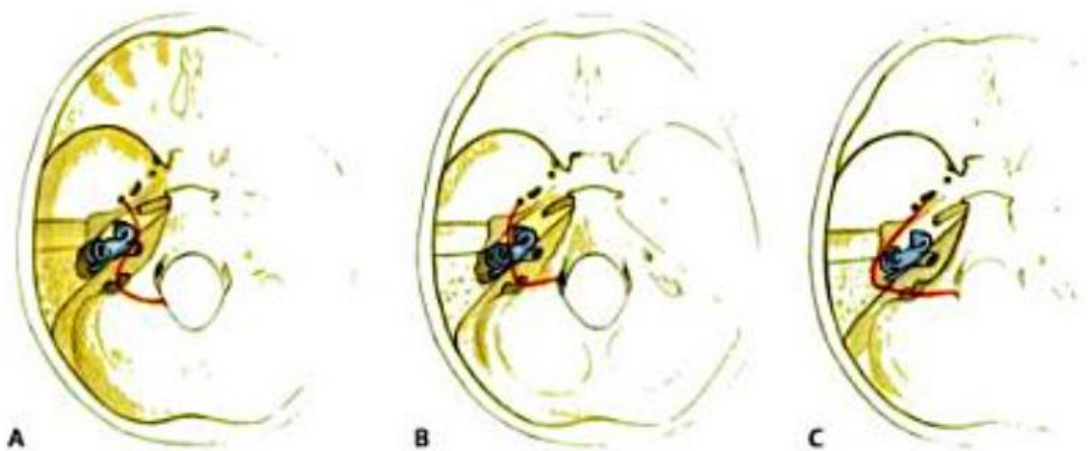
▪ **Fractures resulting from lateral (temporal) blows:**

- Involve squamous and tympanic portions of TB.
- Starting-point:
 - Originate through cortical areas of weakness (EAC, mastoid air cells).
- Course:
 - Propagate medially to pass through the middle ear.
 - Parallel to the long axis of the petrous bone (longitudinal).
 - Deflected anteriorly by otic capsule to reach foramen lacerum.
 - Inner ear and fallopian canal are spared.
- Ending-point:
 - Most fractures terminate in floor of MCF or in sphenoid bone.
 - One third extend across midline to join a contralateral fracture.
 - Minority extend anteriorly to exit skull through:
 - Anterior cranial fossa floor laterally.
 - Midline through the cribriform plate.



▪ **Fractures resulting from posterior (occipital) blows:**

- Involve mastoid and petrous portions of TB.
- Starting-point:
 - Cortical origin of the fracture is generally in the occipital bone.
- Course:
 - Extends to foramen magnum and disrupts the foramen magnum ring.
 - Propagate anteriorly across petrous bone.
 - At right angles to the long axis of petrous bone (transvers).
 - May pass lateral to, through or medial to the otic capsule:
 - Most common path is directly through otic capsule, disrupting inner ear.
 - Rarely, the fracture spares otic capsule by passing medial to it, which puts CN-VII and VIII at risk as the fracture traverse the IAC.
 - Also rare, the fracture spares otic capsule by passing lateral to it through the middle ear.
- Ending-point:
 - Terminates in floor of MCF.



Traditional Classification of TB Fractures:

- Based on their anatomic relationship to the long axis of petrous bone.
- Divided into:
 - 1. Longitudinal Fractures (80%)**
 - 2. Transverse Fractures (20%)**
- Disadvantages:
 - Most fractures are mixed type and follow mixed planes.
 - Does not predict neur-otologic complications.
 - Does not help with selecting surgical approach when necessary.

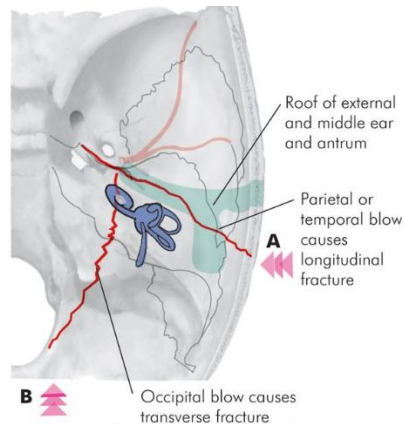


Table 14-2. Differences in longitudinal and transverse fractures of temporal bone

	Longitudinal	Transverse
• Frequency	More common (80%)	Less common (20%)
• Type of injury	Parietal blow	Occipital blow
• Fracture line	Runs parallel to long axis of petrous pyramid. Starts at squamous part of temporal bone to end at foramen lacerum	Runs across the petrous Starts at foramen magnum or jugular foramen towards the foramen spinosum
• Bleeding from ear	Common, due to injury to tegmen and tympanic membrane	Absent because tympanic membrane is intact. Haemotympanum may be seen
• C.S.F. otorrhoea	Present, often mixed with blood	Absent or unmanifested
• Structures injured	Tegmen, ossicles and tympanic membrane	Labyrinth or CN VIII
• Hearing loss	Conductive	Sensorineural
• Vertigo	Less often; due to concussion	Severe, due to injury to labyrinth or CN VIII
• Facial paralysis	Less (20%), delayed onset. Nerve is injured in tympanic segment, distal to geniculate ganglion	Most common (50%). Immediate onset. Injury to nerve in meatal or labyrinthine segment proximal to geniculate ganglion.

Characteristic	Longitudinal Fractures	Transverse Fractures
Prevalence	75%	25%
Fracture Line	parallel to long axis of petrous pyramid	perpendicular to long axis of petrous pyramid
Vector Force	temporoparietal blow	frontal or occipital blow
Relation to Otic Capsule	fracture remains anteriomedial to otic capsule	fracture may transect otic capsule and IAC
Hearing Loss Type	CHL (ossicular disruption, blood in the EAC)	SNHL (labyrinthine or nerve injury)
Signs and Symptoms	blood in EAC, Battle's sign, EAC step-off, rare vestibular signs, hemotympanum	hemotympanum, nystagmus, CSF otorrhea
Facial Nerve Injury	less likely	more likely

* Pure longitudinal and transverse fractures are **not common**, most fractures are **mixed types** and follow mixed planes.

New Classification of TB Fractures:

- Based on their involvement to the otic capsule.
- Divided into:
 - 1. Otic Capsule Sparing Fractures (95%)**
 - 2. Otic Capsule Disrupting Fractures (5%)**
- Advantages:
 - Superior prediction of neur-otologic complications.
 - Helps with selecting surgical approach when necessary.

Feature	Otic Capsule Sparing	Otic Capsule Disrupting
Incidence	Approximately 95%	Approximately 5%
Mechanism	Temporal or parietal trauma	Occipital trauma
Line of fracture	Anterolateral to the otic capsule	Through the otic capsule
Pathway	▪ Squamosa portion of temporal bone ▪ Posterosuperior wall of the external auditory canal and tympanic membrane commonly involved ▪ Also, mastoid air cells and middle ear	▪ Foramen magnum, petrous pyramid, and otic capsule ▪ Also jugular foramen, internal auditory canal, and foramen lacerum ▪ Tympanic membrane and external auditory canal not usually affected
CSF leak	Middle cranial fossa (tegmen mastoideum, tegmen tympani, middle ear, and external auditory canal or eustachian tube)	Posterior cranial fossa (middle ear, eustachian tube) 4-fold higher
Ossicular chain involvement	Common	Rare
Hearing loss	Conductive or mixed	Sensorineural Almost always
Facial paralysis	Less common 6% to 14%	Common 30% to 50%



FIGURE 145-2. Axial-cut high-resolution computed tomography scan demonstrating a longitudinally oriented fracture that has spared the otic capsule. Arrows point along the fracture line.

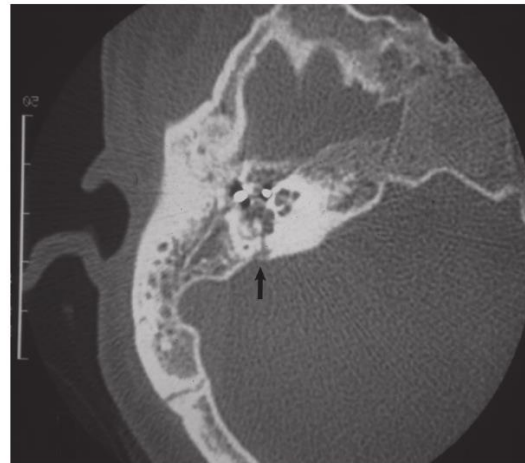


FIGURE 145-3. Axial-cut high-resolution computed tomography scan demonstrating a transverse-oriented fracture that resulted from a gunshot injury and disrupted the otic capsule. The arrow points to the fracture line.

- **Evaluation of Temporal bone fracture:**
- Frequently associated with other life-threatening injuries.
- Initial evaluation must follow ATLS/ACLS guidelines.
- **Focused History and Physical Examination:**
 - Cause:
 - Blunt vs. Penetrating.
 - Lateral vs Occipital.
 - Assess Facial nerve:
 - Partial vs. Complete.
 - Immediate vs. Delayed
 - Hearing loss:
 - Tuning forks if stable.
 - Dizziness:
 - Nystagmus.
 - Peripheral or central.
 - BPPV most common in trauma.
 - **Fistula test is not performed in presence of CSF leak.**
 - Incidence of vertigo does not correlate with severity of trauma
 - Most posttraumatic vertigo resolves spontaneously
 - Presence of other neurological deficits.
 - Presence of cranial nerve palsies.
 - Otoscope examination:
 - Auricle or EAC lacerations
 - TM perforation.
 - CSF otorrhea.
 - Hemotympanum.
 - **Battle's sign:**
 - Ecchymosis over Mastoid process.
 - Indicates fracture of middle cranial fossa.

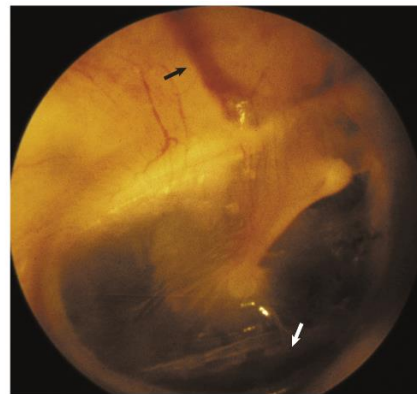


FIGURE 145-6. Otoscopic image demonstrating a nondisplaced fracture along the scutum (black arrow). Blood is layering out inferiorly (white arrow).

- **High-Resolution CT of Temporal Bone:**

- Imaging of choice for temporal bone fractures.
- Fractures are best seen on axial imaging of thin cuts HRCT.
- Evaluates:
 - Site and type of the fracture.
 - Facial nerve involvement.
 - Otic capsule involvement.
- Trauma patients usually underwent CT Brain to assess for intracranial hemorrhage.
- **Indications of requesting HRCT of TB:**
 1. Facial paralysis
 2. CSF fistula
 3. Disruption of superior wall of EAC or scutum.
 4. Suspected vascular injury.
 5. Before any surgical intervention to manage otologic complication.
- **Indications of requesting CT Angiography:**
 1. Neurologic deficits.
 2. Displaced fracture through carotid canal.

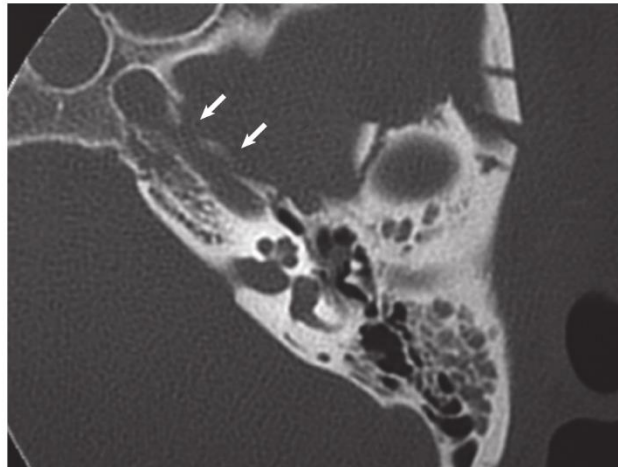


FIGURE 145-10. Axial-cut high-resolution computed tomography scan demonstrating a fracture along the carotid canal. Arrows point to the fractured carotid canal.

- **Audiogram:**

- **Indications:**
 1. After 4-8 weeks of injury to allow for spontaneous resolution of HL.
 2. Prior to any surgical intervention.

Complications of TB fractures and their Management:

**TABLE
150.1**

INCIDENCE OF COMMON COMPLICATIONS OF TEMPORAL BONE FRACTURES IN THE GENERAL AND PEDIATRIC POPULATIONS

Complication	General (%)	Pediatric (%)
Facial nerve injury	7	6
CSF fistula	17	28
Meningitis	2	0.7
Hearing loss	24	33
Conductive HL	21	43
Sensorineural HL	57	52
Mixed HL	22	5

1. External Ear Injuries:

- Auricle:
 - Lacerations are closed after cleaning & debridement.
 - Drain hematomas.
 - Pressure bolsters are sutured in place to close dead space.
- EAC:
 - Better examined with microscope after stabilization.
 - Examined aseptically (gloves & clean instruments).
 - Inspected for:
 - Lacerations
 - Blood/CSF otorrhea
 - Brain herniation.
 - **EAC debridement with suction (irrigation is contraindicated).**
 - Packing should not be inserted unless required to control EAC bleeding:
 - If profuse bleeding, control should be done at operating room or angiography suite.
 - Severely traumatized EAC:
 - Needs close follow-up with meticulous debridement to prevent EAC stenosis and external canal cholesteatoma formation.
 - If progressive stenosis is started, stenting with wicks or merocell sponges serially for 3-6 months.
 - Canalplasty is done if complete EAC stenosis.
- TM:
 - More common with longitudinal fractures.
 - Small traumatic perforations generally heal spontaneously.
 - Tympanoplasty for persistent perforations.

2. Hearing Loss:

- Conductive HL:

- More common with otic-sparing fractures.
- Causes:
 - TM perforation.
 - Blood or CSF in middle ear.
 - Ossicular chain disruption (20%):
 - Incudostapedial subluxation (80%)
 - Incus dislocation (55%)
 - Stapes crura fracture (30%)
 - Ossicular fixation in epitympanum (25%)
- Treatment:
 - 80% of CHL resolve without intervention.
 - Residual CHL following resolution of hemotympanum and healing of TM, suggests ossicular discontinuity.
 - **Exploration and Ossiculoplasty:**
 - Indication:
 - CHL >30 dB that persists for >2 months in the absence of fluid or blood.
 - Contraindications:
 - Active infection.
 - Only hearing ear.
 - Mixed HL:
 - If BC threshold > 30 dB worse than the contralateral ear.
 - Even with excellent closure of ABG, ossiculoplasty will provide minimal subjective improvement.
 - Patient would still require HA in the surgical ear.
 - Ossiculoplasty in this condition can be done only to enhance the HA post-op.
 - Outcome:
 - Superior results compared to CSOM patients.
 - Closure of ABG to 10 dB in 80% of patients.
 - Complete closure of ABG in 45% of patients.

- SNHL:

- More common with otic-capsule disrupting fractures.
- Result in severe to profound SNHL.
- Mechanism:
 - Disruption of membranous labyrinth.
 - Injury to CN-VIII.
 - Interruption of cochlear blood supply.
 - Hemorrhage into cochlea.
 - Perilymphatic fistulae.
- Outcome:
 - Patients with moderate-severe SNHL have some recovery.
 - Extremely poor prognosis for profound deafness.

3. Facial Nerve Injury:

- Pathophysiology:
 - Injury of facial nerve in TB fractures can result from:
 - Compression from bone fragments.
 - Intra-neural hematomas
 - Entrapment from compression and loss of continuity.
- Risk of FN paralysis:
 - Up to 15% in otic-capsule sparing fractures.
 - Up to 50% in otic-capsule disrupting fractures.
 - Due to perpendicular path of fracture with respect to Facial nerve.
- Sites of injury:
 - Most common site of facial nerve injury in temporal bone is in peri-geniculate region (distal labyrinthine segment and geniculate ganglion), due to:
 - Small size and lack of fibrous supporting tissue.
 - Traction between GSPN and geniculate ganglion.
 - Watershed areas of vascularization.
- Types of onset:
 - **Immediate-onset (30%):**
 - Evaluated in ER upon admission and before administration of muscle relaxants.
 - Unknown-onset:
 - Occurs when the patient is intubated before examination of facial function.
 - Painful stimuli will elicit grimace.
 - Should be considered and treated as immediate-onset facial paralysis.
 - Associated with poor prognosis.
 - **Delayed-onset (70%):**
 - Initial normal facial motion followed by deterioration.
 - Latency of facial palsy onset ranges from 1-16 days.
 - Secondary to post-traumatic edema and ischemia.
 - Associated with complete recovery (95%).
 - **Not an indication for surgical exploration and decompression of facial nerve.**

- Electro-diagnostic Tests:
 - Indication:
 - **Complete immediate-onset facial nerve paralysis.**
 - Highest risk for crushed, partially severed and transected nerves which requires surgical decompression.
 - Tests:
 - **ElectroNeuronography (ENoG):**
 - Most accurate electrodiagnostic test for prognostic information.
 - Useful only from 3-21 days after onset of injury.
 - Required to determine timing and necessity of surgical intervention.
 - Interpretation:
 - **≥ 90% degeneration within indicates 50% chance of poor recovery.**
 - < 90% degeneration indicates excellent recovery.
 - **ElectroMyography (EMG):**
 - Useful only after 14 days after onset of injury.
 - Interpretation:
 - **Voluntary action potentials:**
 - Indicates at least partial continuity of nerve.
 - Very high probability of good recovery.
 - **Fibrillation potentials:**
 - Confirm the presence of degenerating motor units.
 - **Poly-phasic potentials:**
 - Indicates re-innervation.
 - May be seen as early as 4-6 weeks after the onset of paralysis.
 - Precedes clinical recovery and predicts a fair to good recovery.
- Prognosis of Facial nerve paralysis:
 - Most important predictive factor for recovery is onset of paralysis:
 - Immediate-onset is associated with poor prognosis.
 - Delayed-onset is associated with full recovery.
 - Poorer prognosis for recovery is associated with:
 1. Immediate onset.
 2. Complete paralysis.
 3. Penetrating mechanism.
 4. Associated infection.

- Management of Facial Nerve Paralysis in TB Fractures:
 - **Conservative:**
 - 2 weeks course of steroids.
 - 60% of patients recover spontaneously within 1 month.
 - 90% of patients recover spontaneously within 3 month.
 - Indications:
 - 1. Delayed-onset:**
 - Any degree of facial nerve paralysis.
 - 2. Immediate-onset:**
 - Incomplete facial nerve paralysis.
 - Complete facial nerve paralysis with ENoG of < 90% degeneration within 14 days.
 - **Surgical Decompression:**
 - Most effective if performed ≤ 14 days of onset.
 - Indications:
 - 1. Immediate-onset:**
 - Complete facial nerve paralysis with ENoG of $\geq 90\%$ degeneration within 14 days.
 - Approaches:
 - Hearing preservation approach (-ve SNHL):
 - Combined trans-mastoid/middle cranial fossa approach.
 - Trans-mastoid/supra-labyrinthine approach.
 - Non hearing preservation approach (+ve SNHL):
 - Trans-labyrinthine total facial nerve decompression.
 - General principles:
 - Nerve is fully exposed.
 - Identify all injured segments.
 - Remove any compression from fracture fragments.
 - Nerve sheath incised and any hematomas must be evacuated.
 - If complete transection, a direct end-to-end anastomosis should be performed
 - Nerve endings should be prepared by sharply cutting at 90 degree angle and re-approximating under no tension with two to three 9.0 nylon sutures through the epineurium.
 - When a direct end-to-end anastomosis creates tension, or when segments of the nerve are missing or severely damaged, inter-positional grafts from greater auricular or sural nerve should be used.

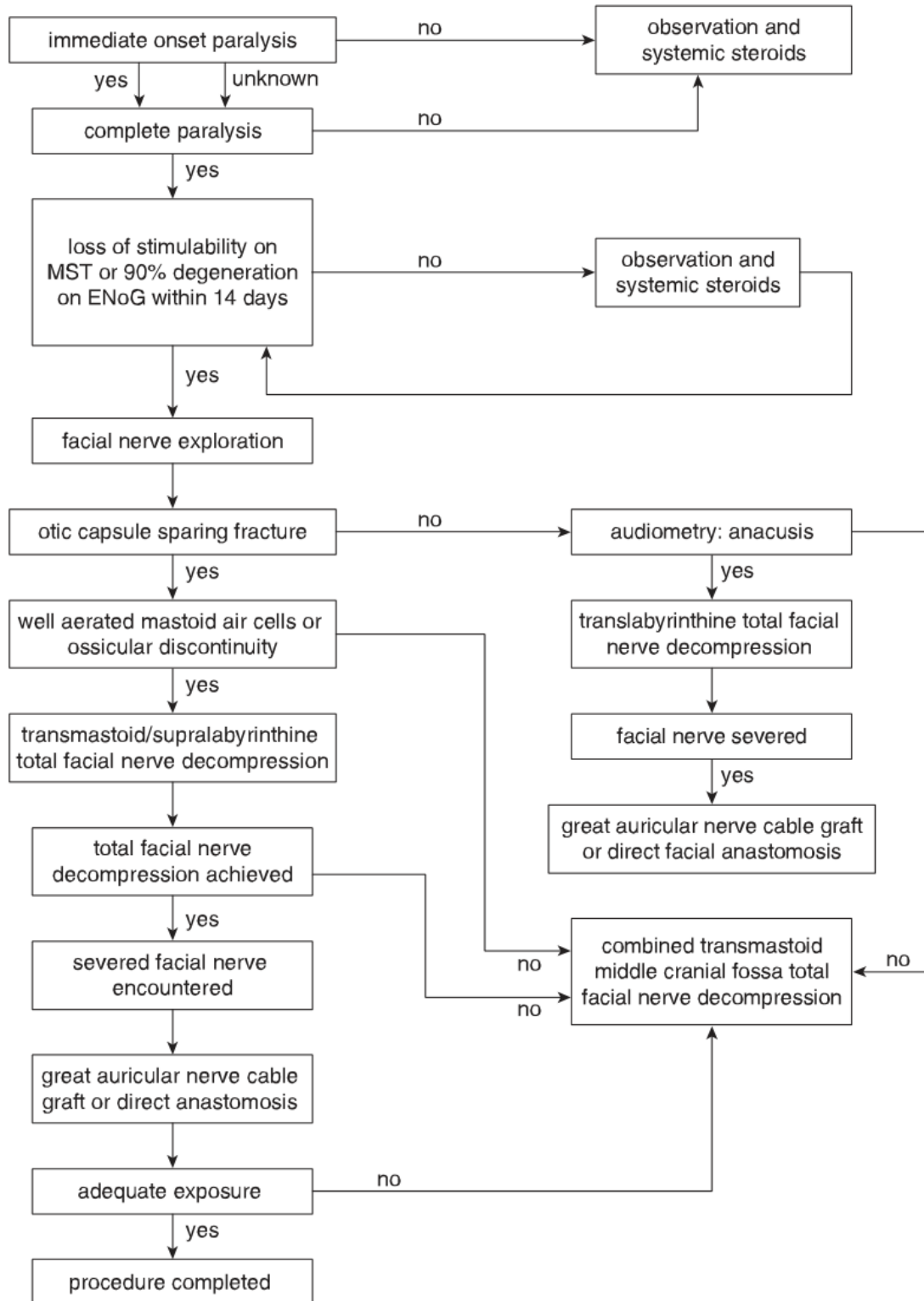


Figure 150.11 Management of traumatic facial paralysis.

4. CSF Leak:

- Occurs in 20% of TB fractures.
- **Otic capsule sparing fracture:**
 - Site of CSF leak:
 - Floor of middle cranial fossa (Tegmen tympani and Tegmen mastoideum).
 - Drains into:
 - Epitympanum, antrum, and mastoid air cells.
- **Otic capsule disrupting fracture:**
 - Site of CSF leak:
 - Posterior cranial fossa.
 - Drains into:
 - Middle ear.
- Pathophysiology:
 - Immediate-onset in majority of patients.
 - Otic capsule fractures does not heal fully and the CSF fistula will continue to leak until fibrosis is formed to close the subarachnoid space or middle ear or mastoid air cell mucosa covers bony defect:
 - This barrier is weak and it will be ruptured by increased ICP (nose blowing or straining) and CSF leak will continue.
 - Delayed-onset (> 1 week) in 30% of patients due to:
 - Resolution of occluding hematoma.
 - Separation of fibers in the dural herniation.
 - Increased ICP.
- Presentation:
 - If reached the ET, CSF rhinorrhea.
 - If TM is perforated, CSF otorrhea.
 - Increases with exertion or leaning forward.
- Diagnosis (Similar to CSF rhinorrhea protocol):
 - **Beta-2 Transferrin:**
 - Detected with protein electrophoresis.
 - Specific for CSF.
 - Only small amount of CSF is required (0.05 mL).
 - **Beta Trace protein:**
 - Synthesized in the meninges.
 - 20-40 fold CSF concentration compared to serum.
 - Less expensive and faster than β 2-transferrin.
 - Detection via nephelometric technique comparable specificity and sensitivity to β 2-transferrin.
 - Recent studies showed 100% positive and negative predictive values of CSF leakage.
 - **High resolution CT:**
 - Shows bony defect in 70% of patients.
 - **Intrathecal fluorescein:**
 - Successful for localizing fistulae when all other methods have failed.

- Management of CSF leak:
 - **Conservative:**
 - Indication:
 - CSF leak after acute trauma.
 - Period:
 - 7-10 days after trauma.
 - Goal:
 - Maintaining CSF pressure gradient below healing tensile strength of the healing barrier.
 - Instructions:
 - Bed rest.
 - Head elevation.
 - Avoid nose blowing, strenuous activity, and constipation.
 - Medications:
 - Diuretic agents (mannitol, acetazolamide, furosemide).
 - Prophylactic antibiotics (controversial)
 - Intervention:
 - Lumbar drain if persistent leak.
 - Prognosis:
 - 60-90% of patients with CSF leak will improve within one week.
 - Complications:
 - **Meningitis:**
 - 5% risk of meningitis in patients active CSF leak of < 7 days.
 - 25% risk of meningitis in patients active CSF leak of > 7 days.

▪ **Surgical Closure:**

- Indications:
 - Failure of conservative management for 7-10 days after acute trauma.
- Goal:
 - Closure of the non-healing CSF fistula.
 - Avoid the increased risk of meningitis after 7-10 days of active leak.
- Approaches:
 - **Non-Hearing preservation approach (+ve SNHL):**
 - Blind sac procedure and fat obliteration of mastoid and middle ear.
 - **Hearing preservation approach (-ve SNHL):**
 - Depends on:
 - Location of fracture.
 - Presence of brain herniation.
 - Status of ossicular chain.
 - Lateral defect in the mastoid at MCF:
 - Mastoidectomy.
 - Temporalis fascia graft over the fistula.
 - Fat obliteration of mastoid cavity.
 - Medial defect in Tegmen tympani with brain herniation (> 1cm):
 - Combined Trans-mastoid/MCF approaches.
 - Trans-mastoid approach used to debride the herniated.
 - MCF approach used for reconstruction with temporalis fascia is placed over floor of MCF followed by bone graft superiorly.
 - Medial defect in Tegmen tympani with ossicular discontinuity and without brain herniation (<1cm):
 - Trans-mastoid approach alone.
 - Cartilage graft followed by Temporalis fascia.
 - Fat obliteration of mastoid cavity.

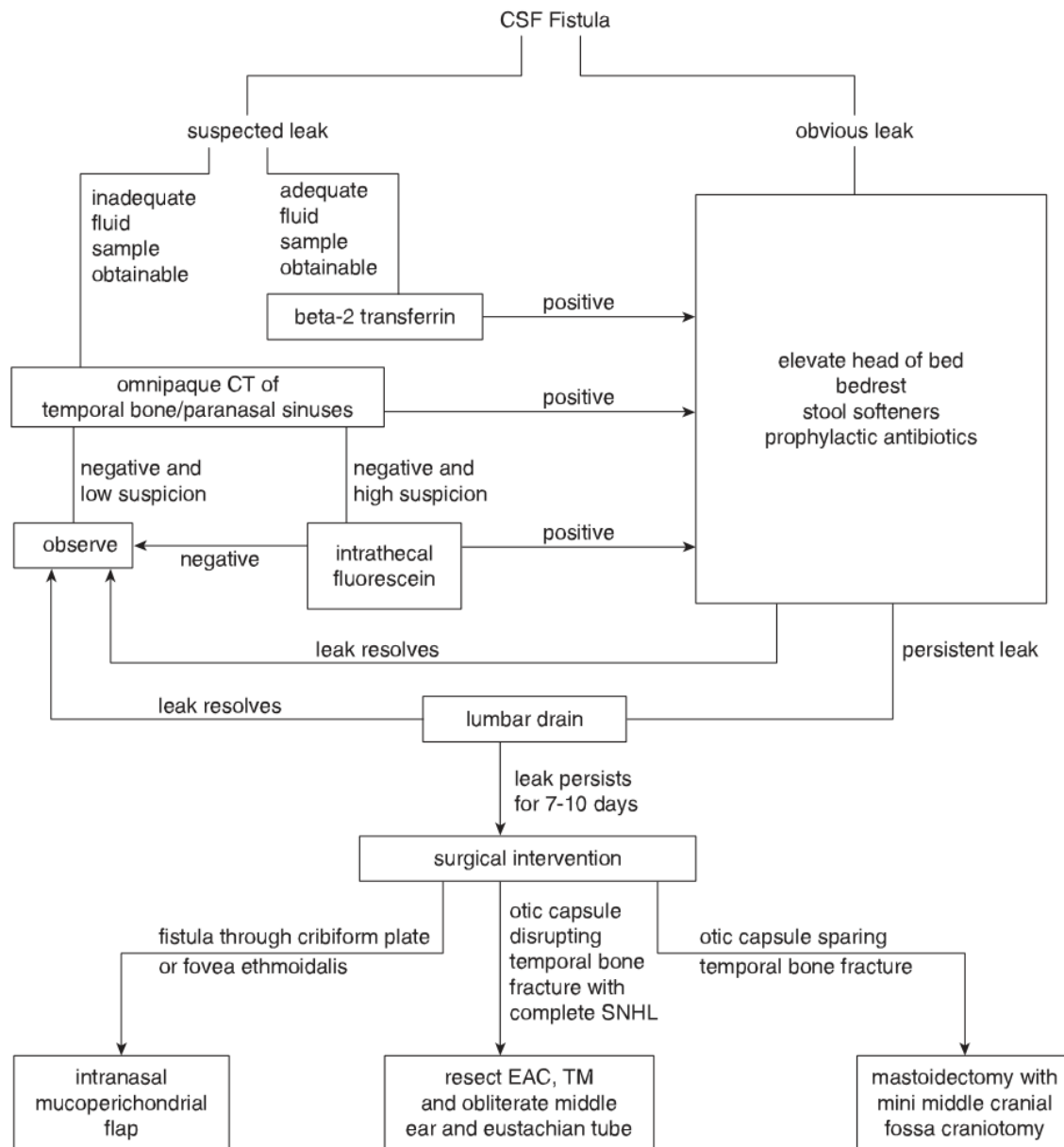


Figure 150.14 Treatment algorithm for the management of CSF fistula.

5. Vertigo:

- Causes:
 - 1.** BPPV (Most common/ 30% of all head traumas)
 - 2.** Labyrinthine concussion
 - 3.** Disruptive injury to the labyrinth.
 - 4.** Disruptive injury to CV-VIII.
 - 5.** Traumatic perilymphatic fistula
 - 6.** Post-traumatic Ménière syndrome

6. Vascular injuries:

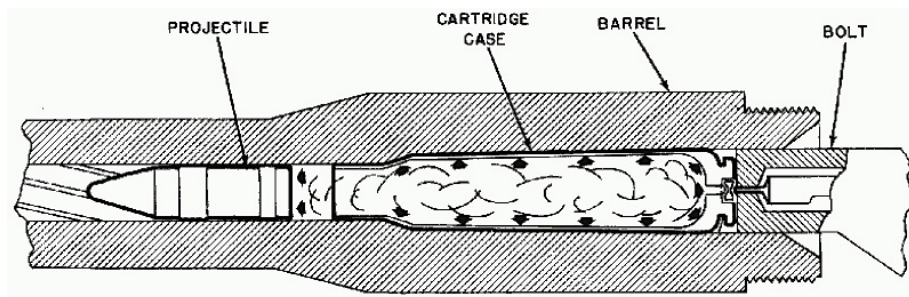
- Rare (1%).
- Intra-temporal carotid injury.
- Present with profuse bloody otorrhea.
- Suspected in patients with fracture of carotid canal on HRCT.
 - 60% sensitivity and 67% specificity.
- CT Angio if actively bleeding with declining neurological status
- OR for ligation or angiography for balloon occlusion

7. Cholesteatoma:

- Late complication.
- Can grow for years without detection
- Mechanisms:
 - 1.** Epithelial entrapment in fracture line (Epitympanum)
 - 2.** In-growth of epithelium through unhealed fracture line
 - 3.** Traumatic implantation of TM skin in middle ear
 - 4.** Trapping of epithelium medial to EAC stenosis

- **Penetrating injury to Facial nerve:**

- Most commonly due to gunshot wounds (GSW).
 - Handguns produce low-velocity injuries.
 - Rifles produce high-velocity ones.
- High-velocity wounds cause injury by crush, laceration and cavitation.
- Cavitation may involve tissues away from bullet's trajectory.
- Internal carotid artery injury due to stretching is common and should be suspected.
- **CN-VII injury occurs in 50% of GSWs to temporal bone.**
 - **More frequently involve labyrinthine or tympanic segment.**
 - **Nerve may be transected, or secondarily injured by kinetic injury from the bullet or from bony fragmentation of the temporal bone.**
 - **Generally, outcome of facial function is much worse with gunshot wounds to temporal bone than with temporal bone fractures.**
- EAC laceration, stenosis and cholesteatoma formation is common.



- **Pediatric Temporal Bone Fractures:**

- Uncommon.
- Same etiologies seen in adults.
- Associated CNS trauma is common (40-75%).
- **CN-VII paralysis is less common in children than adult (3-10%).**
- Temporary CSF leak is common but resolves spontaneously.
- Management of pediatric TB fractures is similar to the adults.

Facial Nerve Paralysis:

- Nerve Fiber Components:

1. Endo-neurium:

- Surrounds each Nerve Fiber (Axons).
- Tightly adherent to Schwann cell layer.
- Provides Endoneurial tube for Regeneration.
- Poorer prognosis for regeneration when disrupted.

2. Peri-neurium:

- Surrounds Fascicles (Group of Axons).
- Provides tensile strength.
- Maintains intrafunicular pressure.
- Protects from infection.

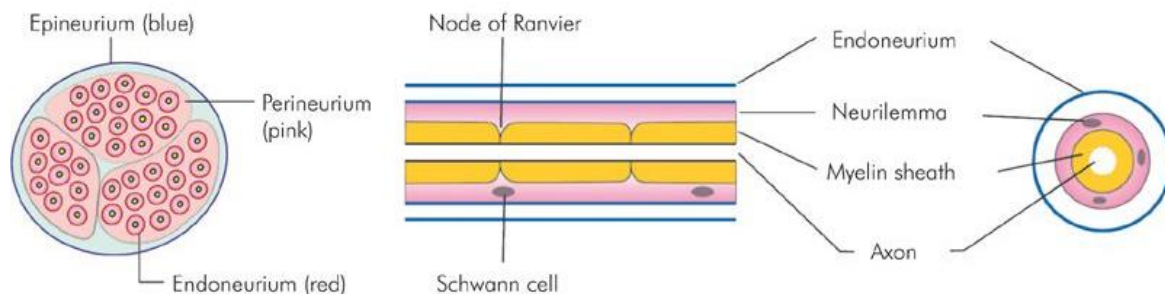
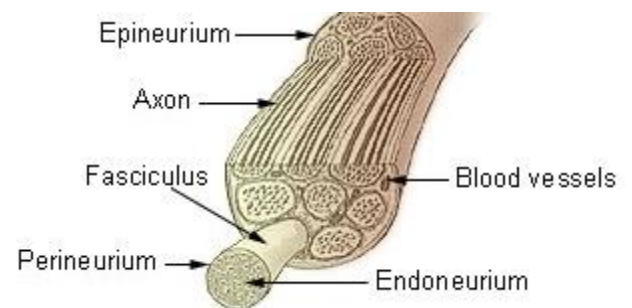
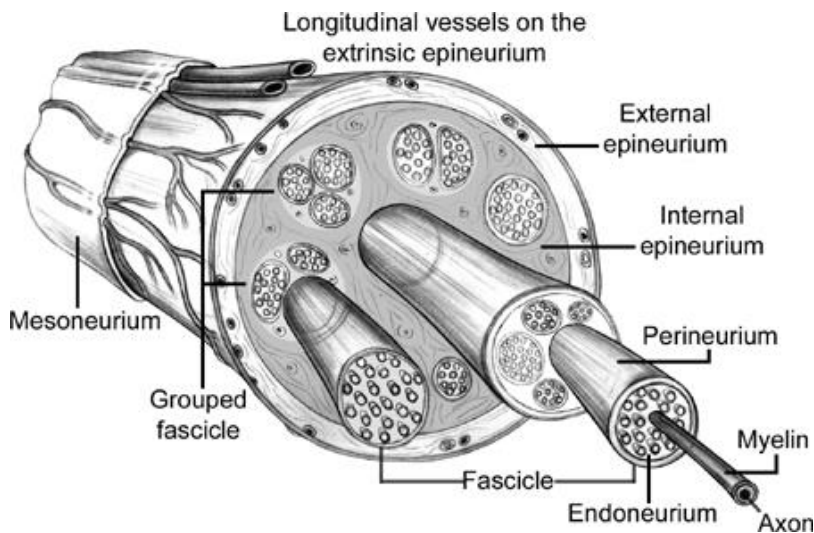
3. Epi-neurium:

- Nerve Sheath.
- Outer layer.
- Surrounds group of Fascicles.
- Contains the Vasa Nervorum for nutrition.

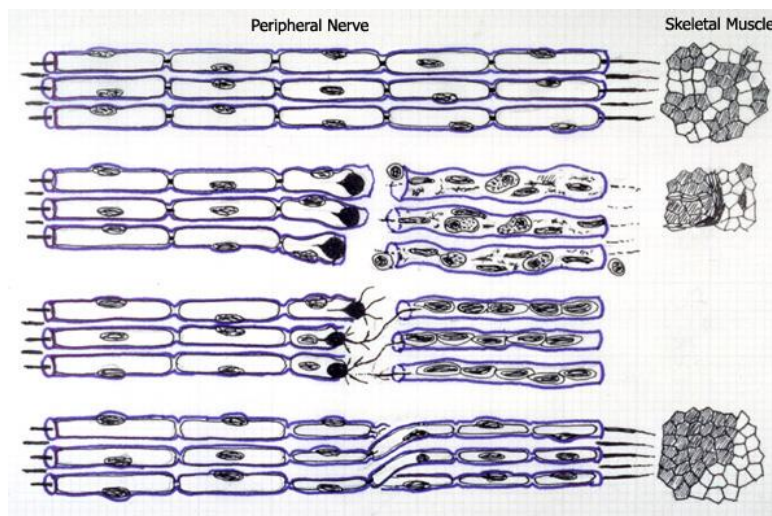
Axon
↓
Myelin sheath
↓
Neurilemma
↓
Endoneurium

Group of Axons
↓
Fascicles
↓
Perineurium

Group of Fascicles
↓
Epineurium



- **Pathophysiology of Nerve injury:**
- When an axon is injured, biochemical and histological changes occur in cell body proximal and distal to site of injury.
- Severity of changes depend upon:
 1. **Distance from the injury to the cell body:**
 - Proximal injuries are more severe than distal injuries.
 2. **Type of injury:**
 - Crush injuries are more severe than clean transections.
 3. **Age of the patient:**
 - Older individuals sustain more severe injury.
 4. **Nutritional and Metabolic status of the patient.**
- **Wallerian Degeneration and Regeneration:**
- Process of degeneration of axons and Myelin sheaths following axonal lesions.
- Begins immediately after injury but progresses slowly.
- Propagates distally from site of injury to motor end plate.
- Propagates proximally to the first adjacent node of Ranvier.
- Prior to degeneration, distal axon stumps tend to remain electrically excitable up to 3-5 days after injury:
 - Electrodiagnostic testing cannot distinguish between neurapraxic and neurodegenerative injuries during this period.
- After injury:
 - → Axonal membrane breaks apart.
 - → Degradation of myelin sheath.
 - → Infiltration by macrophages and Schwann cells to clear the debris from the degeneration.
 - → Axon's Neurolemma does not degenerate and remains as a hollow tube.
 - → Axon sprouts are send out of distal end of proximal part of the nerve towards those tubes.
 - → Grows into the tubes and advances **1 mm/day**.
 - → Reinnervation of target tissue.



- **Sunderland Classification of Nerve Injury:**

1. First Degree (Neuropraxia):

- Intact Axons.
- Partial interruption of Axoplasmic flow due to compression.
- No morphological changes are seen.
- No Wallerian degeneration.
- Will conduct neural discharge if stimulated distal to the block.
- Recovery of function is complete.
- Seen in viral and inflammatory disorders.

2. Second Degree (Axonotmesis):

- Loss of Axons.
- Intact Endoneurial tubes.
- Interrupted Axoplasmic flow.
- Seen in viral and inflammatory disorders.
- Wallerian degeneration occurs.
- Axons regenerate through intact neural tubes @ **1 mm/day.**
- Complete recovery anticipated.

3. Third Degree (Endoneurotmesis):

- Loss of Axons.
- Loss of Endoneurial tubes.
- Seen in viral and inflammatory disorders.
- Wallerian degeneration occurs.
- Regenerating axons:
 - Enter the wrong endoneurial tubes.
 - Fail to enter endoneurial tubes.
- Results in:
 - Synkinesis:
 - Abnormal mass movement of muscles which do not normally contract together.
 - Incomplete recovery.

4. Fourth Degree (Perineurotmesis):

- Partial Nerve Transection.
- Loss of Perineurium (in addition to above).
- Only Epineurium is intact.
- Seen in surgical or accidental trauma or in neoplasms.
- Scarring will impair regeneration of fibers.
- Greater risk for synkinesis and incomplete recovery.

5. Fifth Degree (Neurotmesis):

- Complete Nerve Transection.
- Loss of Epineurium (in addition to above).
- Seen in surgical or accidental trauma or in neoplasms.
- Risk of Neuroma from nerve sprouts outside of nerve sheath.
- Without surgical repair, No chance of regeneration and recovery.

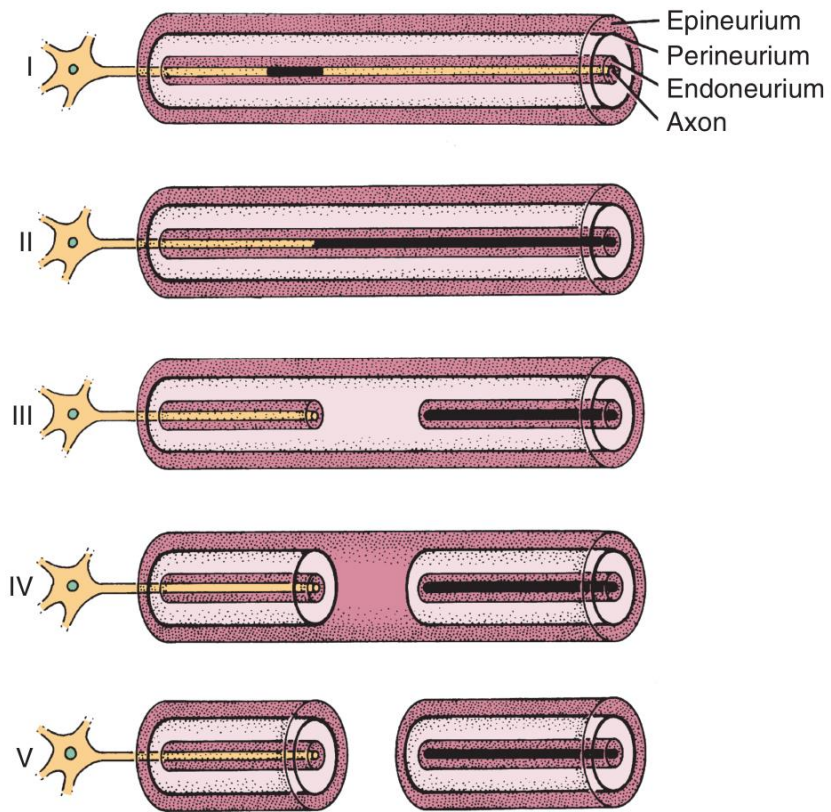


FIGURE 169-2. Sunderland histopathologic classification of peripheral nerve injury. Roman numerals I to V denote the Sunderland class that corresponds to the degree of injury depicted in the diagram.

- **Evaluation of Facial Nerve Paralysis:**

- **History:**

- **Character of Facial weakness:**

- Onset (Sudden vs Delayed).
- Duration.
- Progression of paralysis (complete vs incomplete).
- Recurrent (Previous history of Facial nerve paralysis).

- **Contributing Factors:**

- Recent infection or illness.
- Trauma (birth trauma in neonates).
- Surgery (Otologic, Parotid, or Neurologic surgery).
- Recent tick bites or outdoor activity.
- History of syphilis, HIV, TB or Herpes.
- Toxin exposure (Lead).
- History of Otologic, Neurologic, Diabetic, or Vascular disorders.

- **Associated Symptoms:**

- Change in taste sensation.
- Drooling.
- Epiphora.
- Vision changes.
- Pain (Auricular, Postauricular, or Facial).
- Ear symptoms (hearing loss, aural fullness, otalgia, vertigo or hyperacusis).
- Neurologic deficits.

- **Physical Exam:**

- **Facial Nerve:**

- Unilateral vs Bilateral weakness.
- Central vs Peripheral.
 - Forehead movement.
- Observe facial symmetry at rest and with movement.
- Hemifacial spasms.
- Facial tics at rest.
- Eye closure.
- Tear production.
- Corneal Reflex.
- Visual acuity.
- **Bell's phenomenon:**
 - Globe turns up and out during attempts to close eyes.
 - Indicates Peripheral lesion.



- **House-Brackmann System of Grading Facial Nerve Paralysis:**

- **Grade I:**
 - Normal.
- **Grade II:**
 - Symmetry at Rest.
 - No/minimal synkinesis.
 - Complete eye closure with Minimal effort.
- **Grade III:**
 - Symmetry at Rest.
 - Obvious synkinesis.
 - Complete eye closure with Maximum effort.
- **Grade IV:**
 - Symmetry at Rest.
 - Obvious synkinesis.
 - Incomplete eye closure.
- **Grade V:**
 - Asymmetry at Rest.
 - Obvious synkinesis.
 - Incomplete eye closure.
- **Grade VI:**
 - Total paralysis.

TABLE 6–8. House-Brackmann System of Grading Facial Nerve Paralysis*

Grading	Function
I	normal function
II	mild dysfunction: weakness on close inspection, normal symmetry and tone at rest, moderate to good facial function (slight mouth asymmetry, complete eye closure with minimal effort)
III	moderate dysfunction: obvious weakness, and/or asymmetry (not disfiguring), contracture, and/or hemifacial spasms, normal symmetry and tone at rest; moderate facial function (weak mouth and forehead function, complete eye closure with effort)
IV	moderately severe dysfunction: disfiguring asymmetry and/or obvious weakness, normal asymmetry and tone at rest, incomplete eye closure, no forehead motion, asymmetric mouth motion with maximal effort
V	severe dysfunction: barely perceptible motion, asymmetry at rest, incomplete eye closure, no forehead motion, slight mouth motion
VI	total paralysis

TABLE 169-1. House-Brackmann Facial Nerve Grading System

Grade	Description	Characteristics
I	Normal	Normal facial function in all areas
II	Mild dysfunction	Gross: Slight weakness noticeable on close inspection; may have very slight synkinesis At rest: Normal symmetry and tone Forehead motion: Moderate to good function Eye motion: Complete closure with minimum effort Mouth motion: Slight asymmetry
III	Moderate dysfunction	Gross: Obvious but not disfiguring difference between two sides; noticeable but not severe synkinesis, contracture, or hemifacial spasm At rest: Normal symmetry and tone Forehead motion: Slight to moderate movement Eye motion: Complete closure with effort Mouth motion: Slightly weak with maximum effort
IV	Moderately severe dysfunction	Gross: Obvious weakness and/or disfiguring asymmetry At rest: Normal symmetry and tone Forehead motion: None Eye motion: Incomplete closure Mouth motion: Asymmetric with maximum effort
V	Severe dysfunction	Gross: Only barely perceptible motion At rest: Asymmetry Forehead motion: None Eye motion: Incomplete closure Mouth motion: Slight movement
VI	Total paralysis	No movement

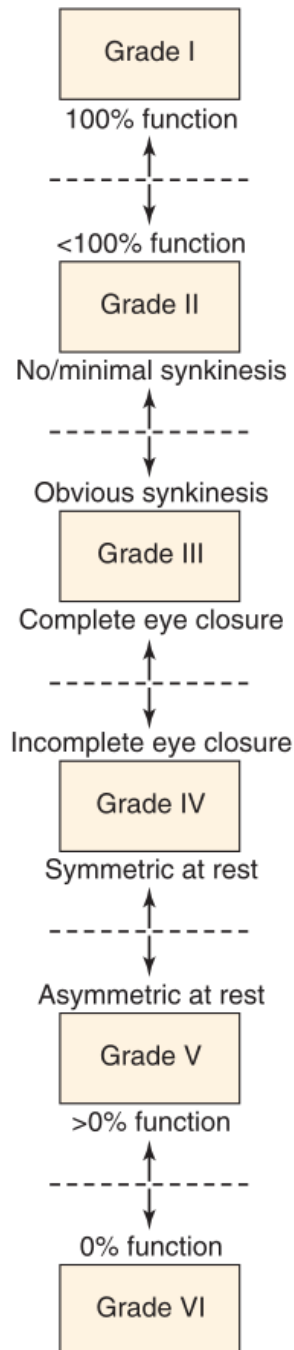


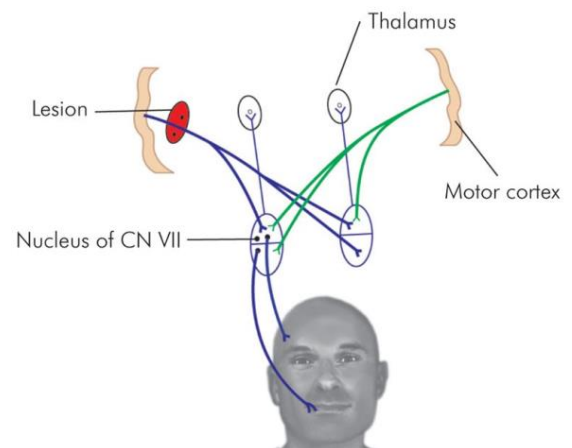
FIGURE 169-1. Schematic diagram of a modified House-Brackmann grading scale using the major functional criteria of absolute movement, synkinesis, eye closure, asymmetry at rest, and absolute paralysis in assigning unambiguous and nonoverlapping degrees of facial paralysis.

- Other Head and Neck Assessment:
 - **Ear examination:**
 - Evaluate for mass or fluid in Middle ear.
 - Presence of vesicles in EAC and concha.
 - **Hitselberger sign:**
 - Hypesthesia of sensory division of Facial nerve at superior posterior concha.
 - **Neck examination:**
 - Parotid masses.
 - Lymph nodes.
 - **Neurological examination:**
 - Other cranial nerve involvement.
 - Other lateralizing signs:
 - Hemiparesthesias.
 - Hemiparalysis.
 - Aphasia.

Localization of Facial Nerve Lesions:

1. Central Facial Paralysis (UMNL):

- Paralysis of lower half of the face on the contralateral side only.
- Forehead is preserved:
 - Receives bilateral innervation.
- Involuntary emotional movements and tone are preserved:
 - Receives fibers from Thalamus.
- Caused by:
 1. CVA.
 2. Tumour
 3. Abscess.



2. Peripheral Facial Paralysis (LMNL):

- Paralysis of all muscles of the face on the ipsilateral side.
- Forehead is involved.
- Lesion at level of Motor Nucleus:
 - Associated paralysis of CN-VI.
- Lesion at CPA:
 - Vestibular and Auditory defects.
 - Associated paralysis of CN-V, CN-IX, CN-X and CN-XI.
- Lesion in Fallopian canal:
 - Localized by Topodiagnostic tests.
- Lesion outside Temporal bone:
 - Affects only motor functions of Facial nerve.

**TABLE
155.1**

ASSESSMENT OF FACIAL PALSY

History

- Onset
- Duration
- Rate of progression
- Recurrent or familial
- Associated symptoms
- Major medical illness or previous surgery

Physical examination

- Complete head and neck evaluation
- Microscopic otoscopy
- Upper aerodigestive tract examination
- Cranial nerve assessment (III–XII)
- Palpation of parotid gland and neck
- Neurologic evaluation
- Cerebellar signs
- Motor
- Facial palsy
- Complete vs. incomplete (paresis)
- Segmental vs. uniform involvement
- Unilateral vs. bilateral
- Schirmer test

Laboratory studies

- Pure-tone and speech audiometry
- Electrophysiologic tests
- Nerve excitability test (NET)
- Maximal stimulation test (MST)
- Electroneurography (ENoG)
- Electromyography (EMG)
- Radiographic studies
- Computed tomography
- Magnetic resonance imaging

Other considerations

- Complete blood cell count and differential with sedimentation rate
- Serum antibody tests (Lyme titers)
- Chest x-ray radiograph
- Lumbar puncture with cerebrospinal fluid assay

Topo-gnostic Tests for Intra-Temporal FN Lesions:

- Based on principle that lesions distal to the site of a particular branch of the facial nerve will spare the function of that branch.
- Rarely used nowadays.
- Not found to be of much use clinically for determining the site of the lesion in facial paralysis or for predicting the outcome.

1. Schirmer's Test:

- Compares lacrimation of the two sides.
- Strip of filter paper is hooked in lower fornix of each eye.
- Amount of wetting of strip is measured.
- After 5 minutes, Length of strip that is moist is compared to the normal side.
- Predict patients at risk for exposure keratitis.
- Abnormal Results (Either):
 - Value of <50% compared to normal side.
 - Total lacrimation less than 25 mm
- Indicates:
 - **Lesions at or proximal to Afferent limb:**
 - Ophthalmic (V1) of Trigeminal nerve.
 - **Lesions at or proximal to Efferent limb:**
 - Greater superficial petrosal nerve (branching at geniculate ganglion).



2. Stapedial Reflex:

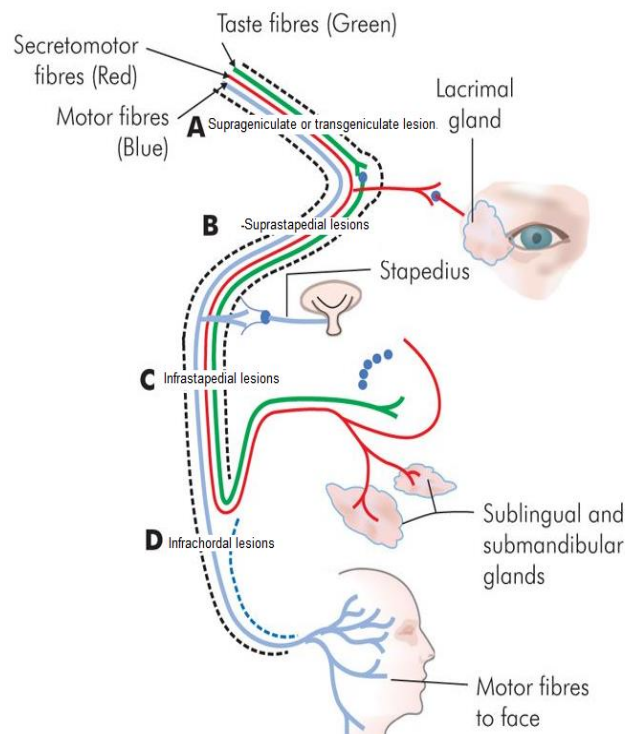
- Most objective and reproducible test.
- Useful in determining facial nerve prognosis.
- Tested by tympanometry.
- Abnormal Result (Either):
 - Absent stapedial reflex.
 - Amplitude of <50% compared to normal side.
- Indicates:
 - **Hearing loss:**
 - Any degree of CHL.
 - Cochlear SNHL > 60 dB
 - Any degree of Retro-cochlear SNHL
 - **Lesions at or proximal to the Efferent limb:**
 - Nerve to stapedius (branching after second genu in the vertical segment).

3. Taste Test:

- Measured by a drop of salt or sugar solution placed on one side of protruded tongue.
- Abnormal Result:
 - Impairment of taste (dysgeusia/ageusia).
- Indicates:
 - **Lesions at or proximal to lingual nerve.**
 - **Lesions at or proximal to chorda tympani:**
 - Branching anywhere in the vertical segment.
- More reliable indicator of interruption of Chorda tympani nerve involves microscopic detection of Absence of taste papillae on the middle 1/3 of involved side of Tongue:
 - Disappear within 10 days post injury.

4. Submandibular Salivary Flow and PH Test:

- Polythene tubes are passed into both Wharton's ducts.
- Measurement of output after five minutes.
- Abnormal Result:
 - Flow rate $\leq 25\%$ compared to normal side.
- Indicates:
 - **Lesions at or proximal to lingual nerve.**
 - **Lesions at or proximal to chorda tympani:**
 - Branching anywhere in the vertical segment.
- Salivary pH may be examined as an indirect measure of flow:
 - As Flow rate decreases, pH decreases.
 - pH of ≤ 6.1 may predict loss of function of Chorda tympani.



- Supraganglionic lesions cause:

1. Decreased lacrimation.
2. Loss of Stapedial reflex.
3. Loss of Taste.
4. Decreased salivation
5. Loss of Motor function.

- Suprastapedial lesions cause:

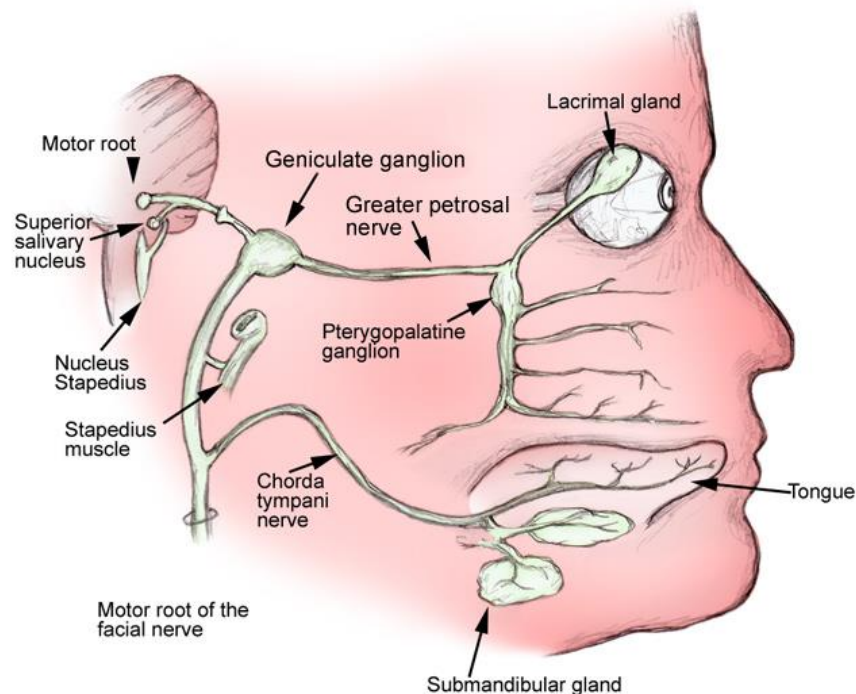
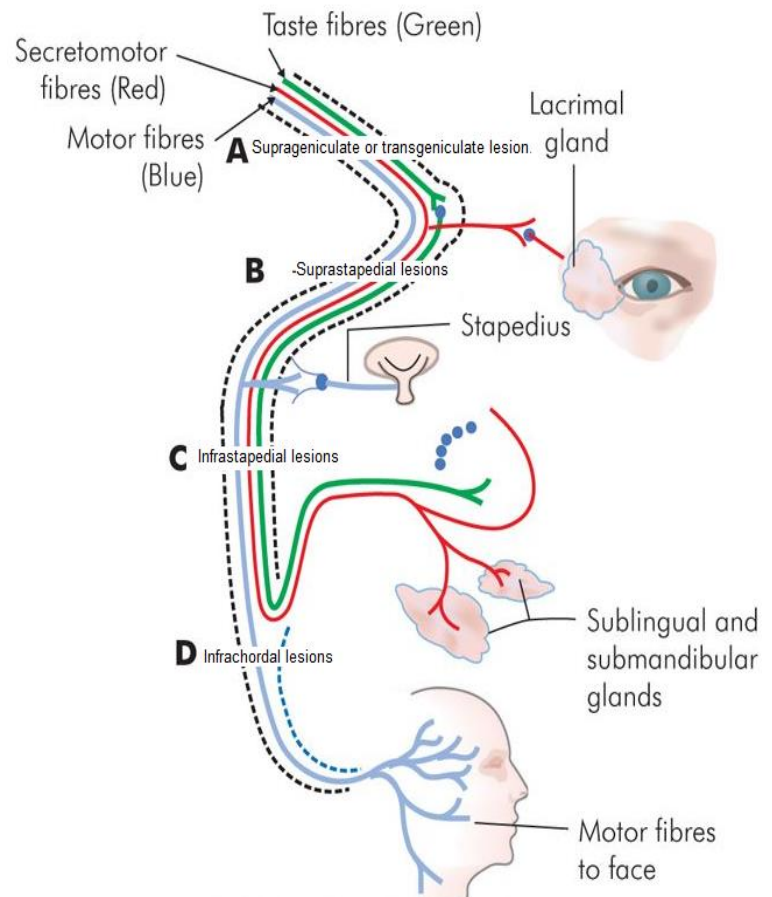
1. Preserved lacrimation.
2. Loss of Stapedial reflex.
3. Loss of Taste.
4. Decreased salivation
5. Loss of Motor function.

- Infrastapedial lesions cause:

1. Preserved lacrimation.
2. Preserved Stapedial reflex.
3. Loss of Taste.
4. Decreased salivation
5. Loss of Motor function.

- Infrachordal lesions cause:

1. Preserved lacrimation.
2. Preserved Stapedial reflex.
3. Preserved Taste.
4. Preserved salivation
5. Loss of Motor function.



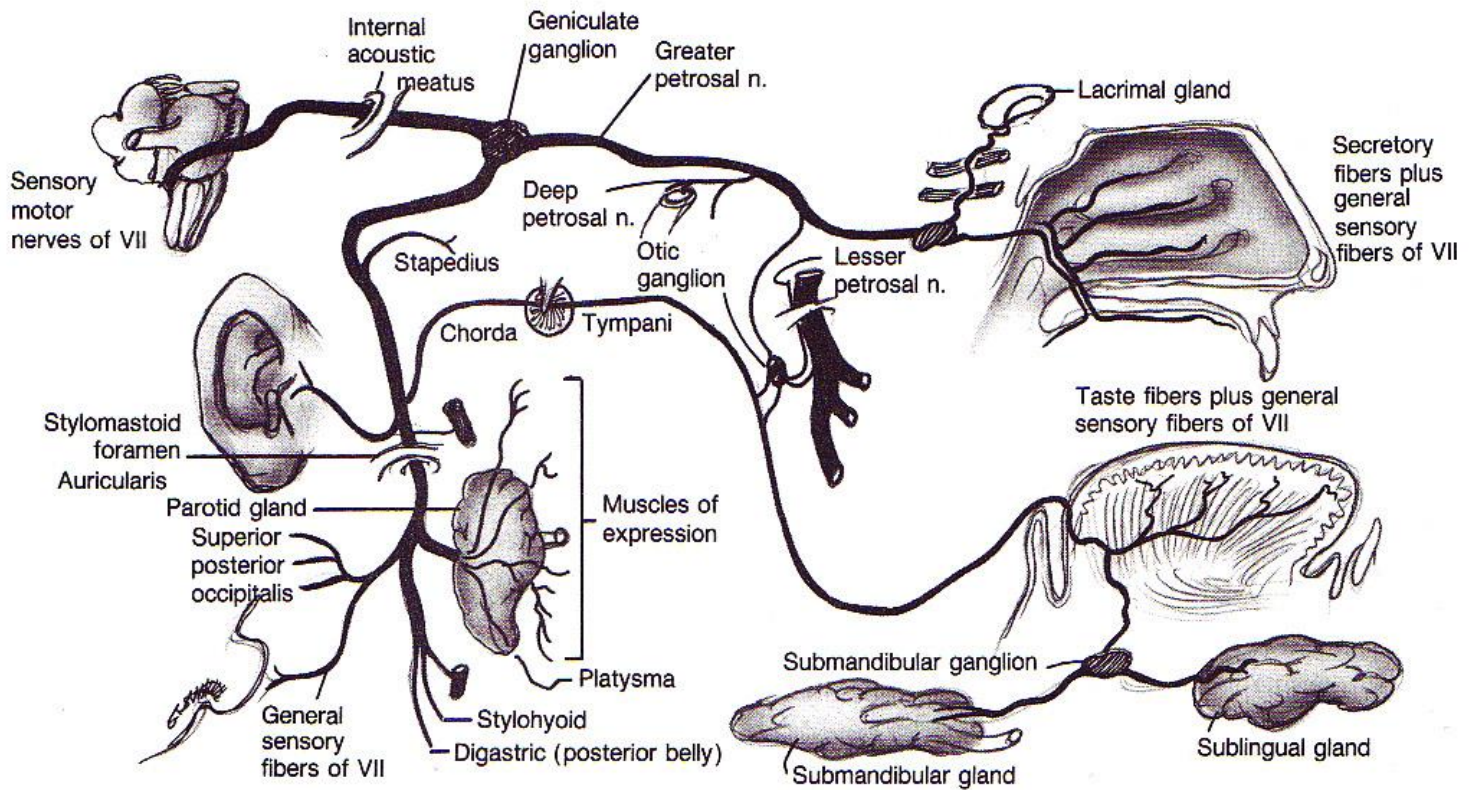


Figure 10.2 Facial nerve. (Modified from The cranial nerves. In: Anderson JE, ed. *Grant's atlas of anatomy*. Baltimore: Williams & Wilkins, 1983:8-1, with permission.)

Ancillary Studies for Evaluation of Facial Nerve paralysis:

- **Audiogram:**
 - Indicated for all Intra-temporal injuries.
 - Preoperative baseline hearing.
- **Topognostic Tests:**
 - Not useful.
 - Replaced with Electrophysiological testing.
- **Blood work:**
 - Indicated for suspected patients with inflammatory diseases.
 - **CBC**
 - **Treponemal Studies:**
 - Lyme titers and VDRL/FTA-ABS
 - **ACE level:**
 - Active sarcoidosis
- **Imaging:**
 - MRI with gad is the most sensitive imaging for evaluation of entire course of facial nerve.
 - CT with contrast can be done for patients contraindicated for MRI.
 - **Indications of imaging in patients with facial paralysis:**
 1. History of recurrent ipsilateral paralysis.
 2. Bilateral facial paralysis.
 3. Gradually developing facial nerve paralysis.
 4. No evidence of recovery after 3 month from onset.
 5. History of Trauma (HRCT Temporal bone).
 6. Concurrent otologic findings (cholesteatoma).
 7. Associated Neurological symptoms.
 8. Other cranial nerve involvements.
 9. Suspected CPA lesions.
- **Electrophysiological Tests:**
 - These tests are complimentary (no single best test).
 - Differentiate between neurapraxia and nerve degeneration.
 - **Indication:**
 - **Patients with complete facial nerve paralysis:**
 - Determine prognosis for return of facial function.
 - Choosing candidates for surgical intervention:
 - Decompression surgery (ENoG).
 - Reanimation surgery (EMG).

- No value of electrophysiological tests in:
 - **Before 3 days of the onset of complete facial nerve paralysis:**
 - Nerve degeneration didn't reach site of stimulation.
 - Facial nerve will remain electrically excitable regardless the type of injury.
 - Results will be misleading.
 - **After 21 days of the onset of complete facial nerve paralysis:**
 - Complete nerve degeneration has occurred.
 - After loss of excitability, tests that require electrical stimulation (NET, MST and ENoG) are no longer useful.
 - **EMG is the only test that give prognostically useful information during this period.**
 - **Patients with incomplete facial nerve paralysis:**
 - Facial movements indicates neurapraxic intact nerve.
 - Good recovery is anticipated.

Types of Electrophysiological Tests:

1. Nerve Excitability Test (NET):

- Rarely used today due to its major drawbacks.
- Method:
 - Facial nerve is stimulated with over stylomastoid foramen.
 - Return electrode is taped to the forearm.
 - Electrical pulses are delivered at steadily increasing current levels until a facial twitch is noted.
 - Lowest current necessary to produce facial twitch (**Excitation threshold**) on normal side is compared with paralyzed side.
 - Difference in thresholds between two sides is calculated.
- Interpretation:
 - **Conduction block (Neuropraxia/Type I):**
 - No difference in excitation threshold between two sides.
 - No degeneration occurs.
 - Good prognosis for complete recovery.
 - **Degenerative injuries (Type II-V):**
 - Difference in excitation threshold of **> 3.5 mA** indicates significant degeneration.
 - Poor prognosis for return of facial function.
- Advantages:
 - Low cost, readily available equipment, and ease of performance.
 - **Used for evaluation of acute facial paralysis (while in the degenerative phase).**
- Disadvantages:
 - **Subjective test** with reliance entirely on a visual end point.
 - Few intact axons can give visible response leading to wrong prediction of good prognosis, while most fibers are degenerated.
 - Used for unilateral paralysis (comparison with normal side).

2. Maximum Stimulation Test (MST):

- Modified version of NET.
- Method:
 - Facial nerve is stimulated with electrode over stylomastoid foramen.
 - Return electrode is taped to the forearm.
 - Electrical pulses are delivered at steadily increasing current levels until a facial twitch is noted.
 - Instead of measuring excitation threshold, the current level which gives maximum facial movement is measured (**Maximum stimulation level**) in the normal side.
 - This maximum stimulation level is then used to stimulate the affected side.
 - Degree of facial contraction in the affected side is subjectively (visually) assessed as either equal, decreased or absent.
- Interpretation:
 - **Good prognosis:**
 - Equal or slightly decreased response on involved side compared to the normal side.
 - Favorable for complete recovery.
 - **Poor prognosis:**
 - Absent or markedly decreased response on involved side.
 - Advanced degeneration.
 - Incomplete recovery.
- Advantages:
 - Low cost, readily available equipment, and ease of performance.
 - Used for evaluation of acute facial paralysis (while in the degenerative phase).
 - MST is considered superior to NET.
 - Stimulating all intact axons.
 - MST response becomes abnormal earlier than NET response.
 - More reliably guide prognosis and treatment than NET.
- Disadvantages:
 - **Subjective test** with reliance entirely on a visual end point.
 - Used for unilateral paralysis (needs comparison with normal side).

3. ElectroNeuronography (ENoG):

- Also called Evoked Electromyography (EEMG).
- Method:
 - Facial nerve is stimulated with electrode over stylomastoid foramen.
 - Muscular response is recorded using bipolar electrodes placed near nasolabial groove.
 - Measures the amplitude of **evoked Compound Muscle Action Potential (CMAP)**:
 - Considered proportional to number of intact axons.
 - The amplitude of evoked CMAP in the involved side is compared to the normal side.
- Interpretation:
 - **Good prognosis:**
 - Degeneration of **< 90%** of axons within 14 days of onset.
 - Amplitude of evoked CMAP on involved side is reduced to more than **10%** of normal side.
 - Expected spontaneous rate of recovery of 80-100%.
 - **Poor prognosis:**
 - Degeneration of **≥ 90%** of axons within 14 days of onset.
 - Amplitude of evoked CMAP on involved side is reduced to **10%** or less of normal side.
 - Faster rate of degeneration occurring in less than 14 days has a poorer prognosis.
 - Poor prognosis for spontaneous recovery without surgical decompression.
- Advantages:
 - Used for evaluation of acute facial paralysis (while in the degenerative phase).
 - Most accurate prognostic test for complete facial paralysis:
 - Objective, qualitative measurement of nerve degeneration.
 - Used to calculate percentage of intact axons.
 - Helps to choose candidates for surgical decompression.
- Disadvantages:
 - Discomfort and cost.
 - Repeated every other day during the first 2-3 weeks following nerve injury to detect accelerating ongoing degeneration.
 - Test-retest variability due to positioning of the electrodes and excitation of the muscles of mastication (V).
 - Used for unilateral paralysis (needs comparison with normal side).

4. ElectroMyography (EMG):

- Evaluates motor activity of facial muscles and presence of functional motor units.
- Method:
 - Direct insertion of needle electrodes into facial muscles (orbicularis oculi and orbicularis oris muscles).
 - Measures spontaneous and voluntary muscle potentials.
 - Normally:
 - Electrical silence in normal muscle at resting state.
 - Diphasic or Triphasic muscle action potentials are generated by voluntary activity.
- Interpretation:
 - **Electrical silence:**
 - Indicates either:
 - Normal muscle in a resting state.
 - Acute facial paralysis in the early stages (before complete nerve degeneration takes place).
 - Severe muscle wasting and atrophy in late stages.
 - **Voluntary action potentials:**
 - Indicates at least partial continuity of nerve.
 - Confirms the integrity of intact axons.
 - Very high probability of good recovery.
 - **Fibrillation potentials:**
 - Spontaneous involuntary action potentials.
 - Electrical sign not clinical sign (not visible).
 - Appears after complete nerve degeneration and loss of excitability (after 14-21 days).
 - Impaired reinnervation yield fibrillation potentials as long as postsynaptic membranes remain electrically active.
 - With persistent denervation, EMG recordings are silent and insertional activity are lost (18-24 months).
 - **Polyphasic potentials:**
 - Indicates re-innervation.
 - After nerve injury, nerve fibers can regrow down the old nerve sheath and can sprout to supply denervated muscles.
 - New sprouts are de-synchronous.
 - Muscle potential is polyphasic (> 5 phases).
 - Observed during voluntary muscle contraction.
 - Appears 4-6 weeks after the onset of paralysis.
 - **Precedes clinical recovery and provide the earliest evidence of recovery.**
 - Predicts a fair to good recovery.

- **Advantages:**
 - Used as complementary for ENoG when $\geq 90\%$ degeneration has occurred and surgery is being considered:
 - If EMG shows voluntarily active facial motor units, the prognosis for a good spontaneous recovery is good.
 - Used for evaluation of chronic facial paralysis (after complete nerve degeneration has occurred).
 - Useful in planning re-animation surgeries:
 - Polyphasic potentials after 1 year of injury:
 - Reinnervation is already taking place.
 - No need for reanimation procedure.
 - Fibrillation potentials after 1 year of injury:
 - Intact motor end plates.
 - No evidence of reinnervation.
 - Need for nerve substitution.
 - Electrical silence after 1 year of injury:
 - Atrophy of motor end plates.
 - Need for muscle transfer procedures rather than nerve substitution.
- **Disadvantages:**
 - Discomfort and cost.
 - Limited value in early evaluation of acute facial paralysis (before complete nerve degeneration takes place).
 - Can't assess degree of nerve degeneration.

TABLE 155.4 ELECTROPHYSIOLOGIC TESTS			
Test	Indication	Interpretation	Limitation
Nerve excitability test	Complete paralysis <2 wk duration	≤ 3.5 mA threshold difference: prognosis good	Not useful in first 3 d after onset or during recovery
Maximal stimulation test	Same as NET	Marked weakness or no muscle contraction: advanced degeneration with guarded prognosis	Subjective
Electroneurography	Same as NET and MST	<90% degeneration: good prognosis >90%: prognosis in question	False-positive results in deblocking phase
Electromyography	Acute paralysis <1 wk duration Chronic paralysis >3 wk duration	Active mu: intact motor axons mu + fibrillation potentials: partial degeneration Polyphasic mu: regenerating nerve	Cannot assess degree of degeneration of prognosis for recovery

NET, nerve excitability test; MST, maximal stimulation test; mu, motor units.

TABLE 169-2. Electrodiagnostic Testing Criteria	
Test	Criteria to Consider in Facial Nerve Decompression
Nerve excitability test (NET)	>3.5-mA threshold difference between sides
Maximal stimulation test (MST)	No response on injured side at maximal stimulation
Electroneuronography (ENoG)	>90% degeneration within 14 days

Causes of Facial Nerve Paralysis:

- Central or Peripheral.
- Peripheral lesions are more common.
 - Bell's palsy is the most common cause (50%) of all cases.
 - Trauma is the second most common cause (20%) of all cases.

TABLE 6-7. Differential Diagnosis of Facial Nerve Paralysis: KITTENS Method

(K) Congenital	Infectious and Idiopathic	Toxins and Trauma	Tumor (Neoplasms)	Endocrine	Neurologic	Systemic/ PSychological
Möbius syndrome Myotonic dystrophy	Idiopathic facial paralysis Melkersson-Rosenthal syndrome Ramsay-Hunt syndrome Otitis media/mastoiditis Necrotizing otitis externa Meningitis Lyme disease Tetanus TB, HIV, EBV, syphilis	Head trauma Temporal bone trauma Iatrogenic injuries Birth trauma	Parotid tumors Facial neuromas Acoustic neuromas Cholesteatoma Gliomas Meningioma Temporal bone tumors	Diabetes mellitus Pregnancy Hyper-thyroidism	Guillain-Barré Multiple sclerosis Myasthenia gravis Stroke	Sarcoidosis Amyloidosis Hyperostoses (Paget's disease, osteopetrosis)

TABLE 170-1. Differential Diagnosis of Facial Paralysis

Acute Paralysis	Chronic or Progressive Paralysis
Polyneuritis • Bell palsy • Herpes zoster • Guillain-Barré syndrome • Autoimmune disease • Lyme disease • HIV infection • Kawasaki disease Trauma • Temporal bone fracture • Barotrauma • Birth trauma Otitis media • Acute bacterial • Chronic bacterial • Cholesteatoma Sarcoidosis Melkersson-Rosenthal syndrome Neurologic disorders • HIV infection • Cerebrovascular disorders, central or peripheral	Malignancies • Primary parotid tumor • Metastatic tumor Benign tumors • Schwannoma • Glomus tumor Cholesteatoma

Table 14-1. Causes of facial paralysis

1. *Central*
 - Brain abscess
 - Pontine gliomas
 - Poliomyelitis
 - Multiple sclerosis
2. *Intracranial part (cerebellopontine angle)*
 - Acoustic neuroma
 - Meningioma
 - Congenital cholesteatoma
 - Metastatic carcinoma
 - Meningitis
3. *Intratemporal part*
 - (a) Idiopathic
 - Bell's palsy
 - Melkersson's syndrome
 - (b) Infections
 - Acute suppurative otitis media
 - Chronic suppurative otitis media
 - Herpes zoster oticus
 - Malignant otitis externa
 - (c) Trauma
 - Surgical: Mastoidectomy Stapedectomy
 - Accidental: Fractures of temporal bone
 - (d) Neoplasms
 - Malignancies of external and middle ear
 - Glomus jugulare tumour
 - Facial nerve neuroma
 - Metastasis to temporal bone
4. *Extracranial part*
 - Malignancy of parotid
 - Surgery of parotid
 - Accidental injury in parotid region
 - Neonatal facial injury (obstetrical forceps)
5. *Systemic diseases*
 - Diabetes mellitus
 - Hypothyroidism
 - Uraemia
 - Polyarteritis nodosa
 - Wegener's granulomatosis
 - Sarcoidosis (Heerfordt's syndrome)
 - Leprosy
 - Leukaemia
 - Demyelinating disease

Box 170-1. CONGENITAL VERSUS ACQUIRED FACIAL PARALYSIS
Congenital

Mononeural agenesis
 Congenital facial paralysis
 Congenital unilateral lower lip palsy
 Facial paralysis with other deficits
 Möbius syndrome (cranial nerves VI and VII; bilateral)
 Hemifacial microsomia
 Oculoauriculovertebral dysplasia
 Poland syndrome (agenesis of pectoralis major muscle)
 Secondary to teratogens
 Thalidomide
 Rubella

Acquired

Birth trauma
 Forceps injury
 Pressure from maternal sacrum
 Pressure from fetal shoulder
 Intracranial hemorrhage
 Idiopathic
 Bell palsy
 Systemic or infectious disease
 Melkersson-Rosenthal syndrome
 Poliomyelitis
 Infectious mononucleosis
 Varicella
 Acute otitis media
 Meningitis

Modified from Harris JP, Davidson TM, May M, Fria T: Evaluation and treatment of congenital facial paralysis. *Arch Otolaryngol* 1983;109:145.

Table 7. Etiologies and clinical features of facial paralysis.

	Condition	Etiologic Agent	Distinguishing Factors
Autoimmune	Guillain-Barré	Autoimmune/infectious	Acute polyneuropathy; ascending paralysis; weakness of hands, feet progressing to the trunk
	Melkersson-Rosenthal syndrome	Unknown	Recurrent facial paralysis, swelling of face/lips, and fissures or folds in tongue
	Multiple sclerosis	Unknown	Abnormal neurologic examination with intermittent symptoms
	Sarcoidosis	Unknown	May be bilateral; laboratory abnormalities including angiotensin-converting enzyme (ACE) level
Congenital	Möbius syndrome	Possibly viral	Age (young), bilateral in nature, unable to move face or eyes laterally
Endocrine	Diabetes	Microvascular disease	Other signs and symptoms of diabetes, laboratory testing
Idiopathic	Acute facial nerve paresis/paralysis	Unknown	Classic Bell's palsy with other etiologies excluded
Infectious	Encephalitis/ meningitis	Fungal, viral, or bacterial	Headache, stiff neck, cerebrospinal fluid abnormalities
	Herpes simplex	Herpes simplex virus along axons of nerve residing in the geniculate ganglion	Fever, malaise
	Human immunodeficiency virus (HIV)	Human immunodeficiency virus (HIV)	Fever, malaise, CD4 count
	Lyme disease	Spirochete <i>Borrelia burgdorferi</i>	May be bilateral, rash, arthralgias
	Mononucleosis	Epstein-Barr virus	Malaise, difficult to distinguish
	Otitis media	Bacterial pathogens	Gradual onset, ear pain, fever, hearing loss
	Ramsay Hunt syndrome	Herpes zoster virus	Pronounced prodrome of pain, vesicular eruption in ear canal or pharynx
	Syphilis	<i>Treponema pallidum</i>	Other neurologic and cutaneous manifestations
Inherited	Heritable disorders	Autosomal dominant inheritance	Family history as high as 4%, may have other neurologic disorders
Neoplastic	Facial nerve tumor, skin cancer, parotid tumors	Multiple carcinomas of the head and neck	May involve only select branches of the facial nerve or other cranial nerves and present as multiple cranial neuropathies
Neurovascular	Stroke	Ischemia, hemorrhage	Forehead sparing most often, extremities often involved
Traumatic	Injury to facial nerve	Trauma, including forceps delivery	Timing of injury coincides with trauma

Bell's Palsy:

- Idiopathic facial paralysis of acute onset and limited duration.
- Most common cause of facial nerve paralysis (70%).
- Epidemiology:
 - Both sexes are affected with equal frequency.
 - Any age group may be affected.
 - Greatest incidence from age 15 to 45.
 - History of previous paralysis in 10% of patients.
 - Family history of Bell palsy in 10% of patients.
- Pathophysiology:
 - **Viral Polyneuropathy:**
 - Most widely accepted theory.
 - Resulted from reactivation of latent herpes simplex virus type 1 (HSV-1) infection.
 - HSV-1 DNA was isolated in the geniculate ganglion using the PCR in 80% of patients with Bell's palsy who underwent decompression surgery.
 - Causing cranial nerve polyneuritis:
 - **Facial paralysis is the most obvious finding.**
 - Facial nerve differs from other cranial nerves by its long bony fallopian canal.
 - HSV-1 infection results in facial nerve edema within the narrowest part of the fallopian canal (**meatal foramen**/junction of meatal segment and labyrinthine segment/0.68 mm) resulting in impaired axoplasmic flow.
 - **Other cranial nerves involvement are relatively minor and transient.**
 - Found in more than 50% of patients with Bell's palsy.
 - **Ischemic Neuropathy:**
 - Vasospasm induced by cold or emotional stress.
 - Ischemia causes increased capillary permeability leading to exudation of fluid, edema and compression of microcirculation of the nerve.
 - **Hereditary:**
 - Fallopian canal is narrow because of hereditary predisposition.
 - Facial nerve is susceptible to early compression with the slightest edema.
 - Positive family history is present in 10% of patients.

TABLE 170-2. Polyneuropathy in Bell Palsy

Symptom	Incidence (%)
Hypesthesia or dysesthesia of the glossopharyngeal or trigeminal nerve	80
Hypesthesia of C2	20
Vagal motor weakness	20
Trigeminal motor weakness	3
Facial or retroauricular pain	60
Dysgeusia	57
Hyperacusis	30
Decreased tearing	17

From Adour KK: Current concepts in neurology: diagnosis and management of facial paralysis. *N Engl J Med* 1982;307:348.

- Histopathology:
 - Most recent studies demonstrated inflammatory infiltrates throughout the course of the facial nerve.
 - Vascular thrombosis is generally not observed.
 - Intra-neural hemorrhage is seen occasionally.
- Diagnosis:
 - Bell's palsy was considered to be a diagnosis of exclusion after ruling out all other possible causes.
 - Recent criteria to diagnose Bell's palsy:
 - 1. Unilateral weakness of all facial muscles.**
 - 2. Sudden onset.**
 - 3. No evidence of otologic disease.**
 - 4. No evidence of CNS disease.**
 - 5. No evidence of CPA lesion.**
- Risk factors for developing Bell's palsy:
 - **Pregnancy**
 - **Obesity**
 - **Hypertension**
 - **Diabetes**
 - **Upper respiratory illness**

- Clinical Features:

- **Unilateral weakness of all facial muscles:**
 - Involving the forehead muscles (LMNL).
 - Left and right sides of the face are equally involved.
 - Bilateral involvement is rare (0.3%).
 - 70% of patients have complete paralysis.
 - 30% of patients have incomplete paralysis.
- **Sudden onset:**
 - Progresses to its maximum severity within 72 hours of onset.
 - Delayed progression over weeks or months is not Bell palsy.
- **Dysgeusia:**
 - Loss of taste.
 - Due to involvement of chorda tympani.
- **Hyperacusis:**
 - Noise intolerance due to stapedial paralysis.

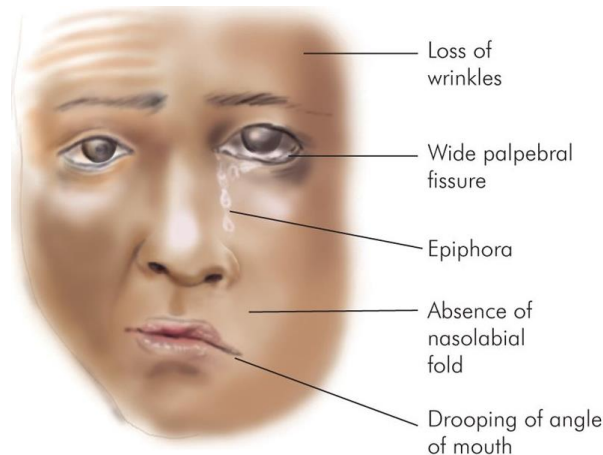


Figure 14.6 Facial paralysis left side. Compare with normal side.

- Investigations:

- **Routine labs:**
 - Not indicated in patients with new onset Bell's palsy.
- **Imaging:**
 - Not indicated in patients with new onset Bell's palsy.
 - MRI with Gad of entire course of facial nerve is the imaging test of choice.
 - Indications of imaging in Bell's palsy:
 1. Recurrent ipsilateral facial paralysis:
 - 20% risk of having tumor.
 2. Bilateral facial paralysis.
 3. Paralysis of isolated branches of the facial nerve.
 4. Other cranial nerve involvements.
 5. No sign of recovery after 3 months.

- **Electrodiagnostic tests:**
 - Indication:
 - Complete facial paralysis.
 - Should not performed in patients with incomplete facial paralysis due to the high chance of recovery.
 - Aim:
 - Identify patients with poor prognosis.
 - Identify candidates for surgical decompression.
 - Period:
 - Done between 3-21 days of onset.
 - Approach:
 - **ENoG is done first:**
 - If degeneration of $< 90\%$ of axons:
 - Indicates good prognosis.
 - Most patients recover normal or near-normal facial movement.
 - If degeneration of $\geq 90\%$ of axons:
 - Indicates poor prognosis.
 - Most patients do not recover.
 - **EMG is then done:**
 - If voluntary muscle potential was recorded:
 - Indicates intact axons.
 - Possible good recovery.
 - If no voluntary muscle potential was recorded:
 - Indicates damaged axons.
 - Poor recovery.
 - May consider surgical decompression.

- **Prognosis for Bell's palsy:**

- Most patients will recover spontaneously without treatment.
- The sooner the recovery, the less likely are the chances that sequelae will develop:
 - 85% of patients will show some recovery within 3 weeks and the ultimate outcome is usually satisfactory.
 - If recovery begins 2-4 months from onset, high risk of permanent sequelae.
- Complete recovery is typically achieved by 2 months:
 - Patients with complete paralysis will have complete recovery after 3-5 months.
- Risk factors associated with a poor outcome:
 1. Complete paralysis:
 - Single most important prognostic factor.
 2. Age greater than 60 years.
 3. Long interval time of before onset of recovery.
 4. Pain in the posterior auricular area.
 5. Hyperacusis.
 6. Decreased tearing.
 7. Diabetes mellitus.
 8. Hypertension.
- Overall rate of spontaneous complete recovery within 6 months:
 - **95%** of patients with incomplete paralysis.
 - **70%** of patients with complete paralysis.
- Patients on steroid therapy has 20-30% better chance of complete recovery compared to untreated patients.
- No difference in prognosis between primary and subsequent attacks of facial paralysis.
- Recovery in bilateral Bell palsy is similar to unilateral palsy.

Management of Bell's palsy:

- Medical Management:

1. Oral Prednisone:

- Dose:
 - 1 mg/kg/day for 5 days, then tapered over 5 days, for total of 10 days.
 - Used within 72 hours from onset of symptoms.
- Strong recommendation:
 - Decrease recovery time.
 - Improving facial nerve functional recovery.
 - Prevents or lessens nerve degeneration.
 - Decreases synkinesis.
- Oral steroids may be considered in pediatric patients.

2. Oral Pantoprazole:

- 40 mg daily.
- Decrease risk of peptic ulcer with oral prednisone.

3. Oral Anti-viral:

- Dose:
 - Acyclovir 400 mg PO 5 times daily for 10 days, or
 - Valacyclovir 500 mg PO BID for 5 days.
 - Used within 72 hours from onset of symptoms.
- Optional:
 - Antiviral therapy combined with oral steroids was not statistically significantly superior to oral steroids alone.
 - Small benefit cannot be completely excluded.
 - Antiviral agents alone provide no benefit.
 - Should not be given as monotherapy in new-onset disease (Strong recommendation against).

4. Eye Care:

- Ophthalmic evaluation to rule out exposure keratitis.
- Strong recommendation:
 - Use of sunglasses.
 - Frequent use of lubricating ophthalmic drops and ointments.
 - Use of a moisture chamber.
 - Eye patching or taping during sleep.

- Surgical Management:
 - **Surgical Decompression:**
 - Controversial.
 - No strong data supporting surgical decompression.
 - Can be considered for patients high risk of poor recovery:
 - Patients with complete facial paralysis and ENoG is showing $\geq 90\%$ axonal degeneration and EMG is showing absent voluntary muscle potential.
 - Period:
 - Surgery must be performed within 14 days of symptom onset for optimal effectiveness.
 - Aim:
 - Decompression of the labyrinthine segment.
 - Decompression of mastoid and tympanic segments no effect on recovery of facial function.
 - Approach:
 - Middle cranial fossa approach.
 - Trans-mastoid approach is not used as the segment involved (proximal labyrinthine segment) is inaccessible through the mastoid.
 - Results:
 - 90% of patients achieved HB outcome of I/II postoperatively compared with only 40% of patients who underwent medical management with steroid-only.
- **No sufficient evidence to recommend for Acupuncture or Physical therapy for management of Bell's palsy.**

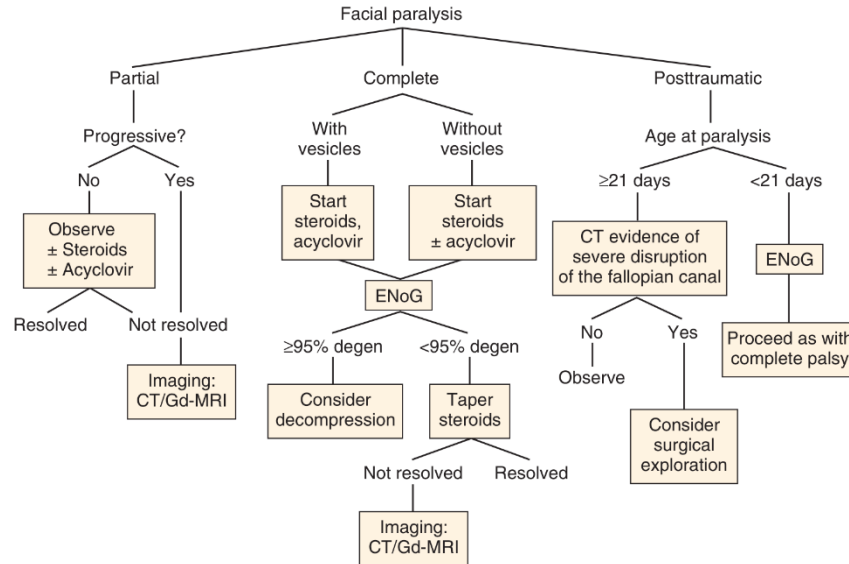


FIGURE 170-1. Management algorithm for facial paralysis. CT, computed tomography; degen, degeneration; ENoG, electroneuronography; Gd, gadolinium enhanced; MRI, magnetic resonance imaging.

Facial Nerve Trauma:

- Second most common cause of facial paralysis (20% of all cases).
- Types:
 - **Temporal bone fractures:**
 - See Temporal bone trauma for more details.
 - **Penetrating facial nerve injuries:**
 - Intra-temporal part:
 - See Temporal bone trauma for more details.
 - Extra-temporal part:
 - **For complete paralysis:**
 - Surgical exploration and repair with an end-to-end anastomosis should be undertaken as soon as the patient is medically stable.
 - Ideally repaired within three days (distal branches may be stimulated up to 3 days after injury).
 - If the wound is contaminated or there is significant tissue loss may consider identifying distal and proximal ends of the facial nerve with plans for a second stage procedure within 3–4 weeks.
 - Electrical stimulation can still be used to help locate the distal branches.
 - **If the penetrating injury is medial to lateral canthus of the eye:**
 - Recover spontaneously.
 - No need for exploration.
 - Nerve endings are small and a rich anastomotic network exists in this area.

**TABLE
155.5****MANAGEMENT OF TRAUMATIC INJURIES WITH COMPLETE FACIAL NERVE PARALYSIS**

Mechanism of Injury	Tests	Critical Result	Management
Intratemporal Injury			
Blunt trauma (temporal bone fracture)	CT	Fracture involving fallopian canal	
	NET	>3.5 mA difference	
	ENoG	>90% degeneration	
	EMG	Absent volitional activity	Surgical exploration if all of above present
Penetrating trauma	Audiometry	Conductive loss	Transmastoid–middle fossa approach
	History	Sensorineural loss—severe	Translabyrinthine approach
	Follow algorithm above, add: Angiography	Delayed onset	Observation
		Suspicion of major vascular injury	Endovascular or open repair
Iatrogenic	History	Immediate onset	Surgical exploration
		Delayed onset (>3 d)	Observation
Extratemporal Injury			
Penetrating trauma	Physical exam		Exploration and repair
Blunt trauma	Physical exam		Observation
Iatrogenic	History	Suspected transection by report	Exploration and repair
		Nerve intact	Observation

NET, nerve excitability test; ENoG, electroneurography; EMG, electromyography.

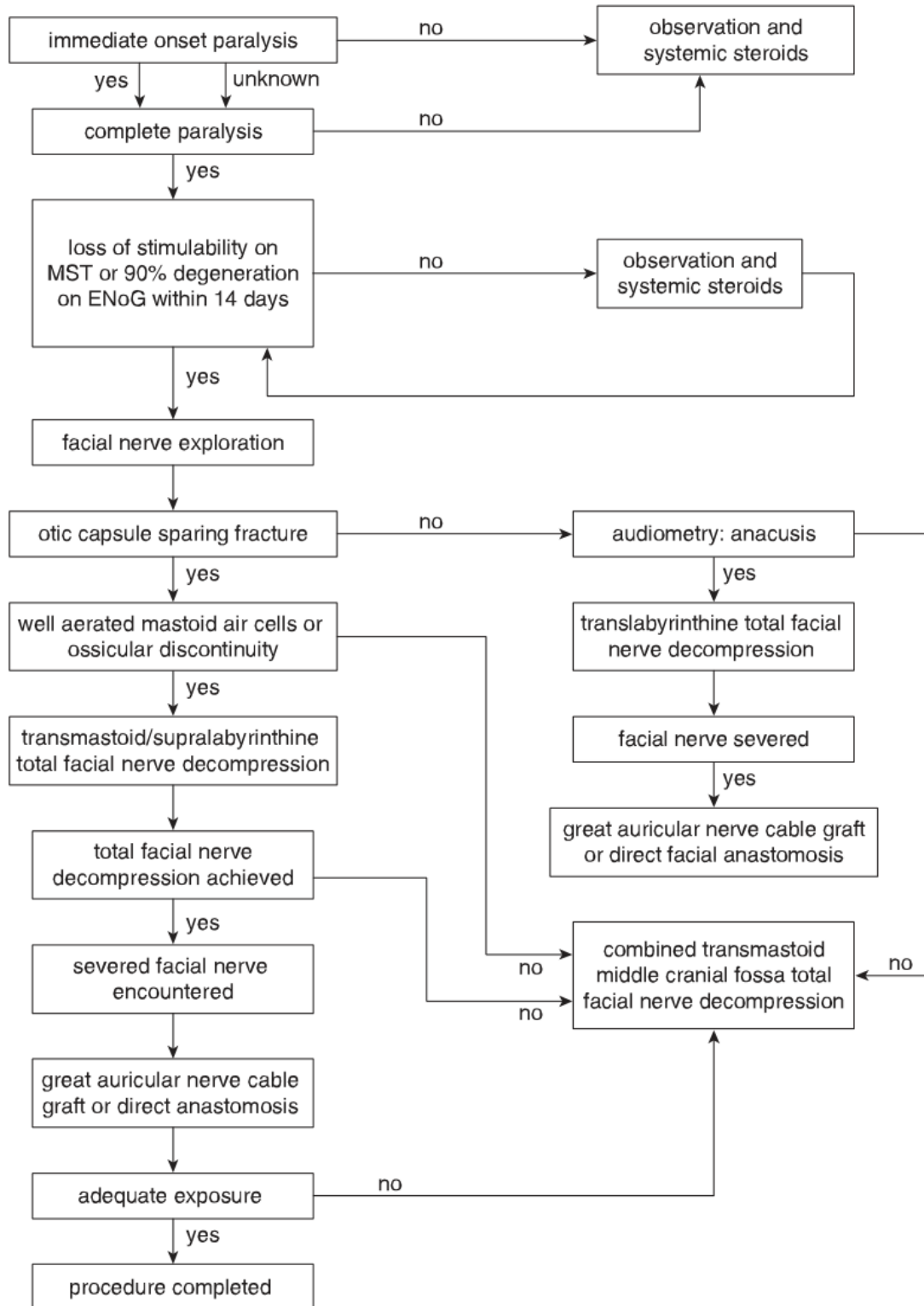


Figure 150.11 Management of traumatic facial paralysis.

- **Iatrogenic facial nerve injuries:**
 - Occurs in < 1% during middle ear or mastoid surgeries.
 - Most common sites of facial nerve injury during ear surgery (most common sites of facial nerve dehiscence):
 - 1. Tympanic segment (80%):**
 - Superior or adjacent to oval window.
 - 2. Vertical segment (1%).**
 - Facial nerve injury may also occur with salivary gland surgery, neck dissections, rhytidoplasties, and branchial cleft excisions
 - Management of iatrogenic injury during ear surgery:
- **Confirmed intraoperative injury:**
 - Injury to fallopian canal with exposure of epineurium with minor abrasion of nerve sheath:
 - No consequence.
 - Transection of < 30-40% of nerve diameter:
 - **Expert colleague must be consulted.**
 - Fallopian canal should be decompressed 3-4 mm proximal and distal to injured segment and epineurium should be incised.
 - Intra-op and post-op steroids.
 - Transection of > 40-50% of nerve diameter:
 - **Expert colleague must be consulted.**
 - Injured segment is resected and repaired with primary anastomosis or cable grafting.
- **Immediate postoperative facial weakness:**
 - Tight dressings and packing should be released immediately to relieve the pressure over facial nerve.
 - Wait for local anesthetic to wear off (2–3 hours) then re-evaluate.
- **If still paralyzed:**
 - Surgeon is confident about Facial nerve preservation intra-op:
 - Expert colleague must be consulted.
 - Corticosteroids.
 - Follow progression with serial electrodiagnostic testing.
 - If advanced degeneration (>90%) is evident:
 - Surgical exploration and Decompression.

- Surgeon is Not confident about Facial nerve preservation intra-op:
 - Surgical exploration and repair as soon as possible by an expert surgeon.
- **Delayed postoperative facial weakness:**
 - More common than immediate-onset paralysis.
 - Occurs after 3 days post-surgery.
 - Usually due to viral reactivation.
 - Treated with steroids.
 - Good recovery is anticipated.
 - In cases of delayed complete paralysis:
 - The patient is followed up with electrodiagnostic tests to determine degree of injury.
 - Loss of electrical excitability > 90% degeneration by ENoG within 14 days of mastoid surgery is an indication for nerve exploration.
- **Operative injuries to Facial nerve can be avoided if attention is paid to the following:**
 1. Anatomical knowledge of Facial nerve course, possible variations and anomalies and its surgical landmarks.
 2. Always working along course of nerve and never across it.
 3. Constant irrigation when drilling, to avoid thermal injury.
 4. Use diamond burr when working near the nerve.
 5. Gentle handling of the nerve when it is exposed, avoiding any pressure of instruments on the nerve.
 6. Not to remove any granulations that penetrate the nerve.
 7. Using magnification; never to work on facial nerve without an operating microscope.

- Surgical Landmarks of Facial Nerve in Ear and Mastoid Surgery:

1. Processus Cochleariformis:

- Geniculate ganglion (1st Genu) lies Anterior to it.
- Tympanic segment of CN-VII starts at this level.

2. Horizontal SCC:

- Tympanic segment of CN-VII runs below Horizontal SCC.
- 2nd Genu of Facial nerve runs Infero-lateral to Horizontal SCC.

3. Oval Window:

- Tympanic segment of CN-VII runs above Oval window (Stapes).

4. Short Process of Incus:

- Tympanic segment of CN-VII lies medial to Short process of Incus at level of Aditus.

5. Pyramid:

- Mastoid segment of CN-VII runs behind Pyramid and Posterior Tympanic sulcus.

6. Facial Recess:

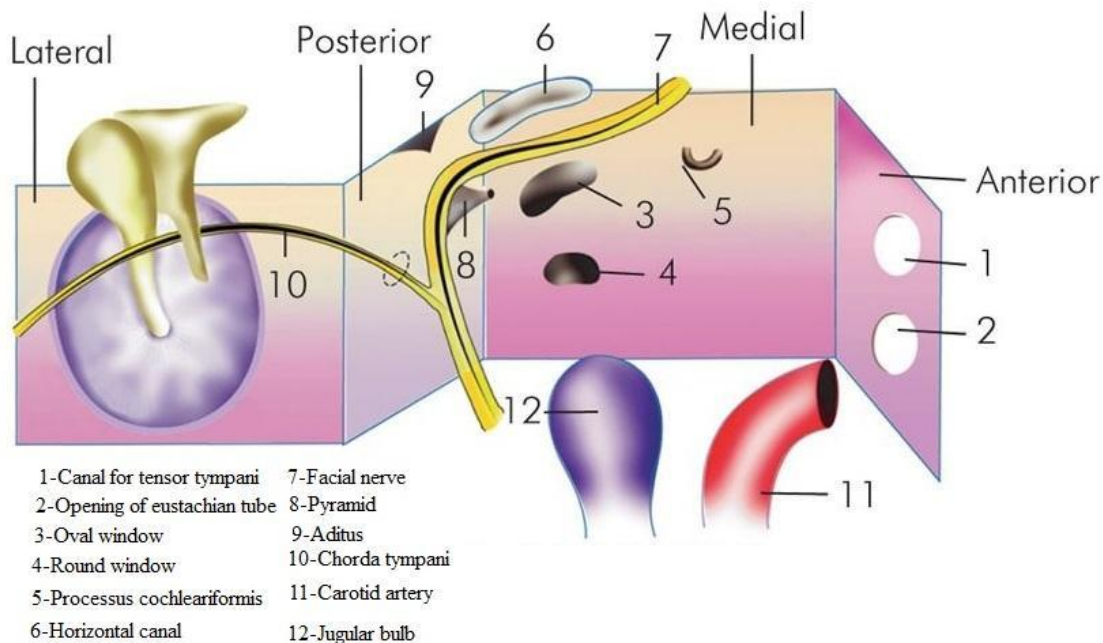
- Long process of Incus points toward Facial recess.
- Chorda tympani nerve serves at Lateral margin of Triangular facial recess.
- Chorda tympani nerve can be exposed along its length and can be followed inferiorly and medially to its takeoff from the main trunk of Facial nerve.

7. TympanoMastoid suture:

- Mastoid segment of CN-VII runs behind this suture.

8. Digastric Ridge:

- Mastoid segment of CN-VII leaves Mastoid at Anterior end of Digastric Ridge.



- **Surgical Landmarks of Facial Nerve in Parotid Surgery:**

1. Cartilaginous Pointer:

- Sharp triangular piece of cartilage of Pinna points to Facial Nerve.
- Extra-Temporal part of Facial Nerve lies 1 cm inferior and medial to the pointer.

2. Tympanomastoid Suture:

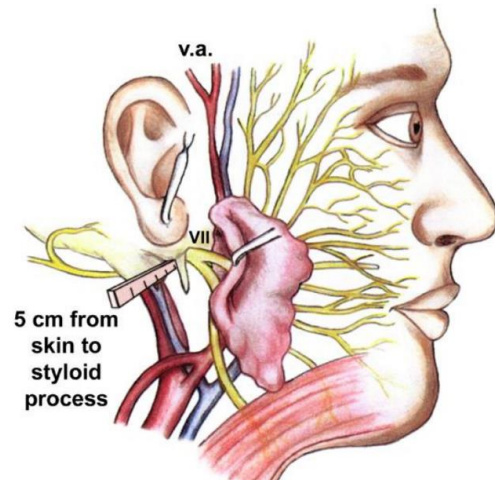
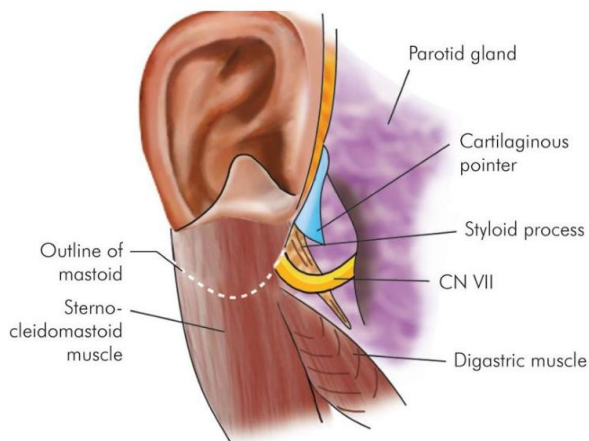
- Between Mastoid and Tympanic part of Temporal bone.
- Facial Nerve lies 6-8 mm deep to this suture in Stylomastoid foramen.

3. Styloid Process:

- Facial Nerve crosses lateral to Styloid process.

4. Posterior belly of Digastric:

- Facial Nerve lies between attachment of Posterior belly of digastric muscle to Digastric groove and Styloid process.



Herpes Zoster Oticus (Ramsay Hunt Syndrome):

- 3rd most common cause of facial nerve paralysis.
- Herpes zoster induced Facial nerve palsy.
- Pathophysiology:
 - o Reactivation of latent Varicella zoster virus (VZV) in Geniculate ganglion of **CN-VII** during a period of decreased cell mediated immunity.
 - o Rising titers of antibodies to VZV.
- Compared to Bell's palsy:
 - o More severe symptoms.
 - o Worse prognosis with residual weakness in 30-50% of patients.
 - o Higher involvement of other cranial nerves.
- Clinical picture:
 - o **Acute Peripheral Facial Palsy.**
 - o **Severe otalgia.**
 - o **Painful vesicular eruption in Concha, EAC or TM:**
 - Occurs simultaneously with the paralysis in most cases.
 - May precedes the paralysis in 25% of cases, and has a higher chance of recovery.
 - o **SNHL and vertigo:**
 - Involvement of **CN-VIII** (30% of cases).
 - o **Herpes zoster ophthalmicus:**
 - Involvement of ophthalmic division (V1) of trigeminal nerve.
 - Include uveitis, keratoconjunctivitis, optic neuritis and glaucoma.



- Treatment:
 - 1. Oral Prednisone:**
 - Relieve acute pain.
 - Reduce vertigo.
 - Decrease incidence of post-herpetic neuralgia.
 - 2. Oral Anti-viral:**
 - **Acyclovir 800 mg PO 5 times daily for 10 days, or**
 - **Valacyclovir 1g PO TID for 5 days.**
 - 3. Eye Care.**

Otitis Media:

- AOM:

- Toxic effects from infectious spread into the nerve sheath results in facial nerve dysfunction.
- Dehiscence in Fallopian canal may serve as portals for direct bacterial invasion and inflammation along the nerve.
- Facial paralysis may begin within a few days of onset of an acute otitis media and is usually incomplete.
- Treatment includes myringotomy, drainage, C/S and Antibiotic.
- Facial palsy associated with acute otitis media generally resolves with aggressive management of the infection.

- CSOM:

- Facial nerve paralysis may occur from compressive effects from a cholesteatoma or from granulation tissue.
- Progressive unilateral facial palsy with suppurative otitis media.
- CT may reveal cholesteatoma or soft tissue compression.
- Tympanomastoidectomy should be performed as soon as possible with facial nerve exploration and decompression without opening the perineurium.

Malignant (Necrotizing) Otitis Externa:

- Progressive unilateral facial palsy due to Facial nerve injury from the effect of skull base osteomyelitis.
- Caused by pseudomonas infection in immuno-compromised patients.
- Clinical picture:
 - Granulation tissue in EAC at bony-cartilagenous junction.
 - Persistent otalgia and otorrhea.
 - Cranial nerve involvement (CN-VII > CN-X > CN-XI)
- Diagnosis:
 - History and Physical exam.
 - Elevated ESR.
 - CT Temporal bone.
 - Technetium 99 bone scan:
 - Evaluates osteoblastic activity.
 - Excellent for Diagnosis of Acute or chronic process.
 - Positive for prolonged period.
 - Gallium 67 bone scan:
 - Evaluates Inflammation
 - Excellent for Follow up the course.
 - Fades if disease resolves.
 - Biopsy with culture of EAC.
- Management:
 - Prolonged IV anti-pseudomonas antibiotics (6-8 weeks)
 - Anti-pseudomonas Antibiotics ear drops.
 - Aggressive Diabetic control.
 - Meticulous cleaning and debridement.
 - Surgical debridement if medical management failed.

Tumors of the Skull Base and Facial Nerve:

- 5% present with facial paralysis.
- Suspected in the following cases:
 1. Slow progression over > 3 weeks.
 2. No return of facial function over time (> 3 months).
 3. Recurrent ipsilateral paralysis.
 4. Concurrent facial twitching with slow paresis.
 5. Multiple cranial nerve deficits.
 6. Presence of neck or parotid mass.
 7. History of cutaneous malignancy.
 8. Chronic ET dysfunction with no history of middle ear disease.
- Most common tumor of the facial nerve is **facial schwannoma**:
 - If in CP angle or IAC, present with hearing loss/vertigo.
 - Extra-temporal rarely have paralysis; parotid mass instead.
 - Geniculate ganglion and labyrinthine most common sites.
 - Need interposition graft; Grade III is the best outcome.
 - If young, operate early; if old, watch it grow.



FIGURE 170-2. Axial computed tomography scan of the right temporal bone shows enlargement of the mastoid segment of the fallopian canal (arrow) in a child with progressive facial paralysis. The lesion was a neuroma of the facial nerve.

- Most common malignancy to cause facial nerve paralysis is **parotid tumors**:
 - Mucoepidermoid carcinoma is the most common tumor to cause facial dysfunction, although adenoid cystic carcinoma has a higher rate of neural involvement.
- If malignant tumors involving the facial nerve:
 - Typically require excision with tumor-free margin.
 - Except (spare the facial nerve intra-operatively):
 - Lymphoma
 - Leukemic invasion
 - Low-grade parotid lesions

Melkersson-Rosenthal Syndrome:

- Idiopathic disorder.
- Begins in 2nd decade of life.
- Symptoms appears sequentially and rarely simultaneously.
- Triad:
 - 1. Recurrent orofacial edema:**
 - Defining feature.
 - Non-pitting edema.
 - Cannot be explained by any other cause.
 - 2. Recurrent facial paralysis:**
 - Occurs in 50% of patients.
 - Unilateral or bilateral.
 - 3. Fissured tongue (Lingua plicata):**
- Diagnosis:
 - **Biopsy of lip:**
 - Non-caseating epithelioid cell granulomas surrounded by histiocytes, plasma cells and lymphocytes.
- Treatment:
 - Multiple treatments have been tried (steroids, metronidazole, dapsone, acyclovir and methotrexate) without a consistent response to therapy.



Bilateral Facial Paralysis:

- Has higher incidence of systemic causes than unilateral palsy.
- Mostly associated with other neurologic signs.
- Requires careful work-up:
 - **Full neurologic examination:**
 - Detect other cranial neuropathy.
 - **Serology:**
 - Blood chemistry.
 - Blood culture.
 - VDRL for syphilis.
 - **Lumbar puncture:**
 - For cytological studies.
 - **MRI:**
 - Exclude space occupying lesions.
- Most common causes:
 - **Guillain-Barré syndrome:**
 - **Most common cause in adults.**
 - Progressive ascending motor paralysis after viral infection
 - Usually affects initially the lower limbs.
 - Diagnosed based on typical clinical picture and presence of elevated spinal fluid protein with normal cell count.
 - **Lyme disease:**
 - **Most common cause in children.**
 - Responsible for half of all cases of pediatric facial paralysis in endemic areas.
 - **Bell's palsy (0.3%)**
 - **Pontine tumor**
 - **Benign intracranial hypertension (BIH)**
 - **Bilateral Neurofibromas (Neurofibromatosis type 2).**
 - **Bacterial meningitis.**
 - **Brainstem encephalitis**
 - **Myasthenia Gravis**
 - **Syphilis**
 - **Leukemia**
 - **Sarcoidosis:**
 - Chronic non-caseating granulomatous disease.
 - Heerfordt syndrome (Uveoparotid fever):
 - Non-suppurative parotitis
 - Uveitis
 - Mild fever
 - Cranial nerve paralysis (Mainly facial paralysis).
 - From direct invasion of facial nerve by the granulomatous process.

Facial Paralysis in Newborns:

- Important to differentiate between true congenital paralysis and birth trauma.
- **Birth Trauma (80%):**
 - Most common cause of unilateral facial paralysis in neonates.
 - Pathophysiology:
 - Extra-temporal part of facial nerve is at risk for compressing injury during delivery as it courses the underdeveloped mastoid process.
 - Intrauterine facial nerve injury can occur from pressure on infant's face by sacral prominence during birth.
 - Risks:
 - Forceps delivery.
 - Prolonged delivery (primiparity).
 - Large infant (BW > 3.5 kg).
 - Clinical picture:
 - Asymmetric crying face
 - Facial contusion
 - Hemotympanum
 - Peri-auricular ecchymosis
 - Diagnosis (Done for complete paralysis):
 - EMG (preserved neuromuscular activity suggests inherited or developmental etiology)
 - Electrophysiological testing within first three days.
 - ABR (evaluate for associated hearing loss)
 - Management:
 - Observation.
 - Prognosis for spontaneous recovery is excellent
- **Congenital Unilateral Lower Lip Palsy:**
 - Most common true congenital cause of facial paralysis.
 - Due to hypoplasia of depressor labii inferioris muscle.
 - Associated with cardiac defects ($\approx 10\%$).
- **Möbius Syndrome:**
 - Wide spectrum of abnormalities secondary to central brain stem and peripheral neuromuscular defects.
 - Bilateral or unilateral Facial and Abducens nerve palsies.
 - Club foot (talipes equinovarus).
 - Tongue weakness.
 - Mixed hearing loss.
 - Mental retardation.
 - External ear deformities.
 - Ophthalmoplegia.

Sequelae and complications of Facial nerve paralysis:

1. Incomplete recovery:

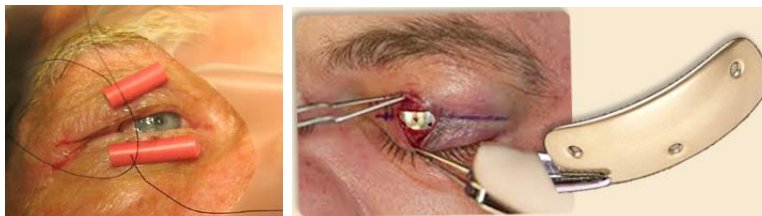
- Largest portion of Facial nerve is composed of Efferent fibers that stimulate muscles of facial expression.
 - Suboptimal regeneration of this portion results in paresis of all or some of these facial muscles.
- Dysgeusia (impairment of taste) or Ageusia (loss of taste) may occur with incomplete regeneration of Chorda tympani.
- Treated with facial re-animation producers.

2. Aberrant re-innervation of the facial nerve:

- Synkinesis is condition in which involuntary movements accompany voluntary movements.
- During regeneration, some neural fibers may take an unusual course and connect to neighboring muscle fibers.
- Produces unusual neurologic pathways.
- When voluntary movements are initiated, they are accompanied by involuntary movements (eg, eye closure associated with lip pursing or mouth grimacing that occurs during blinking of the eye).
- Improved with Botox injections.

3. Exposure keratitis:

- Eye cannot be closed, tear film from the cornea evaporates causing dryness, exposure keratitis and corneal ulcer.
- Worse when tear production is also affected.
- Prevented by:
 - **Eye care:**
 - Artificial tears every 1-2 hours
 - Eye ointment and proper cover for the eye at night.
 - **Tarsorrhaphy:**
 - Upper and lower eyelids are partially sewn.
 - Indicated temporarily for severe cases.
 - **Upper eyelid gold-weight implant:**
 - Indicated for patients with long-term complete facial paralysis to improve eye closure.
 - Upper eyelid gold-weight implant sutured to the tarsal plate deep to levator palpebrae muscle.



4. Contractures:

- Result from fibrosis of atrophied muscles or fixed contraction of a group of muscles.
- Affect movements of face but facial symmetry at rest is good.

5. Bogorad's syndrome (Gustatory Lacrimation/Crocodile tears):

- Unilateral lacrimation during mastication.
- Etiology:
 - **Insult to facial nerve.**
 - Causes:
 - **Trauma (temporal bone trauma)**
 - **Bell's palsy**
- Pathophysiology:
 - **Insult proximal to geniculate ganglion (Most common):**
 - Aberrant cross-reinnervation between preganglionic secretomotor parasympathetic fibers to submandibular and sublingual glands (Chorda Tympani) and the preganglionic parasympathetic fibers to lacrimal gland (Greater superficial petrosal nerve).
 - **Insult distal to geniculate ganglion:**
 - Aberrant cross-reinnervation between preganglionic secretomotor parasympathetic fibers to parotid gland (Lesser petrosal nerve) and the preganglionic parasympathetic fibers to lacrimal gland (Greater superficial petrosal nerve).
- Treatment:
 - **Botox injection into lacrimal gland.**
 - Surgical management (Vidian neurectomy).

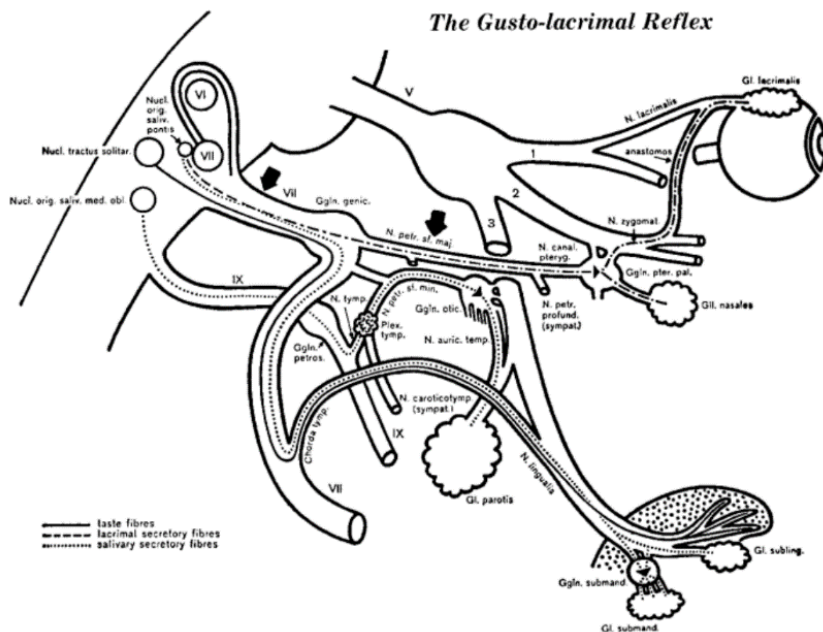


Fig. 17-26. *Gustatory lacrimation*. Diagram shows two possible mechanisms for production of the syndrome. Most often, lesion is proximal to geniculate ganglion and in regeneration, fibers destined for the submandibular and sublingual salivary glands may become interchanged with those for the lacrimal gland. The lesion may also be distal to the geniculate ganglion and involve interchange of fibers of the greater and lesser superficial petrosal nerves. [From A Axelsson and JE Laage-Hellman, *Acta Otolaryngol* (Stockholm) 54: 239, 1962.]

6. Frey's syndrome (Gustatory sweating):

- Auriculo-temporal nerve syndrome.
- Unilateral facial skin flushing and sweating during mastication.
- Occurs within first year of insult, but may be delayed more.
- Etiology:
 - **Insult to the auriculo-temporal nerve.**
 - Causes:
 - **Iatrogenic (Parotid surgery):**
 - Most common cause.
 - Occurs in 30-60% of patients.
 - Symptomatic Frey syndrome occurs in 10% only of these patients.
 - **Blunt trauma**
 - **Infection (Bell's palsy, Herpes zoster).**
- Pathophysiology:
 - Aberrant cross-reinnervation between postganglionic secretomotor parasympathetic fibers to parotid gland (Auriculotemporal nerve) and the postganglionic sympathetic fibers that supply the sweat glands of skin.
 - Occurs as both sympathetic and parasympathetic fibers use acetylcholine neurotransmitters.

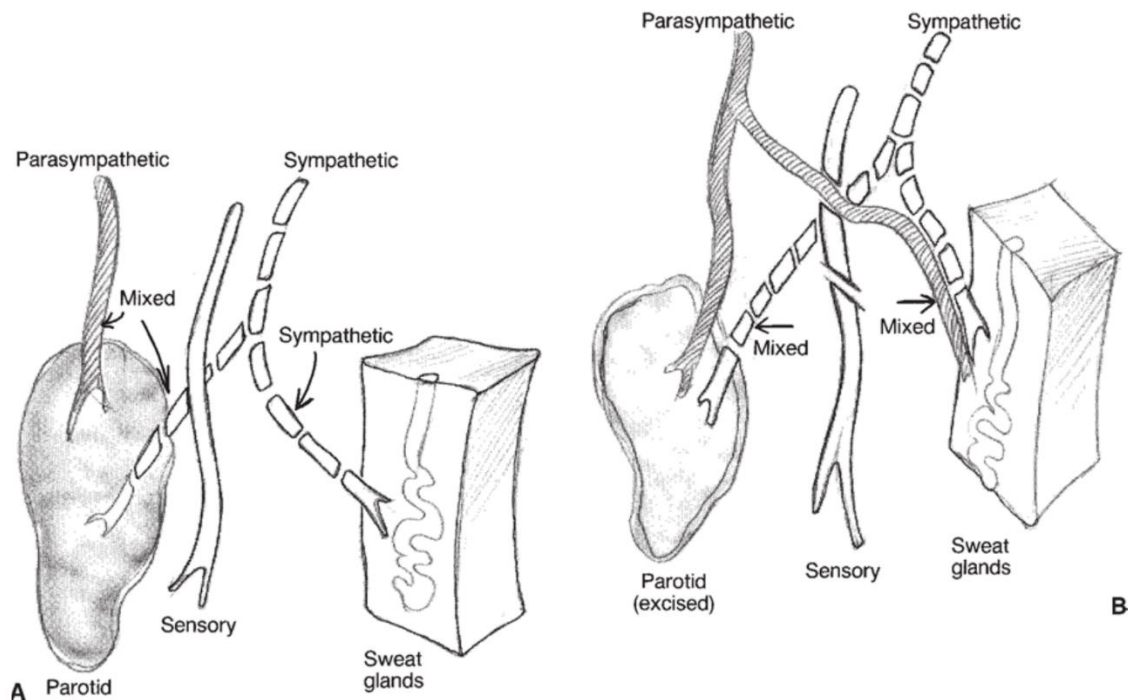


Figure 115.21 A: Normal innervation of parotid and sweat glands. B: Proposed mechanism of Frey syndrome.

○ Diagnosis:

▪ **Minor starch-iodine test:**

- Objective test to confirm the diagnosis.
- Method:
 - Ipsilateral side of face and neck is painted with iodine solution and allowed to dry.
 - Starch powder is dusted over the painted area.
 - Patient is allowed to chew on a sialagogue such as a lemon wedge, for several minutes.
 - Appearance of **dark blue spots** as a result of the reaction of dissolved starch with iodine confirms the diagnosis.
- Positive in 96% of patients underwent parotid surgery even if they didn't complain clinically.



○ Treatment:

▪ **Observation:**

- For non-bothersome symptoms.

▪ **Medical management:**

- Topical Anti-perspirant
- Topical Anticholinergic (Glycopyrrolate lotion)
- Botox injections (excellent control for prolonged periods).

▪ **Surgical management (rarely indicated):**

- Placement of inter-positional barriers:
 - Fat, acellular dermal grafts or muscle flap.
- Tympanic neurectomy:
 - Controversial.
 - Jacobson's nerve section via tympanotomy approach.
 - High incidence of recurrence.

○ Prevention during parotid surgery:

- Elevation of a thick skin flap.
- Performance of partial parotidectomy.
- Placement of inter-positional barriers (fat, acellular dermal grafts or muscle flap).

Facial Nerve Rehabilitation (Repair and Reanimation)

- Goals of Facial Rehabilitation:
 - **Functional:**
 - Protect the eye from corneal exposure.
 - Improve sphincteric function of eye and mouth
 - Improve drooling and articulation.
 - **Cosmetic:**
 - Provide a balanced smile.
 - Provide symmetry at rest.
- General principles:
 - Whenever the continuity of facial nerve has been disrupted, every effort should be made to restore its continuity.
 - Early exploration and nerve repair is performed when possible.
 - If concomitant injuries prohibit immediate direct repair, the wound should be explored and nerve ends tagged for future exploration.
 - Nerve injuries medial to the lateral canthus do not require repair because of extensive crossover.
 - Eye care should be continued during the planning time for surgical correction.

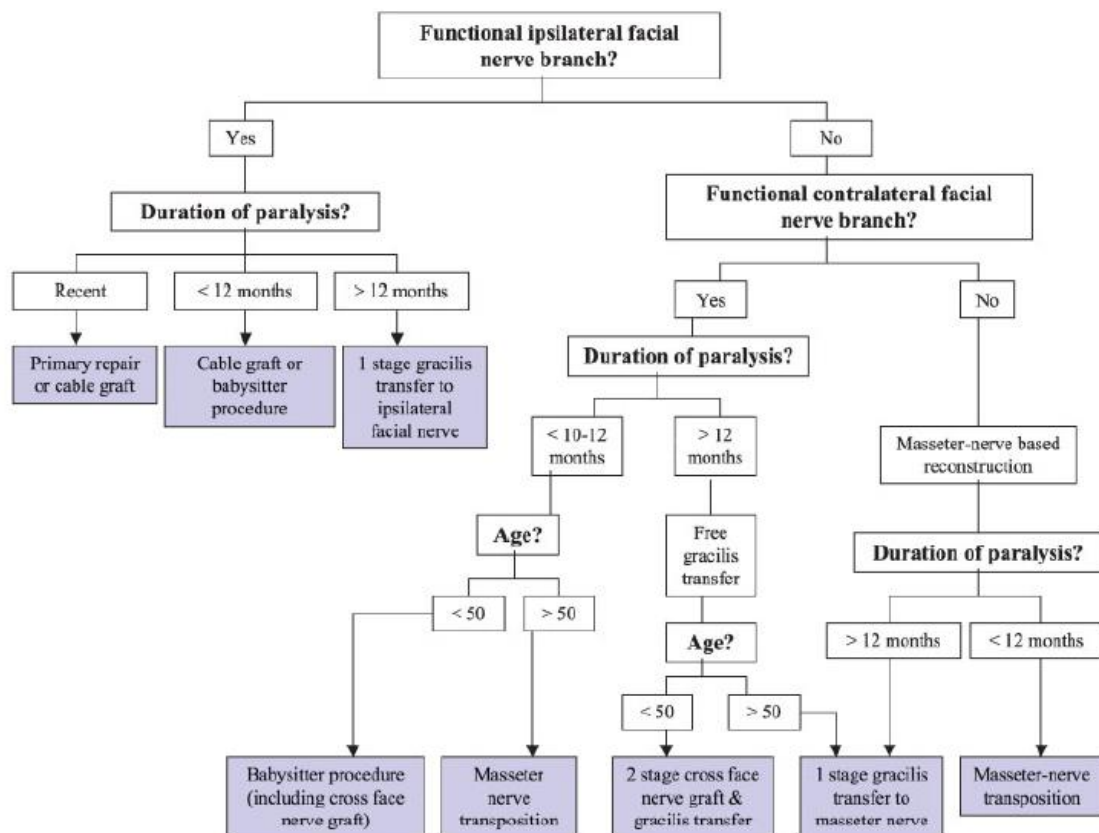
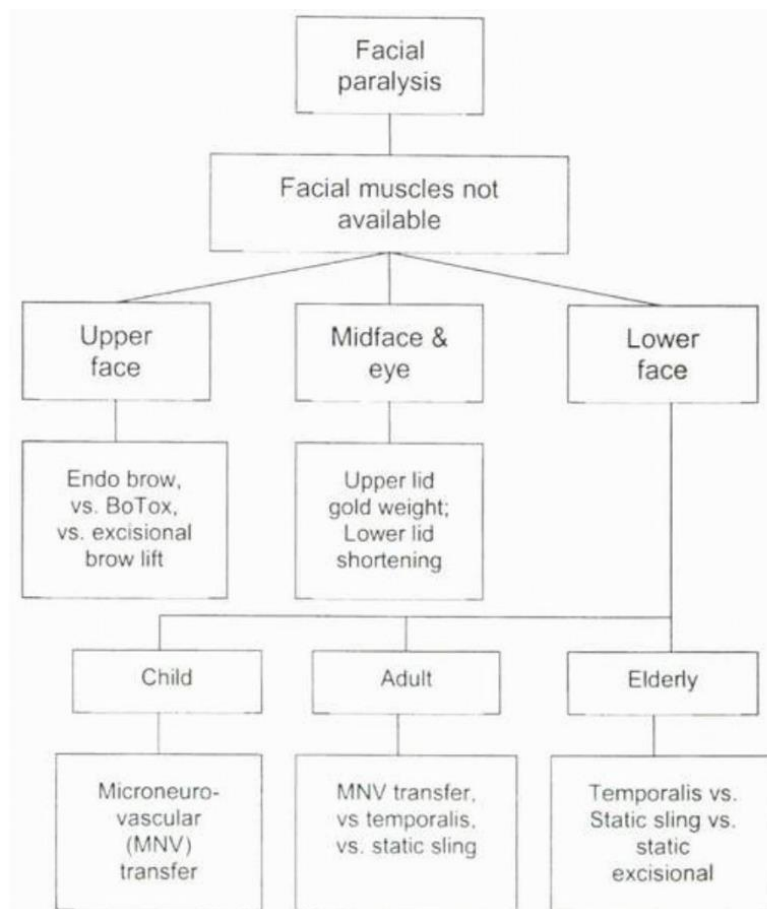
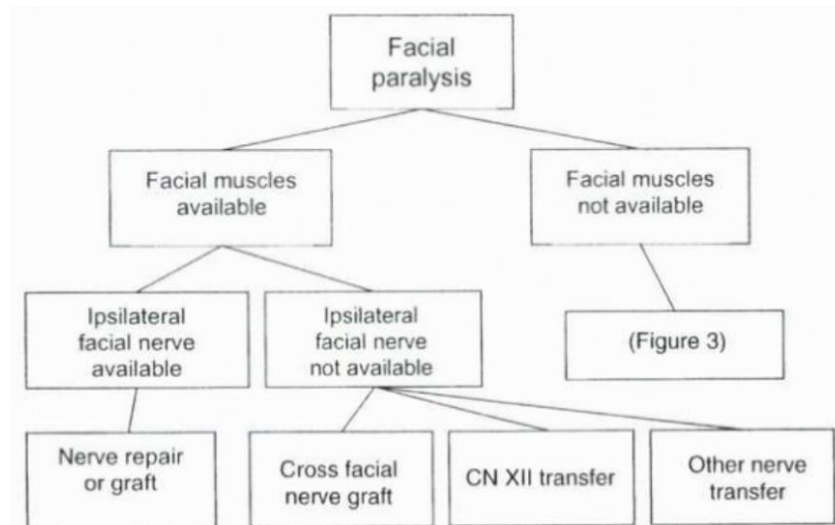


Figure 6 Suggested algorithm for dynamic reconstruction of facial paralysis.

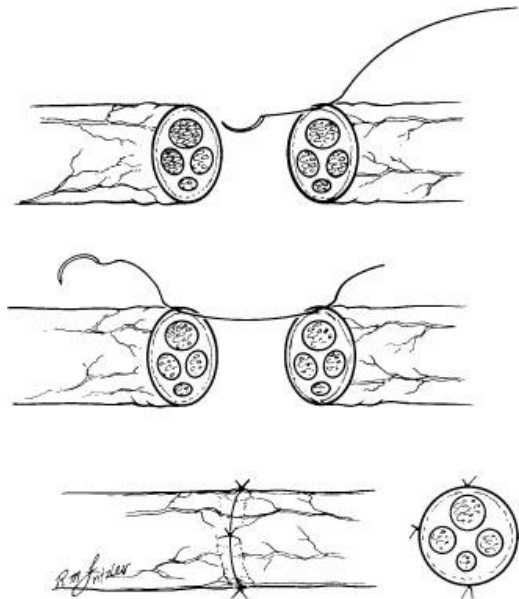
- **Early management of facial paralysis (<2 years):**
 - Ideally nerve repair should be done within 3 days of injury as the distal branches can be stimulated to help in identification.
 - Re-innervation is best within 6 months post-paralysis.
 - All nerve repair methods produce synkinesis but sphincteric function of the mouth and eye are restored.
 - Best result for any nerve repair surgery is HB grade III.
 - In Electrodiagnostic tests:
 - EMG is showing fibrillation potentials.
 - Muscles are not de-innervated.
 - No muscle atrophy or loss of motor end plates.
 - Methods of early facial re-innervation:
 1. Primary end-to-end anastomosis (Neurorrhaphy)
 2. Interposition (cable) graft
 3. Cross-facial nerve graft
 4. Nerve transfer
- **Delayed management of facial paralysis (>2 years):**
 - In Electrodiagnostic tests:
 - EMG is showing Electrical silence.
 - Muscles are atrophied.
 - No functioning motor end plates.
 - Important to identify which aspects of facial paralysis are most troublesome for the patient, so you can treat each region of the face separately:
 - Methods of Delayed facial rehabilitations:
 1. **Dynamic procedures:**
 - Muscle transfer:
 - Indicated for adults (<50 years) with average motivation.
 - Free Nerve Muscle Transfer:
 - Indicated for adults (<50 years) with high motivation.
 2. **Static procedures:**
 - Indications:
 - Old patients (>50 years).
 - Adults with low motivation.
 - Types:
 - Static slings.
 - Static excisional procedures.



- **Methods of Early Facial Rehabilitation:**

1. Primary End-to-end Anastomosis (Neurorrhaphy):

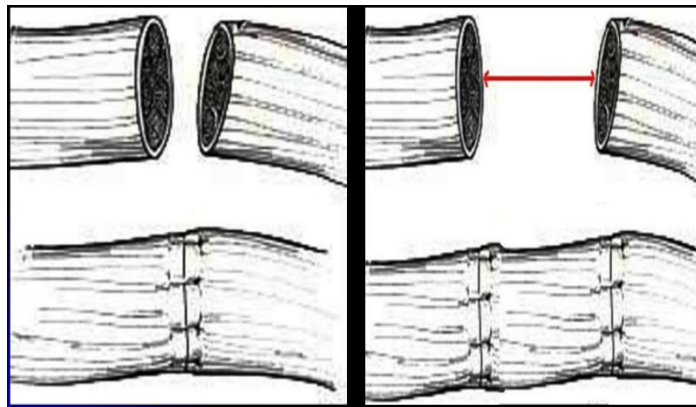
- Indications:
 - **Recent injury and tension-free closure is possible:**
 - Proximal and distal ends should be presents.
 - Preferred to be done within 3 days of injury.
 - Can be done up to 3 weeks of injury.
- Method:
 - Injured ends of the nerve should be freshened at 45-degree angle to expose more neural tubules and improves regrowth of the nerve.
 - Microsurgical anastomosis of epineurium with non-absorbable 10-0 nylon sutures.
 - Application of glue without sutures can be done if the nerve within the fallopian canal and there was is no chance of nerve end movement.
 - Avoidance of tension is essential which may require releasing proximally and distally or rerouting at the mastoid segment.
 - Priority of repair must be given to zygomatic and buccal branches if facial nerve was sacrificed during parotid surgery for malignancy.
- Advantages:
 - Provides best chance of rehabilitation (facial muscle movement with the least synkinesis).



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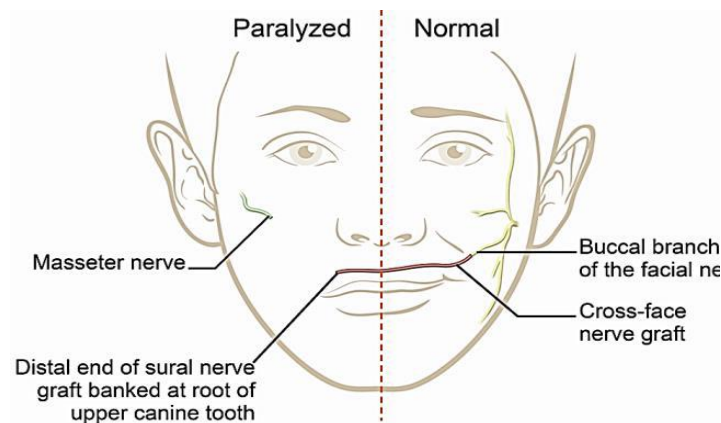
2. Interposition (Cable) Grafting:

- Indications:
 - **Recent injury and tension-free closure is NOT possible:**
 - Proximal and distal ends should be present.
 - Gap > 1-2 cm between the two ends.
 - Preferred to be done within 3 days of injury.
 - Can be done up to 3 weeks of injury.
- Method:
 - Nerve graft interposed between facial nerve endings.
 - Options:
 - **Greater auricular nerve** is the preferred graft as it is readily available near operative field and has same diameter of facial nerve.
 - **Sural nerve** graft is used if length > 10 cm is required.
 - Follow same principles of primary anastomosis.
- Advantages:
 - Provides resting muscle tone and spontaneous facial expression.
- Disadvantages:
 - Nerve grafting produces more synkinesis.



3. Cross-Facial Nerve Graft (CFNG):

- Indications:
 - **Nerve injury with unavailable proximal stump and intact motor end plates:**
 - Contralateral facial nerve is available.
 - Preferred to be done within 6 months of injury.
 - Can be done up to 2 years of injury.
- Method:
 - **1st stage:**
 - Proximal branches of facial nerve (mainly buccal branch) from normal side are coapted to distal end of sural nerve graft.
 - Tunneled subcutaneously under the upper lip to reach the opposite side.
 - Banked in the upper buccal sulcus just past the midline.
 - **2nd stage:**
 - Performed 6-12 months after 1st stage.
 - Distal end of the sural nerve graft then coapted to corresponding branches supplying specific muscle groups on the paralyzed side.
- Advantages:
 - Provides spontaneous animation.
- Disadvantages:
 - Risk of synkinesis.
 - Violation of the normal facial nerve.
 - **Axonal regeneration takes a long time to cross the face and could lead to irreversible muscle atrophy:**
 - Mainly if done after > 6 months of injury.
 - Needs to keep the facial muscles alive by **baby setting** them to another nerve.
 - Mainly nerve used is the ipsilateral nerve to masseter (branch from V3 of CN-V).
 - Done during the 1st stage of CFNG.



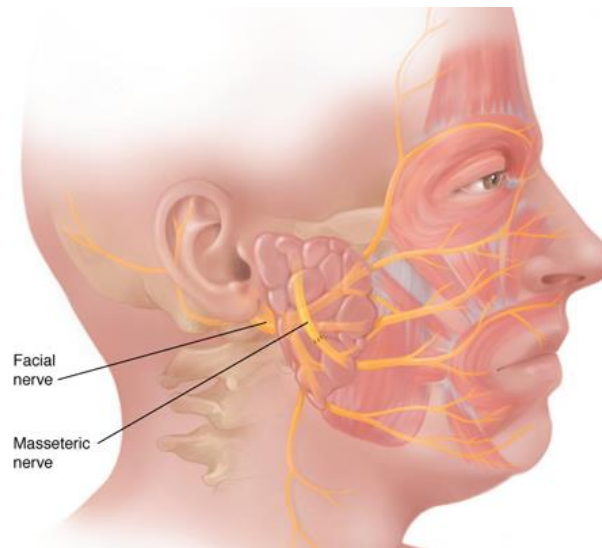
4. Nerve Transfer:

- Indications:
 - **Nerve injury with unavailable proximal stump and intact motor end plates:**
 - Preferred to be done within 6 months of injury.
 - Can be done up to 2 years of injury.
- Method:
 - Uses branches of other nerves to innervate the ipsilateral distal stump of facial nerve.
 - Types of nerve transfer:
 - **Hypoglossal-Facial Nerve Transfer:**
 - Most commonly used procedure.
 - Different methods:
 1. Entire CN-XII transection and coaptation to main trunk of the facial nerve.
 2. Split CN-XII transection (40%) and coaptation to lower division of the facial nerve.
 3. End-to-side neurorrhaphy between CN-XII and donor cable nerve graft (great auricular nerve) and coaptation to main trunk of the facial nerve.



- **Masseteric-Facial Nerve Transfer:**

- It is increasingly being used for facial reanimation.
- Good option due to its minimal donor morbidity.



- Advantages:

- Excellent muscle tone
- Provides potentially greater number of axons.
- Restores some voluntary motion and resting tone typically by 6 months.
- Can be used as a "babysitter" (to maintain muscle innervation) during the long period of axonal regeneration through a cross-facial graft.

- Disadvantages:

- Lack of spontaneous expression.
- Alteration of expression with chewing.
- Requires patient re-education of motor coordination.
- Risk of hemitongue weakness.
- Risk of significant synkinesis.

- **Methods of Delayed Facial Rehabilitation:**

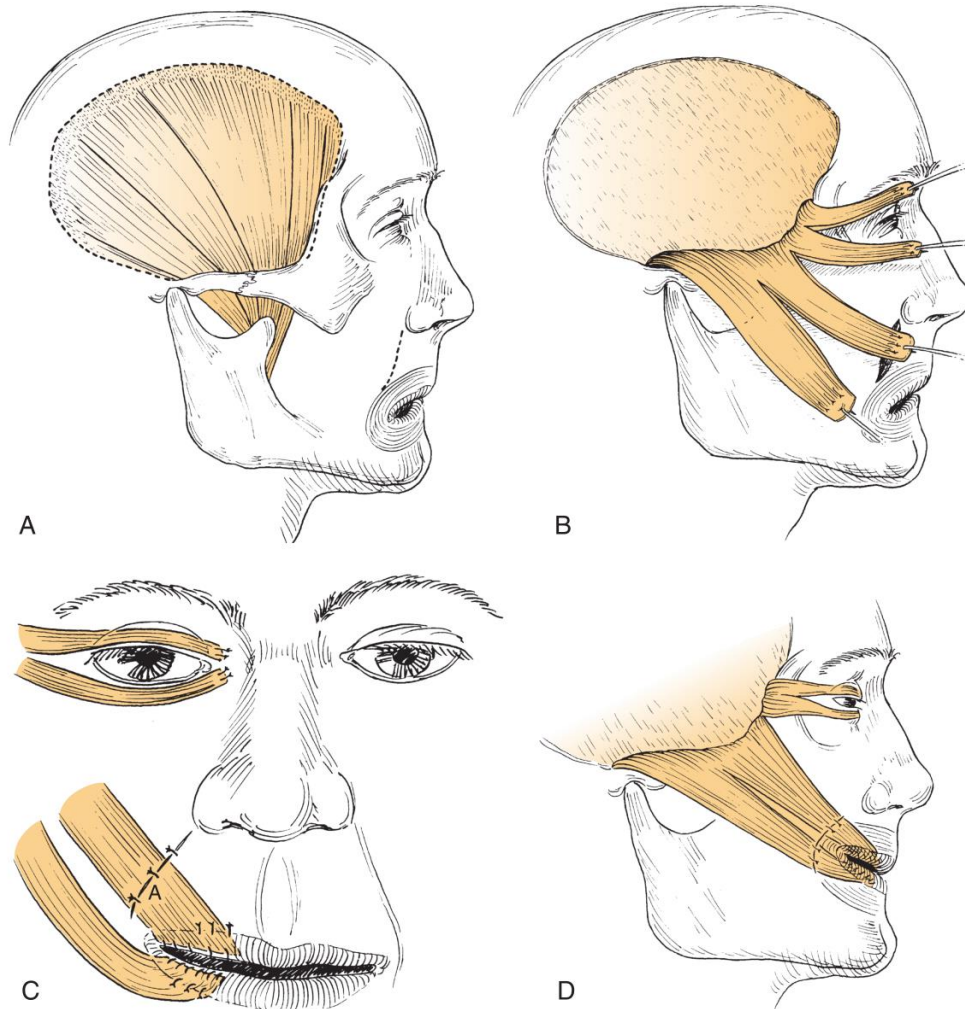
1. Dynamic Procedures:

- Indications:
 - **Prolonged nerve injury with unavailable proximal stump and loss of motor end plates:**
 - Done after 2 years of injury.
- Aim:
 - Restore symmetry both at rest and while smiling.
- Types:
 - **Muscle Transfer:**
 - Indication:
 - Adults (<50 years) with average motivation.
 - Advantages:
 - One stage procedures.
 - Disadvantages:
 - Patients must clench the teeth to produce a smile.
 - **Free Nerve Muscle Transfer:**
 - Indication:
 - Adults (<50 years) with high motivation.
 - Advantages:
 - Spontaneous animation of the smile.
 - Potential of achieving individual segmental contractions.
 - Reduction of synkinesis.
 - Disadvantages:
 - Lengthy procedures with two stages.
 - Requires innervation.
 - Requires postoperative physiotherapy.

- **Common types of Muscle Transfer:**

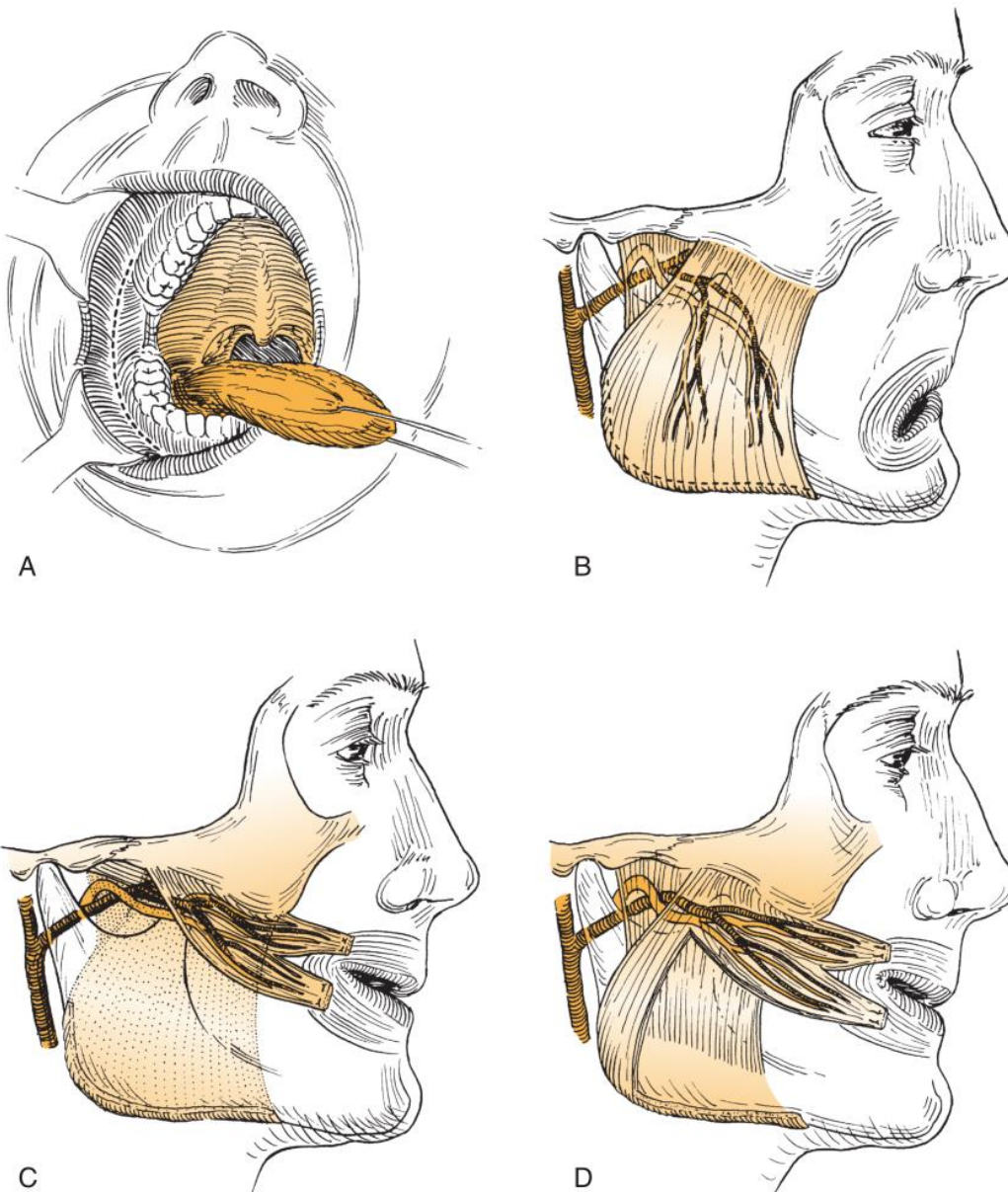
1. Temporalis Muscle Transfer:

- Innervated by CN-V.
- Methods:
 - Temporalis is detached from temporal fossa and turned over the zygomatic arch to reach the oral commissure.
 - Used mainly to reanimate the mouth using the horizontal vector.
 - Superior vector is used to reanimate eye.
- Disadvantages:
 - Patients must clench the teeth to produce a smile.
 - Not very effective for eyelid closure.
 - Produces significant temporal hollow and bulge over the zygomatic arch.



2. Masseter Muscle Transfer:

- Innervated by CN-V.
- Used when temporalis muscle is not opted.
- May be preferred due to avoidance of large facial incision.
- Disadvantage:
 - Less available muscle compared to temporalis
 - Vector of pull on oral commissure is more horizontal than superior/oblique like temporalis.



- **Common types of Free Nerve Muscle Transfer:**

1. Gracilis Muscle Flap:

- Workhorse for free muscle transfer.
- Long, thin muscle in medial thigh.
- Methods:
 - Can be done as one or two stages depending on the method of innervation.
 - One-stage procedure:
 - Gracilis muscle transfer.
 - Vascular anastomosis to the facial artery and vein or to superficial temporal vessels.
 - Obturator nerve of gracilis connected to either:
 - Ipsilateral CN-VII (if present).
 - Masseter nerve.
 - Two-stage procedure:
 - **1st Stage:**
 - Cross-face nerve graft with sural nerve graft.
 - **2nd Stage:**
 - Performed after 6-12 months of CFNG.
 - Gracilis muscle transfer performed after neural ingrowth of graft.
 - Vascular anastomosis to the facial artery and vein or to superficial temporal vessels.
 - Obturator nerve of gracilis connected to distal end of sural nerve graft.

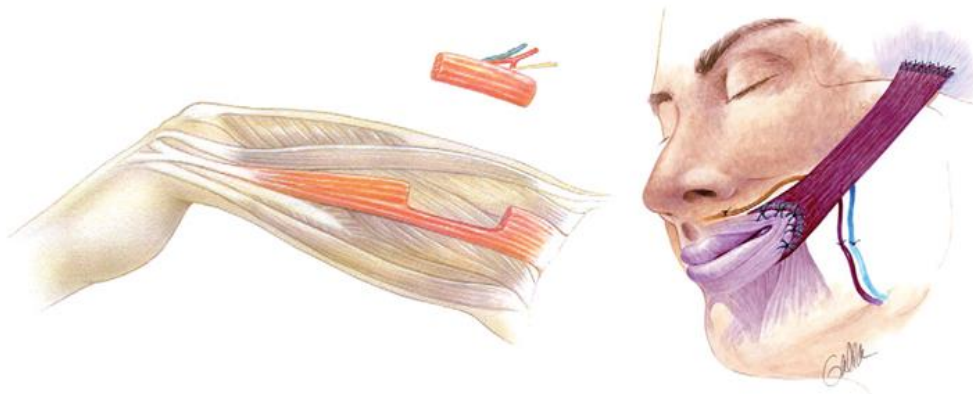


Table 11.6 Muscles available for microneurovascular transplantation

Gracilis
 Pectoralis minor
 Rectus abdominis
 Latissimus dorsi
 Extensor carpi radialis brevis
 Serratus anterior
 Rectus femoris
 Abductor hallucis

Table 11.7 Postoperative regimen following muscle transplantation

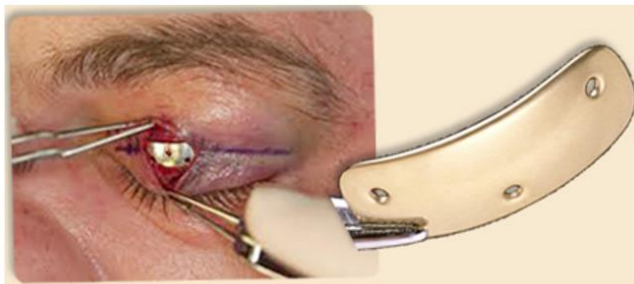
Fluids to soft diet as tolerated
 Bed rest day 1 then up in chair with assistance and gradual guarded ambulation
 Cefazolin in appropriate age-related dosage for 3 doses
 Morphine PRN for 48 hours
 Tylenol scheduled maximum dose for 3 days
 No nicotine for 6 weeks
 No caffeine for 6 weeks
 No pressure on surgical site
 Restrict sports or rough activities that may lead to trauma on surgical site for 6 weeks
 After muscle begins to function, active exercises may be helpful with biofeedback to increase excursion, achieve symmetry, and facilitate spontaneity

2. Static Procedures:

- Indications:
 - **Prolonged nerve injury with unavailable proximal stump and loss of motor end plates:**
 - Done after 2 years of injury.
- Aim:
 - Restore symmetry at rest only.
- Indication:
 - Old patients (>50 years).
 - Adults with low motivation.
- Types:
 - **Static slings:**
 - Fascial, allograft, or synthetic (Gore-Tex).
 - Commonly used:
 - Palmaris longus tendon.
 - Tensor fascia lata.
 - **Static excisional procedures.**
- Regions:
 - **Eye brow procedures:**
 - Used to correct eye brow ptosis.
 - Static excisional procedures:
 - **Brow lift (Most common).**

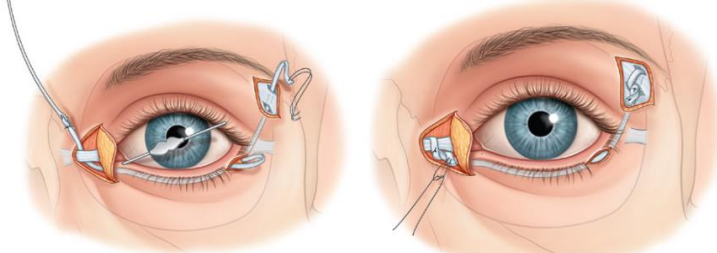


- **Upper eyelid procedures:**
 - Used to correct lagophthalmus.
 - **Most common is gold weight and tarsorrhaphy.**



- **Lower eyelid procedures:**

- Used to correct lid eversion and ectropion.
- Static excisional procedures:
 - **Horizontal lid shortening (Most common).**
- Static slings:
 - **Palmaris longus tendon.**
 - **Tensor fascia lata.**

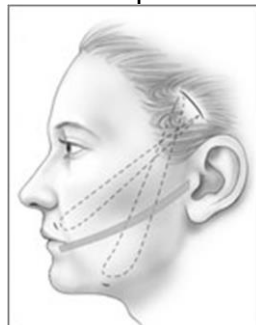


- **Nasal valve procedures:**

- Used to correct nasal valve collapse.
- Static excisional procedures:
 - **Septoplasty.**
- Static slings:
 - **Palmaris longus tendon.**
 - **Tensor fascia lata.**

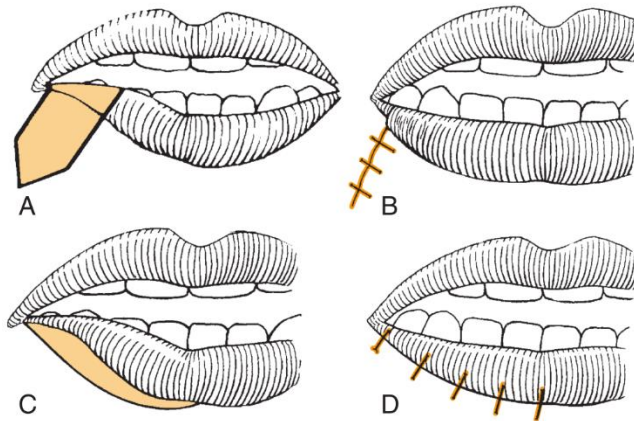
- **Upper lip and cheek procedures:**

- Used for smile reconstruction.
- Static slings:
 - **Palmaris longus tendon**
 - **Tensor fascia lata.**
- Principles:
 - Slings weaved from oral commissure and upper lip to temporalis fascia or zygoma.
 - Over correction is made to allow for stretching.
 - Usually 3 slings applied.
 - Can be combined with dynamic procedures.



- **Lower lip procedures:**

- Used to correct the inability to depress, lateralize, and evert lower lip.
- Static excisional procedures:
 - **Wedge excision of affected side.**
 - **Resection of normal side depressor labii inferioris muscle**
- Botox injection to normal side to weaken the depressor labii inferioris.



CSF Otorrhea

- Leakage of (CSF) through the ear structures is a rare but potentially life-threatening situation that requires rapid intervention.
- The presence of an abnormal communication of the sterile subarachnoid space with the flora of the sinonasal tract places the patient at great risk for meningitis.
- An episode of meningitis may be the presenting problem for a person with an otologic CSF leak.
- Actual leakage of the fluid from the ear is not always present.
- CSF otorrhea occurs only if a perforation in the eardrum or a defect in the external ear canal is present. (trauma, surgery)
- However, in the absence of such a defect, the fluid flows down the eustachian tube and manifests as a clear rhinorrhea.
- (Remember; just because the leak is through the nose does not mean that an otologic source is not a possibility.)

Etiology:

- Violation of the bony and meningeal barriers that separate the subarachnoid space from the middle ear and mastoid
- Defect must exist not only in the bone, but also in the dura mater

***: Traumatic (most common)**

- Accidental (80%) – 90% will close spontaneously (manitoba)
- Surgical/iatrogenic (16%)

***: Nontraumatic (4%)**

- High pressure – Tumors, Hydrocephalus
- Normal pressure – Congenital, Spontaneous, Osteitis/Osteomyelitis/otologic disease itself

Sometimes broadly be categorized into acquired and congenital

Spontaneous cerebrospinal fluid leak

- Leakage that occurs without an obvious antecedent pathology.
- Spontaneous leakage represents the greatest challenge to the clinician because the source of the leakage is not readily obvious
- Rare, with less than 500 cases having been reported in the literature worldwide
- More common in children (72%) than adults.
- Such leakage is usually due to some **congenital** defect in the temporal bone.
- Although congenital sources are more common in children, they can occur in people of any age and can even be observed in the geriatric population
- Bony dehiscences are most commonly found on the floor of the middle fossa, along the tegmen plate.
- Continuous pressure of the CSF over the years results in formation of a meningocele or encephalocele.
- Defects in the middle fossa are associated with 88% of spontaneous leaks in adults.
- Most common Sites of Middle Ear CSF Leak
 1. Tegmen
 2. Fallopian canal
 3. Hyrtl's fissure
- The remainder of spontaneous leaks are due to posterior fossa defects and arachnoid granulations
- Spontaneous leaks have recently been shown to be often associated with increased intracranial pressure.
- This is often manifested by a partially empty sella on the MRI scan.

Congenital causes of CSF Otorrhea

1: Bony Tegmen defect

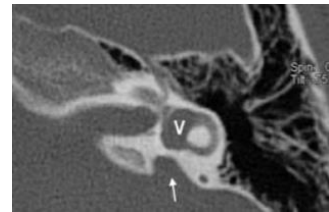
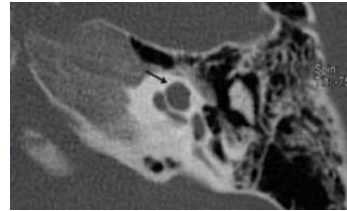
(Most common congenital, but present later in life)

2: Arachnoid granulations

("spontaneous", also present later in life)

3: Mondini

- Autosomal D
- There is incomplete partition between the scalae apical and middle turn due to absence of osseous spinal lamina.
- Condition is unilateral or bilateral.
- Triad of IP type II with a minimally dilated vestibule and large vestibular aqueduct
- This deformity may be seen in Pendred, Waardenburg, Branchio-oto-renal, Treacher-Collins and Wildervanch syndromes
- Have a patency at the lateral aspect of their internal auditory canal, allowing direct movement of CSF into the inner ear.
- A second defect, usually of the annular ring of the stapes footplate (**OVAL W**) then results in drainage of CSF into the middle ear.
- This usually causes loss of the remainder of hearing.



4: Enlarged Cochlear Aqueduct or vestibuler

5: Enlarged Fallopian Canal

6: Patent Hyrtl's fissure

- A congenital fusion plane found between the otic capsule and the jugular bulb
- (A bony cleft inferior to the round window niche and running towards the posterior fossa)
- (Also termed the tympanomeningeal fissure).
- Such leakage may not be associated with any inner ear abnormalities, and the patient may have no evidence of a sensorineural hearing loss.



7: Petromastoid canal fissure

- Normally carries the subarcuate artery.
- Such patients may present with recurrent bouts of meningitis associated with what appears to be a unilateral serous otitis.
- Frequently, the true nature of the problem is not realized until a myringotomy is performed

Acquired cerebrospinal fluid leak

- Result from temporal bone trauma, surgery, or infectious or neoplastic causes
- Far more common than
- Congenital leakage.

Postoperative leakage following surgery is probably the most common cause of acquired CSF leaks.

- It is a recognized complication of acoustic neuroma removal and other skull base surgery.
- Leaks as a postoperative complication of skull base surgery represent the bulk of cases.
- This complication has been reported to occur in 6-12% of such cases
- These leaks are usually evident in the early postoperative period.
- They rarely occur more than 2 months postoperatively.
- Mastoid surgery for chronic ear disease is also a potential cause of an acquired CSF leak.
- When the dura is violated intraoperatively, the defect should be repaired immediately if possible.
- Frequently, however, the dura is not injured, but a defect is left in the bony plate of the tegmen.
- Over the years, the continuous pulsations of the CSF cause the dura to thin, allowing the arachnoid or brain to prolapse through this defect. (encephalocele)
- This dura may become thin and spontaneously rupture, resulting in a leak of CSF

Temporal bone fracture

- Leakage as a result of temporal bone fracture is the second most common etiology and occurs in 17- 21%
- Otic capsule sparing fracture (Longitudinal):
 - CSF from floor of middle cranial fossa.
 - Drains into epitympanum, antrum, and mastoid air cells
- Otic capsule disrupting fracture (Transverse):
 - CSF from Posterior cranial fossa.
 - Drains through Disrupted otic capsule into Middle ear.
- Otic capsule does not heal fully
 - Fibrous tissue and periosteal bone may seal off the fracture.
- Immediate vs delayed leaks (> 1 week)

Similar problems may result from middle ear disease, most notably **cholesteatoma**

Presentation

- CSF otorrhea
- CSF rhinorrhea
- Salt taste
- Leak with position or strain
- Hearing loss
- Meningitis
- Seizure

- CSF leakage through the ear is present is a clear watery drainage from the ear.
- This, however, is not always present and does not occur unless the eardrum or canal is in some way violated.
- Leakage may be evident as a clear watery nasal discharge.
- This discharge may be Positional or intermittent in nature
- May only become apparent during straining or leaning forward.
- Strange salty taste in the back of the throat.

- Unilateral hearing loss. (The nature of the hearing loss is important)

- **Sensorineural** hearing loss suggests an associated abnormality of the inner ear (associated with spontaneous leaks in children and is present 82% of the time)

- **Conductive**, suggesting a leak elsewhere in the temporal bone (OME)
May only be recognised at the time of myringotomy (gusher)
If that happened:
 - The ear canal is often packed to prevent continuous drainage from the ear and allow the eardrum to heal
 - Other authors think that packing may cause stasis and provide a nidus for infection, hastening meningitis, and they recommend only a loosely placed, frequently changed sterile cotton ball placed in the conchal bowl.
 - Then conservative Tx measures

- Unexplained episode of **meningitis**. (A common presentation)
- It is found in 93% of children and 36% of adults with spontaneous CSF leakage.
- A history of unilateral hearing loss or chronic ear disease may suggest which side is involved.
- Absence of otologic symptoms and a history of sinonasal disease may suggest an anterior source of leak.
- In situations of leakage following surgery and trauma, the source of the leak usually is readily apparent

Physical examination

- Complete otologic, neurologic, and head and neck examination
- Microscopy to examine the ears.
- Pneumatic otoscopy may be helpful in demonstrating fluid within the middle ear space
- Tuning fork evaluations
- Audiometric evaluation.

An important part of the physical examination is an attempt to demonstrate the leak. (Straining, leaning)

Dandy maneuver, the patient leans forward with the head pointed down while performing a Valsalva.

The side of the nostril from which the leakage occurs usually agrees with the side of the otologic source.

Investigations:

Done if obvious cause like trauma or surgery not found:

- **Halo sign**
- Test for **Glucose** and protein content
- **B2 Transferrin** (Most sensitive measure of identifying CSF)
(differentiate from perilymph by amount and rate)
- Beta trace protein
- High resolution **CT** or CT Cisternography
 - Unless an otologic source is certain, the scan should cover all 3 cranial fossae.
 - Check the otic capsule for abnormal morphology, such as a Mondini deformity.
 - Note the sizes of the vestibular and cochlear aqueducts.
 - Check the tegmen plates of the posterior and middle fossae for defects
 - CT scanning may be enhanced with the use of intrathecal contrast, such as iopamidol or iohexol.
 - The presence and location of pneumocephalus on CT scanning may help to identify and localize a CSF leak

- **MRI**

- Spinal fluid, bright on T2 sequences
 - In cases where a tegmen defect is observed on CT scanning, MRI may demonstrate whether or not brain tissue is prolapsed into the middle ear
 - Because this is important information for surgical planning, an MRI is a critical adjunct
 - A partially empty sella has recently been recognized as a possible sign of increased intracranial pressure.
 - The increased CSF pressure causes infiltration of the sella with CSF and displacement of the pituitary tissue
 - Has been shown to occur in 71% of patients with spontaneous CSF leaks.
- Intrathecal **Fluorescein** (0.1 cc of 10% Fluorescein in 10 cc ofCSF)

Management:

Conservative:

- Leak secondary to recent **surgery** or **trauma** can often be treated conservatively
- Treatment Options for CSF leak
 - Bedrest with Head Elevation, avoid increased ICP (avoid straining, stool softeners)
 - Compression dressing (Postmastoidectomy)
 - Lumbar Drain or serial lumbar taps if conservative measures fail
 - Diuretics (eg, furosemide, hydrochlorothiazide),
 - Carbonic anhydrase inhibitors (eg, acetazolamide)
 - Steroids.
- Prophylactic antibiotics is controversial but needed in case of drain or pack placed, immunosuppressed and when soilage of the central nervous
- Antibiotics should be withheld unless signs and symptoms of meningitis occur

A 25-year Meta-analysis by Brodie (1997) suggested statistically significant reduction in meningitis using prophylactic antibiotics.

However, a Cochrane review in 2011 showed no benefit and does not support prophylactic antibiotic use in patients with Basilar Skull Fractures, whether there is evidence of CSF leakage or not.

- Spinal fluid leaks following acoustic neuroma surgery respond to this treatment 80% of the time
- Leaks associated with temporal bone fractures, which almost always seal in 3-4 weeks with this conservative therapy

Surgical Therapy:

Indications for Open Exploration of CSF leak

- Persistent leak despite Medical Management (10-14 days)
- Large Defect
- Brain/Meningeal Herniation
- Brain Penetration by bony Spicule
- Persistent Pneumocephalus
- Recurrent Meningitis
- ? Gravity dependant defect location
- Primary treatment of a spontaneous otogenic spinal fluid leak

Approaches:

1) Transcanal approach

- Spontaneous leaks in children with otic capsule defects
- Such as Mondini
Because rarely any hearing is present, a stapedectomy is usually performed and the oval window obliterated with soft tissue
- Can also be used in some cases of CSF leakage due to a patent Hyrtl fissure

2) Transmastoid approach

- If the leak is related to a small (< 1 cm) defect in the bone of the tegmen or exact site not really known
- Exposure of a mastoidectomy usually allows excellent visualization of the leakage site.
- The site can often be repaired with a small amount of fascia supported by Gelfoam
- The fascial repair can be supported with a tragal cartilage graft placed between the intact bony edges and the dura
- Occasionally, a fat or muscle graft may be needed.

- Leaks occurring from defects of the posterior cranial fossa anterior to the sigmoid sinus present a special problem. This is the area of the basal cistern, where no arachnoid mesh is present. Leakage from this area is explosive and profuse and is not well controlled with fascia alone. A large fat graft obliterating the mastoid is usually required

3) Transmastoid with Obliteration

- Rare cases, the exact site of leakage is not found, and **diffuse** leak is observed from multiple mastoid air-cell tracts.
- mastoid may need to be obliterated with fat.
- Obliteration of the middle ear and eustachian tube may also be required, especially if the leakage is not limited to the mastoid

4) Blind sac surgery

- Previously undergone **CWD** mastoidectomy (or EAC eradicated by disease)
- In these situations, the external ear and mastoid epithelium must be completely removed and the ear canal sewn over.
- Abdominal fat is then used for obliteration

5) Middle fossa/transmastoid combined approach

- If a leak is due to a large (**>1 cm**) defect in the floor of the middle fossa
- A mastoidectomy is performed first to identify the site of leakage.
- Do not attempt to reduce herniated brain tissue.
- Such encephaloceles do not contain functioning brain tissue and should be excised using bipolar cautery.
- Once the defect is identified, use the middle fossa approach for repair.
- The middle fossa affords excellent visualization of the defect and an opportunity to use the intact bony edges of the defect to hold any repair material in place.
- Recommended repair of such large defects is with a 3-layer technique. A layer of calvarial bone (from the craniotomy) is sandwiched between 2 layers of fascia.
- Fibrin glue is a very useful adjunct (not alone)

Grafts:

- Facia
- Muscle
- Cartilage
- Bone
- Biocompatible (hydroxy apatite, bpvin pericardium)
- Synthetic (silicone sheeting, Marlex mesh, titanium plates, and Methylmethacrylate), high rate of extrusion

Postoperative Details:

- Continuous lumbar spinal fluid drainage is an important adjunct to surgical repair of otogenic CSF leakage

Drainage should be accomplished by draining a specific amount every **hour, usually about 10 mL**. Occasionally, this causes severe headache, in which case a smaller amount can be removed or the fluid can be removed less frequently (eg, 5 mL every half hour)

Check spinal fluid drainage every 2 days with a Gram stain and a culture.

The lumbar drain is usually left in place for 2-3 days postoperatively. If no sign of leakage is present, it is clamped and the patient is observed for an additional 24 hours. If no further leakage is observed, the drain is then removed

- Observed in an ICU (any neurologic changes)

Perform hourly neurologic checks for the first 24 hours after surgery and then every 2 hours until the patient is well enough to leave the unit

ICU may not even be necessary. These include cases treated by a transcanal approach that do not require lumbar drainage

- Patients should remain on bed rest. Use position changes and compression boots to prevent bedsores and deep venous thrombosis.

Complications:

- Intracranial bleeding
- Cerebral edema
- Hydrocephalus
- Stroke
- Meningitis

Change in mental status in the postoperative period must be rapidly evaluated

An immediate CT scan of the brain performed without contrast usually demonstrates any bleeding, edema, stroke, or hydrocephalus.

Clamp the drain until the CT scan findings are shown to be normal.

Hearing Aids and Implantable Hearing Devices

HEARING AIDS

- The most common treatment for sensorineural hearing loss (SNHL) is the use of hearing aid amplification
- Hearing aids are the principal means of auditory rehabilitation for patients with sensorineural hearing loss (SNHL).
- Hearing aids may also play a role in the management of conductive hearing losses (CHLs), particularly those that result from pathologies not amenable to medical and surgical treatment
- Goal: increase magnitude of acoustic signal at output Vs. input
- Basic system has three **components**:

- o **Microphone** : converts signal from acoustic to mechanical to electrical
- o **Amplifier** : boosts electrical signal
- o **Receiver/ speaker** : converts electrical back to acoustic signal

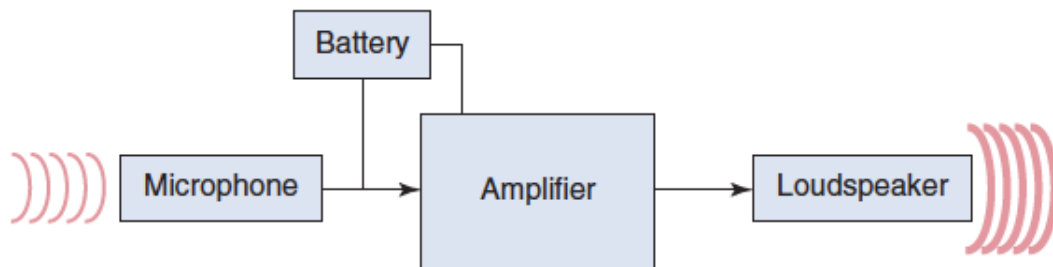


FIGURE 162-3. Schematic representation of the components of a hearing aid.

- Three **categories** of devices currently on market

1. Nonprogrammable analogue devices
 - Constant analysis and modification of input stimulus
2. Digitally programmable analogue systems
 - Digital memory used to generate several different HA responses that can be selected for different listening conditions
 - Different processing strategies allow better electro-acoustic adjustments and flexibility in adjusting gain and output
3. Digital signal processing (DSP) devices:
 - Sound is represented as a string of numbers (analog-to-digital conversion)
 - Sound is then manipulated by DSP using math formulae
 - New string of numbers are converted back to sound (digital-to-analog conversion)
 - Smaller and cheaper to make than analogue counterparts
 - Tailor output to individual loss
 - Currently 50% of market;

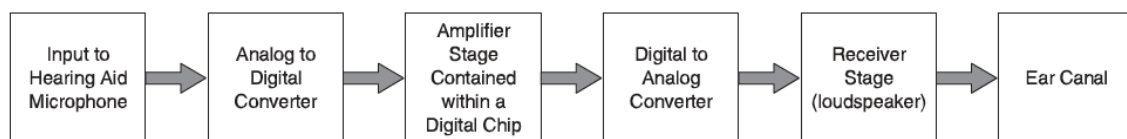
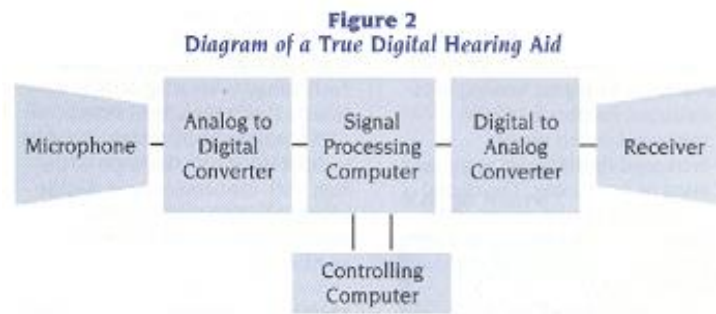


Figure 164.1 Schematic of the basic components of a hearing aid.

Gain: amplification; can be applied in linear or non-linear · (compressive or expansive) fashion

· **Types** of Gain

o **Linear:** direct relationship between input and output

o **Non-linear:**

· **Compressive** decreases gain with increasing input intensity (good for those with small range between functional hearing and discomfort i.e. narrow “dynamic range”)

· Compression HA function as linear devices for sound intensities below compression threshold (CP)

o High CP-limits max output level, while providing linear amplification for most speech inputs

o Low CP-nonlinear amplification for most levels

· **Expansive** decreases gain for decreasing amplitude sounds (to reduce background noise)

Another goal addressed by nonlinear amplification strategies is to make speech and other important environmental sounds both audible and comfortable

Understanding speech in noise

- Most common cause of poor compliance
- **Signal to noise ratio:** difference between level of sound you want to hear and that of background noise
- Pt with HL require higher Signal-to-Noise Ratio (SNR) to hear in noise

o **Omnidirectional** microphones do not improve SNR in comparison to the unaided ear

o **Directional microphones** use multiple microphones ports to allow SNR improvements based on spatial separation between signal and noise

- Signal of interest-front hemisphere
- Noise-rear hemisphere
- Reduced effectiveness in reverberant (reflective) environments and when the signal is located at a great distance
- SNR improvement is **3-6 dB**
- Full-time directional HA is not recommended

- **Acoustic feedback** occurs when the amplified sound that emanates from a receiver is directed back into the microphone of the same amplifying system.
- Physical separation of the microphone and receiver is the most effective way to reduce feedback
- Causes of feedback:
 - Poor fit
 - Over amplification
 - EAC plugged
 - Mic too close to speaker

Digital Signal Processing-Based Features

• **Digital noise reduction (DNR) scheme:** decrease gain in ranges with no detectable speech; detects speech based on amplitude modulations; shown to improve comfort, but has no speech recognition benefit

• **Digital feedback suppression (DFS) scheme:** allow for increased gain under constant coupling constraints (same HA shell and vent size); can be helpful in those with occasional feedback, and to allow for a bigger **vent** to improve sound quality of wearer's voice

Styles and Shapes of Hearing Aids

Behind-the-ear HA (BTE)

- o Replaced body worn HA
- o More cosmetic appeal; no body baffle effect (on torso-increased low frequency amplification and decreased amplification in high frequencies); no noise from clothes rubbing against microphone
- o custom-fitted earmold, a standard coupler, or a receiver in the canal
- o telecoil



In-the-ear (ITE)

- o Benefit of normal acoustics provided by the pinna increased gain in the high frequencies
- o Pinna acoustics also important for sound localization cues
- o smaller and fits into the ear canal is known as an **in-the-canal (ITC)**



Completely-in-the-canal

- o It is even smaller and fits deeply in the canal
- o Maximize pinna and concha effects high-freq gain and reduced occlusion effect
- o Fitting is difficult



Figure 164.3 Schematic of a deep insertion completely-in-the-canal hearing aid. Drawing courtesy of Starkey Laboratories, Inc.



	Type of Hearing Aid	Advantages	Disadvantages
Riyadh et al. Notes	Behind-the-ear	Range of power sufficient to reach severe and profound hearing loss. Allows adequate space to accommodate gain, output, and frequency response controls on analogue instruments for adjustment by dispenser. Allows for placement of directional microphone.	In cases of severe or profound hearing loss, earmold must fit snugly in the ear canal to eliminate feedback problems. May be slightly more difficult to place on ear in cases of manual dexterity problems. For those with small pinnae, particularly small children and infants, accessory devices may need to be considered to hold in place. However, custom instruments are inappropriate for small children and infants due to frequent changes in ear size and geometry.
	In-the-ear	Cosmetically appealing. Placement of microphone takes advantage of pinna and concha effects enhancing amplification in the high frequencies. Microphone placement improves sound source localization as head is moved. May be easier to insert because only one component is involved. Allows for placement of directional microphone.	Because of close proximity of microphone and receiver, the amount of real ear gain may be limited because of feedback problems, especially in cases of severe profound high-frequency hearing loss.
	In-the-canal	More cosmetically appealing because it fits almost entirely in the ear canal. Microphone placement takes advantage of pinna and concha effects boosting gain in the high frequencies. Microphone placement improves localization to sound source as head is moved.	Amount of gain is sufficient for no more than moderate hearing loss. Due to small size and small battery, may be difficult to handle for individuals with manual dexterity problems. Small size of instrument limits the number of output/response controls in analogue instruments. Venting options limited. Deep placement precludes use of directional microphone.
	Completely-in-the-canal	Most cosmetically appealing because canal hearing aid is not visible when placed in the ear. Its deep insertion potentially takes full advantage of pinna and concha effects, boosts real ear gain (especially) in the high frequencies and reduces occlusion effect.	Amount of gain is sufficient for no more than moderate hearing loss. Due to small battery, may be difficult to handle for individuals with manual dexterity problems. Small size of instrument limits the number of output/response controls in analogue instruments. Deep placement precludes use of directional microphone. Venting options very limited. Deep tight fit requires precision, and is not possible on all patients due to ear geometry.

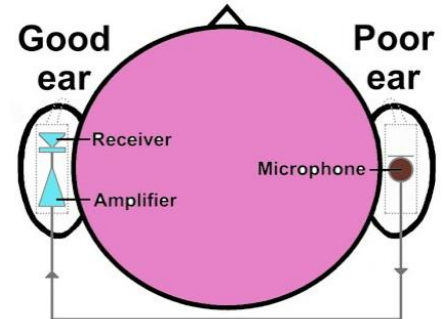
	Behind-the-Ear (Standard and Mini)	In-the-Ear	In-the-Canal	Completely in-the-Canal
Fitting range	Full range	Mild-to-moderate (with some feedback control options, can fit more severe losses)	Mild-to-moderate	Mild-to-moderate
Number of features*	Very flexible	Depends on ear canal size	Depends on ear canal size	Few options
Advanced signal processing	Available	Available	Available	Available
Manipulation	Easy	Easy	Can be difficult	Difficult
Care	Minimal	Cerumen	Cerumen	Cerumen
Cosmetics	Depends on hair style (slim tube has made this more attractive to individuals)	Visible	Visible	Least visible
Telephone use	Good	Good	Poor	Good
Coupling to ALDs	Good	Fair	Poor	None

*Features refer to physical features such as volume control, program button, and multiple microphones. ALDs, assistive listening devices.

Other styles:

Contralateral routing of signal (CROS)

- o No usable hearing in 1 ear and normal hearing or a minimal HL in the other ear
- o Microphone is placed on the impaired side and transmitted to the good ear via an open ear mold or a polyethylene tube
- o Disadvantages: user must be motivated to hear sounds arriving at both sides equally (not natural), 2 devices, poor sound quality, poor cosmetics



BiCROS

- o No usable hearing in 1 ear and minimal HL in the other ear
- o Amplification to the better ear AND contralateral routing of signal from bad ear

Transcranial CROS

- o Single deep-fitting custom instrument to the impaired ear; allows mechanical coupling between HA shell and bony portion of the EAC
- o transmission of sound to contralat cochlea through both AC and BC

Bone-conduction aid

- o BC receiver is placed on the mastoid and held in position by **headband**
- o More energy is required to stimulate the ear via BC-only useful for milder SNHL
- o More useful for CHL (traditional bone conducting hearing aid)

Bone-anchored HA (BAHA)

- o HA transducer is coupled to titanium screw in upper mastoid region of TB via a titanium abutment screw
- o Indications: CHL, cannot use conventional HA, unilat SNHL

Middle ear implants

- o Pure SNHL and mixed losses
- o Piezoelectric, electromagnetic, electromechanical properties used to drive an output transducer mounted on the ossicular chain

Electroacoustic Characteristics of Hearing Aids

- Three most important are: gain; output sound pressure level with 90 dB input (OSLP90) ; frequency response
- **Gain**
 - **Difference in the output relative to input**
 - Full-on gain: amount of amplification with maximum volume control setting
 - High-frequency average (high-frequency full-on gain): gain measured at 1000, 1600, 2500 Hz (recommended by ANSI)
 - Reference test gain: amount of amplification when volume control is adjusted such that the average gain at 1000, 1600, and 2500 Hz is 17 dB below OSPL90 or full on if HA has mild gain
 - Use gain (as-worn gain): gain measurement with volume control adjusted to its normal use position
- **OSPL90**
 - Saturation sound pressure level
 - Maximum amount of amplification provided by the instrument
 - As input level increased, output level increases until a certain point, above which further increases in input do not change output **saturation**
 - OSPL90: 90-dB input signal and output measured across freq range
- **Frequency response**
 - Gain of HA across a range of frequencies
 - Range of frequency of gain is usually limited
 - Frequency response: measure reference test gain, subtract 20 dB; line is drawn parallel to the abscissa until it intersects the low and high frequency ends of the curve

Output Limiting

- **TD** = threshold of discomfort (aka LDL, or loudness discomfort level)
- **Dynamic range of hearing**: between level at which sound is barely detectable, up to TD; can be small for SNHL
- OSPL90 should be set below TD
- **Peak clipping**: a way to limit output in linear systems; introduces harmonic and intermodulation distortion as peaks are clipped; compression is better

Hearing Aid Candidacy

- Indication based on: degree and configuration of HL, communication disorder, motivation of Pt and attitude toward HA
- Audiometry alone can't tell you who will benefit; trial is essential
- Also consider **motivation** for aiding; **acceptance** of loss (they won't wear it if sent in by a spouse) and **expectations**
- Another audiometric indicator of hearing aid use is suprathreshold speech-recognition ability

(If it is sufficiently poor, hearing aid amplification will contribute to audibility but may not be satisfactory overall.)

- Success measured objectively and subjectively; reduction of handicap
- The majority of patients who choose to use hearing aids have hearing loss that is at least moderate in degree.

TABLE 155.2

CONSIDERATIONS IN SELECTING HEARING AID CANDIDATES

- The first step in any hearing aid selection process is to determine the degree and type of hearing loss and determine the need for medical intervention.
- Secondly, how does the hearing loss impact on the individual's daily activities?
- Hearing loss of 25–35 dB HL may only be needed on a part-time basis (e.g., meetings or conferences for adults and educational environments for children), or a full-time basis.
- For those with moderate hearing loss (41–55 dB HL) communication is more difficult, particularly at distances of more than 4 feet, and amplification should be considered for many social and work-related environments.
- Individuals with hearing loss greater than 55–70 dB HL find communication with others very difficult without the use of amplification.
- For hearing losses greater than 70 dB HL, hearing aids are indispensable.
- What is the patient's word recognition ability?
- Are there any difficulties with loudness tolerance?
- Acceptance of hearing loss. Denial of the hearing problem reduces the motivation to wear the instrument and lessens the acknowledged benefit derived.
- Patient motivation. When an individual is not motivated toward the use of a hearing aid, the chances of achieving a successful hearing aid fitting is diminished. Counseling may improve the patient's attitude.
- Patient expectations. If expectations are too high the patient should be counseled appropriately so that the end result does not reduce the enthusiasm for wearing the instrument.

- As a general rule, if the hearing impairment is enough to cause a problem in communication, the patient is a candidate for hearing aids.
- If a patient has a sensorineural or other nontreatable hearing loss that is causing a communication disorder, the patient is a candidate for amplification

Benefits of Bilateral Hearing or Amplification

1. Release from masking (AKA binaural squelch): improved speech intelligibility due to phase differences of the signal and noise occurring between the two ears
2. Sound localization better with binaural hearing
(Brain relies on differences in time and intensity of signals between the ears to localize the source)
3. Eliminates head shadow effect
4. Binaural summation: sum; get louder sound & better amplification
5. Better sound quality

Unilateral hearing aid fitting places the unaided ear at a relative disadvantage, which may have a long-term negative effect on the speech perception ability of the unaided ear

Selecting Hearing Aids

- Must perform assessment of communication needs
- Can include: hearing thresholds and TD; middle ear function; EAC shape, geometry and movement; speech recognition in quiet and in noise; hearing aid expectations; perceived hearing handicap; specific listening environments in which the patient has trouble

Verification

- Fitting and verification is a process, not an event
- Real ear insertion gain **REIG**: measured at TM with mold and aid in place, rather than the 2 cm³ artificial test; good because it can account for all factors (pinna, head diffraction and EAC resonance)
- Real ear aided response **REAR**: used in children
- Real ear coupler difference **RECD**: used in younger children
- Aided soundfield threshold measurements not good, because:
 - Time consuming
 - Poor frequency resolution
 - Poor test-retest reliability
 - Misleading information in case of severe/profound, or mild HL
- Orientation: teach them how to use and maintain it
- Validation: keep testing over time to make sure it's working

Problems with HA:

- Bad fitting
- Feedback
- Headaches and Tinnitus
- Improper sound level and quality
- Sweating and wax impaction
- **Insertion loss:** a certain amount of hearing loss occurs with occlusion of the ear canal while the hearing aid is in place (for patients who have normal low-frequency hearing)
- **Occlusion effect:** Occlusion of the ear canal also causes a phenomenon wherein a patient's own voice is perceived as loud and echoing

One solution to occlusion problems is the placement of a **vent**, which is simply a passageway for exchange of air and sound around or through a hearing aid or earmold

Other hearing assisting devices:

Assistive Listening Devices

- Mild impairments
- Types of communication needs
 - Interpersonal communication and media
 - Telecommunications
 - Signals such as wake-up alarms, fire alarms, telephones
- ALDs are for specific listening situations
 - TV or radio amplifiers
 - Flashing alarms
 - Amplified telephone
- ALDs can help with speech in noise (improve the SNR in noise and reverberations)

FM Wireless System

- Adv: acceptable SNR for speech understanding in noise (educational environment for children)
- Talker wears a transmitter/ microphone around the neck; frequency-modulated signal is broadcast to an FM receiver unit worn by the listener
- External receivers are typically coupled to ear molds, earphones
- Coupling of FM system to personal HA
 - Electrical coupling
 - Inductive loop arrangement

Soundfield Systems

- Speech signal is delivered through a loudspeaker placed strategically in the classrooms

Infrared Systems "Signal transmission via infrared light"

- Acoustic signal is converted to electric signal; **infrared transmitter** modulates it into infrared light frequencies emitted from radiators
- Several light-emitting radiators are placed throughout the room

Other Assistive Listening Devices

- Telephone amplifiers (telecoil or telephone amplifiers)
- Television/radio amplifiers
- Signaling devices (high-intensity door bell)
 - Alerting devices visual (strobes, bright lights); tactile (vibration, airstream)

Training

- Speech reading
- Auditory training
- Speech conservation
- Sign language

LIMITATIONS OF AUDITORY REHABILITATION USING TRADITIONAL HEARING AIDS

PHYSICAL FACTORS

Limited in their ability to amplify sound without imparting distortion or generating feedback

- **Insufficient Gain**
- **Acoustic Feedback**
 - Worst for CIC aids, and for ears with mastoid bowls, in which an air seal is difficult to obtain or when very high amplification
 - Fitting hearing aids tightly into the external auditory canal can decrease feedback but at the cost of increased discomfort and at the risk of otitis externa, autophony, and blockage of natural sound input
- **Distortion of Spectral Shape and Phase Shifts**
 - HA are limited in the frequency range over which they amplify sound in the speech band (500 to 2000 Hz)
 - Isolated severe hearing loss at low frequencies, as in Meniere disease, or at high frequencies, as in presbycusis or ototoxic exposure, can be difficult to remediate with traditional aids without overamplifying the midrange frequencies
 - Digital signal processing technology has greatly improved spectral shaping options compared with analog aids, but these fundamental limitations persist.
- **Nonlinear Distortion**
 - Hearing aids are designed under the assumption of linearity—that is, if a given sound input at the microphone leads to some output at the speaker
 - For high-intensity speaker output, this assumption breaks down as the speaker is driven into the range of movements for which it begins to saturate or clip.
 - An input sinusoid can appear at the output as a waveform with blunted peaks.
 - This nonlinear distortion imparts aberrant spectral components into the sound percept, giving it an artificial or robotic character.
 - Whereas digital signal processing can mitigate distortion effects, remains a fundamental limitation of traditional hearing aids.

- **Occlusion Effects**
 - To minimize feedback
 - Canal occlusion has several undesirable effects:
 - 1) it can be uncomfortable because of pressure on canal skin
 - 2) it increases the likelihood of otitis externa as a result of disturbance of wax egress and air circulation, and some patients with chronic suppurative otitis media cannot tolerate hearing aids because of exacerbation of otorrhea
 - 3) it causes autophony and a sense of aural fullness that can worsen with changes in ambient barometric pressure
 - 4) it blocks the normal pathway for sound entry to the ear
 - 5) it disrupts the spectral shaping that normally occurs as a result of external auditory canal resonances
- **Poor Appearance**
- **Poor Transduction Efficiency**
 - Loss of energy as a result of impedance mismatching and transduction
 - Impedance-matching transformer by virtue of the relative areas of the tympanic membrane and stapes footplate and by the lever action of the ossicular chain
 - When this middle ear apparatus is malfunctioning—as occurs in otosclerosis, tympanic membrane perforation, or a CWD mastoidectomy— traditional hearing aids must overcome the impedance mismatch.
 - The result is either reduced effective gain or increased distortion, or both

Box 157-1. NONIDEAL FEATURES OF CONVENTIONAL HEARING AIDS

Insufficient amplification
Acoustic feedback
Spectral distortion
Nonlinear/harmonic distortion
Occlusion of external auditory canal
Appearance/visibility
Lack of directionality

HUMAN FACTORS

- **Recruitment and Compression of Dynamic Range**
- **Disordered Perception of Pitch and Sound Localization**

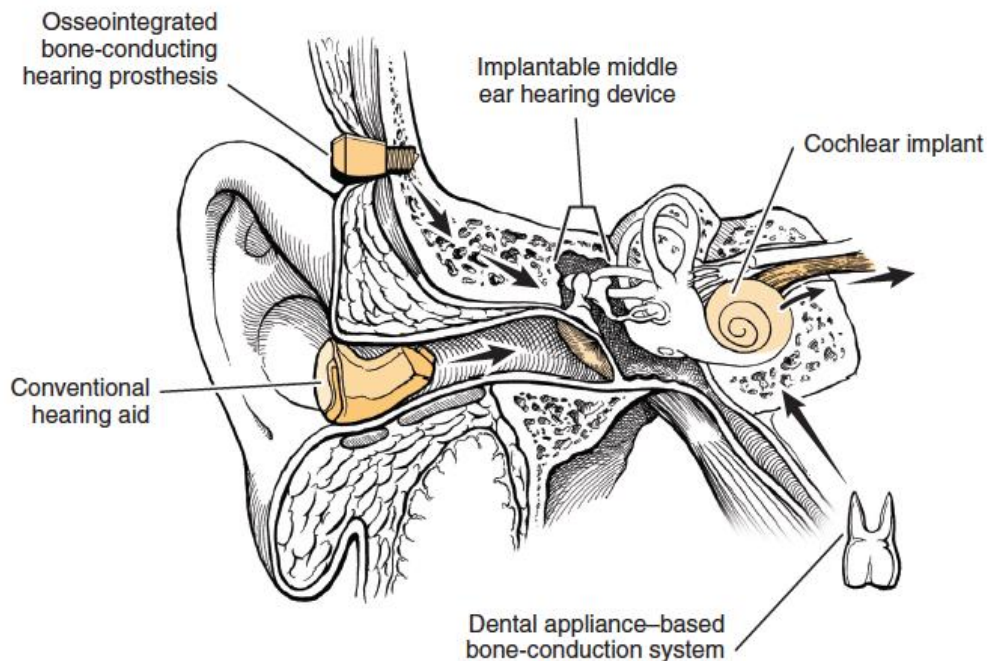
THE PROMISE OF IMPLANTABLE HEARING DEVICES

Implantable hearing devices offer the patient with hearing loss several potential advantages over conventional hearing aids. These include

- **Increased gain** and dynamic range
- **Reduced feedback** and distortion
- Improved **sound quality**
- Better **directional** hearing
- Reduced **maintenance**
- Improved **appearance**
- Freedom from **ear canal** occlusion

Downsides of their use, which include

- Risk of surgical implantation
- Difficulty of maintenance
- Increased initial device cost



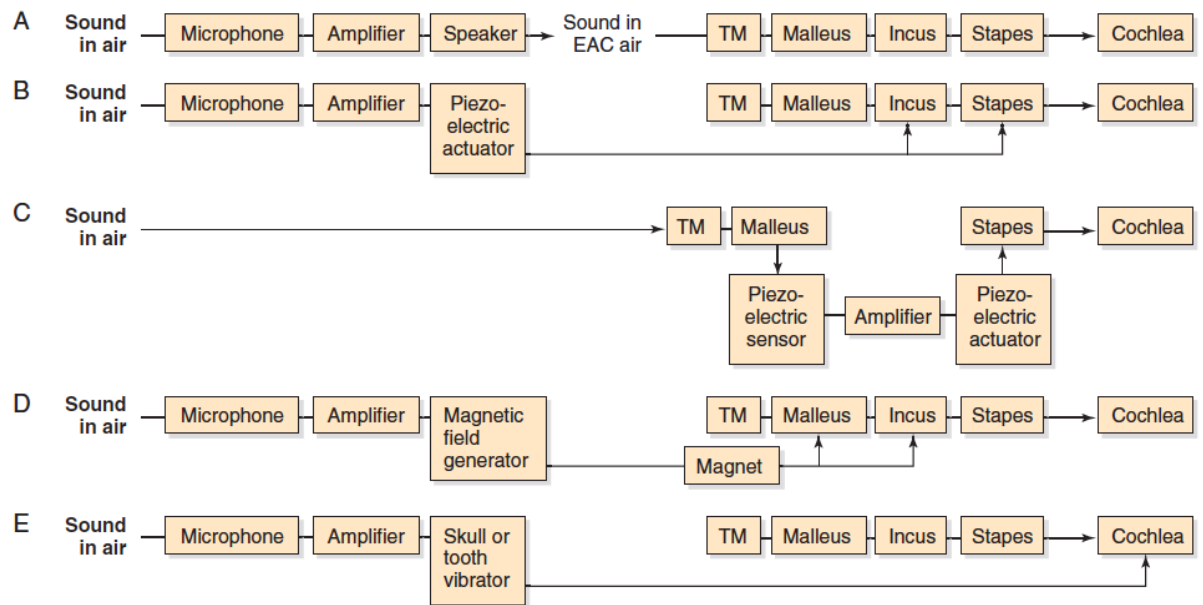


FIGURE 157-2. Sound transduction and coupling to the auditory system for different classes of hearing aids. **A**, Conventional air-conducting hearing aid. **B**, Implantable aids with piezoelectric actuator. **C**, Implantable aids with piezoelectric sensor and actuator. **D**, Implantable aids with magnetic actuator. **E**, Osseointegrated or dental bone-conducting hearing prostheses. EAC, external auditory canal; TM, tympanic membrane.

OSSEOINTEGRATED BONECONDUCTING HEARING PROSTHESES

- **Titanium** had a unique ability to become firmly anchored in bone
- Back ground: was initially applied to dental implants, then to provide an anchor for facial prosthetics (pinna, eye, or nose) and an attachment for a bone-anchored hearing device
- Indicated in: **CHL**, **MHL** and **unilateral SNHL** (as in CROS)

Criteria for **CHL** or **MHL**

- A minimum average bone conduction threshold of 45-dB HL or 55-dB HL
- For bilateral fitting, symmetric bone conduction thresholds are recommended
- The BAHA Cordelle II body worn device is recommended for severe mixed hearing losses, but the average bone conduction thresholds of the implanted ear should be less than 65-dB HL
- Pt with CHL or mix not suitable surgical candidates for correction of there disease (otosclerosis, tmpanosclerosis, canal atreseaa...etc)
- Unable to tolerate a conventional hearing aid (discomfort from sound, large mastoid, meatoplasty, discharging ear, canal closure like in blind sack or extensive base of skull surgery)

Criteria for unilateral **SNHL**

- Profound hearing loss in the implanted ear
- Hearing in the contralateral ear is better than or equal to 20-dB HL

TABLE 157-2. Indications and Contraindications for Baha

Indications	Contraindications
Any patient using a conventional BC hearing aid	Age <5 years
Any AC hearing aid user with chronic otorrhea	Emotional instability, drug abuse, development delay
Any AC hearing aid user experiencing too much discomfort because of chronic otitis media or otitis externa	PTA BC thresholds (0.5 to 3.0 kHz) worse than 65 dB HL, SDS >60%
Any AC hearing aid user experiencing uncontrollable feedback because of a radical mastoidectomy or large meatoplasty	
Otosclerosis, tympanosclerosis, canal atresia with a contraindication to repair (e.g., an only-hearing ear)	

AC, air conduction; BC, bone conduction; HL, hearing loss; PTA, pure tone average; SDS, speech discrimination score.

OSSEOINTEGRATION: BIOLOGY AND HISTOLOGY

- **Direct contact** must be made between the bone matrix and the implant
- **No interposed** fibrous or soft tissue
- **No** evidence of **inflammation** should be present at the implantation site (includes infiltrates or osteolysis)
- **No** evidence of a connective tissue **capsule** on the implant surface

RADIOLOGIC IMPLICATIONS

- A chief disadvantage of any metallic implant is interference with subsequent radiologic studies
- IN BAHA This scatter effect has relatively little impact on the quality of **CT** images produced.
- **MRI** is not contraindicated with the presence of a titanium implant.
- Any attached components must be removed prior to the scan

BAHA components:

- (a) Titanium fixture (implant)
- (b) Titanium abutment
- (c) Sound processor

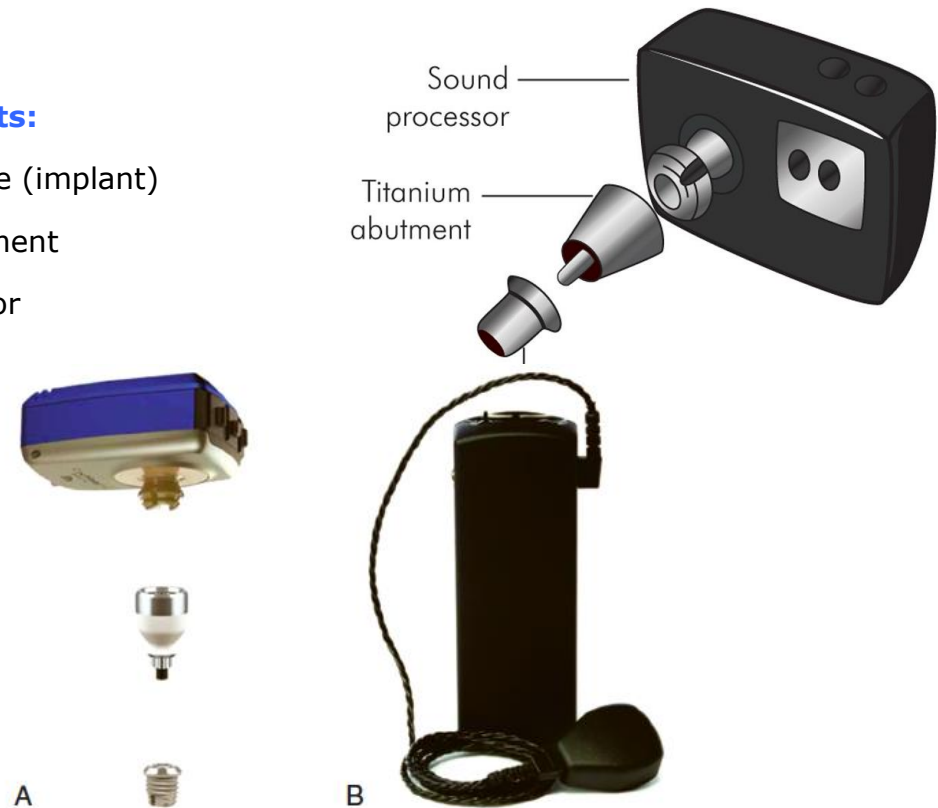


FIGURE 157-10. Osseointegrated Baha. **A**, Baha 4 Connect System is shown with its schematic attachment to the abutment and titanium implant. **B**, For patients with a more severe sensorineural component, the Cordelle II offers increased amplification. (Courtesy Cochlear Americas.)

OPERATIVE TECHNIQUE

- In adult now done as **single** stage
- In children done in 2 stages (6 months interval) give time for integration
- Implant must be performed meticulously and as gently as possible to minimize thermal damage to surrounding bone
- The skin penetrated by the implant must be hairless to facilitate keeping the implant site clean
- Subcutaneous tissue should be aggressively thinned to minimize skin mobility in relation to the implant
- Place the device 50-60mm from the ear canal
- Drill a whole in mastoid 3-4mm
- At the end place a healing cap onto the abutment
- Sound processor is loaded after 12 weeks (in pediatric post single stage place the processor 16 weeks)

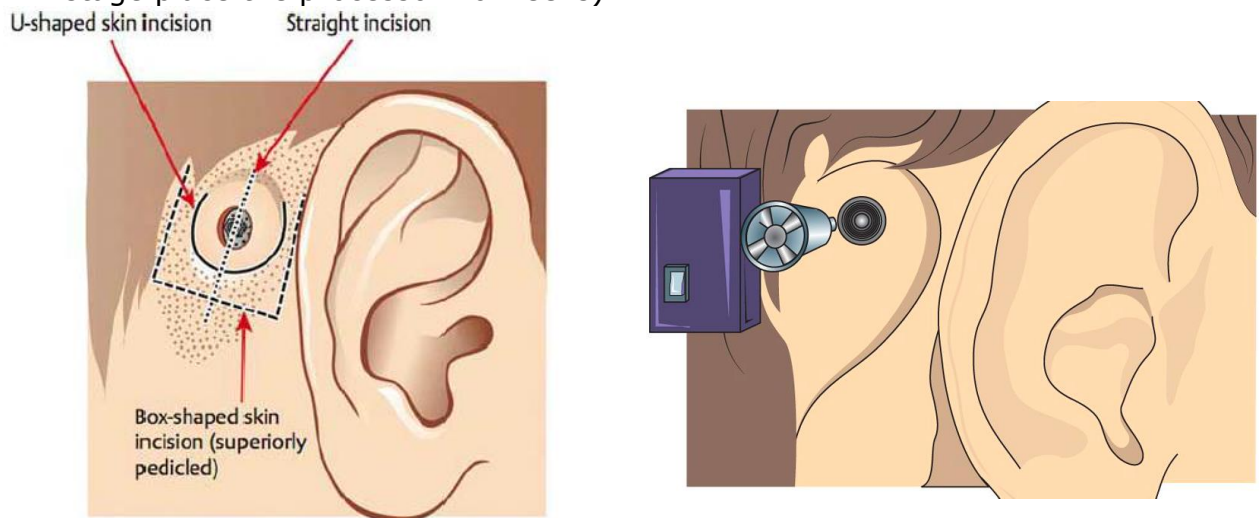


Figure 163.11 Skin incision options for placement of a BAHA. The straight incision has emerged as the preferred option used in clinical practice today.

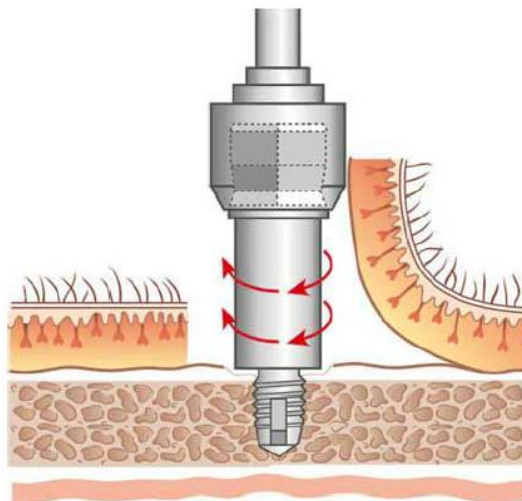
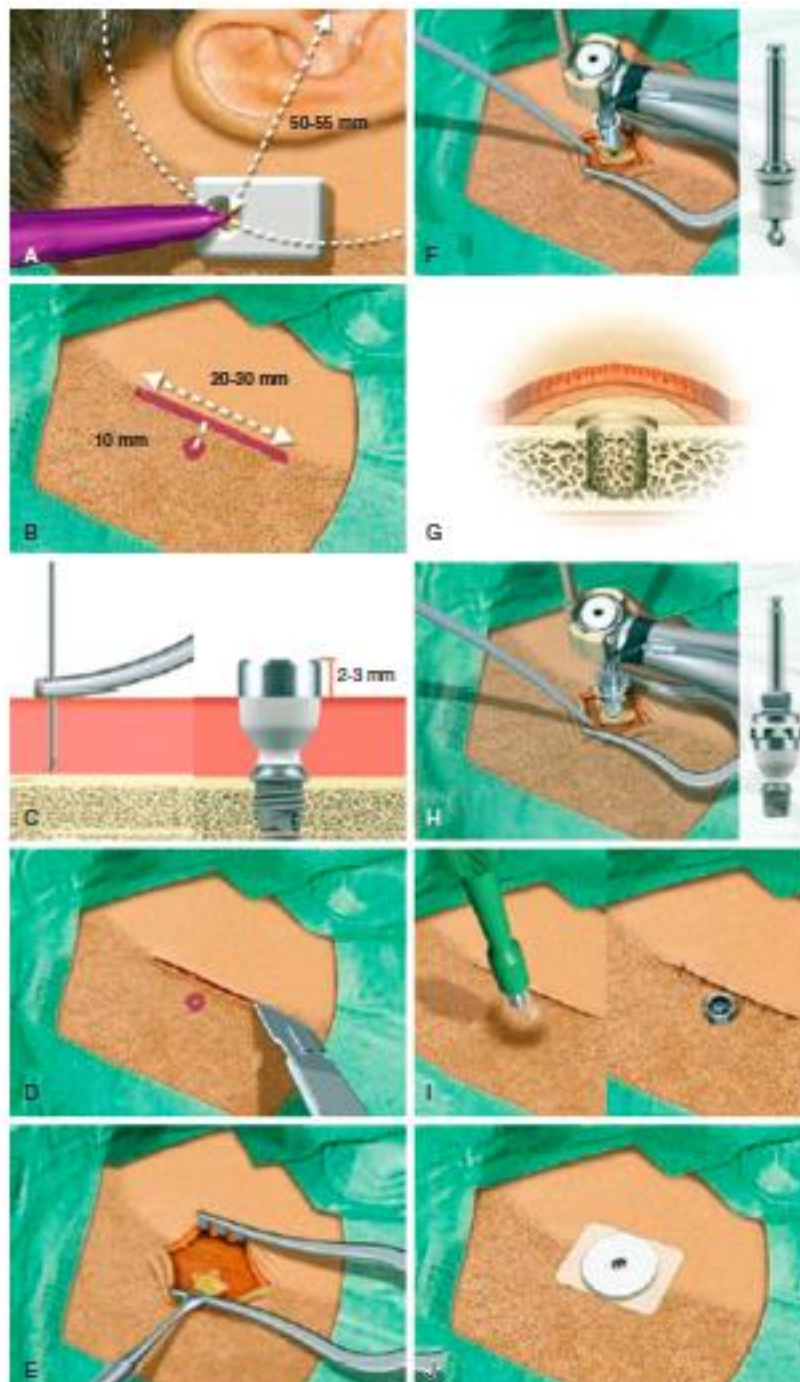


Figure 163.12 Placement of the implant into the skull. First, the thickness of the skull has to be assessed via 3- and 4-mm guide drills. Then, the site for the implant is created by enlarging the initial hole. The implant is then threaded using a torque sensitive drill.



BAHA IN YOUNG CHILDREN

- Baha recommends the osseointegration procedure in children over the age of 5 years.
- For children under the age of 5 years in need of bone-conduction hearing, the Baha Softband is available
- Higher incidence of nonintegration (avoided by 2 stage surgery)
- New studies showed even better results with staged surgery in <5 years old



COMPLICATIONS

- **Soft tissue reaction**
 - Skin overgrowing the abutment (**most common complication**)
 - Redness
 - Infection
 - Necrosis
 - Moisture

Avoided by extensive intraoperative soft tissue reduction. Also, the skin can be tacked down to the periosteal layer to immobilize this region.

- **Osseointegration failure (second most common complication)**
- **Need for revision surgery**

BAHA RESULTS

Recommended that to be in consideration for a "high success rate" with the Baha, patients should have a PTA below 45 dB

BAHA FOR UNILATERAL SENSORINEURAL HEARING LOSS

Compared to CROS:

- Sound localization poor in both
- BAHA has better speech recognition
- BAHA has better amplification
- BAHA has better hearing in noise

Alternatives to BAHA

Transcutaneous Bone-Anchored Hearing Devices

- Created to avoid the complications of having a percutaneous abutment
- Instead The implant is magnetically coupled to the external oscillator and transmits vibrations to the skull
- Same indications as BAHA

1. Alpha 2



2. Bonebridge (Med-El)



Dental Appliance-Based Bone-Conduction System

- The **Sound-Bite**
- Uses a BTE receiver and sound processor that sends signals to a custom-made, removable, in-the-mouth piezoelectric transducer coupled to the buccal surface of the maxillary molars



ITM hearing device inserted in-the-mouth



Custom fits around the upper back teeth



ITM is nearly invisible when worn



MIDDLE EAR IMPLANTS

- 2 types of transducers available
 - **Piezoelectric**
 - **Electromagnetic**
- 2 types of implant:
 - **Partial**
 - external microphone and speech processor
 - transmitter with an external coil that transmits electric energy transcutaneously
 - internal device
 - **Total**
 - house all of the abovementioned internally (including the battery pack).
 - Slightly larger in size and complexity, but less visible (all under)
 - require reoperation at approximately 5-year intervals to exchange the battery
- must adapt to the mechanics of the middle ear and ossicular chain
- Couple to one of the three middle ear bones in one of a number of ways to directly drive the ossicular chain.
- Convert the electric signal into mechanical energy that is then coupled directly to the ossicular chain
- Some designs will also couple directly to the oval or round window.

The Classical indication to MEI is mild-to-moderate sensorineural hearing loss with intact speech discrimination abilities

ELECTROMAGNETIC MIDDLE EAR IMPLANTABLE HEARING DEVICES

VIBRANT SOUNDBRIDGE (MED-EL)

Indicated in:

- Moderate to severe **SNHL**
- **CHL** and **MHL** (when placed in oval or round window or even on ossicle prosthesis "*Vibroplasty*")

Components:

- Implantable component (internal receiver and processor)
- Vibrating ossicular prosthesis (**VORP**) typically attach to long process of incus
- Floating mass transducer (**FMT**)
- External microphone and receiver (similar to HA)

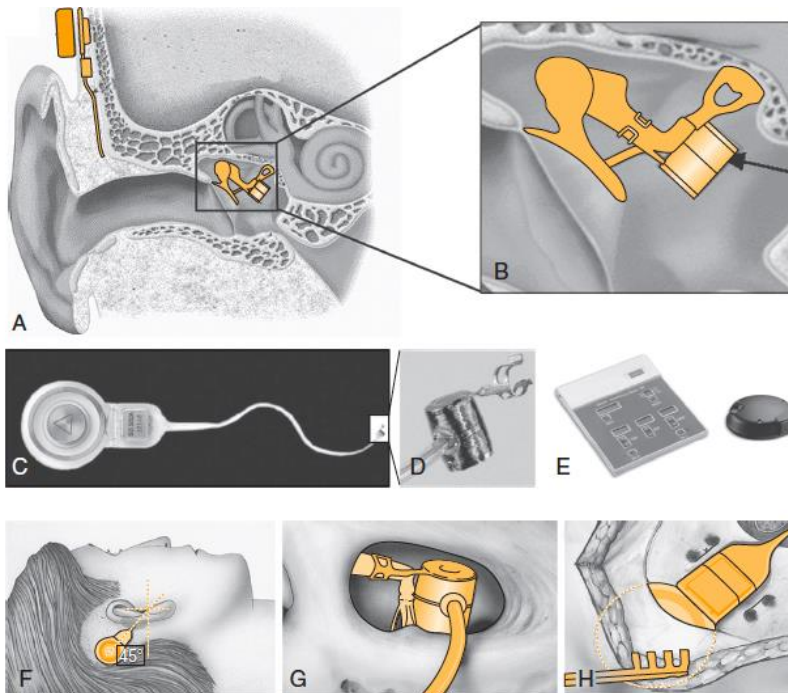
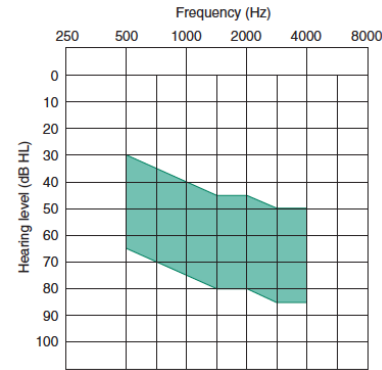


Figure 163.13 FMT of the Vibrant Soundbridge hearing system. The clip on the upper right-hand side attaches to the long process of the incus. In case of alternative placement sites such as the round window, this clip can be removed.

FIGURE 157-3. The Med-El Vibrant Soundbridge semimplantable hearing device. **A** and **B**, External microphone couples via an inductive transcutaneous link to the actuator; a vibrating ossicular prosthesis (VORP) that couples to the incus long process (**B**). **C**, Implanted component of the device. **D**, VORP actuator. **E**, Programming unit and external circuitry. **F** and **G**, Inductive link is implanted against retromastoid cortical bone, similar to a cochlear implant (**F** and **H**). VORP is clipped to the incus via a facial recess approach (**G**).

- Implanted by standard **trnsmastoid facial recess** approach
- Only need to be near the ossicles (not directly attached)
- The ferromagnetic device attached to the ossicles will transmit the auditory signal to the ossicles.
- The VORP is crimped into the **long process of the incus**
- The internal receiver is placed in a bony trough in the retrosigmoid bone

- Because of the confined space between the eardrum and the incus, the dimensions and mass of the FMT are limited, which restricts its output.
- Drawback is **erosion** of the incus long process because of **ischemia** at the clip attachment site
- FMT includes a magnetic component, implantation of this device prevents a patient from undergoing magnetic resonance imaging (MRI) without device removal
- **Other** than incus: stapes, oval window, round window, prosthesis or piston

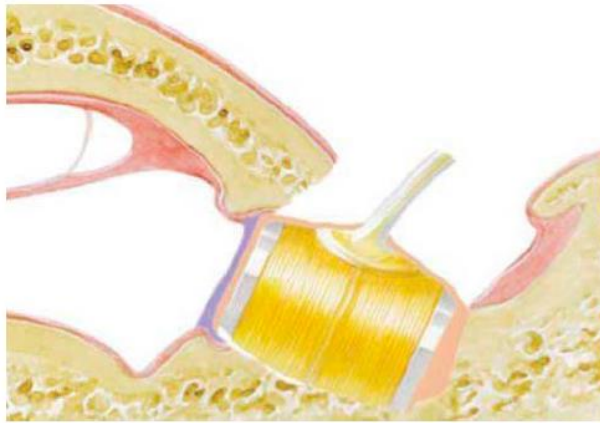


Figure 163.16 Alternate placement site of the Vibrant Soundbridge onto the round window membrane for direct cochlear stimulation.

PIEZOELECTRIC MIDDLE EAR IMPLANTABLE HEARING DEVICES

- Function by connecting the ossicles directly to an amplifier using a piezoelectric crystal vibrator.
- Piezoelectric materials are dielectric materials with coupled electric and mechanical properties
- Larger than electromagnetic devices
- Advantage of this type of transducer is its ability to deliver more distortion-free amplification directly to the ossicular chain
- Attach to body of **incus** body (carina)
- Attach to **malleus and stapes** head (Esteem from envoy)

CARINA

- Fully implanted
- No external component
- Implanted through **atticotomy** and a hole in body of incuse (**laser**)
- Has battery that is chargeable

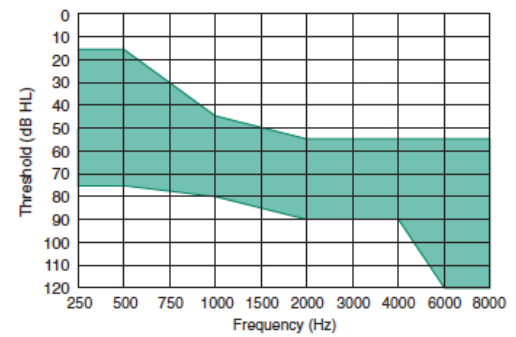


FIGURE 157-7. Audiologic selection criteria for OTOlogics middle ear transducer and Carina. The shaded area of the figure corresponds to the pure tone audiometric implant criteria. HL, hearing loss.

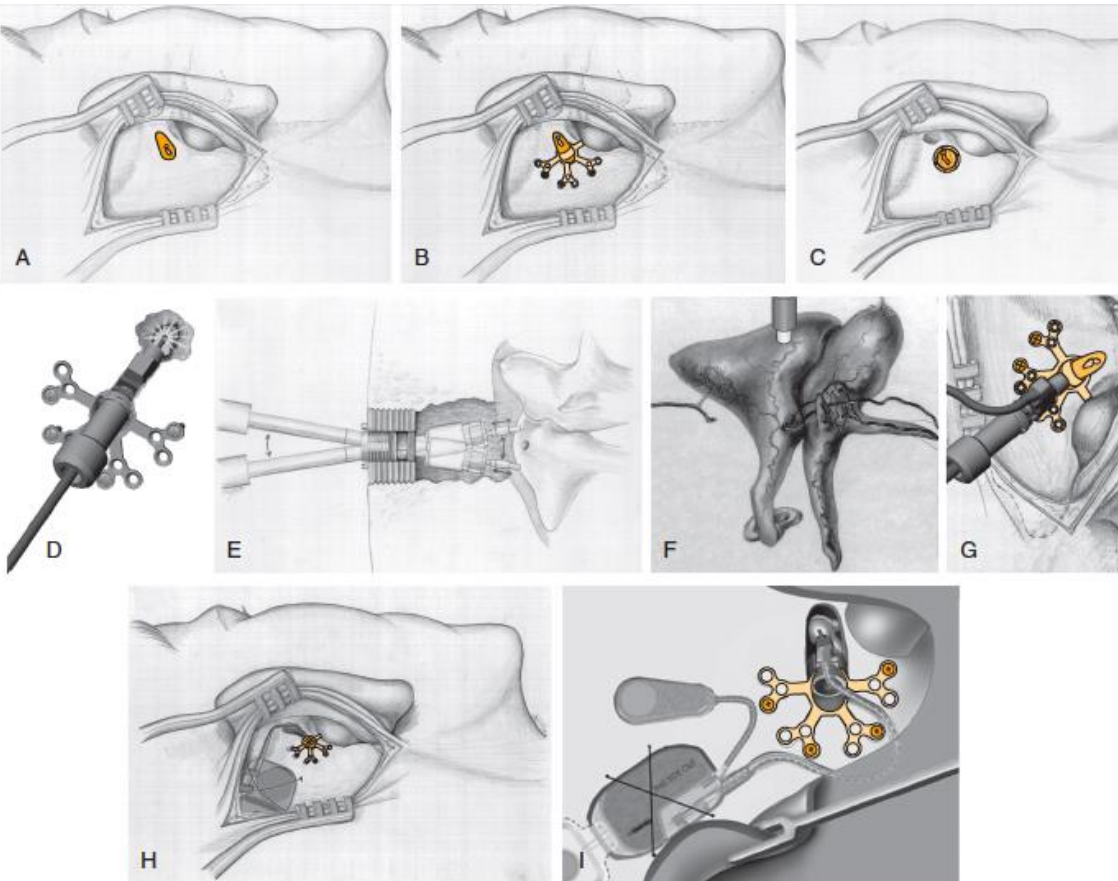


FIGURE 157-6. Surgical implantation of the OTOlogics middle ear transducer (MET) or Carina begins with a limited antrostomy (A) that spares cortical bone ledges for attachment of a metallic stage (B and C), which is used to steady a laser (D) for creation of a small hole in the posterosuperior incus (E). The laser is removed and replaced with the MET actuator, the tip of which inserts into the incudotomy (F). The actuator is secured to the stage (G), and the remainder of the implanted device is secured to retromastoid cortical bone (H). In the Carina totally implantable version of this device (I), a subcutaneous microphone beneath the skin covering the mastoid replaces the external microphone and inductive link used in the MET, but the implantation procedure is otherwise similar.



FIGURE 157-5. OTOlogics Carina fully implantable system. (Courtesy OTOlogics, Boulder, CO.)

Variations on the size and length of the ossicular coupling have expanded the application of the Carina to patients with aural atresia and ossicular discontinuity via direct attachment of the device on the stapes capitulum, stapes footplate, and round window

ESTEEM

- Fully implantable
- Designed for mild to severe HL
- A piezoelectric bimorph placed on the **malleus** head can effectively use the tympanic membrane as a microphone
- Uses a piezoelectric crystal to convert malleus vibrations, the result of sound waves impinging on the tympanic membrane, into a voltage encoding sound.
- Requires partial removal of the incus to prevent feedback from the actuator to the sensor. (This results in a maximal CHL)
- Then output transducer, which converts the applied voltage to mechanical vibration coupled to the **stapes**
- Battery designed to last for 5 years (non chargeable)

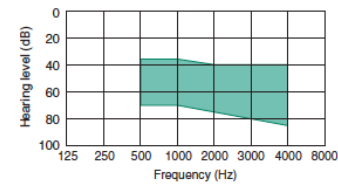


FIGURE 157-9. Audiologic selection criteria for the Envoy Esteem, which was designed for patients with moderate to severe hearing loss (35 to 85 dB) between 500 and 4000 Hz in the implanted ear that is equal to or worse than the hearing loss in the nonimplanted ear when tympanometry is normal and speech discrimination is 60% or greater. The shaded area of the figure corresponds to the pure tone audiometric implant criteria.

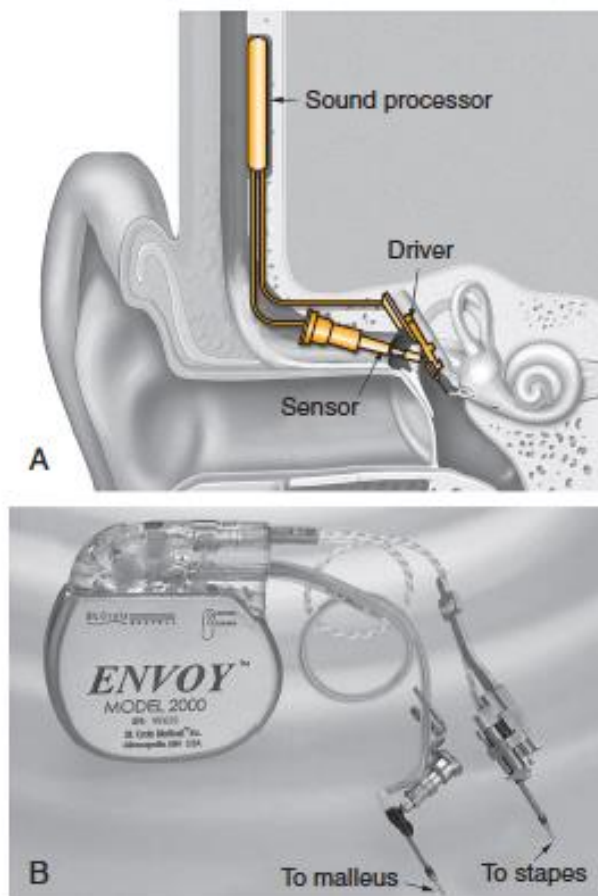


FIGURE 157-8. The Envoy Esteem piezoelectric totally implantable hearing device. **A**, Implantation schematic. Instead of a microphone, sound enters the device via a piezoelectric transducer coupled to the malleus and tympanic membrane. A piezoelectric actuator produces amplified stapes motion. The incus has been removed to prevent feedback. **B**, Complete device.

Complications of MEI

Complications from transmastoid and facial recess drilling.

Specifically, most devices require a large recess => **chorda tympani injury**

Others:

- Skin breakdown
- Postoperative infection
- Extrusion
- Improper device placement
- Inadequate hearing benefit.
- Inadvertent activation with various electromagnetic fields such as a microwave
- Device failure

COCHLEAR IMPLANTS

- A device designed to convert environmental sound into electrical impulses that are delivered along a multiple electrode array situated in close proximity to the cochlear (auditory) nerve
- It has been reserved for patients with significant degrees of hearing loss that are not easily rehabilitated with conventional amplification

Cochlear Implant Systems

- Essential components:
 - **External device** (microphone, speech processor, headpiece)
 - **Internal implanted device** (receiver -stimulator, multichannel electrode array)
 - **Microphone**: convert acoustic to electrical energy
 - **External speech processor**: processes signal (convert it to digital electric signal); processing of the electric signal involves:
 - Filtering; to separate the information into discrete freq. band that are delivered to the appropriate regions of the cochlea to provide spectral information about the speech signal
 - Amplification
 - Compression; the information must be compressed to fit the narrow dynamic range of electric stimulation (It is necessary to map the wide dynamic range of the normal acoustic environment onto the limited dynamic range of electrical stimulation)
 - **Transmitter**
 - **Implanted receiver-stimulator**: decode the signals and sends it to electrode array
 - **Electrode array**: implanted in the cochlea; delivers signal
- **Spiral ganglion cells or axons** are stimulated directly; damaged or missing hair cells are bypassed with a cochlear implant system
- Multielectrode systems are designed to take advantage of **the tonotopic** organization of the cochlea

TABLE
163.1

CURRENT FDA APPROVED COCHLEAR IMPLANT DEVICES

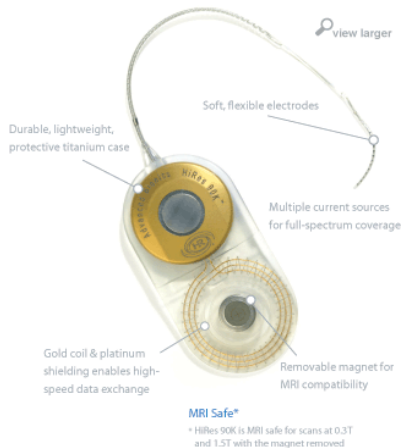
	Advanced Bionics	Cochlear	MedEl
Electrode arrays	Conforming (helix) –15.5 mm/16 contacts Lateral wall (1J) –17 mm/16 contacts	Conforming (contour advance) –15 mm/22 contacts Lateral wall (straight)* –16.4 mm/22 contacts	Conforming (NA) Lateral wall (Standard, Medium, Compressed)* –26.4, 20.9, 12.1 mm/24 contacts
Receiver stimulator	Silastic titanium (90 K)	Silastic titanium (system 5 and freedom)	Silastic titanium (concert and sonata) Ceramic (pulsar)
Processors	Ear level (harmony, auria) Body worn (platinum)	Ear level (system 5)-modular Body worn (NA)	Ear level (opus 1 & 2)-modular Body worn (NA)
Coding strategy	CIS Hi fidelity 120	ACE 1589	Continuous interleaved sampling (CIS) Fine structure processing (FSP)
MRI compatibility	Magnet removal required	Magnet removal required	0.2 Tesla MRI
Rechargeable battery	Available	Available	Available



- **Nucleus 24** cochlear implant system
 - Made by **Cochlear corporation** (also make Nucleus 22)
 - 22 intracochlear electrodes; 2 extracochlear ground electrodes
 - Three programming strategies: SPEAK; ACE and CIS



- **Clarion** or (HiRes 90K) cochlear implant system (**Advanced Bionics corporation**)
 - HiRes Bionic Ear is the newest of their models
 - 16 intracochlear electrodes
 - Can use continuous (CIS) programming strategy, or a variety of other combinations



- **C40+** cochlear implant system/ **Sonata**
 - **Med-El corporation**
 - Combi 40+ system (C40+)
 - 12 electrode pairs; can be inserted deep into apical region



Pathologic Basis for Success

Cochlear implantation usually results in a loss of residual acoustic hearing, presumably from:

- intracochlear **trauma** induced through cochleostomy or electrode insertion
 - **disruption** of the endocochlear **potential**
 - or **delayed** reaction to the **foreign** body.
-
- Trauma and bone dust may also induce further changes in the inner ear including fibrosis and osteoneogenesis.
 - These changes might also reduce future abilities to reimplant the cochlea or the patients to consider other biologic therapies

Patient Selection

Patient's history followed by a physical examination.

Otologic **history** includes

- Onset and progression of hearing loss
- Etiology of hearing loss
- History of amplification use (recommendation use at least 3 months pre-implant)
- History of meningitis
- Ear infections—past/current
- Previous otologic surgeries

Exam

Active infection

Perforations

Tympanostomy tubes

Absolute contraindications to cochlear implantation include those patients without either a cochlea (Michel aplasia) or a cochlear nerve.

Relative contraindication might include

- Active middle ear disease
- severe anesthetic risk
- too much residual hearing
- those who are unwilling to tolerate the surgical risks.

Preexisting mastoid cavity must be either obliterated with blind-sac closure, or the post canal wall must be rebuilt

There are clearly patients who may require adjustment of expectations through

- (a) cochlear disorders such as extensive cochlear obstruction from previous meningitis, otosclerosis, or inner ear malformation
- (b) auditory neuropathy / desynchronize (50%)
- (c) CNS disorders (stroke, dementia, MS, infection, tumor)

Candidacy

➤ Adults

(**>=18 years**) are required to have bilateral moderate-to-profound hearing loss without medical contraindications and the **desire** to be a part of the hearing world. And low aided **speech perception**

**TABLE
163.2**

DEVICE LABELING BASED ON SPEECH RESULTS IN ADULTS

Current **adult** selection criteria in the most recent clinical trials include

- 1) severe or profound hearing loss with a pure-tone average of 70 dB hearing level (HL)
- 2) use of appropriately fitted hearing aids or a trial with amplification
- 3) aided scores on open-set sentence tests of less than 60%,
- 4) no evidence of central auditory lesions or lack of an auditory nerve
- 5) no evidence of contraindications for surgery.

Advanced Bionics Cochlear

MedEL

Test materials	HINT Q	HINT Q	HINT Q
Presentation level	Not specified	70 dB SPL	Not specified
Aided condition	Appropriately fit	Binaural best-aided	Best-aided
Score	≤50%	≤50% implanted ear ≤60% best-aided	≤40% best-aided

HINT Q, Hearing in noise test administered in quiet. Presentation level 70 dB SPL.

➤ Prelingual children

Can be as young as **12** months of age (FDA), with bil profound HL gain limited benefit from amplification, while being enrolled in an early intervention program.

acquire speech and language through central plasticity that results from stimulation by auditory prostheses.

➤ **Earlier implantation has risk of:**

- None accurate diagnostic test (most accurate after 6 months)
- Anatomical challenges (temporal thickness, mastoid development, facial nerve)
- Risk of bleeding (blood volume, immature mastoid bone bleed)
- Difficult language development with HA or after implant

➤ **Clear indication of early implant:**

- After meningitis or temporal or cochlea trauma (before ossification)
- In blind child

Older children with some degree of speech perception should also have specific speech perception testing results that are obtained while wearing appropriate amplification

**TABLE
163.3**

DEVICE LABELING BASED ON SPEECH PERCEPTION TEST RESULTS IN CHILDREN

	Advanced Bionics	Cochlear	MedEL
Pure-tone thresholds (HL)	≥90 dB HL	Bilateral profound	≥90 dB PTA (@ 1 kHz)
Hearing aid trial	3 mo if <2 y 6 mo if >2 y (NA if ossified)	3–6 mo	3–6 mo
Rehabilitation	Not mentioned	Yes	Yes
Open-set test	<20% MLNT (<4 y) <12% PBK (>4 y) <30% HINT (>4 y) (@ 70 dB SPL MLV)	≤30% MLNT	<20% MLNT or LNT

Cochlear implant evaluation for **children** can be considered when

- 1) unaided thresholds are greater than 90 dB HL at 2000 Hz and above in the better ear, even if hearing levels at 250 and 500Hz are better
- 2) aided thresholds in the better ear are greater than 35 dB HL, especially at 4000 Hz
- 3) no response is found for auditory brainstem response testing in both ears or no response is found for one ear and responses are at elevated levels in the other ear;
- 4) parents are disappointed with their child's development of auditory and/or communication skills
- 5) progressive hearing loss is found with detection levels at or near the profound range at 2000 Hz and above;
- 6) evidence of severely impairing AN/D is present.

- **Audiologic assessment**

Adults

- Unaided and aided thresholds for pure tones
- Minimum Speech Test Battery (MSTB)
- Used at many cochlear implant centers to assess pre and postimplant performance
- Set of compact disc recordings for standardization
- Includes the following:
 - Consonant-Nucleus-Consonant (CNC) Monosyllable Word Test
 - Arizona Biomedical (AzBio) Sentences (in quiet and in noise)
 - Bamford-Kowal-Bench Sentences in Noise (BKB-SIN)
 - hearing in noise test (HINT) sentences were previously part of the MSTB but have fallen out of favor due to ceiling effect

Children

- **ABR** and **OAEs**
- Implant candidates typically have no response at limits of the testing equipment
- Findings/implications in auditory neuropathy (-ve ABR/+ve OAE)
- Unaided and aided thresholds for pure tones
- Speech perception tests
- Meaningful Auditory Integration Scale (**MAIS**)
questionnaire for family of children too young to participate in speech perception tests
- Early Speech Perception (**ESP**) Test
word is spoken without visual cues, patient selects correct object or picture of the stimulus
- Lexical Neighborhood Test (LNT), multisyllabic LNT (MLNT)
LNT—50 monosyllabic words, ranging from “easy” (high frequency, few lexical neighbors) to “hard” (low frequency, many lexical neighbors)

MLNT 2-3 syllable words; open-set test

Pre op imaging

Imaging of the temporal bone and brain:

- (a) identify the etiology of hearing loss
- (b) define surgical anatomy and the potential for complications or squeal
- (c) identify factors that negatively impact upon prognosis for performance using the device.

Pre-op CT Scans

1. Location of large mastoid emissary veins
2. Height of jugular bulb
3. Thickness of parietal bone
4. CNS disorders
5. IAC
6. Inner ear morphology
7. Cochlear patency (calcified)
8. Position of fallopian canal
9. Size of facial recess

Pre-op MRI

- Consider in cases of possible ossification or when CT shows IAC < 3 mm diameter to demonstrate a cochlear nerve
- cochlear patency (fibrosis)

(Hypoplastic cochlear nerve can be checked with Ecog intra-op)

- **Psychological assessment**
 - Family dynamics, support structure and motivation assessed
 - Must rule-out early psychosis and MR

Vaccination

- Pediatric and adult patients with cochlear implants are at increased risk of acquiring *S. pneumoniae* meningitis
- Note:
 - **Pprevnar** pneumococcal 7-valent conjugate (conjugated) ears

**Box 159-2. ADVISORY COMMITTEE ON
IMMUNIZATION PRACTICES RECOMMENDATIONS
FOR THOSE RECEIVING A COCHLEAR IMPLANT**

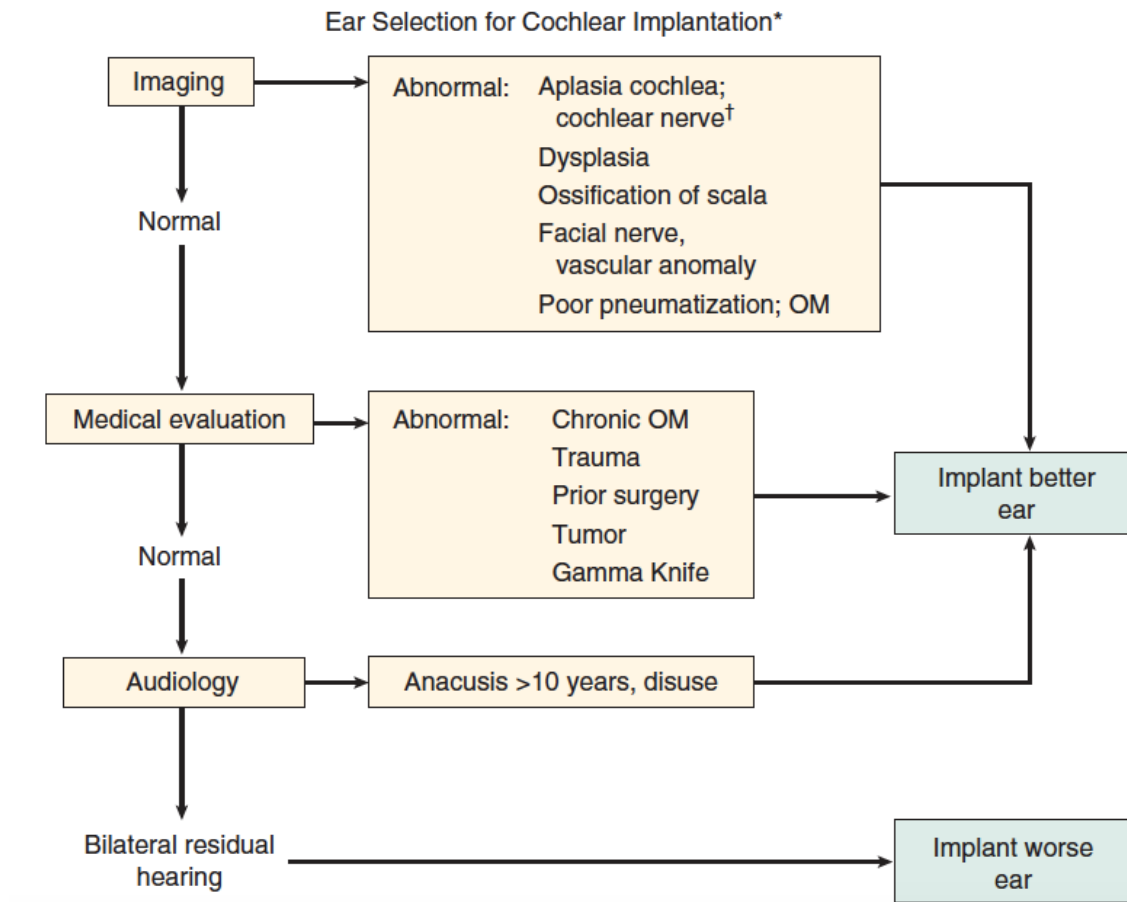
- Children <2 years with cochlear implants should receive PCV-13, as is universally recommended.
- Children who have previously received PCV-7 should receive PCV-13 based on the number of prior doses of PCV-7 received. At least one supplemental dose of PCV-13 should be received, even with complete PCV-7 vaccination. Older children (2-5 years) with no previous PCV-7 vaccination should receive two doses of PCV-13.
- Children >2 years who have completed the PCV-13 series should receive PPSV-23 2 or more months after the final vaccination with PCV-13. Revaccination should be performed 5 years after the initial PPSV-23 dose.
- Pneumococcal vaccine-naïve adults should receive PCV-13 with a dose of PPSV-23 to follow 8 weeks later. A second PPSV-23 dose should be received 5 years after the initial dose if the patient is >65 years.
- Adults who have received at least one dose of PPSV-23 should receive one dose of PCV-13 >1 year from their most recent PPSV-23 dose.
- Persons planning to receive a cochlear implant should be up to date on age-appropriate pneumococcal vaccination ≥ 2 weeks before surgery if possible.

Surgery for Cochlear Implantation

Ear Selection

- Physical characteristics
 - Presence of cochlea and auditory nerve, degree of dysplasia, degree of ossification, prior surgical procedures, CN 7 anomalies, chronic OM
- Residual hearing level
 - Worse ear favoured if opposite ear can be aided
 - Also consider duration of deafness (if >10-15 yrs, implant better hearing ear)

Clear middle ear space should be confirmed through:
Otoscopy, tympanometry, and/or temporal bone imaging



- Perioperative antibiotics are given 30 minutes before skin incision. (A first-generation cephalosporin)
- GA
- Facial monitoring
- local anesthetic with vasoconstrictor is infiltrated post auricular area
- lazy S incision (don't go lower than mid of pinna => superficial facial nerve)
- cortical mastoid and facial recess approach opened maximally

very important to recognize that limited opening of the facial recess can result in an inferior view towards the hypotympanum with a resulting look at the air cell system rather than the promontory and round window region.

- The round window niche overhang is initially identified (located 1-2mm inferior to the oval window niche and anterior-inferior to the stapedius tendon).
- niche overhang is removed to expose the round window membrane, the primary landmark to scala tympani.

Alternately **Cochleostomy placed anteroinferior to round window**

- position of the receiver stimulator usually is significantly superior and posterior to the pinna (subperiosteal pocket is created)
- A bony depression can be created according to the device templates
- Open the round window membrane
- A smooth, resistance-free insertion of the proposed electrode array in to patent scala tympani is the goal of most implantations.
- Fascia or muscle placed around opening for sealing

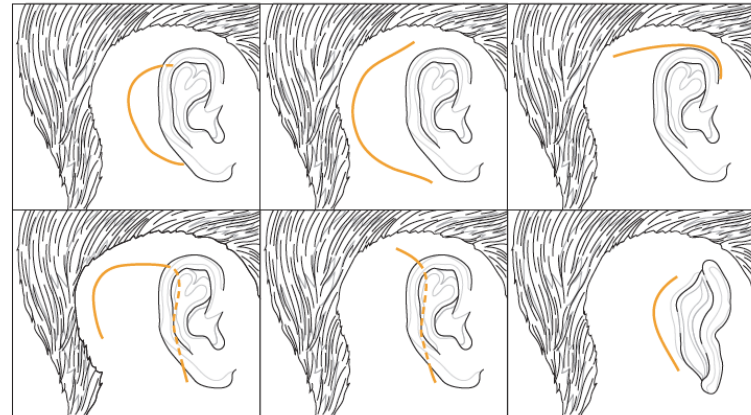
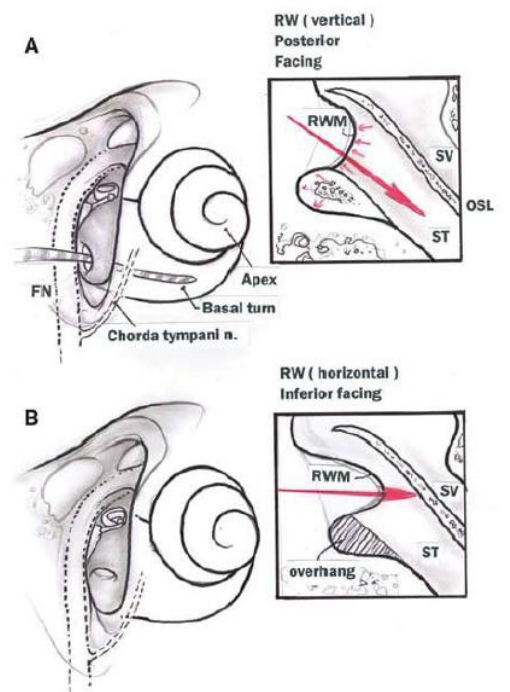
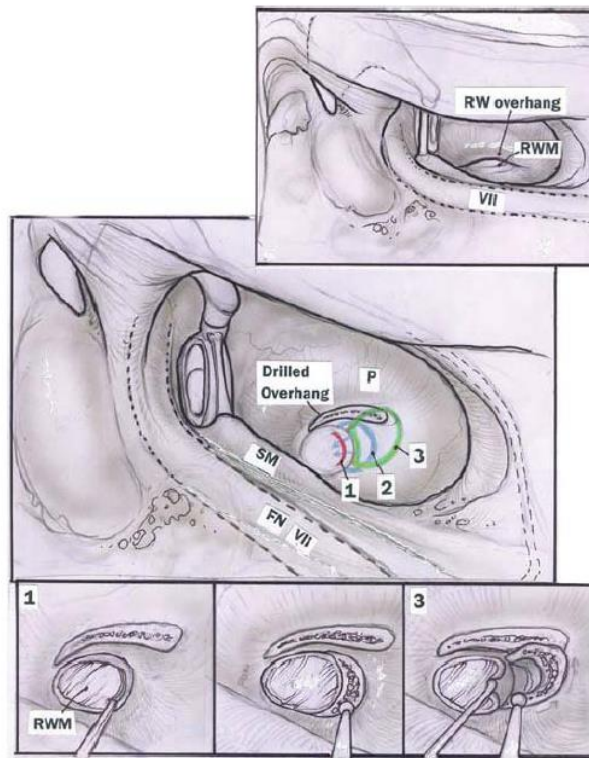


Figure 163.4 Typical skin incision used for cochlear implantation. A similar incision is used for active MEIs. Larger incisions have been used in the past but are currently not used anymore.



Neural response telemetry "NRT"

- Performed during the CI surgery and at the follow-up appointments as needed
 - Gives confirmation that the CI is effectively stimulating the hearing nerve fibres in the inner ear
 - NRT measurements assist in selecting and optimizing initial programming parameters — speeding and simplifying the programming of young children
- electrode impedances (detect open circuit)
 - electrically evoked compound action potentials.

Special Surgical Considerations

For inner ear anomaly and chronic ear (go to specific chapters)

1- Inner Ear Obstruction

- inner ear inflammation from meningitis
- immune-mediated ear disease
- otitis media with labyrinthitis
- severe otosclerosis
- trauma.

cochlear lumen can be narrowed or obliterated with fibrous tissue or bone. Preoperative imaging with MRI and CT will usually reveal the extent to which such obstruction exists and the approach needed for implantation.

In cases of ongoing meningitis, intervention should be considered in the early stages so as to avoid total obstruction

Bilateral implant in initial surgery

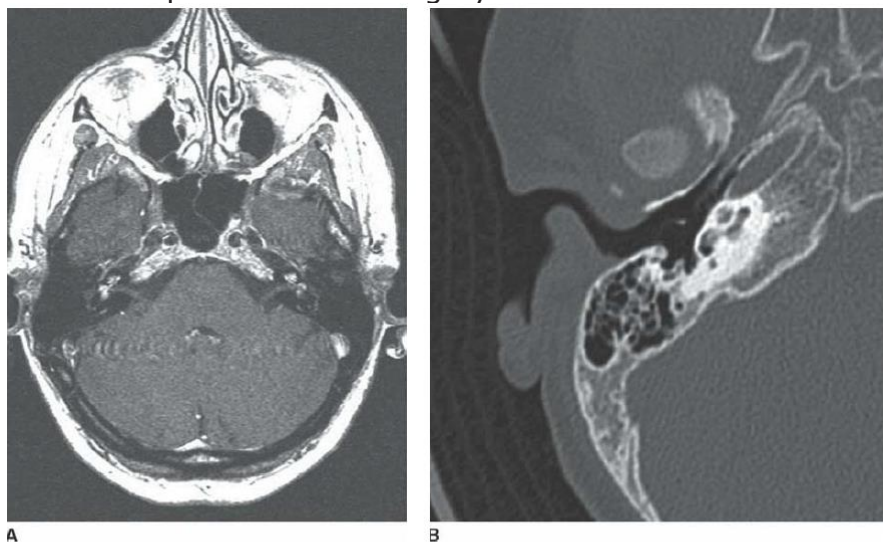


Figure 163.3 Labyrinthine obstruction. **A:** Axial-enhanced T1-weighted MRI demonstrates bilateral labyrinthine enhancement. **B:** Axial bone-window CT demonstrates postmeningitic ossification within the basal cochlear turn.

- In cases of short segment obstruction resulting from either fibrosis or bone (1 to 3 mm), a typical round window-related cochleostomy can be created by drilling through the short segment of obstruction.
- Array insertion can usually proceed in a normal fashion.
- If more than 3 mm of drilling fails to reveal a patent cochlear lumen, a scala vestibuli insertion should be considered by moving superiorly, above and anterior to the round window

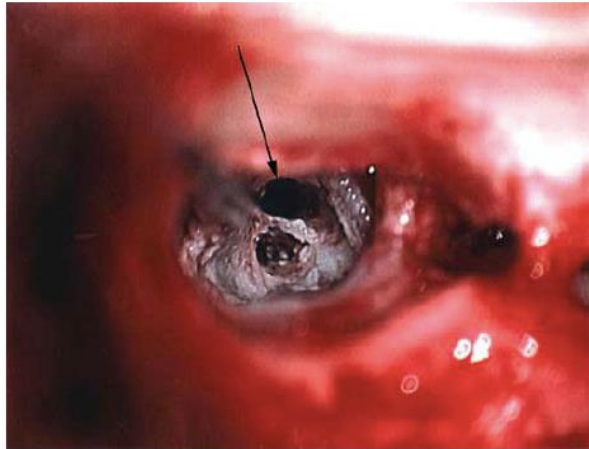


Figure 163.8 Surgical view (left ear) through the facial recess in a case of postmeningitic ossification. A cochleostomy (arrow) into the basal turn's scala vestibuli has been furnished superior to the initial attempt into scala tympani, which revealed ossification of the lumen.

- An apical cochleostomy is also created for an upper retrograde insertion.
- This opening is made anterior to the oval window inferior to the cochleariform process.
- communication with the individual programming the device is critical to insure that proper pitch assignment is made for the retrograde electrodes.

Ossification can happen post meningitis as early as few months
And can happen after 10 years of infection

2- Otitis Media

- Active otitis media is a contraindication to cochlear implantation
- Can result in suppurative labyrinthitis with subsequent meningitis and ossification.
- Device colonization can result in subsequent wound infection, breakdown, and hardware loss
- In cases of acute otitis media, delayed intervention is justified.
- If recurrent disease is probable. tympanostomy tube insertion is reasonable with delayed implantation.
- There are some advocates for subtotal petrousectomy with blind sac closure in the setting of otitis media to avoid future considerations for this problem.
- In cases of serous effusion. the incidence of bacterial colonization is probably very low.
- But in cases of mucoid effusion, delayed intervention is wise.

Pre op OME => place a VT

Intra op found OME can proceed with some difficulty intra-op

Intra op VT => can be removed if no longer needed but if left in place no harm

Post implant OME => can be treated with VT or leave it

Cochlear implantation is associated with neither increased incidence of otitis media nor complications such as labyrinthitis

Post-op follow up

- Mastoid X-ray first day post-op
- Tests of performance:
 - MSTB (Minimum Speech Test Battery)
 - Standardized set of comprehensive tests of pre- and post-op speech recognition
 - Components
 - HINT (Hearing In Noise Test)
 - CNC (consonant/nucleus/consonant test)
- Follow up with audiologist for programming
 - At least q/1 yr for adults
 - At least q/6 mo for kids
 - Also need post-implant rehab (to improve perception and language skills)

Complications

**TABLE
163.4**

COMPLICATIONS OF COCHLEAR IMPLANTATION

Device-Related	Medical/Surgical
Internal Hard Failure (i.e., dead device)	Flap wound (infection, extrusion)
Internal suspected malfunction (i.e., soft failure)	CSF gusher or leak
Speech processor failure	Dural or vascular injury
Accessories malfunction (cables, headpiece, etc)	Incomplete electrode insertion
Open electrode circuits	Electrode extrusion
	Facial nerve injury

- **Hard failures** result from internal, receiver-stimulator problems and **require revision surgery**
- **Soft failure** despite the presence of auditory percepts, the patient experiences aversive symptoms such as pain, shocking, atypical tinnitus, or a decrement in function, device changed only after **confirming location in imaging and changing external device showed no benefit**
- With surgical advancements, these issues have become decidedly rare.
- Minimal approaches to skin incisions and proper device location and immobilization have been key in reducing such complications.

Other complications include

- taste disturbance from chorda tympani nerve manipulation or sacrifice (-10%)
- transient dizziness (-10% of adults and less common in children)
- complete vestibular deficit is uncommon
- subcutaneous seroma (10%) (resolve spontaneous)

Meningitis in Cochlear Implant Recipients

The incidence of meningitis in cochlear implant recipients is greater than that of an age-matched cohort in the general population

Though overall rate is low (<0.6%)

Risk factors in this population include

- young age
- the presence of inner ear malformations
- the use of a two-part electrode system.

The risk of meningitis also appears to be higher among individuals with sensorineural hearing loss without a cochlear implant although this risk appears lower than that of implanted

The most common organism identified in postimplantation meningitis is *S. pneumoniae*.

All potential routes of spread (middle ear, inner ear, hematogenous) should be considered in such a patient.

Precautions:

- create a relatively atraumatic cochlear opening, insert the electrode array without trauma
- adequate seal at the cochleostomy with connective tissue
- pneumococcal vaccination in implanted is recommendation by the FDA and implant manufacturers
- Early and adequate treatment of otitis media

Results

Some of the factors that should be considered include

- Age
 - age of onset of hearing loss (deafness)
 - duration of deafness
 - medical comorbidities,
 - etiology of deafness
 - residual hearing
 - anatomic cochlear and cochlear nerve status
 - cognitive abilities, behavioral condition
 - psychological factors
 - parental desires and involvement
 - educational setting and training
-
- Very wide range of results; very patient dependent
 - Most important factor is duration of deafness; longer = poorer results
 - Speech recognition remains the standard metric of cochlear implant candidacy and performance
 - Adult:
 - Depends on age at implantation and duration of deafness
 - Average post-op sound field detection thresholds for warble-tone stimuli are approximately 25 to 30 dB HL for frequencies between 250 and 4000 Hz
 - Children:
 - Spoken word recognition
 - Similar to adults but continual development of speech perception and spoken word recognition abilities over a long time course
 - In children, initial reports suggested that implantation before the age of 3 years provides distinct advantages over later implantation in cases of early-onset deafness.
 - More recent data suggest that speech recognition and language skills are facilitated with implantation in earlier toddler and infancy stages.
 - Outcomes related to educational success, quality-of-life impact, and cost effectiveness after early cochlear implantation generally relate to a child's ability to acquire oral language as a result of auditory sensitivity and enhanced speech recognition.

- Screening for other **handicapping** conditions, particularly conditions that would impair the acquisition of receptive and productive communication skills, helps determine candidacy and directs rehabilitative strategies.
- In the absence of handicapping conditions, a generic analysis of the literature on cochlear implant outcomes yields the conclusion that more than **85%** of implant recipients are able to use their device to gain meaningful engagement with the hearing world.
- Postimplantation **rehabilitation** can be important for some adult implant recipients, but it is crucial for children to optimize the usefulness of an implant.

A second cochlear implant, implanted sequentially or simultaneously, improves sound localization and speech recognition in noise over a single implant.

**TABLE
163.5**

**ADVANTAGES AND DISADVANTAGES
OF BILATERAL COCHLEAR
IMPLANTATION**

Bilateral Cochlear Implants

Advantages	Disadvantages
Always implant better ear	Two surgeries
Hearing in quiet	One or two anesthetics
Hearing in noise	Loss of acoustic hearing
Never off the air	Bathtub hearing
	CI limited frequency spectrum
	Future therapies
	Vestibular effects
	Double programming
	Economics

- **Finally**, cochlear implants are being placed in patients with progressively greater degrees of residual hearing using special electrode arrays and surgical techniques that allow for preservation of residual acoustic hearing.
- The combination of electric and acoustic stimulation in the same ear (electro acoustic stimulation or **hybrid** implantation



With the benefits of binaural hearing increasingly being recognized, candidacy decisions now include bilateral implantation, cochlear implantation for single-sided deafness, and implantation of ears with low-frequency hearing with hearing preservation.

Revision CI

Non-Device-Related Indications

- Infections
- Cholesteatoma
(remove the cholesteatoma remove the disease and cut the electrode in the cochlea, reimplant in second look with blind sac)
- Allergic reactions
- Extracochlear electrodes (most common after device related)

Device-Related Indications

- most common cause of reimplantation
- Facial nerve stimulation
- Device failure (hard or soft)

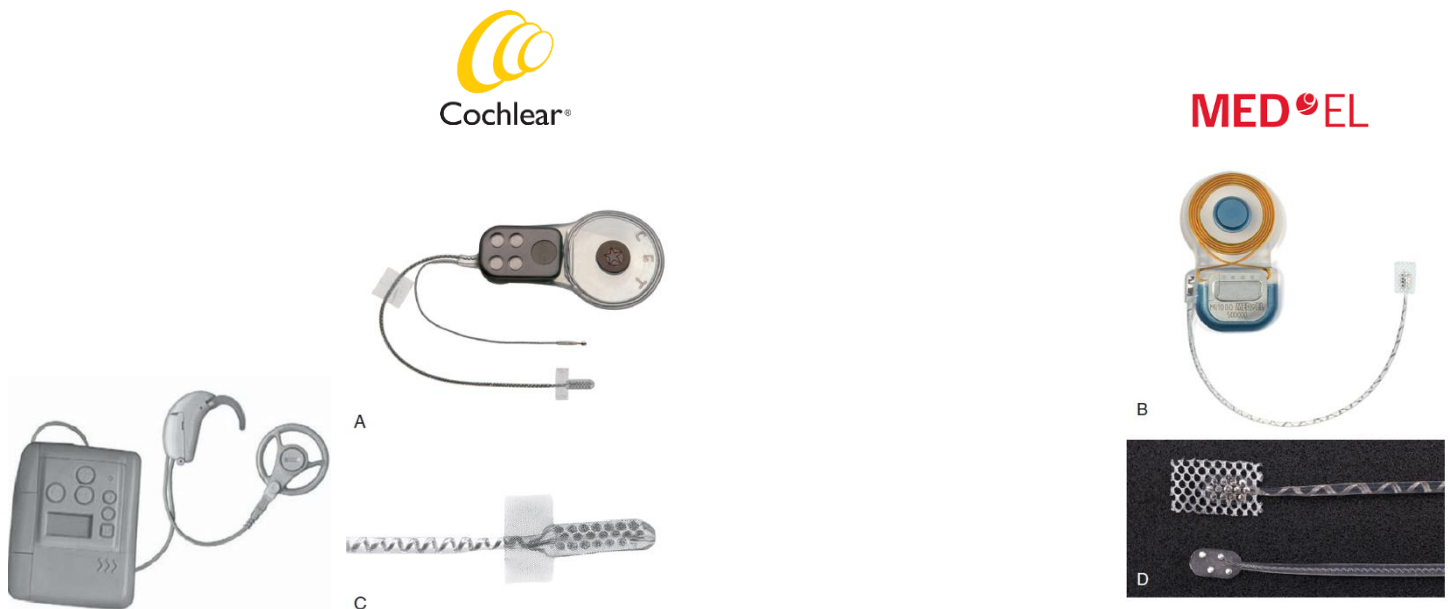
AUDITORY CNS IMPLANTS (Auditory Brainstem Implant)

- convert environmental sound into electrical impulses that are delivered along a multiple electrode array situated in close proximity to the cochlear nucleus in the brainstem.
- The **ABI** is indicated for patients in whom the cochlear nerve is absent or unavailable for stimulation bilaterally.

(Most commonly, patients with hearing loss secondary to neurofibromatosis type II with bilateral acoustic tumors)

Principles of Electric Stimulation of the Auditory System and Coding Strategy

- ABI was developed from the cochlear implant; the basic tenants of stimulation are similar
- The tonotopic arrangement of the **cochlear nucleus**. however, is somewhat more complex and less accessible
 - cochlear nerve fibers transmitting **low-frequency** sounds project mainly to the **ventral** portion of the cochlear nucleus
 - while basal cochlear nerve fiber transmitting **high-frequency** sounds project to the **dorsal** portions of each nuclear subdivision



Indications:

Profound hearing loss and no available cochlear nerve to stimulate bilaterally.

Designed for the purposes of helping patients with neurofibromatosis type II this remains the most common indication and the only FDA-approved.

There remain a number of other clinical scenarios that might be amenable to ABI, and these include

- Congenital cochlear nerve aplasia.
- bilateral temporal bone fracture with cochlear nerve disruption
- severe inner ear malformation (cochlear aplasia, internal auditory canal aplasia)
- dense ossification secondary to previous meningitis or advanced otosclerosis

In these cases, cochlear nerve integrity might be unclear and a trial of cochlear implantation might be justified prior to undertaking an ABI operation

Surgery

❖ Approach

generally carried out through a posterior fossa craniotomy-either

- **translabrynthine**

Or

- **rettosigmoid.**

distinct advantages of the **translabrynthine** approach for ABI are

- (a) incision location that is more natural for receiver-stimulator placement.
- (b) a lack of cerebellar retraction
- (c) direct line of site to the lateral recess of the 4th ventricle afforded by the presigmoid exposure
- (d) electrode path through the immobile temporal bone rather against the pulsatile cerebellar hemisphere

However, it is limited in some cases by a high jugular bulb, making lower cranial nerve and recess visualization somewhat more tangential and difficult at times

❖ **Cranial nerve monitoring**

Should include facial muscle electrodes, as well as palatal and trapezius electrodes to monitor cranial nerves 9, 10, and 11.

❖ **ABR monitoring electrodes**

critical for appropriate device placement

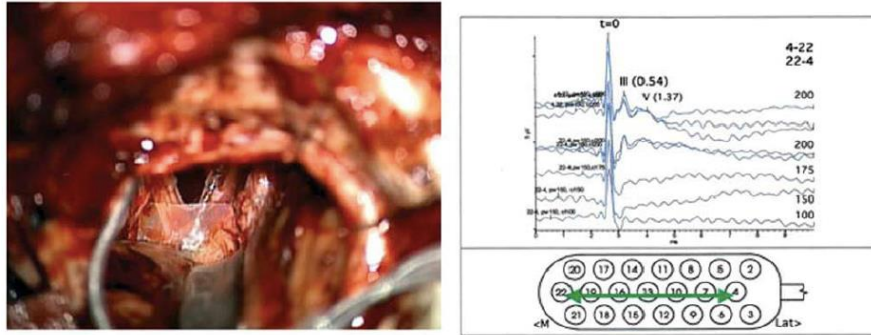


Figure 163.20 Intraoperative image (left) and electrically evoked auditory brainstem response (E-ABR) demonstrating proper placement of an ABR. The recording example shows evoked potentials when stimulating across the array (demonstrated below).

- ❖ The **receiver-stimulator** should be placed in a region similar to the cochlear implant. in a subperiosteal pocket Given
- ❖ The **lateral recess of the fourth ventricle** (i.e., foramen of Luschka) is the target for device placement.
Landmarks include the 8th and 9th cranial nerve root entry zones
 - 8th superior boarder of recess
 - 9th inferior boarders of the recess
- ❖ A postoperative **CT** also confirms array location



Figure 163.21 Axial head CT shows the expected postoperative appearance after placement of an ABR into the left lateral recess.

Complications:

Most complications described in these patients result from the posterior fossa surgery indicated for removing the patient's acoustic neuroma rather than from the device placement.

Include

- bleeding.
- Infection
- facial nerve injury
- loss of residual hearing
- **CSF** leakage (along the electrode carrier is more common among ABI patients than typical acoustic tumor patients)
- Electrode dislodgment or difficult to find the recess
- Experience non auditory sensation (tingling, dizziness, jittering of eye, choking, pain, vertigo, facial twitching, and potential changes in heart rate, blood pressure, and respiratory efforts
- Stroke
- transient dysphonia aspiration related to lower cranial nerve dysfunction.
- death.

Results:

- Wide range of results
- The ABI often provides environmental sound awareness and acts as an aid to lip reading
- however, open-set speech perception can be achieved
- Better speech-perception outcomes have been demonstrated in **non tumorous** NF2 (non-NF2) recipients with conditions such as postmeningitic ossification or traumatic auditory nerve avulsion.

Cochlear Implantation in Chronic Ear

- Cochlear implant are Auditory neural stimulators for patients with preserved auditory nerve populations and are usually placed longitudinally within the cochlear lumen, in Close proximity to the distal nerve fibers.
- Through auditory electrical stimulation, these devices have allowed patients with congenital and acquired deafness to obtain sound awareness and enhanced speech understanding in both quiet and noise, thereby drastically improving educational and employment opportunities, social integration, and overall quality of life.

Absolute contraindications to CI

- Without either a cochlea (Michel aplasia) or cochlear nerve.

Relative contraindication might include those patients with

- Active middle ear disease,
- Severe aesthetic risk
- Too much residual hearing

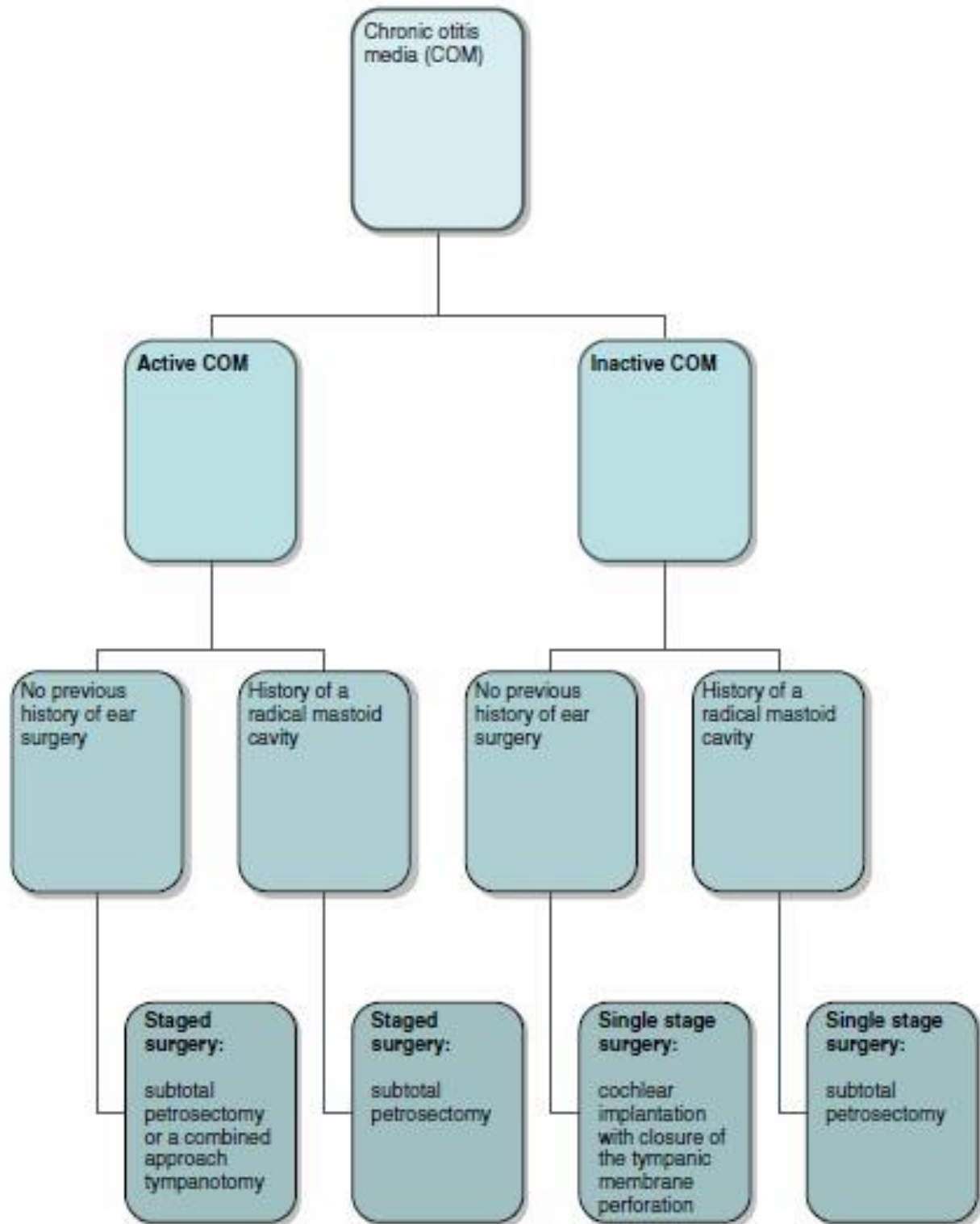
Cochlear implantation in a chronic diseased ear may lead to

- Implant colonization and subsequent implant extrusion
- Meningitis as a consequence of inserting an electrode through an infected mastoid or middle ear cavity into a space with intracranial communication.

The degree of the activity of the disease has influence on the cochlear implant strategy in patients with a chronic diseased ear.

COM can be distinguished on clinical and radiological characteristics into

- **Active** (with or without cholesteatoma)
- **Inactive** form



Inactive form either

- 1. Simple dry perforation**
- 2. A pre-existing radical mastoid cavity without Hx of active disease for 6/12**

1- Inactive form with a simple dry perforation, **placement of the cochlear implant and closing of the dry tympanic membrane perforation can be performed as a single stage procedure**

2- In patients having a pre-existing radical mastoid cavity without evidence for an active inflammation, **cochlear implantation is performed with a subtotal petrosectomy (Blind Sac) as a single stage procedure**

Reports on complications which required surgical treatment after CI in patients with a stable COM are rare although some cases can be found

The complications include:

- Recurrence of cholesteatoma
- Explantation of the implant due to severe inflammation
- Wound breakdown
- Retraction pocket exposing the electrode array
- Extrusion of the implant side due to flap difficulties

All of these cases were reported as a consequence of flaring up of the infection which all required subsequent surgical treatment

Interestingly, all complications occurred after either single-staged or staged procedures

In case of an active infection (with or without cholesteatoma) Either:

- **With pre-existing radical mastoid cavity with active disease**
- **Non-operated with active disease**

In all patients with COM the main issues of concerns are:

- Recurrence of a cholesteatoma
- Flaring-up of the infection

The options for active disease are mainly **staged procedure** either

- Combined approach tympanotomy with posterior tympanotomy followed by CI in 3-6 months
- Subtotal petrosectomy (blind sac procedure) followed by CI in 3-6 months
- Middle cranial fossa approach (to bypass the infected medium)

Subtotal petrosectomy (blind sac procedure):

- Complete exenteration of all accessible air-cell of the temporal bone
- Sealing the Eustachian tube orifice
- Closure of the external meatus (blind sac)

This may be followed by obliteration of the tympanomastoid cleft with a pedicled temporalis flap or with abdominal fat

Drawbacks:

- Risk of cholesteatoma recurrence if epithelial cells were left behind which may lead to asymptomatic destruction (need to take utmost care)
- Mucocoele may developed (if not all mucosa removed although its rare)
- Difficulty to facilitate radiological imaging and a second look to detect a recurrent cholesteatoma (To overcome this issue the tympanomastoid cavity can be left open)

Middle fossa approach

- Access to the cochlea bypassing the possible infected conventional route for cochlear implantation
- The electrode is inserted through a basal turn cochleostomy created in the floor of the middle cranial fossa

The possible drawbacks of this procedure are

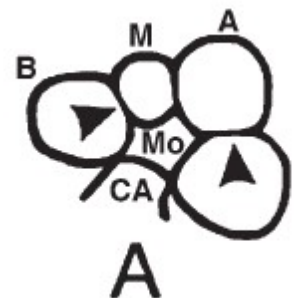
- Risks of a craniotomy and compression of the temporal lobe
- Vascular injuries
- Facial nerve injury

Cochlear Implantation in Inner Ear Malformations

- Inner ear malformations constitute about 20% of congenital sensorineural hearing loss.
- At the beginning of the cochlear implant (CI) era, inner ear malformations were regarded as a contraindication to surgery.
- In the present day, CI surgery in inner ear malformations is accepted as a standard surgical approach.
- There are two major surgical problems in CI surgery in this patient population:
 1. Cerebrospinal fluid (CSF) gusher and meningitis
 2. Facial nerve anomalies
- **Classification of cochlear malformations**
- Jackler et al. (1987) made the first classification of inner ear malformations.
- They separated the anomalies into five groups:
 1. Michel deformity (labyrinthine aplasia)
 2. Cochlear aplasia
 3. Common cavity
 4. Cochlear hypoplasia
 5. Incomplete partition (IP)
- Sennaroglu and Saatci (2002) later defined the radiological features of two completely different types of IP anomalies of the cochlea, as IP-I and IP-II
- Recently X-linked deafness has been recognized as a third type of IP, IP-III

Normal cochlea

- The most important section to determine the normal cochlear architecture and IP anomalies is the **midmodiolar section** (Figure 1A)
- Midmodiolar view demonstrates the modiolus as a quadrangular or pentagonal structure in the centre of the basal turn.
- Interscalar septa are thicker partitions between the inner wall of the cochlea and the modiolus, which appear to separate the normal cochlea into 2.5 (or 2.75) turns; the basal, middle and apical turns
- The cochlear aperture (or cribriform area) is the central bony defect at the base of the modiolus transmitting the cochlear nerve and blood vessels.



- **A section inferior to the midmodiolar view**, passing through the area of the round window niche (Figure 1B), shows the basal, middle and apical cochlear turns.
- The basal turn is in continuity at this section.
- It is important to see the interscalar septum between the middle and apical turns.



Cochlear malformations

Labyrinthine aplasia (Michel deformity)

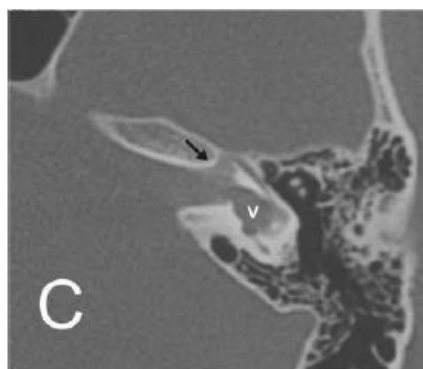
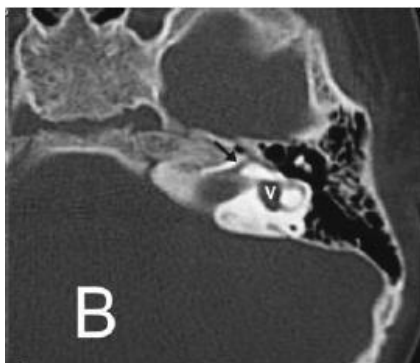
- The cochlea, vestibule, vestibular aqueduct and cochlear aqueduct are absent.
- In the majority of patients, the internal auditory canal (IAC) consists of only a bony facial canal. Nerves other than the facial nerve are not present.

Figure: only stapes present



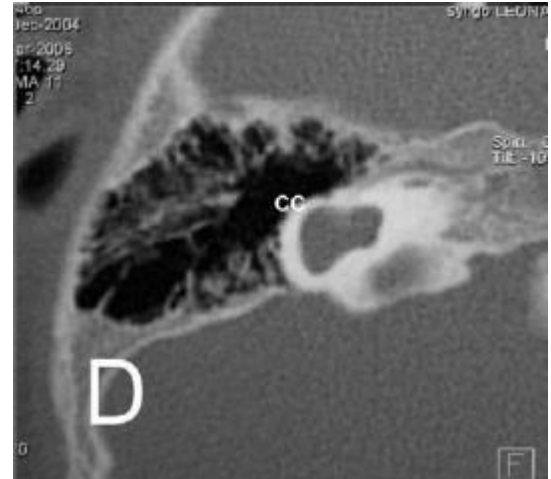
Cochlear aplasia

- This is absence of the cochlea. The accompanying vestibular system may be normal (Figures c and B) or there may be an enlarged vestibule. (Figures D and C).



Common cavity

- In this malformation, the cochlea and vestibule are represented by a single chamber (Figures E and D).
- Theoretically, this structure has cochlear and vestibular neural structures
- Presence of the cochlear or cochleovestibular nerve should be demonstrated by MRI before cochlear implantation.
- It can be an ovoid or round structure.
- Semicircular canals (SCCs) or their rudimentary parts may accompany a common cavity
- The IAC usually enters the cavity at its center.
- A common cavity is more anteriorly located than cochlear aplasia with vestibular dilatation.
- Cochlear implantation in a common cavity may result in good acoustic stimulation but in cochlear aplasia with vestibular dilatation there will be no stimulation with a CI.



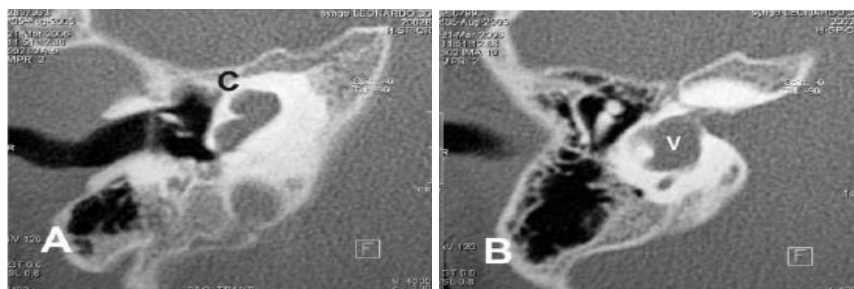
Incomplete Partition (IP) of the cochlea

Three different types of IP cases were identified according to the defect in the modiolus and the interscalar septa.

IP type I

This is the type of cochlea already described in 'cystic cochleovestibular malformation'

- In this group, the cochlea lacks:
 - the entire modiolus
 - interscalar septa
- It looks like an empty cystic structure. (Figure A)
- It is accompanied by a large dilated vestibule (Figure B).

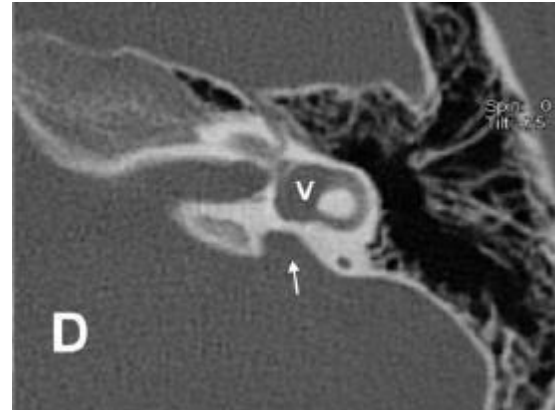
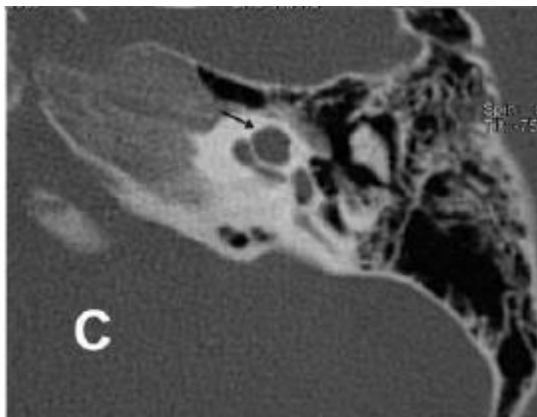


Cochlea (C), without the modiolus and interscalar septa, looks like an empty cystic structure (A), is accompanied by dilated vestibule (v)

- Due to the defective development of the cochlear aperture and absence of the modiolus, there is a defect between the IAC and the cochlea.
- The cochlea is located in its usual location in the anterolateral part of the fundus of the IAC

IP type II

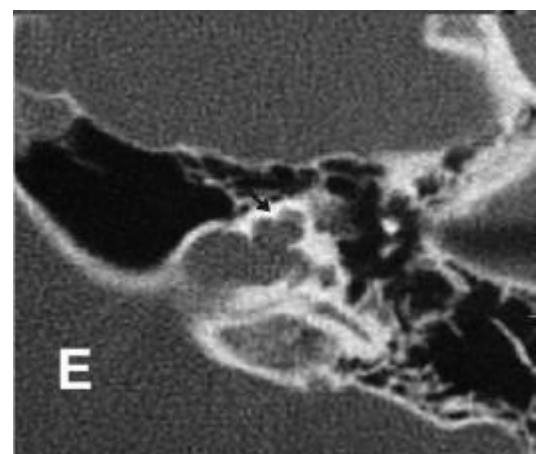
- In a type II cochlea only the basal part of the modiolus is present (Figures G and C).
- This is the type of cochlea originally described by Carlo Mondini
- IP type II with a minimally dilated vestibule and large vestibular aqueduct constitutes the triad of the **Mondini deformity**. (Figure D)



- The apical part of the modiolus and the corresponding interscalar septa are defective.
- This gives **the apex of the cochlea a cystic appearance** due to the confluence of the **middle** and **apical** turns

IP type III

- This is the type of cochlea observed and reported in X-linked deafness
- In this deformity, the interscalar septa are present but the modiolus is completely absent (Figures H and E).
- The cochlea is placed directly at the lateral end of the IAC instead of its usual anterolateral position. (open immediately to IAC)
- This gives the cochlea a characteristic appearance



Hypoplasia

- This is the cochlea with dimensions less than normal.
- In smaller cochlea, it is usually difficult to count the number of turns with computerised tomography (CT) and/or MRI.
- the definition 'cochlea with 1.5 turns' should be used for hypoplasia (particularly type III), rather than for IP type II cochlea
- Three different types of cochlear hypoplasia can be identified.

Type I (bud-like cochlea)

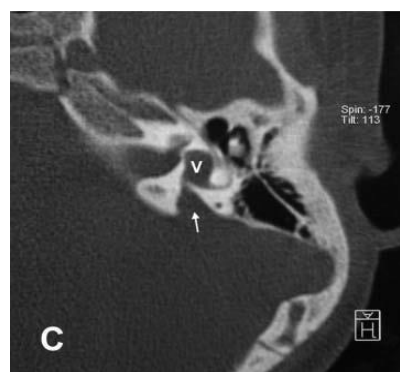
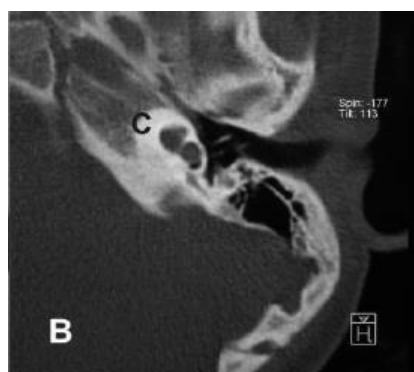
- The cochlea is like a small bud arising from the IAC (Figures I and A).
- Internal architecture is severely deformed; no modiolus or interscalar septa can be identified.

Cochlea (C) is like a small bud arising from the IAC. Modiolus and interscalar septa cannot be identified



Type II (cystic hypoplastic cochlea)

- The cochlea is smaller in its dimensions with no modiolus and interscalar septa, but its external architecture is normal (Figures 1J and 4B).
- There is a wide connection with the IAC. The vestibular aqueduct is enlarged and the vestibule is minimally dilated (Figure 4C).
- This is the type of hypoplasia where a gusher and unintentional entry of the electrode into IAC are possible.



Type II (Cystic hypoplastic cochlea): Cochlea (c) is smaller in dimensions with no modiolus and interscalar septa, but external architecture is normal (B).

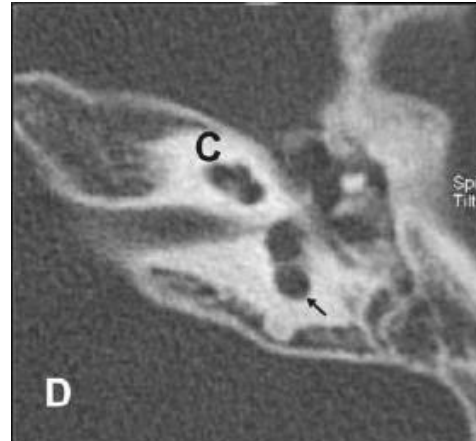
Vestibular aqueduct (white arrow) is enlarged and vestibule (v) is minimally dilated (c)

Type III (cochlea with less than two turns)

- The cochlea has a shorter modiolus and the overall length of the interscalar septa is less, resulting in a smaller number of turns (less than two turns)
- The internal and external architecture (modiolus, interscalar septa) **is similar to that of a normal cochlea, but the dimensions are less** and hence the number of turns is less (Figures K and D).
- The vestibule and the SCCs are hypoplastic.

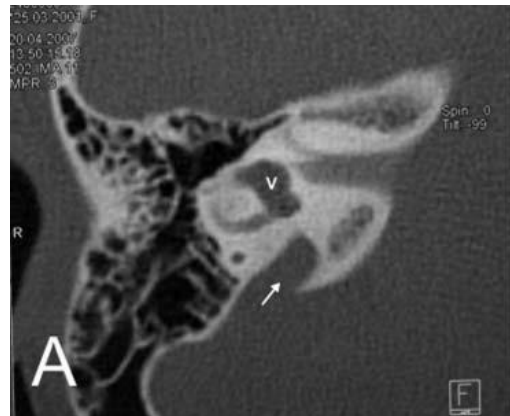


Cochlea (c) with fewer number of turns. Internal and external architecture (modiolus, interscalar septa) is similar to normal cochlea, but the dimensions are less and hence the number of turns is less. Ampulla of the posterior semicircular canal is dilated (black arrow).



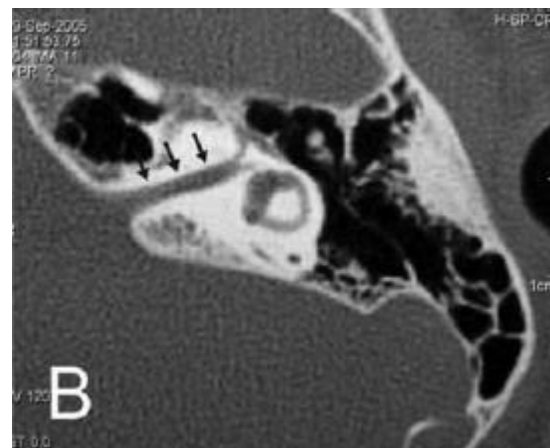
Large vestibular aqueduct syndrome (LVAS)

- This is the presence of an enlarged vestibular aqueduct (the midpoint between the posterior labyrinth and operculum is larger than 1.5 mm)
- In the presence of normal cochlea, vestibule and SCCs (Figure 5A).



Narrow IAC

- The width of midpoint of the IAC is smaller than **2.5 mm** (Figure 5B).
- In cases of a narrow IAC, there is a possibility of an absent or hypoplastic cochlear nerve and this must be checked with MRI.



Preoperative radiological evaluation to diagnose the presence of an inner ear malformation, to determine its type and to identify any other abnormality in the temporal bone that may be encountered during surgery such as :

- Type of malformation,
- Pathologies in the middle ear and mastoid, □ Presence of the cochlear nerve.
- Sclerotic mastoid
- Narrow facial recess
- Facial nerve anomaly
- Defect in the IAC with potential CSF gusher

Radiology may guide the decision to offer a CI in patients with inner ear malformations

The decision to offer a CI is taken earlier in patients who are less likely to achieve benefit from hearing aids, such as:

- Common cavity
- IP-I
- IP-III.

In some patients hearing level may be suitable for hearing aids like

- IP-II
- LVAS

In some patients directly evaluated for auditory brainstem implantation (ABI):

- Labyrinthine aplasia
- Cochlear aplasia

HRCT

HRCT of the temporal bone should be obtained in axial and coronal sections This gives very good bony details of the temporal bone. Classification can easily be done following HRCT

MRI

MRI is important to diagnose:

- Presence of nerves in the IAC
- Cochlear fluids (indirectly can also show Cochlear fibrosis)

The cochlear nerve can be absent in inner ear malformations and this is a contraindication to CI surgery.

Aplasia of the cochlear cranial nerve needs to be ruled out with MRI, especially in

- Common cavity abnormality
- Narrow IAC visualized on CT scan (i.e. <2 mm in diameter)
- CHARGE syndrome

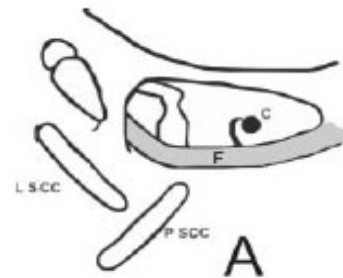
Recurrent meningitis is not rare in this patient group (and MRI will show indirectly cochlear fibrosis by demonstrating a decrease in signal intensity coming from the inner ear fluids.

Surgical approach

The majority of CI operations in malformations can be done via the classical transmastoid-facial recess approach.

Facial recess approach done through the triangular space between:

- Facial canal
- Fossa incudis
- Ear canal



There are two situations where facial recess approach cannot be used:

1. Abnormal location of the facial nerve in the facial recess

- Course of the facial nerve may be altered in certain malformations such as IP-I, common cavity or cochlear hypoplasia.
- It usually lies over the oval and round windows and the surgeon may be unable to use the facial recess approach.

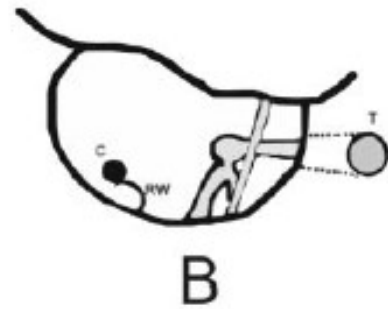
2. Unfavorable cochlear anatomy through the facial recess area

- In certain inner ear malformations, the cochlear promontory may not be fully developed
- The round window and other necessary landmarks may not be visualized and cochleostomy cannot be performed.

Transcanal approach

Being used in standard CI surgery in some centers

- When the anatomy of the inner ear is not severely distorted, a transcanal approach can also be used for cochlear implantation
- In severe anomalies, such as common cavity and hypoplastic cochlea, it may be difficult to use this approach



The electrode cable can be then transferred to the mastoid cavity after making a cut in the bony ear canal.

Transmastoid labyrinthotomy

By taking a direct transmastoid labyrinthotomy approach to the common cavity and avoiding the facial recess and promontory dissection it may be possible to implant the electrode array with maximum visualisation and minimal risk to the facial nerve

Canal wall-down mastoidectomy with blind sac closure

This is a preferred approach when there is:

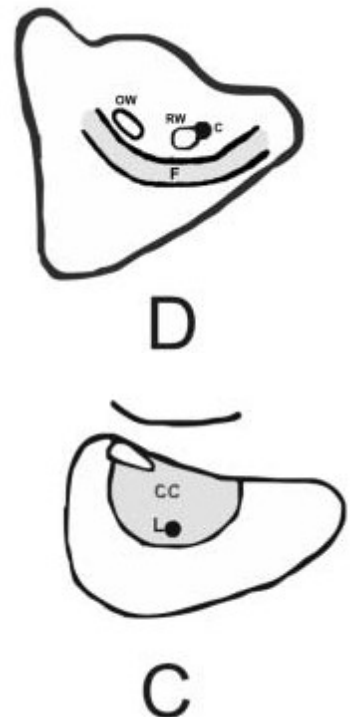
1. **Facial nerve location** prevents the standard facial recess approach,
2. In patients with **uncontrollable gusher and recurrent meningitis**

Using a canal wall-down procedure provides better visualization of the promontory, and oval and round windows

There are two disadvantages of this procedure:

1. The **duration** of the operation is increased because the whole skin of the ear canal and the tympanic membrane must be completely removed.
2. There is a possibility of **leaving some squamous epithelium** in the cavity which could become cholesteatoma within a period of a few months.

This may create a surgical problem because MRI cannot be used to differentiate between cholesteatoma and other soft tissue masses in patients with a CI.



Facial nerve anomalies in malformations

The facial nerve has a complex route in the temporal bone.

One of the most important factors determining the final position of the facial canal is the development of the inner ear.

Common cavity and cochlear aplasia and hypoplasia cases

Anteromedial migration of the labyrinthine segment of the facial nerve may occur in

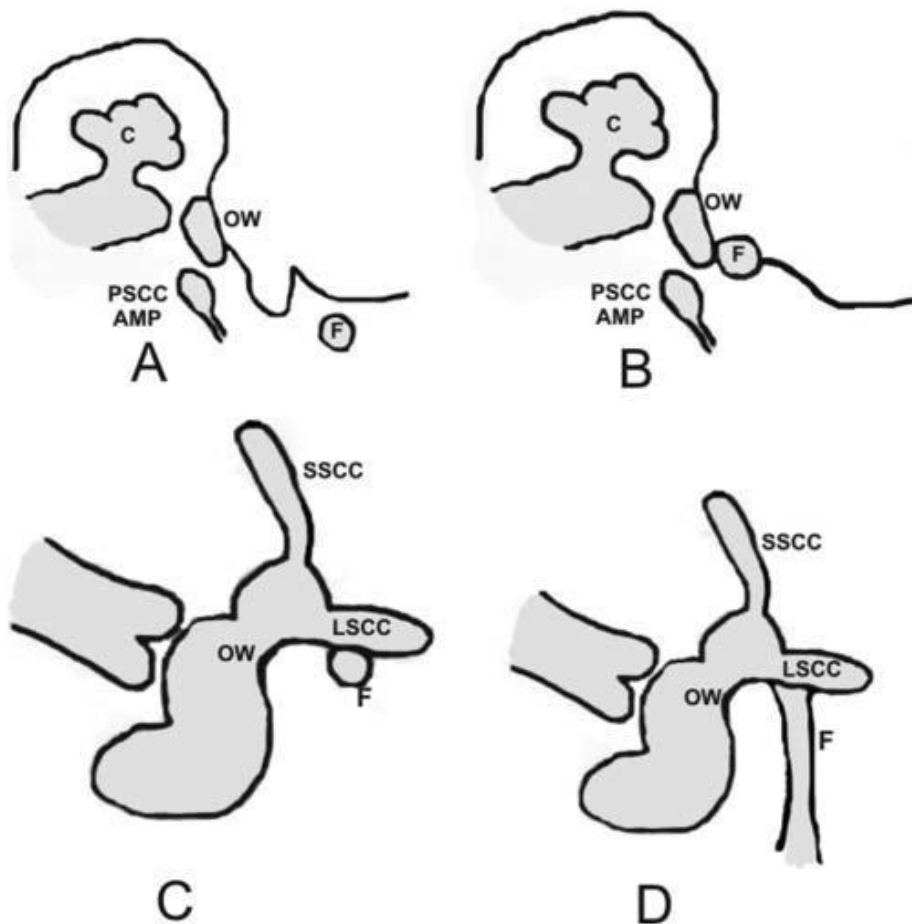
Mondini malformation the facial nerve is at the usual location

Hypoplasia or aplasia of the lateral SCC may cause a change in the location of the second genu

Large Vestibular Aqueduct, no change in the course of the facial nerve is expected

For CI surgery, the most important parts of the facial canal are the second genu and the vertical segment

IP-I has severe facial nerve anomalies preventing facial recess access are possible



Facial nerve: normal course and abnormalities as seen on HRCT.

A: Normal HRCT. Axial section showing the relationship between the facial nerve (F) and the oval window (OW). Vertical segment of the facial nerve is located lateral and posterior to the oval window.

B: Anteromedial dislocation of the facial nerve. Axial section showing the facial nerve which is dislocated anteriorly and medially lying adjacent to the oval window.

C: Normal HRCT. Coronal section showing the relationship between the facial nerve (F) and the oval window (OW). The tympanic segment of the facial nerve lies inferior to the lateral semicircular canal (LSCC).

D: Anterior dislocation of the facial nerve. Instead of the horizontally lying tympanic segment, vertical segment is seen on the coronal section inferior to the LSCC.

CSF gusher

Definition: The term 'gusher' is generally used in the literature to describe the egress of profuse clear fluid upon making an opening into the inner ear.

This is common during surgery in inner ear malformations either:

- Cochlear implant cochleostomy
- Stapedectomy

In cases of gusher, the fluid coming from the inner ear cannot be perilymph, as the inner ear contains only a few microliters of perilymph.

The profuse fluid coming from the inner ear is **CSF** and is due to a defect of variable size between the malformed inner ear and the IAC.

Incidence

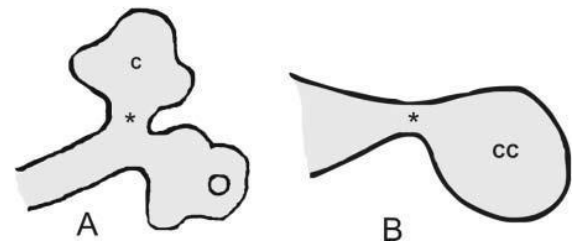
The majority of the papers reported the incidence of gusher to be between 40-50% of their patients with inner ear malformations.

Types of gusher

A gusher of CSF is the result of an abnormal bony defect at the lateral end of the internal auditory meatus (Figure A and B).

Normally, the CSF in the subarachnoid space extends laterally into the IAC as far as the fundus, where it is separated from the perilymph by the bony plate of the lamina cribrosa.

In some congenitally dysplastic ears, there is a defect at the lateral end of the IAC, allowing direct confluence of CSF and perilymph.



Two types have been described:

- Oozing (A gentle flow of clear fluid)
- Gusher (a profuse flow)

Oozing:

Result of a small defect between the malformed inner ear and the IAC.

Intermittent flow of CSF in small quantities

Usually stops after a few minutes.

Small defect between the IAC and the malformed ear

Easily controlled CSF outflow with soft tissue packing around the electrode.

More common in type II and LVAS.

Gusher:

Result from wide communication between the subarachnoid space and inner ear.
Profuse CSF outflow upon making the cochleostomy.

Lasts between 10 and 20 minutes.

This is the more serious type with more risk of causing postoperative meningitis.

With IP-I and some patients with IP-II.

IP-III is the least frequent form of IP anomaly

Radiology

Demonstrates the defect at the lateral end of the IAC.

HRCT is the best method to demonstrate the defect at the lateral end of the IAC

Treatment

It is very important to firmly pack a piece of muscle around the electrode lead at the level of the cochleostomy to prevent CSF fistula in the postoperative period.

If the seal is not watertight, there is a risk of permanent CSF leakage with the potential risk of meningitis.

Size of the cochleostomy

Small cochleostomy:

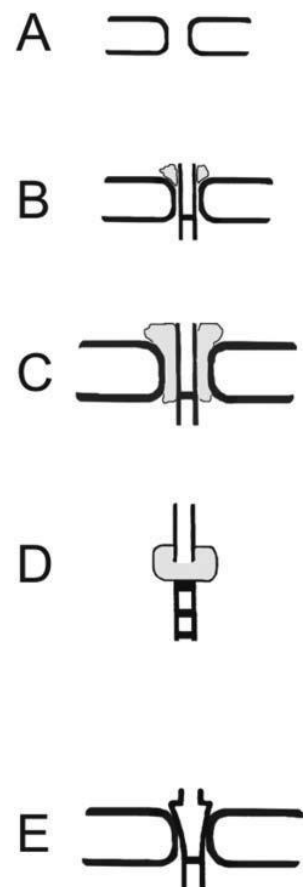
Allowing the electrode cable to partly block the flow of CSF, reinforced with connective tissue, muscle and fibrin glue (Figure A and B).

It was observed that it was very difficult to effectively insert pieces of muscle or fascia around the electrode inside the cochleostomy.

Large cochleostomy:

Allows easy insertion of the electrode and seems to make introduction of muscle around the electrode reasonably easier (Figure C).

Proper application of muscle tissue around the cochleostomy represents the first line of barrier against CSF leakage and the flow of CSF stops quickly when a proper seal has been established.



Helpful maneuvers:

- Elevating the head of the operating table reduces the flow from the gusher.
- Putting the tip of the suction on the bone at the edge of the hole keeps the area round the hole and also that part of the middle ear clear of fluid

Continuous lumbar drainage (CLD)

Although rarely required, this method should be applied if the gusher cannot be controlled effectively during surgery or the child develops rhinorrhoea after surgery.

Usually CLD is kept in its place for four to five days

The purpose of CLD is to lower the pressure of CSF

It is also important to temporarily close the Eustachian tube orifice with Surgicel to prevent CSF leakage into the nasopharynx in the postoperative period

Use of soft tissue around the electrode

Application of soft tissue (muscle or fascia) is important in cases of gusher.

Different ways:

- Fascia or muscle can be applied as small pieces separately around the electrode.
- Harvest a small round piece of fascia, about 5–6 mm in diameter. The electrode is passed through a hole made in the centre of the fascia block (Figure D). When the electrode is inserted the fascia is advanced to the level of the cochleostomy around the electrode.

D



Eustachian tube obliteration

After sealing the cochleostomy, the Eustachian tube is temporarily blocked to prevent escape of the CSF into the nasopharynx.

Oxidised cellulose (SURGICEL) is the material usually preferred for this purpose.

Subtotal Petrosectomy (BLIND SAC)

In cases of intractable CSF leakage, more secure sealing of the leak can be achieved by subtotal petrosectomy

This operation has important steps:

1. The lateral part of the skin of the external meatus is everted and closed as a blind sac.
2. The medial part of the skin of the ear canal and the tympanic membrane are completely removed.
3. The canal wall is taken down, and the facial canal and oval and round windows are exposed.
4. The Eustachian tube is blocked.
5. The mastoid cavity is obliterated with abdominal fat. Obliteration is necessary in patients with gusher.

Additional measures:

Head elevation

48 hours of bed rest decrease CSF leakage.

Acetazolamide can be used to decrease CSF production

Avoid heavy lifting

CSF fistula and meningitis

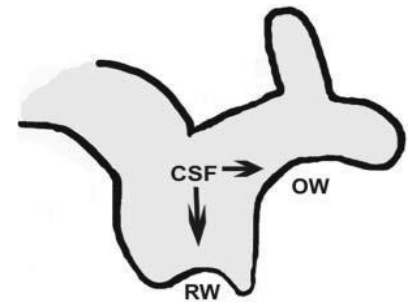
CSF fistula in inner ear malformations is associated with considerable morbidity and mortality.

It may easily lead to recurrent meningitis unless there is a surgical intervention to repair the fistula.

The majority of CSF fistulae are located at the stapes footplate and occasionally encountered at the cochleostomy site

Pathogenesis:

Inner ear malformations with a wide defect in the **lamina cribrosa** and modiolus may cause CSF to come into contact with the medial surface of the oval and round windows. Continuous CSF pressure may cause erosion and then a defect and fistula at the stapes footplate (Figure 10).



Site of fistula:

Majority of spontaneous CSF fistulae are reported to be located in the oval window.

Sometimes the leak site can be the cochleostomy.

Contralateral non implanted site can be the source.

Radiology:

Radiology is the most important tool to diagnose CSF leaks preoperatively and the predisposing anatomic factors.

The presence of inner ear malformations, a defect in the lamina cribrosa or a bone fracture on HRCT may be the aetiology of a CSF gusher.

CT cisternography:

- can show CSF fistula in patients with CSF otorhinorrhoea and unilateral hearing loss
- Which is an invasive method, demonstrates the fistula reliably in patients with a CI.

A non-invasive method for confirming this finding is a FSE T2-weighted MRI sequence through the region

MRI (using 3DFSE T2WI and 3D FIESTA sequences) a useful technique in the assessment of patients with CSF fistulae

Treatment

Middle ear exploration

Once the diagnosis is made, the middle ear should be explored to diagnose the site of the leak and treat the fistula.

Oval window leak:

After removing the ossicles, the vestibule is packed through the oval window with a layer of muscle or fascia followed by an injection of fibrin glue

If the stapes crurae can be preserved during surgery, then they may be helpful in stabilising the soft tissue graft in the oval window.

Another layer of muscle or fascia may be used on top.

Intraoperative CLD and lumbar drainage continued postoperatively

Cochleostomy site leak:

This should be sealed by Temporalis fascia packing around the electrode array and into the vestibule until the leakage stopped.

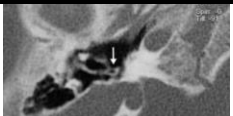
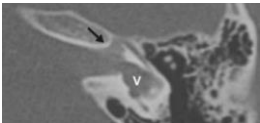
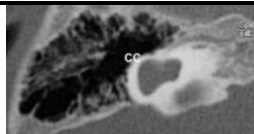
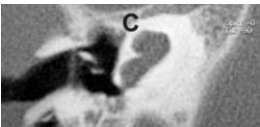
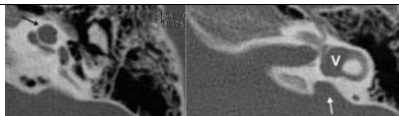
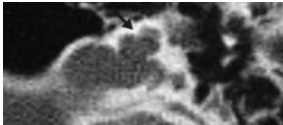
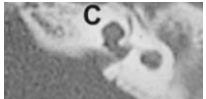
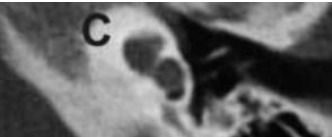
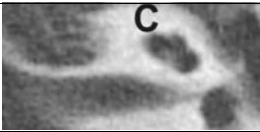
A piece of temporalis muscle then placed in the middle ear space on top of the fascia. The wound then closed, and a lumbar drain inserted.

Subtotal petrousectomy

In recurrent meningitis some recommended subtotal petrousectomy with removal of the middle ear mucosa and closure of the Eustachian tube and ear canal

Inner Ear Malformations

Riyadh et al. Notes

	Cochlea	Vestibule	SSC	VAD	IAM	Schematic	CT
Labyrinthine Aplasia (Michel)	X	X	X	X	Narrow contains FN only	-	
Cochlear Aplasia	X	Normal/Dilated Posteriorly located	✓	✓	Narrow contains FN and vestibular only		
Common Cavity	Single chamber Anteriorly located IAM enters at its center		Rudimentary	✓	✓		
Incomplete Partition – I (Cystic cochlea)	- No Mod - No ISS	Dilated	✓	✓	Normal, Defect between IAM and Cochlea (Gusher)		
Incomplete Partition – II (Mondini)	- No Apical Mod - No Apical ISS (Cystic cochlear apex)	Dilated	✓	Dilated	✓		
Incomplete Partition – III	- No Mod - Present ISS Located at lateral end of IAM	Dilated	✓	Dilated	✓ (Gusher)		
Cochlear Hypoplasia Type-I	- No Mod - No ISS (Bud-like cochlea)	✓	✓	✓	✓ (Gusher)		
Cochlear Hypoplasia Type-II	- No Mod - No ISS (Cystic hypoplastic cochlea)	Dilated	✓	Dilated	Normal, wide connection with IAM (Gusher)		
Cochlear Hypoplasia Type-III	- Small Mod - Small ISS (1.5 turn Cochlea)	Hypoplastic	Hypoplastic	Dilated	✓		

Labyrinthine Ossificans

- **What is Labyrinthine Ossificans (LO)?**
 - Pathologic formation of new bone within the lumen of the otic capsule
- **What are the causes of LO?**
 1. Bacterial meningitis leading to Suppurative labyrinthitis (Most common cause)
 2. Suppurative labyrinthitis associated with AOM.
 3. Advanced Otosclerosis
 4. Temporal bone trauma
 5. Autoimmune inner ear disease
 6. Vascular ischemia of labyrinthine artery
 7. Leukemia
- **What is the most common cause of LO?**
 - Bacterial meningitis.
 - Leads to Suppurative labyrinthitis
 - 80% of patients with profound postmeningitic SNHL have LO.
 - Complete ossification is associated with a very poor prognosis for residual hearing.
- **What are the most common organisms causing meningitis?**
 - *S pneumoniae* (Most common)
 - *Neisseria meningitidis*
 - *H influenzae*
- **Does vaccination in children reduce the risk of meningitis?**
 - *S. pneumoniae* vaccination has decreased the incidence of **severe** *S.pneumoniae* infections.
 - *Haemophilus influenzae* vaccination has decreased the incidence of *Haemophilus influenzae* meningitis.
- **Which organism is the most common to cause ossification?**
 - Post-meningitis SNHL greatest with *S pneumoniae* (**20-30%**).
 - Lowest with *Haemophilus influenzae* (**5%**).

- **What are the risk factors for post-meningitis SNHL?**
 - Disease Factors:
 1. S. pneumonia
 2. High ICP
 3. Low CSF Glucose
 4. Long hospital stay > 14 days
 5. Seizure
 - Treatment Factors:
 1. Delay treatment
 2. Partial antibiotic treatment
 3. No steroid therapy
- **What are the stages of post-meningitis SNHL?**
 1. Acute stage:
 - Purulence fills the perilymphatic space.
 - Begins within days after the onset of meningitis.
 - Seen in MRI as cochlear enhancement (Earliest finding).
 2. Fibrous stage:
 - Fibroblastic proliferation within the perilymphatic space.
 - Begins 2 weeks after the onset of meningitis.
 - Seen in MRI (More sensitive) but not in CT (False negative results in 60%).
 3. Ossification stage:
 - Bone formation in the basal turn of cochlea with obliteration of perilymphatic and endolymphatic spaces.
 - Begins as early as 2 months after the onset of meningitis.
 - Seen in both CT and MRI.
- **Where is the earliest site of involvement in LO and why?**
 - Scala tympani of the basal turn due to its relative decreased blood flow.
- **When will be the onset of hearing loss following meningitis?**
 - Ossification has been noted to occur within a year after meningitis, although the hearing loss may occur as early as 48 hours after infection.
- **What is the recommended protocol Post-meningitis?**
 - Audiological evaluation for all post-meningitis patients should be done as soon as the clinical condition of the patient allows.
 - Radiological evaluation for all post-meningitis patients should be done with Gad-MRI within the 1st month after meningitis (to evaluate for cochlear enhancement).

- **What are the findings on MRI in patients with post-meningitis SNHL?**

- Gad T1 MRI:

- **Cochlear Enhancement:**

- **Earliest imaging finding of post-meningitis SNHL.**
- Seen as early as 1st day of infection till 8 weeks after infection.
- Indicates active inflammation and found to be related to the occurrence of SNHL.
- Accentuating the necessity for rapid cochlear implantation.

- T2 MRI:

- **Cochlear Obliteration:**

- Detect cochlear lumen obliteration caused by fibrosis or ossification.
- Seen as early as 2 weeks of infection..
- Most sensitive imaging tool (90-100% accurate in predicting intra-op difficulty in insertion of CI electrode).

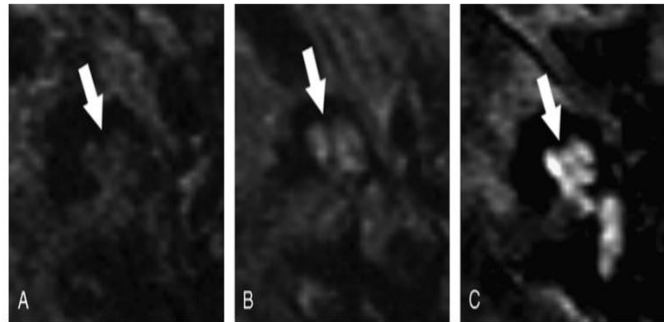


FIG. 1. A-C, Grading of cochlear enhancement. Axial gadolinium-enhanced T1-weighted MRI showing different grades of cochlear enhancement after contrast admission (arrows). A, Grade 0: no enhancement of the cochlea (Patient 14). B, Grade 1: moderate enhancement (Patient 9). C, Grade 2: severe cochlear enhancement (Patient 13).

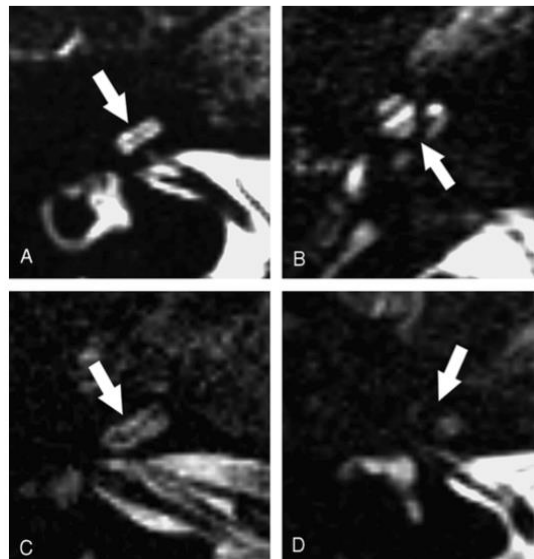
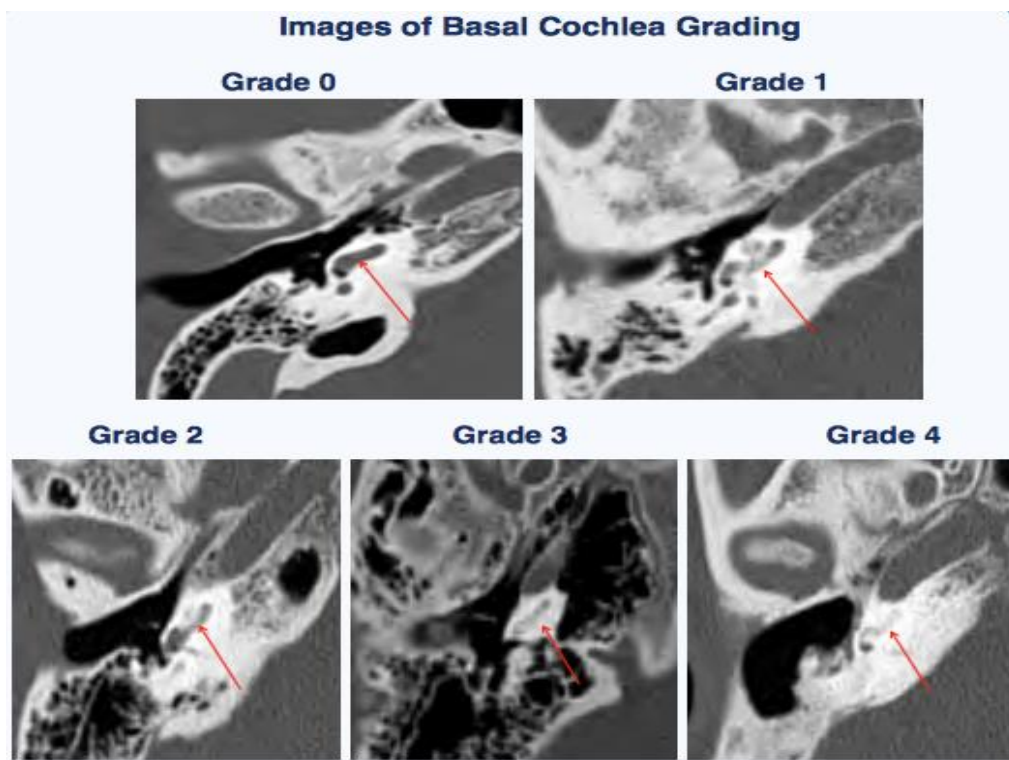


FIG. 2. A-D, Grading of cochlear patency. Axial 3D heavily T2-weighted MRI images showing different grades of intensity or fluid distribution within the cochlea (arrows). A, Grade 0: hyperintense or no loss of fluid (Patient 12). B, Grade 1: partial and irregular loss of fluid (Patient 2). C, Grade 1: moderate hypointensity (Patient 9). D, Grade 2: severe hypointensity or severe loss of fluid (>50%) (Patient 13).

- **What is the finding on CT in patients with post-meningitis SNHL?**
 - Labyrinthine ossification of basal turn (Most common).
 - Seen as early as 2nd month of infection
 - Sensitivity of 70% to detect cochlear obliteration (only detects ossification not fibrosis).
- **What is the grading of Basal turn LO on CT scan?**

Grading Score	Ossification Assessment
0	None
1	< 25%
2	25-50%
3	50-75%
4	>75%



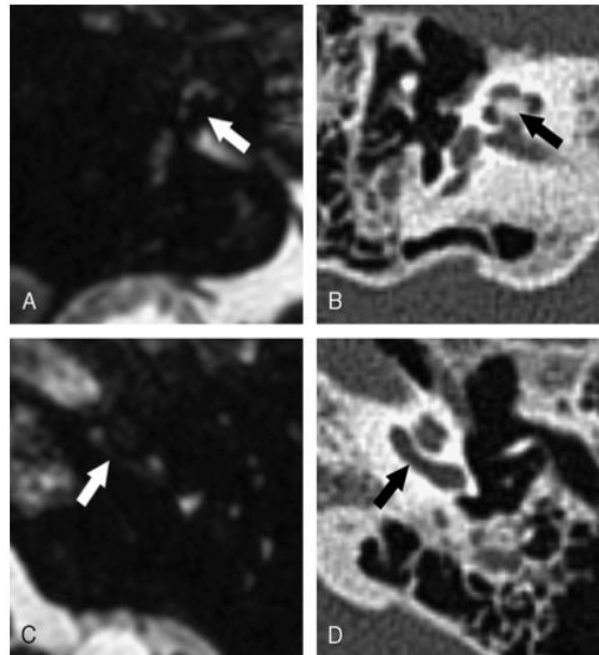


FIG. 7. A–D, MRI corresponds better with the surgical findings than CT. Axial 3D heavily T2-weighted MRI and HRCT through the modiolus of the right cochlea (A and B) and through the basal turn of the left cochlea (C and D). T2MRI shows a severe loss of both sides (white arrows, image A+C). Corresponding HRCT shows, however, extensive calcifications around the modiolus of the right cochlea (black arrow, image B), whereas the basal turn of the left cochlea displays no calcification (black arrow, image D). HRCT can be misleading as this cochlea was full of fibrosis during surgery and only a double array placement was possible (Patient 8).

852

M. C. VAN LOON ET AL.

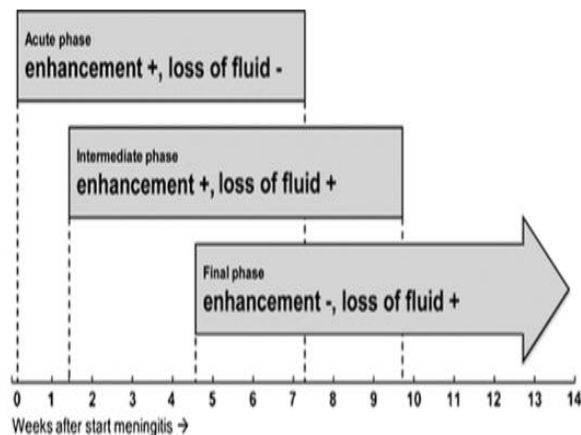
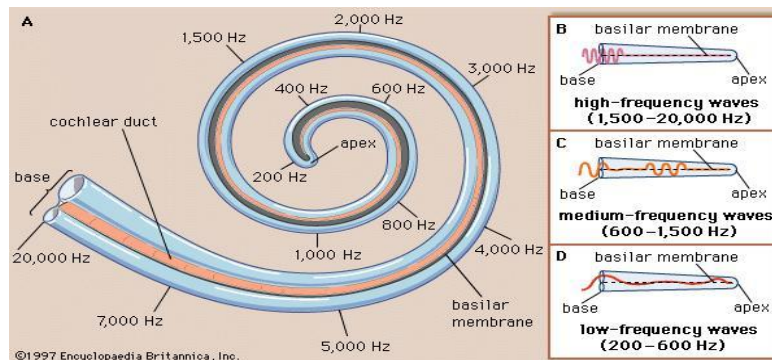
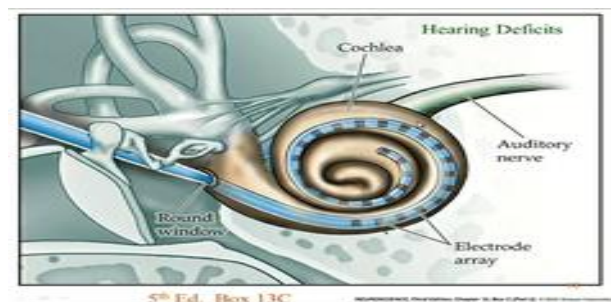


FIG. 6. Different phases of postmeningitic cochlear inflammation as seen on MRI. In the acute phase, MRI can show enhancement of the cochlea from the first day after meningitis [data validated on our series and Kopelovich et al. (9)]. Enhancement can last until 68 days (end intermediate phase). Loss of fluid is seen in our series as early as 10 days after start of the meningitis (start intermediate phase) and will most likely be permanent. Loss of fluid can be caused by fibrosis which, over a period, can partly or completely be transformed into ossification. Ossification can be seen on CT as early as 14 to 30 days after start of the meningitis [Aschendorf et al. (11) and Casselman et al. (19)].

- **What are the proven medications to prevent post-meningitis SNHL?**
 1. Ceftazidime:
 - First-line agent for the prevention of otogenic and meningogenic labyrinthitis.
 2. Steroid:
 - Reduce incidence of SNHL loss associated with bacterial meningitis.
 3. NSAIDs:
 - Reduce incidence of SNHL when administered early course of meningitis.
- **What are the options of hearing rehabilitation in unilateral post-meningitis SNHL?**
 1. Hearing aid
 2. Contra Lateral Routing of Signals (CROS)
 3. Bone Anchored Hearing Aid (BAHA)
 4. CI
- **What are the options of hearing rehabilitation in bilateral post-meningitis SNHL?**
 1. CI
- **What is the topographic representation of frequency in the cochlea?**



- **What is the topographic representation of frequency in the CI electrode?**



- **What is the appropriate timing of CI in post-meningitis SNHL?**
 - Exact timing remains controversial.
 - Preferred to be earlier to avoid the possibility of severe ossification.
- **What is the surgical approaches for CI in LO?**
 - Standard mastoidectomy.
 - Facial recess approach.
- **1. Round window Approach:**
 - Indicated for minimal ossification at the distal part of the basal turn.
 - Requires initial drilling until a lumen is identified.
 - Insertion of arrays from Scala Tympani of Basal turn toward the apex.
- **2. Scala Vestibuli Cochleostomy (Upper Retrograde Approach):**
 - Indicated for complete ossification of entire basal turn.
 - Performed by doing Scala Vestibuli (Apical) cochleostomy:
 - Anterior to the oval window and inferior to cochleariform process.
 - Insertion of arrays from Scala Vestibuli of Middle turn toward the base.
 - Risk of injury to the labyrinthine segment of the facial nerve.
- **3. Double/Split Array Approach:**
 - Indicated for complete ossification of entire cochlea.
 - Performed by doing 2 Cochleostomies:
 - **Scala Tympani Cochleostomy:**
 - Anterior and Inferior to RW
 - Allows insertion into Scala Tympani of Basal turn.
 - **Scala Vestibuli Cochleostomy:**
 - Anterior to OW and inferior to cochleariform process.
 - Allows insertion into Scala Vestibuli of Middle turn.



Auditory Neuropathy

- Auditory neuropathy/auditory dyssynchrony (AN/AD) is a condition that affects the neural processing of auditory stimuli.
- Patients with this disorder are able to respond to sounds appropriately, but their ability to decode speech and language is hindered.
- AN/AD has only recently been described.
- In the late 1970s, clinical investigators began to describe groups of patients with normal or slightly elevated audiogram pure tone thresholds accompanied with absent or severely abnormal auditory brainstem responses (ABRs).
- With the advent of the otoacoustic emissions (OAEs) in the mid 1980s, these groups of patients were found to have normal cochlear function.
- The finding of normal cochlear function accompanied with abnormal brainstem responses was defined in 1996 as auditory neuropathy (AN).
- Whether this represents a true auditory nerve neuropathy is debatable.
- Further investigations led to the conclusion that AN may truly represent a dyssynchronous auditory nerve rather than a neuropathy.
- This finding gave rise to the newer term of auditory dyssynchrony (AD).[1] For the purposes of this summary, AN and AD are considered synonymous (ie, AN/AD).

Pathophysiology

- The term auditory neuropathy/auditory dyssynchrony (AN/AD) describes a diagnosis that affects a small group of patients with hearing loss and speech intelligibility scores out of proportion with presumed hearing loss.
- Many authors have suggested that the abnormalities that cause AN/AD reside within the lower auditory system.
- Specifically, the spiral ganglion cells, auditory nerve, or the auditory brainstem nuclei have all been implicated.
- The combination of a dysfunctional auditory nerve with preservation of cochlear function can theoretically be caused at several different points along the lower auditory pathway.
- The following abnormalities have been proposed:
 - Injury to the synaptic junction between inner hair cells of the cochlea and dendrites of spiral ganglion neurons
 - Direct damage to the dendrites of the spiral ganglion neurons
 - Direct injury to the spiral ganglion neurons
 - Direct axonal damage to the auditory nerve that causes a cascade of damage to the lower auditory nuclei
- Several risk factors have been speculated to contribute to AN/AD. Those include the following:
 - Neonatal history of anoxia;
 - Neonatal history of hyperbilirubinemia;
 - Neonatal history of mechanical ventilation, hypoxia, or both;
 - Congenital brain abnormalities;
 - Low birth weight < 1.5kg
 - Extremely premature birth (< 28 wk);
 - Genetics or family history of AN/AD

- In addition, AN/AD has been reported in association with viral diseases, seizure disorders, and high fever.
- AN/AD can occur with or without accompanying neurologic disorders.
- Friedrich ataxia, Stevens-Johnson syndrome, Ehlers-Danlos syndrome, and Charcot Marie-Tooth syndrome are all disorders with peripheral neuropathies that have been associated with AN/AD.
- Although a complicated perinatal history is common among most patients with AN/AD, one third of patients have no predisposing factors that led to the development of AN/AD.

Epidemiology

- Some authors have suggested that the prevalence is 2-15% of children with known hearing loss.
- In a 2002 review of the prevalence, Sininger suggested that approximately 1 in 10 children with hearing loss and severely affected ABR test results have AN/AD.
- Two thirds of the children with auditory neuropathy/auditory dyssynchrony (AN/AD) demonstrate risk factors associated with perinatal hearing loss.

History

- AN/AD should be suspected in any child with slightly-to-severely abnormal hearing thresholds and severe speech and language delay out of proportion with the presumed hearing loss.
- Most children affected by AN/AD continue to display abnormal pure tone averages and ABR test results that requires a lifelong commitment by the child, family, speech pathologist, and audiologist.
- Further evaluation should include auditory brainstem response (ABR) and otoacoustic emission (OAE) testing to rule out the presence of AN/AD or other retrocochlear processes.

Diagnostic Considerations

- These include the following:
 - Ototoxicity
 - Auditory dyssynchrony
 - Central auditory processing deficits
 - Hyperbilirubinemia

Laboratory Studies

- No hematologic workup is necessary to diagnose auditory neuropathy/auditory dyssynchrony (AN/AD).
- History and audiologic testing establish the diagnosis

Imaging Studies

- Imaging studies are not necessary in the newborn period.
- Once the diagnosis is made correctly, conservative treatment can be initiated.
- If the parents choose surgical intervention, high-resolution computed tomography scanning of the temporal bones should be performed.
- This test helps the otologist or neurotologist determine the possibilities of inner ear malformations that might contribute to the disorder.
- In addition, the inner ear can be visualized and preparations for cochlear implantation can be made.
- Typically, magnetic resonance imaging (MRI) has no role in AN/AD.

Criteria for the diagnosis of AN/AD:

- All of the following must be present in newborns to diagnose AN/AD:
 - Absent or severely abnormal ABR test results at maximal stimulus (100 dBnHL).
 - Normal outer hair cell function as determined by OAEs or CMs
 - Absent or elevated stapedial reflex thresholds
- Suspect AN/AD in older children or adults with the following audiologic findings:
 - Pure tone thresholds are abnormal.
 - The entire range of abnormalities, from near-normal to profound, may be seen.
 - A more severe loss is usually displayed in the lower frequency thresholds.
 - Poor speech discrimination scores are out of proportion with the level of loss suspected based on the pure tone average.
- The audiogram findings may vary some, but the overall milieu usually remains unchanged.

Treatment:

- Treatment of patients with auditory neuropathy/auditory dyssynchrony (AN/AD) starts with the parents.
- Information should be made available to all parents of children with hearing loss.
- Once this is done and the condition is thoroughly understood, the proper supportive adjuvant therapies can begin which include:
 - Speech pathology, hearing aid placement, and use of other hearing devices.

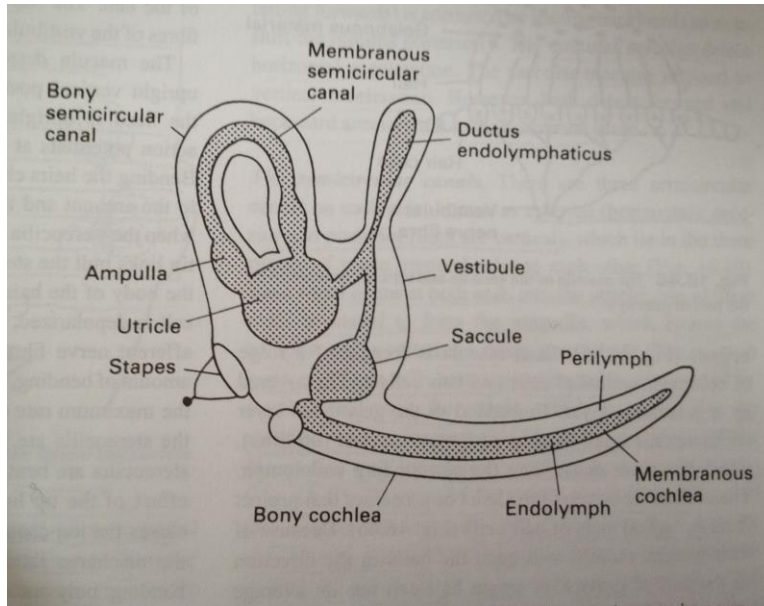
- The use of hearing aids can begin with children at around age 3 months.
- Children with AN/AD were once thought not to benefit from hearing aid amplification; however, recent studies demonstrate that 50% of children can benefit from placement of an amplification device.
- When children with AN/AD were tested with hearing aids, their speech discrimination scores improved and were more consistent with the degree of hearing loss expected via their pure tone audiometry scores.
- The use of hearing aids prior to cochlear implantation is currently recommended.
- Once the child is aged approximately 6 months, behavior audiometry thresholds should be obtained.

- Surgical Treatment:

- In 2001, the use of cochlear implantation was expanded to include children with AN.
- A literature review by Fernandes et al indicated that in children with AN spectrum disorder, cochlear implants lead to improvements in hearing skills similar to those associated with cochlear implants in children with sensorineural hearing loss.
- Study by Liu et al found that children with AN spectrum disorder who received cochlear implants prior to age 24 months tended to show better development of auditory and speech skills than did children who received the implants at a later age.
- If cochlear implantation fails, another option may exist in AN/AD, with brainstem implantation having been reported.

Physiology of Vestibular System:

- Peripheral Receptors:



- **1. Cristae:**

They are located in the ampullated ends of the three semicircular ducts.

These receptors respond to angular acceleration

All hair cells in each crista are oriented in the same direction

- **2. Maculae**

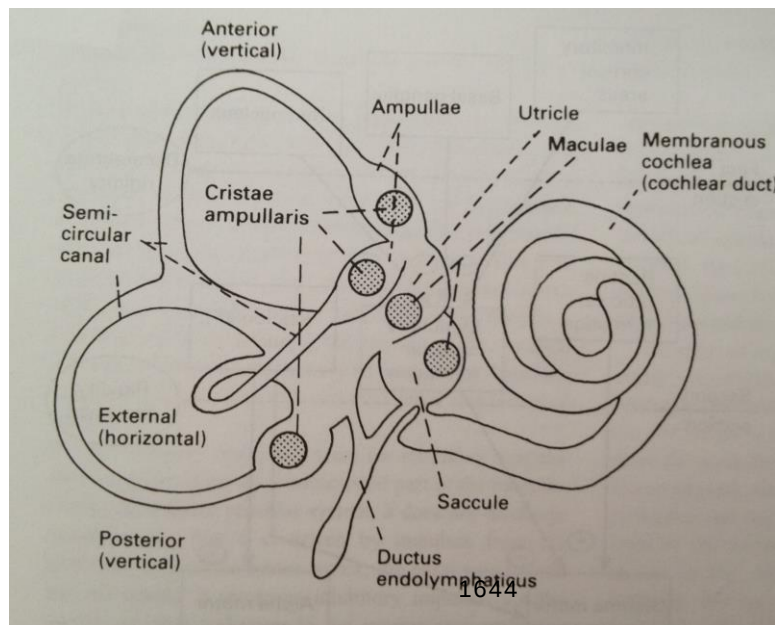
They are located in otolith organs (i.e. utricle and saccule).

Macula of the utricle lies in its floor in a horizontal plane.

Macula of saccule lies in its medial wall in a vertical plane.

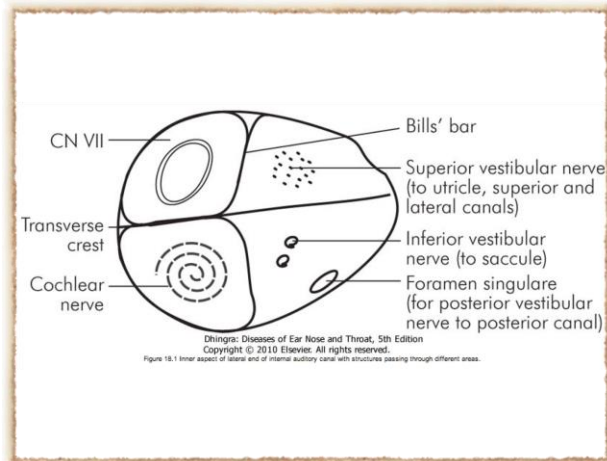
They sense position of head in response to gravity and linear acceleration

Hair cells in the macula oriented in all different directions



Vestibular Nerve

- Part of VIII cranial nerve
- Mialen sheet covering the part inside the IAC (out side the canal covered by dura, medial to porus acusticus)



- Vestibular or Scarpa's ganglion is situated in the lateral part of the internal acoustic meatus.
- It contains bipolar cells.
- The distal processes of bipolar cells innervate the sensory epithelium of the labyrinth while its central processes aggregate to form the vestibular nerve

Central Vestibular Connections:

- The fibers of vestibular nerve end in vestibular nuclei and some go to the cerebellum directly
- Vestibular nuclei are four in number, the superior, medial, lateral (deiters) and descending (inferior, spinal).
- Afferents to these nuclei come from:

(i) Peripheral vestibular receptors (semicircular canals, utricle and saccule)

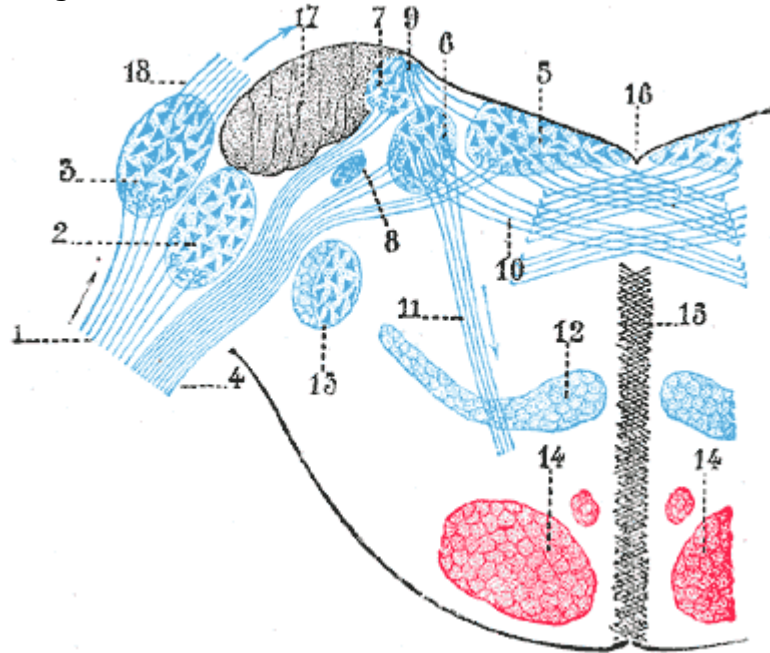
(ii) Cerebellum (some fibers pass directly to it)

(iii) Reticular formation (reticulospinal tract)

(iv) Spinal cord (vestibulospinal tract medial and lateral))

(v) Contralateral vestibular nuclei. (**Vestibular commissural** system (contralateral discharge rate inhibits ipsilateral firing; basis of "push-pull" connection – paired canals complement one another by canceling out inherent asymmetries))

(Thus, information received from the labyrinthine receptors is integrated with information from other somatosensory systems)



1. Cochlear nerve, with its two nuclei. 2. Accessory nucleus. 3. Tuberculum acusticum. 4. Vestibular nerve. 5. **Internal nucleus**. 6. **Nucleus of Deiters**. 7. **Nucleus of Bechterew**. 8. Inferior or descending root of acoustic. 9. Ascending cerebellar fibers. 10. Fibers going to raphe. 11. Fibers taking an oblique course. 12. Lemniscus. 13. Inferior sensory root of trigeminal. 14. Cerebrospinal fasciculus. 15. Raphe. 16. Fourth ventricle. 17. Inferior peduncle. Origin of striæ medullares. (Testut.)

- Efferents from vestibular nuclei go to:

(i) Nuclei of **CN III, IV, VI** via medial longitudinal bundle. It is the pathway for **vestibulo-ocular reflexes** and this explains the genesis of nystagmus.

(**Medial longitudinal fasciculus** and the **ascending tract of Deiters**)

(Correct eye movement & help fixing objects seen in the visual field when head is rotating)

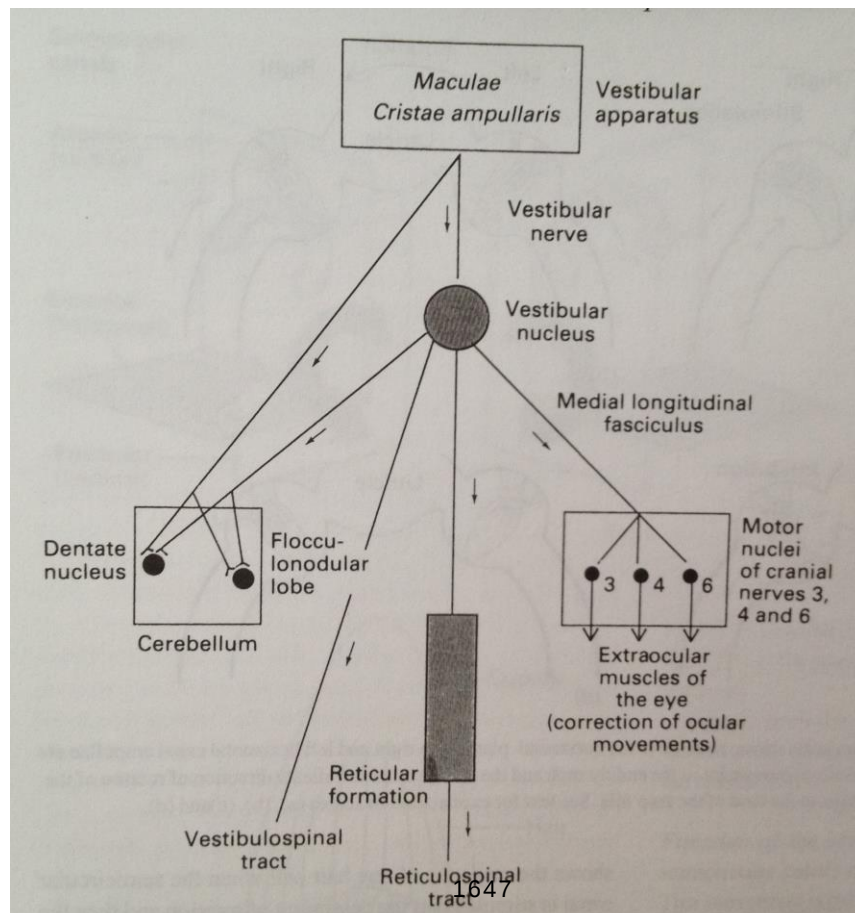
(ii) Motor part of spinal cord (**vestibulospinal fibres**). This coordinates the movements of head, neck and body in the maintenance of balance.

(iii) Cerebellum (**vestibulocerebellar fibres**). It helps to coordinate input information to maintain the body balance.

(iv) **Autonomic** nervous system. This explains nausea, vomiting, palpitation, sweating and pallor seen in vestibular disorders (linked to motion sickness & BP control)
(e.g. Meniere's disease).

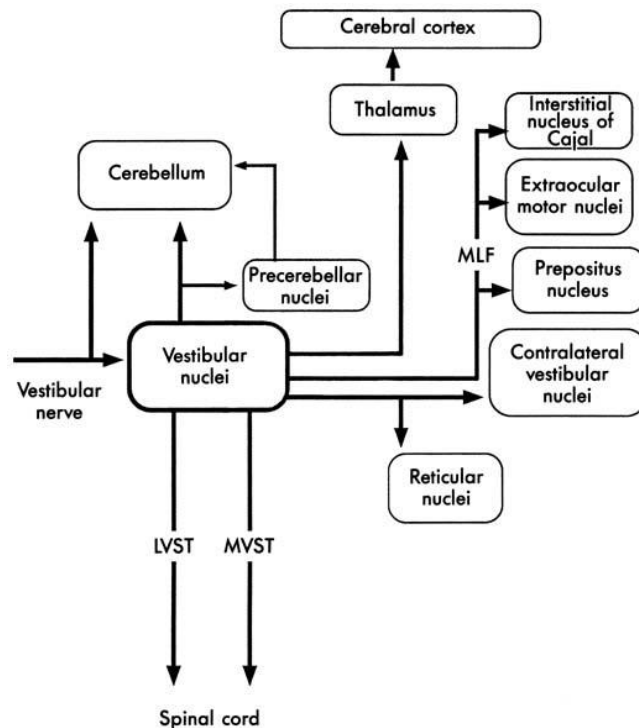
(v) Vestibular nuclei of the **opposite side**.

(vi) Cerebral cortex (**temporal lobe**). This is responsible for subjective awareness of motion.



Cummings

- The superior vestibular nucleus is involved in VOR pathways. Neurons in this nucleus fire in relation to eye movements as well as head movements and a prominent efferent projection of this nucleus is to the oculomotor nucleus via the medial longitudinal fasciculus. This tract is particularly important in the control of the VOR
- The lateral vestibular nucleus (of Deiters) can be subdivided on anatomic and functional grounds into two subnuclei: the dorsal lateral vestibular nucleus (or Deiters' nucleus) and the ventral lateral vestibular nucleus
- The dorsal lateral vestibular nucleus give rise to the lateral vestibulospinal tract, and the ventral lateral vestibular nucleus give rise to vestibuloocular pathways, the medial vestibulospinal tract, and vestibulothalamic pathways
- The lateral vestibulospinal tract terminates directly on the ventral horn cells. This tract is particularly important for vestibulospinal and vestibulocolic reflexes
- The "rostral" medial vestibular nucleus, like the superior nucleus, contains many neurons that project to the oculomotor nuclei and whose firing behavior is related to eye movements
- The medial vestibular nucleus also gives rise to the medial vestibulospinal tract, which descends in the medial longitudinal fasciculus to terminate on interneurons in the cervical spinal cord and ascends to terminate in the eye motor nuclei. This tract is particularly important for cervicovestibuloocular reflexes
- The inferior (or descending) vestibular nucleus contribute to the vestibulospinal pathways, but the major projections from this area are to the cerebellum and reticular formation



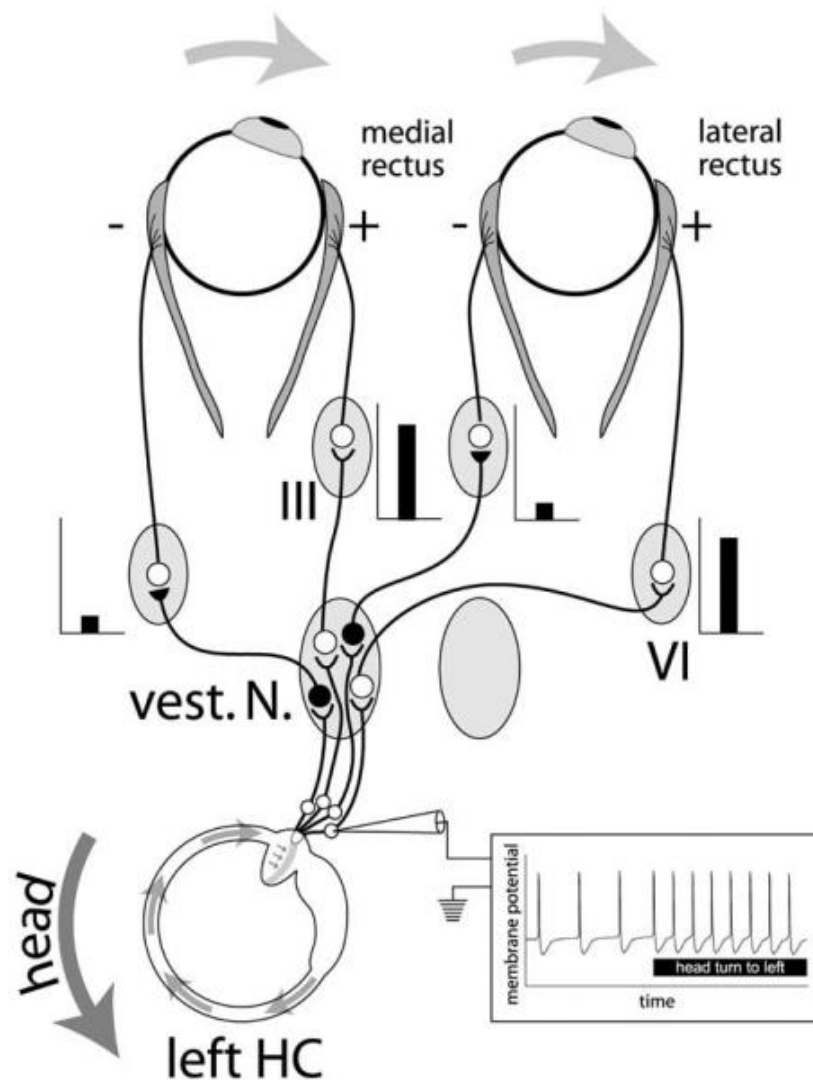


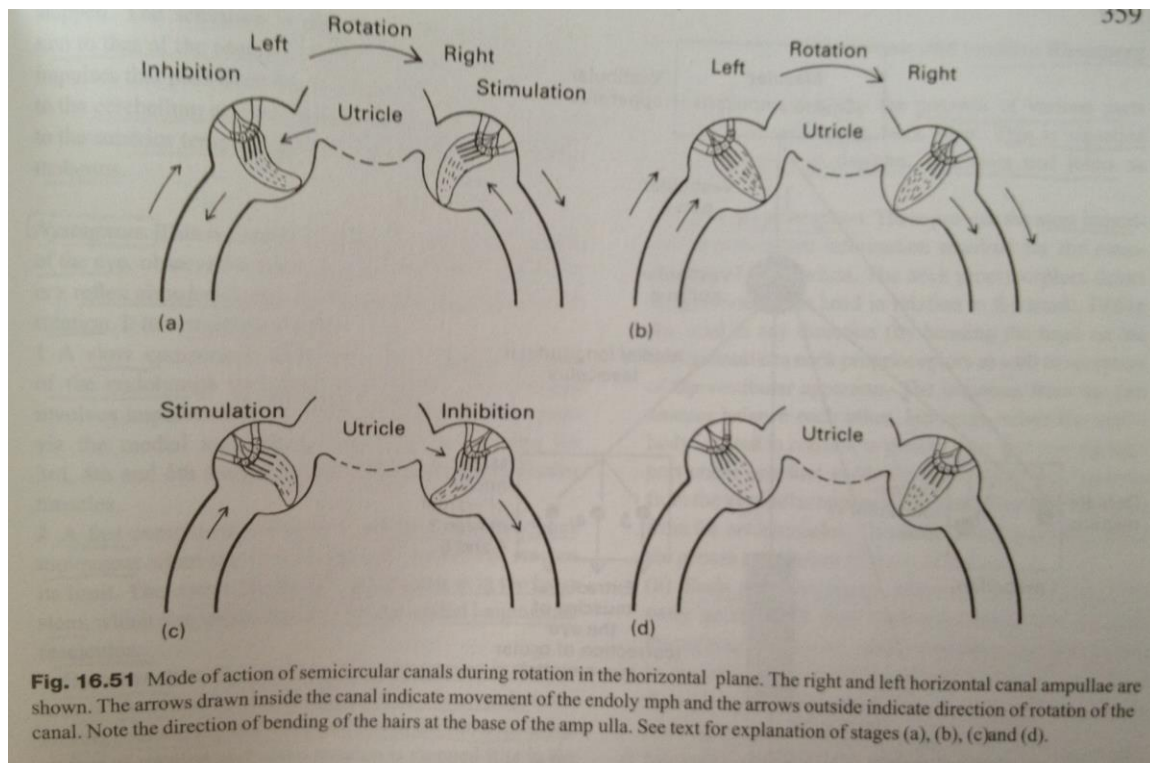
Figure 139-15 Neural connections in the direct pathway for the VOR from excitation of the left horizontal canal (left HC). As seen from above, a counterclockwise head rotation (head) produces relative endolymph flow in the left HC that is clockwise and toward the utricle. The cupular deflection excites the hair cells in the left HC ampulla, and the firing rate in the afferents increases (inset). Excitatory interneurons in the vestibular nuclei (vest. N.) connect to motor neurons for the medial rectus muscle in the ipsilateral IIIrd nucleus (III) and lateral rectus muscle in the contralateral VIth nucleus (VI). Firing rates for these motor neurons increase (bar graphs). The respective muscles contract and pull the eyes clockwise—opposite the head—during the slow phases of nystagmus. Inhibitory interneurons in the vestibular nuclei connect to motor neurons for the ipsilateral lateral rectus and contralateral medial rectus. Their firing rates decrease (bar graphs), and these antagonist muscles relax to augment the eye movement.

Vestibular system is conveniently divided into:

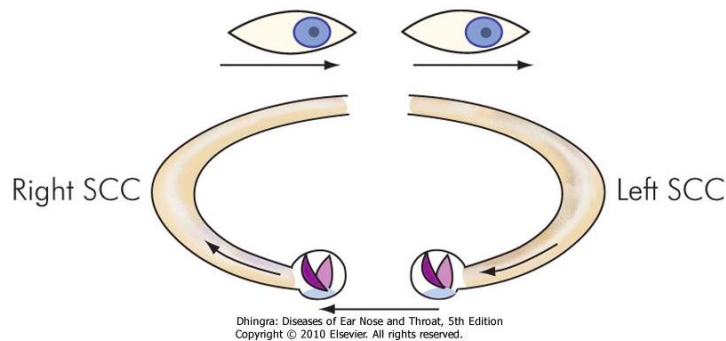
- (a) Peripheral**, which is made up of membranous labyrinth (semicircular ducts, utricle and saccule) and vestibular nerve.
- (b) Central**, which is made up of nuclei and fiber tracts in the central nervous system to integrate vestibular impulses with other systems to maintain body balance

Semicircular Canals (angular acceleration: head rotation in any direction)

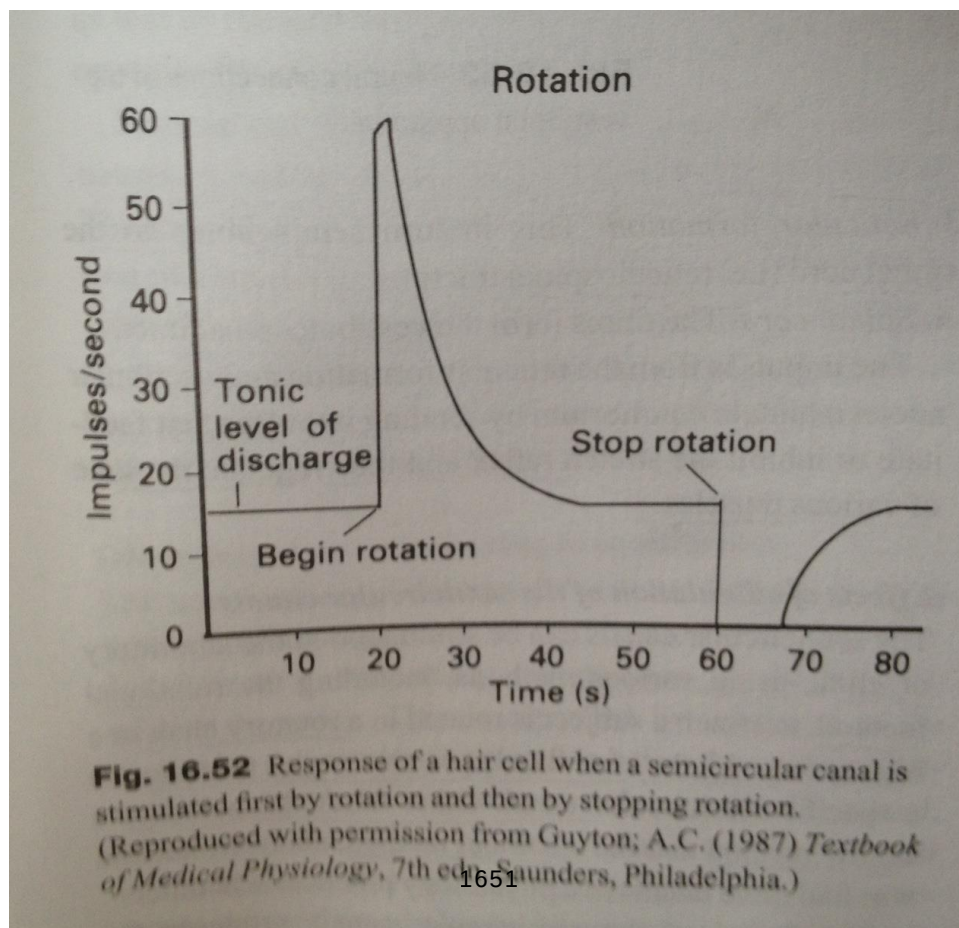
- During rest there is continuous impulses from both sides (200/s)
- At the beginning of rotation endolymph remains stationary due to inertia
- They respond to angular acceleration and deceleration.
- The one, which lies at right angles to the axis of rotation, is stimulated the most.
- Thus horizontal canal will respond maximum to rotation on the vertical axis and so on.
- Due to this arrangement of the three canals in three different planes, any change in position of head can be detected.
- Constant rates of rotation stimulate the SCC for a maximum 30s after which the discharge goes down to tonic level
- They detect only beginning of rotation, end of it and any change in rate of rotation



- Stimulation of semicircular canals produces nystagmus and the direction of nystagmus is determined by the plane of the canal being stimulated (Thus, nystagmus is horizontal from horizontal canal, rotatory from the superior canal, and vertical from the posterior canal)

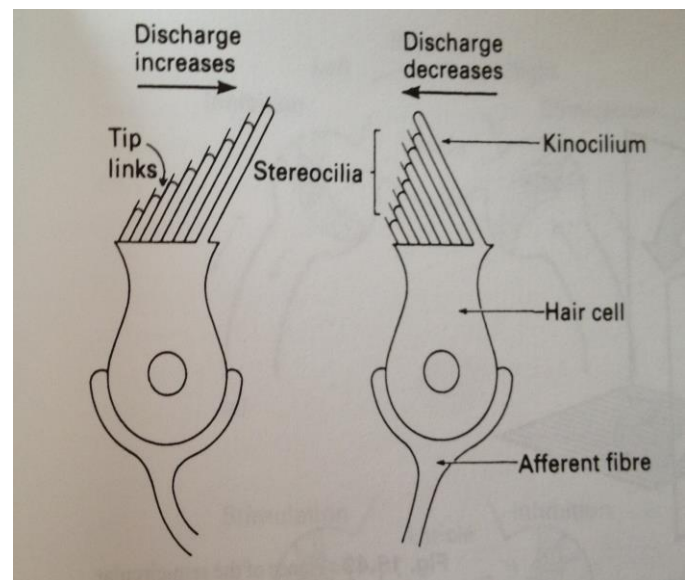
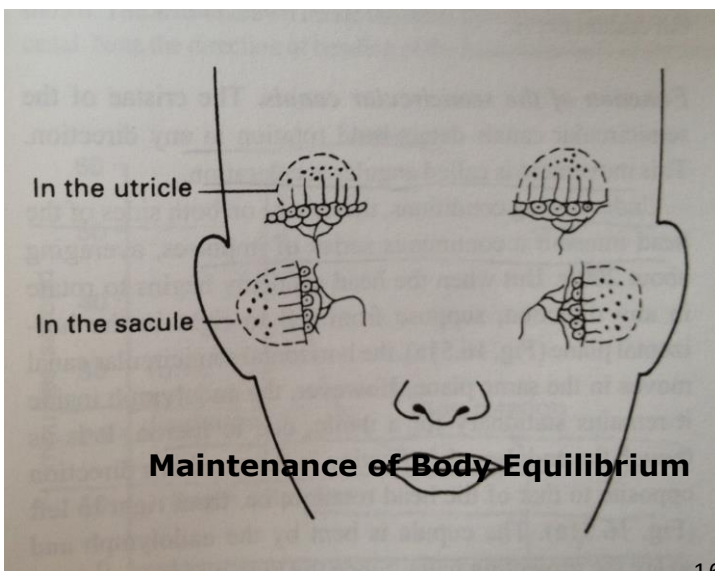


- The stimulus to semicircular canal is flow of endolymph, which displaces the cupula.
- The flow may be towards the cupula (ampullopetal) or away from it (ampullofugal), better called utriculopetal and utriculofugal.
- Ampullopetal flow is more effective than ampullofugal for the horizontal canal.
- The quick component of nystagmus is always opposite to the direction of flow of endolymph.
(Thus, if a person is rotated to the right for sometime and then abruptly stopped, the endolymph continues to move to the right due to inertia (i.e. ampullopetal for left canal), the nystagmus will be horizontal and directed to the left)



Utricle and Saccule (detect static tilt and linear acceleration)

- Utricle is stimulated by linear acceleration and deceleration or gravitational pull during the head tilts.
 - The sensory hair cells of the macula lie in different planes and are stimulated by displacement of otolithic membrane during the head tilts. (Horizontal, forward and backward)
 - The function of saccule is similar to that of utricle as the structure of maculae in the two organs is similar but experimentally (he saccule is also seen to respond to sound vibrations)
(Vertical, forward and backward)
 - The vestibular system thus registers changes in the head position, linear or angular acceleration and deceleration and gravitational effects.
 - This information is sent to the central nervous system where information from other systems-visual, auditory, somatosensory (muscles, joints, tendons, skin)-is also received.
 - All this information is integrated and used in the regulation of equilibrium and body posture
 - Cerebellum, which is also connected to vestibular end organs further coordinates muscle movements in their rate, range, force and duration and thus helps in the maintenance of balance
- **Tilt vs. linear acceleration:** otoliths alone can't tell the difference between linear accelar and tilt up with gravity pulling down; SCC input helps at high rotational frequency, and other sensory cues can help (vision); fighter pilots taking off in dark may perceive linear accelar as tilt up; they may compensate by nosing the aircraft down, potentially plunging into the water



- A useful clinical approach to understand the physiology of equilibrium is to imagine that the balance system (vestibular, visual and somatosensory) is a two-sided push and pull system.
- In static neutral position, each side contributes equal sensory information (push and pull system of one side is equal to that of the other side)
- If one side pulls more than the other, balance of the body is disturbed. During movement (turning or tilt, there is a temporary change in the push and pull system which is corrected by appropriate reflexes and motor outputs to the eyes (vestibulo-ocular reflex), neck (vestibulocervical reflex), and trunk and limbs (vestibulospinal reflex) to maintain new position of head and body, but if any component of push and pull system of one side is disturbed for a longer time due to disease, vertigo and ataxia will develop.

Nystagmus:

- A series of jerky oscillatory movements of the eye
- It is a reflex aimed at fixing objects in the visual field during rotation
- 2 components:
 1. Slow (in the direction of endolymph) (vestibulo-ocular reflex)
 2. Fast (compensatory flick-back movement) (from brainstem)

(The direction of eye movement in nystagmus is identified by the direction of the fast component)

- During rotation nystagmus in the same direction of the **rotation**
- When the rotation stop it is in the opposite direction of the rotation

Vertigo and Dizziness

- False sensation of spinning
- Disorientation in space causes vertigo or dizziness and can arise from disorders of any of the three systems, vestibular, visual or somatosensory.
- Normally, the impulses reaching the brain from the three systems are equal and opposite.
- If any component on one side is inhibited or stimulated, the information reaching the cortex is mismatched, resulting in disorientation and vertigo.
- The vestibular inhibition on one side (e.g. acute vestibular failure, labyrinthectomy, Meniere's disease, VIIIth nerve section) causes vertigo.
- Similarly, stimulation of labyrinth by thermal or rotational stimulus causes vertigo.
- Dizziness can similarly result from the ocular causes, e.g. high errors of refraction or acute extraocular muscle paralysis with diplopia.

Motion Sickness

- Nausea, vomiting, pallor and sweating during sea, air, bus or car travel in certain susceptible individuals characterizes it.
- It can be induced by both real and apparent motion and is thought to arise from the mismatch of information reaching the vestibular nuclei and cerebellum from the visual, labyrinthine and somatosensory systems.
- It can be controlled by the usual labyrinthine sedatives.
- The passenger may feel much better if he/she sits in front seat or keeps looking out watching the moving visual field

Other receptors contributing equilibrium: (visual, vestibular, somatosensory)

1. Position in body in relation to space (visual and auditory receptors)
2. Position of various parts of the body
 - a. Neck proprioception
 - b. Body proprioception
3. Information concerning the direction of support (Cutaneous pressure receptors) (eg. Soles of feet)

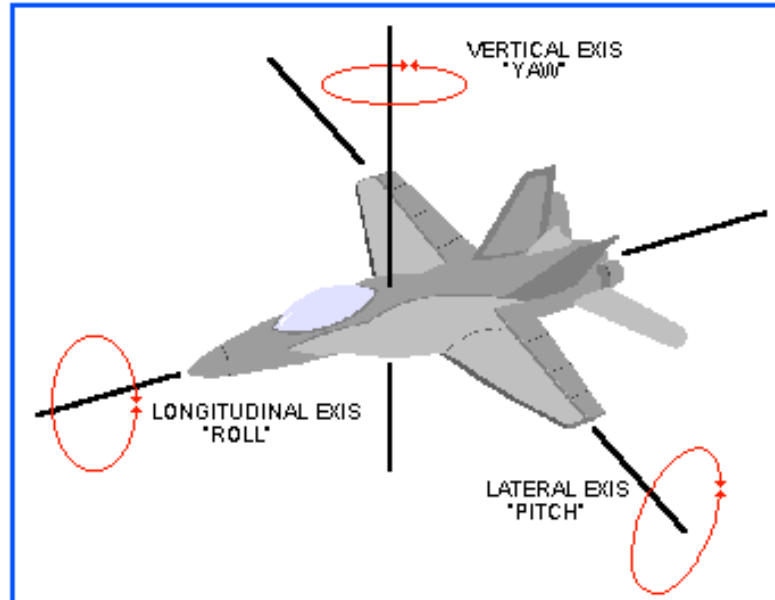
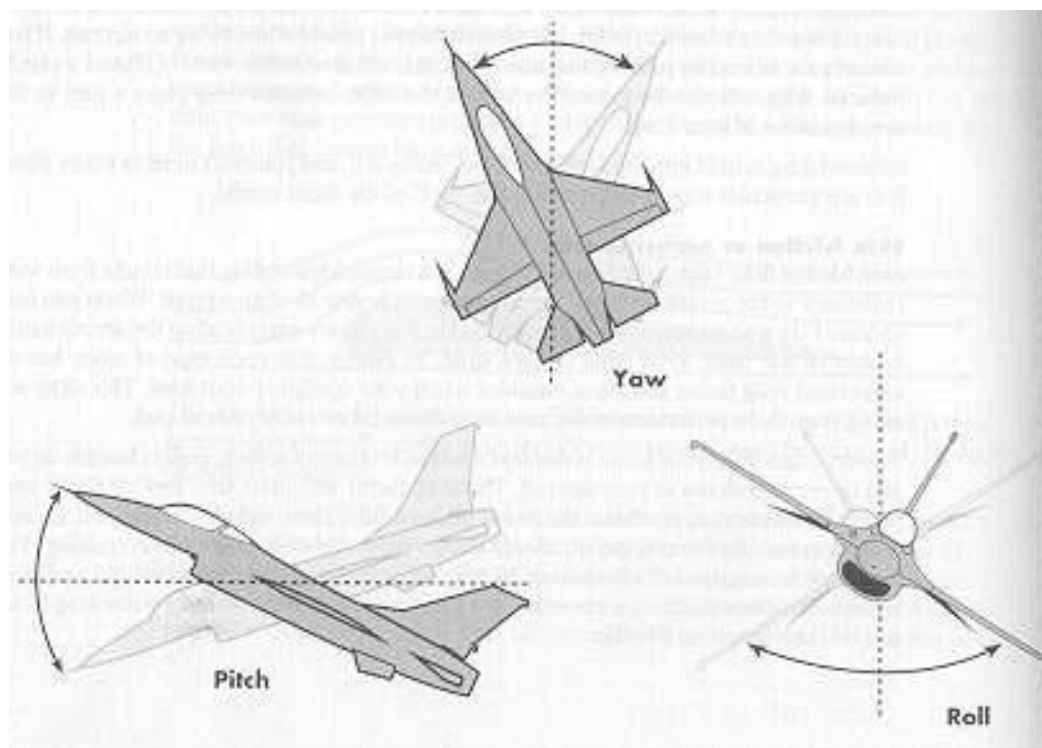


Figure 5-1 The axis of rotation.



Evaluation of a Dizzy Patient:

- Three primary sensory inputs responsible for balance:
 - Vestibular pathway
 - Visual pathway
 - Somatosensory (proprioceptive) pathway
- Vestibular labyrinth is divided into:
 - **Semicircular canals (SCCs):**
 - Detect **angular** head movement.
 - Produce a compensatory eye movement via vestibuloocular reflex (VOR) for stabilization of visual images on the retina and maintenance of acuity during head movement.
 - **Otolith organs (utricle and saccule):**
 - Detect **linear** acceleration (including gravity).
 - Produce compensatory postural changes in response to transient linear movements and changes via gravity through vestibulospinal reflex (VSR) pathway.
- Maintenance of balance requires integration of:
 - Appropriate detection of environmental sensory inputs
 - Accurate CNS integration of all sensory input
 - Accurate performance of the correct muscle response for maintenance of postural control and gaze stability

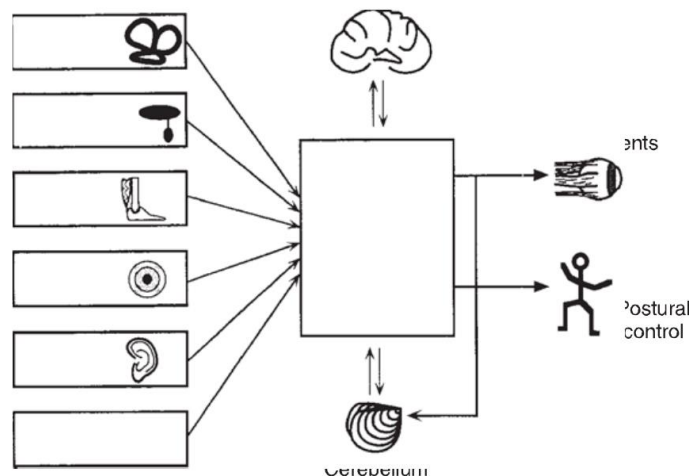


Figure 165.1 An integrated approach to the diagnosis of dizziness and imbalance. Maintaining balance requires a patient to sense the environment (via sensory afferent input), to integrate the environmental information via central processing, and to calculate the correct motor response (delivered via motor efferent output). Accurately locating the abnormality within this integrated system is crucial when a patient presents with dizziness, imbalance, or vertigo. (From Hughes GB, Pensak ML, eds. *Clinical otology*, 2nd ed. New York: Thieme Medical Publishers, 1997:44, with permission.)

- **Dizziness** is a term used to describe any of a variety of sensations that produce spatial disorientation.
- In a majority of dizzy patients, a specific diagnosis may be made by an adequate history and physical examination.

HISTORY:

- Describe Dizziness:

○ **Vertigo:**

- Illusion of rotational or tilting movement (spinning, whirling, turning) of the patient or the surroundings.
- In most instances, patients will sense that environment is in motion around them (objective vertigo).
- Minority of patients feel that they are in motion relative to a stationary world (subjective vertigo).
- Indicates a problem in the peripheral vestibular system (labyrinth or CN-8).

○ **Disequilibrium:**

- Difficulty maneuvering within the environment without experiencing an illusion of motion.
- Sensation of instability of body positions, imbalanced or fear of falling.
- Symptoms worse while standing or ambulating.
- Indicate CNS, peripheral neuropathic or musculoskeletal disorders.

○ **Light-headedness:**

- Sense of impending faint or presyncope.
- Indicate migraine, vascular, metabolic, drug-induced, endocrine or psychogenic causes.

○ **Oscillopsia:**

- Inability to focus on objects with motion, such as reading a sign while walking,
- Seen with loss of vestibulo-ocular reflex (VOR) in bilateral vestibular dysfunction.

- Duration of each vertigo attack:

○ **Lasts for seconds:**

- BPPV

○ **Lasts for minutes to hours:**

- Meniere disease
- Vestibular migraine.

○ **Lasts for days to weeks:**

- Vestibular Neuritis
- Labyrinthitis

○ **Constant vertigo:**

- Central vertigo.

- Contributing factors:
 - **Trauma:**
 - Direct mechanical trauma:
 - Head trauma or ear surgery.
 - Lead to:
 - Labyrinthine concussion
 - Temporal bone fracture
 - Perilymphatic fistula
 - BPPV
 - Barotrauma:
 - Blast injuries, scuba diving, hyperbaric oxygen treatments, straining, changes in altitude (airplane flights or driving in the mountains).
 - **Infection:**
 - Recent URTI:
 - Vestibular neuritis
 - CSOM:
 - Labyrinthitis
 - Perilymphatic fistula
 - Others:
 - Herpes zoster oticus (Ramsay Hunt syndrome)
 - Otosyphilis
 - TB
 - HIV
 - Lyme disease
 - **Medical condition:**
 - Cardiac arrhythmias
 - Atherosclerosis
 - Anemia
 - Hypothyroid
 - Diabetes
 - Neurological disorders
 - Migraine
 - Wegener granulomatosis
 - Systemic lupus erythematosus
 - **Medications:**
 - Topical or systemic ototoxic medications:
 - Aminoglycosides, Chemotherapy, Loop diuretics.
 - Medications affect blood pressure:
 - Diuretics, B-blockers, Calcium channel blockers.
 - Medications affect CNS:
 - Sedatives, Neuroleptics, Antidepressants.

- Aggravating factors:
 - **Head movement:**
 - Peripheral vestibular disorders are aggravated by head movements and gravity.
 - Patients are advised to keep their head as still as possible to avoid sudden movements.
 - Examples:
 - Posterior canal BPPV:
 - Vertigo with rolling over in bed or tilting the head backward and toward the affected ear.
 - Horizontal canal BPPV:
 - Vertigo with lying supine and turning the head to the side.
 - **Body movement against gravity:**
 - Rising from a bed or chair.
 - Indicates orthostatic hypotension.
 - **Life style induced:**
 - Vestibular migraine:
 - Food-induced dizziness (caffeine, alcohol, cheese)
 - Stress-induced dizziness
 - Motion simulation
 - Meniere's disease:
 - Food induced dizziness (Salt).
 - Superior semicircular canal dehiscence:
 - Sound-induced vertigo (Tullio phenomenon)
 - Pressure-induced vertigo (Hennebert sign)
 - Valsalva maneuvers
- Associated symptoms:
 - **Otological symptoms:**
 - Meniere's disease:
 - Hearing loss, tinnitus and aural fullness.
 - SCDS:
 - Hearing loss and autophony.
 - **Migraine symptoms:**
 - Photophobia, phonophobia and heightened sense of smell.
 - Indicates vestibular migraine.
 - **Neurological symptoms:**
 - Dysphagia, dysphasia, limb weakness, ataxia and LOC.
 - Indicates CNS involvement.
 - **Psychogenic symptoms:**
 - Anxiety, panic attacks and agoraphobia.

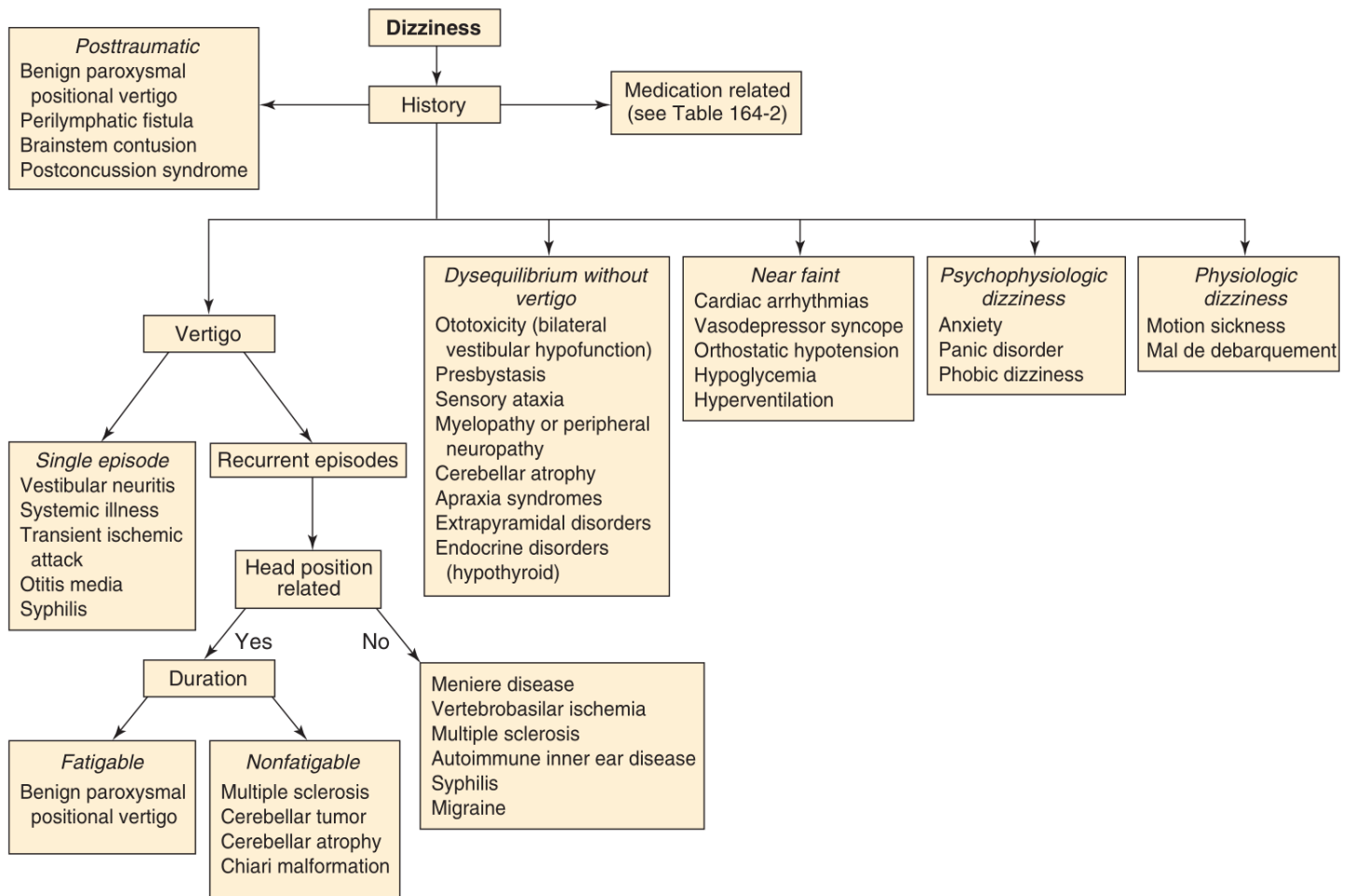


FIGURE 164-4. Algorithm for the differential diagnosis of dizziness based on information from the patient's history. (Modified from Baloh RW, Fife TD, Furman JM, Zee DS: The approach to the patient with dizziness. In Mancall EL, editor: *Continuum: lifelong learning in neurology*, Cleveland, 1996, Advanstar Communication, pp 25-36.)

**TABLE
165.1****THE HISTORY OF PRESENT ILLNESS IN A PATIENT WITH DIZZINESS****Components of the Interview****Key Historical Features**

What is the character of the dizzy sensation?	Vertigo, disequilibrium, or light-headedness
What is the dizziness pattern?	Continuous or episodic
What is the time course of dizzy episodes?	Seconds, minutes, hours, days, or longer
Was there an event associated with dizziness onset?	Head/inner ear trauma, barotrauma, upper respiratory infection, ear infection, systemic illness or infection, ototoxic medication
What associated symptoms accompany the dizzy episodes?	Hearing loss, tinnitus (continuous vs. pulsatile), aural fullness, conductive hyperacusis, diplacusis, dysacusis, autophony, oscillopsia, otorrhea, otalgia, headache, facial or limb weakness, dysphagia, dysphasia, visual changes, photophobia, phonophobia, loss of consciousness, seizure
What exacerbates the dizziness?	Rapid head movement, particular head positions, increased pressure (Hennebert sign), sounds (Tullio phenomenon), hyperventilation, strong environmental stimuli (bright lights, odors, etc.), food triggers
What medications (past and present) could be involved? ^a	Antibiotics, antineoplastic, analgesics, antihypertensives, neuroleptics, antidepressants, sedatives
What is the patient's past medical history?	Migraine, endocrine (e.g., diabetes), rheumatologic, cancer, cardiovascular, systemic infections
What is the patient's past surgical history?	Focusing on otologic or brain surgery
What is the family history?	Migraine, endocrine (e.g., diabetes), rheumatologic, cancer, cardiovascular, systemic infections, genetic mutations

**TABLE
165.2****MEDICATIONS THAT MAY CAUSE DIZZINESS**

Drug Class	Mechanism	Symptoms
Ototoxic		
Aminoglycosides <i>Gentamicin, neomycin, tobramycin, amikacin, kanamycin</i>	Damages vestibular type I sensory cells, outer hair cells in the organ of Corti, and cochlear and vestibular neurons Loop diuretics and vancomycin can potentiate ototoxicity	Oscillopsia from symmetric vestibular hypofunction Vertigo from asymmetric vestibular loss, sensorineural hearing loss, and tinnitus
Antineoplastic agents <i>Cisplatin</i>	Damages vestibular hair cells, outer hair cells in the organ of Corti, stria vascularis, and spiral ligament	Same symptoms as aminoglycoside damage
Analgesic agents NSAIDs <i>(aspirin, naproxen, indomethacin, and ibuprofen)</i> <i>Acetaminophen/hydrocodone</i>	Reversible outer hair cell function changes and reduced blood flow via vasoconstriction Permanent hair cell damage	Reversible sensorineural hearing loss, tinnitus, and rarely vertigo Rapidly progressive, permanent sensorineural hearing loss, tinnitus, and rarely vertigo
Systemic Blood Flow		
Antihypertensives <i>Diuretics, β-blockers, vasodilators, CCB α-adrenergic blocker</i>	Systemic vasodilation leading to decreased peripheral vascular resistance	Light-headedness, syncope, visual changes, and fatigue
Central Nervous System		
Neuroleptics Antidepressants <i>Tricyclics, MAOIs, SSRIs</i> sedatives	Target central dopamine, serotonin, GABA, and acetylcholine neurotransmitter pathways	Light-headedness, vertigo, and ataxia

CCB, calcium channel blocker; GABA, γ -aminobutyric acid; MAOIs, monoamine oxidase inhibitor; NSAIDs, nonsteroidal anti-inflammatory drugs; SSRI, selective serotonin reuptake inhibitor.

TABLE 6–5. General Characteristics of Peripheral and Central Causes of Vertigo

Characteristic	Peripheral	Central
Intensity	severe	mild
Fatigability	fatigues, adaptation	does not fatigue
Associated Symptoms	nausea, hearing loss, sweating	weakness, numbness, falls more likely
Eye Closure	symptoms worse with eyes closed	symptoms better with eyes closed
Nystagmus	horizontal, may be unilateral, rotary	vertical, bilateral
Ocular Fixation	suppresses nystagmus (may not suppress during acute phase)	no effect or enhances nystagmus

TABLE 166-1. Differentiation of Central and Peripheral Vertigo

Feature	Central Origin	Peripheral Origin
Imbalance	Severe	Mild to moderate
Neurologic symptoms	Frequent	Rare
Nystagmus	Changes direction on lateral gaze No change with fixation	Unidirectional Decreases with fixation
Hearing loss	Rare	Frequent
Nausea	Variable	Severe
Recovery	Slow	Rapid

TABLE 167.1**DISTINGUISHING CENTRAL FROM PERIPHERAL CAUSES OF DIZZINESS**

	Nausea and Vomiting	Ability to Walk	Hearing Loss	Focal Neurologic Signs and Symptoms	Associated Autonomic Symptoms (Sweating, Pallor)
Peripheral	Severe	Unsteadiness but able to walk	Common	Rare	Pronounced
Central	Moderate	Nearly incapable of walking	Rare except AICA infarct	Common	Less pronounced

**TABLE
166.1**

**BASIC PRESENTATIONS
OF PERIPHERAL VESTIBULAR
DYSFUNCTION**

1. Episodic disruption of unilateral vestibular function
 - Lasting minutes-hours*
 - Ménière's disease
 - Perilymph fistula
 - Lasting >24 h*
 - Vestibular neuritis
 - Labyrinthitis
 - IMIED
 - Lasting variable periods of time*
 - Migrainous vertigo
 2. Brief/episodic excitation of unilateral vestibular function
 - BPPV
 - SCD syndrome
 - Perilymph fistula
 3. Chronically inadequate vestibular function
 - Unilateral*
 - Unilateral vestibular hypofunction following vestibular neuritis, trauma, etc.
 - Acoustic neuroma
 - Bilateral*
 - Aminoglycoside toxicity
 - Chemotherapy
 - Familial
-

PHYSICAL EXAM:

- **No clinical test measures directly the function of the peripheral vestibular system.**
- H&N and General Physical Exam:
 - o General condition and vital signs.
 - o Neurological and cranial nerve examination.
 - o Examination for orthostatic hypotension:
 - Reduction of **systolic BP > 20** or **diastolic BP > 10** within 3 minutes of standing.
- Otoscopy:
 - o **Inspection of TM:**
 - AOM, OME, CSOM or masses.
 - o **Pneumatotomography:**
 - Fistula test:
 - Induce nystagmus by applying intermittent pressure on tragus or by using Pneumatic Otoscopy.
 - Normally, the test is negative because the pressure changes in EAC cannot be transmitted to the labyrinth.
 - **Positive Fistula Test:**
 - o Nystagmus to same side (irritative).
 - o Indicates:
 - Perilymph fistula (50%).
 - SCDS
 - **False Positive Fistula Test (Hennebert's sign):**
 - o Nystagmus to same side (irritative) in the absence of Perilymph fistula.
 - o Indicates:
 - Meniere's disease (25%) due to fibrous bands connecting utricular macula to the stapes footplate.
 - Congenital syphilis due to hypermobility of stapes footplate.
- Tuning fork examination:
 - o **512-Hz fork:**
 - Auditory stimulus for Weber and Rinne testing.
 - Tullio Phenomenon:
 - Noise-induced vertigo and nystagmus in perilymphatic fistula or SCDS.
 - o **256-Hz fork:**
 - Malleolar test:
 - Patient can perceive sound in affected ear while the fork is placed on the ankle due to "third-window effect" in perilymphatic fistula or SCDS.

- Bed side vestibular tests:

1. Nystagmus:

1. Spontaneous Nystagmus
2. Gaze-Evoked Nystagmus
3. Saccades
4. Smooth Pursuit

2. Head Thrust (Impulse) Test

3. Head Shake (Heave) Test

4. Dix-Hallpike Maneuver

5. Limb Coordination Tests

6. Fukuda Stepping Test

7. Tandem Romberg Test

8. Gait Test

9. Hyperventilation Test

- **Nystagmus:**

- Rapid involuntary oscillatory movement of the eyes.
- Results from imbalance between the bilateral vestibular system.
- Direction of nystagmus:
 - Slow phase is the pathologic component.
 - Determined by the corrective fast phase.
- Types of nystagmus:
 - **Irritative Nystagmus:**
 - Nystagmus beating toward the affected ear.
 - Due to increased excitability of vestibular system at the affected ear.
 - Examples:
 - Perilymphatic fistula
 - Serous labyrinthitis
 - Post-stapes surgery
 - **Paralytic Nystagmus:**
 - Nystagmus beating away from affected ear.
 - Due to decreased excitability of vestibular system at the affected ear.
 - Examples:
 - Ménière's disease
 - Suppurative labyrinthitis
 - Dead ear
- Alexander's Law:
 - Peripheral nystagmus will increase when gaze is in the direction of fast phase, and will decreased when in the direction of slow phase.
- Ewald's Law:
 - Peripheral nystagmus is in the plane of SCC being stimulated and its direction (fast phase) is opposite to the direction of endolymphatic flow.
 - Stimulating/excitatory response are greater than inhibitory response.

- **Fixation suppression:**
 - Peripheral nystagmus is suppressed with visual fixation.
 - Absence of suppression suggests central lesion.
 - Suppression can be prevented by:
 - Frenzel glasses.
 - Darkness.

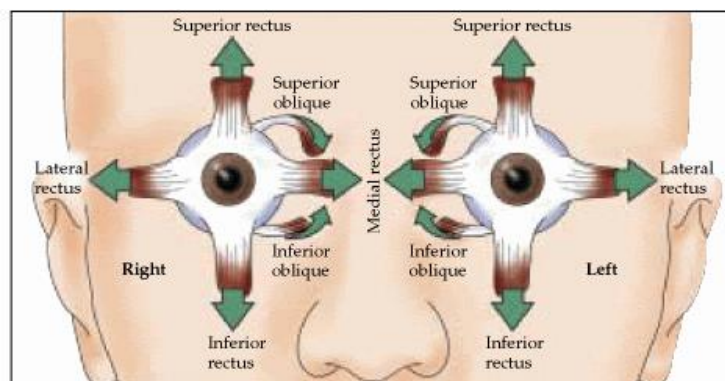
Table 6-1. Degree of nystagmus*

1st degree	It is weak nystagmus and is present when patient looks in the direction of fast component.
2nd degree	It is stronger than the 1st degree nystagmus and is present when patient looks straight ahead.
3rd degree	It is stronger than 2nd degree nystagmus and is present even when patient looks in the direction of the slow component.

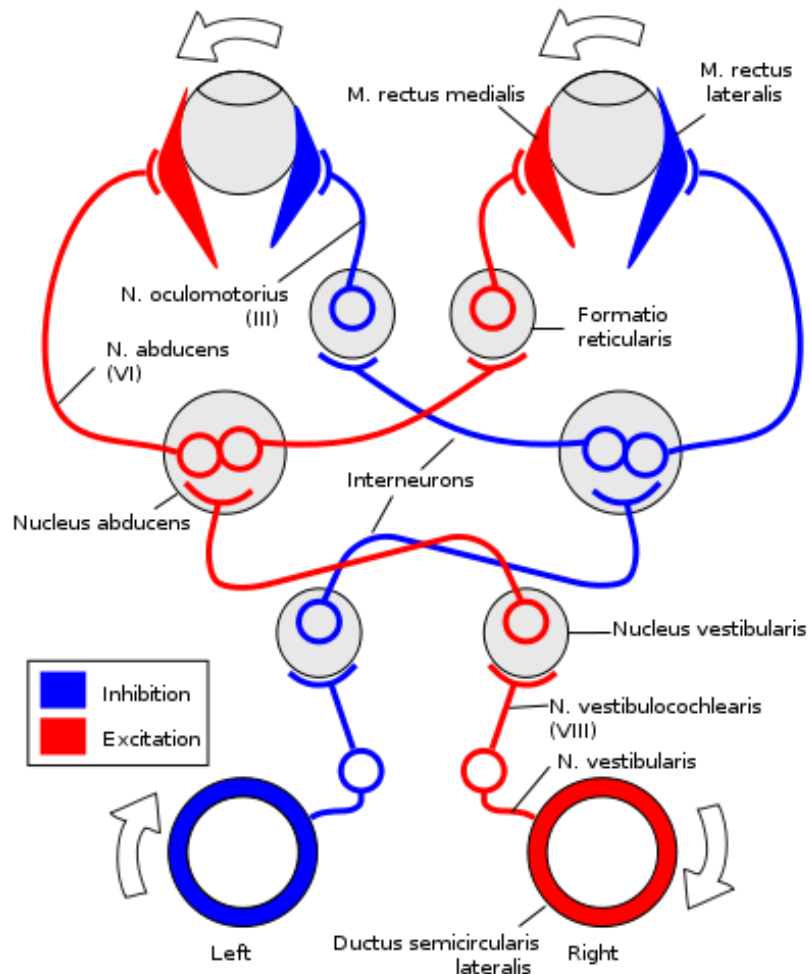
*These degrees are according to Alexander's law and may not hold true in case of nystagmus of central origin.

**TABLE
167.2****DISTINGUISHING CHARACTERISTICS OF PERIPHERAL VERSUS CENTRAL NYSTAGMUS**

	Type of Nystagmus	Effect of Fixation	Effect of Eye Position	Positional Nystagmus	Other Abnormalities on Testing
Peripheral: Labyrinth or vestibular nerve	Horizontal and torsional	Inhibited by fixation	Follows Alexander law	Latency, fatigues, torsional upbeat nystagmus, <1 min	May be associated with asymmetry of caloric response or of the VOR
Central: Brainstem or cerebellum or interconnections	Can be pure vertical, horizontal, or pure torsional	Unaffected by fixation	May be unidirectional, but if direction-changing is always central	No latency, does not fatigue, oftentimes downbeat or reversal of direction	Often associated with abnormalities of smooth pursuit, optokinetic nystagmus OKN, or saccades



- Vestibulo-Ocular Reflex (VOR):
 - Head rotation in a plane results in equal but opposite eye rotation to allow retina to maintain fixation on an object.
 - Head motion 10° to left produces eye movement 10° to right.
- Vestibulospinal Reflex:
 - Otolith organs modulate the anti-gravitational muscles in a similar way as VOR modulates the eye to allow for postural control and position (simultaneous contraction of extensor muscles with contralateral flexor muscles to maintain posture).
 - Motion of muscles and joints may also modify the vestibulospinal reflex via receptors found in intervertebral joint receptors (upper cervical).



- **Spontaneous Nystagmus:**
- Nystagmus present without visual or vestibular stimulation.
- Important sign in the evaluation of vestibular system.
- **Method:**
 - Patient is seated in upright position and fixates on a stationary target with best-corrected vision (with glasses if applicable).
 - Eyes are observed for nystagmus with Frenzel glasses.
 - If nystagmus was observed, the following characteristics should be noted:
 - **Nystagmus waveform:**
 - Jerk nystagmus (slow and fast phase).
 - Pendular nystagmus (equal movement).
 - **Type:**
 - Horizontal
 - Vertical
 - Torsional
 - **Direction:**
 - Right
 - Left
 - Up-beating
 - Down-beating
 - **Effect of fixation:**
 - Suppressed
 - No affected
 - **Effect of gaze (degree of nystagmus):**
 - Increased
 - Decreased
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Jerk nystagmus.
 - Fixed direction.
 - Horizontal or torsional.
 - Enhanced with:
 - Frenzel glasses.
 - Gazing in the direction of fast phase.
 - Suppressed with:
 - Visual fixation.
 - Gazing in the direction of slow phase.
 - **CNS pathology:**
 - Jerk or pendular nystagmus.
 - Direction-changing.
 - Pure vertical or torsional.
 - No suppressed visual fixation.
 - Not affected by gazing in different directions.

- **Gaze-Evoked Nystagmus:**
- Abnormal persistent nystagmus that present with eye gazing between 0-30° from midline.
- End-point nystagmus is a transient NORMAL gaze-evoked nystagmus that present with gaze > 30° from midline.
- **Method:**
 - Patient is seated in upright position and fixates on a stationary target with best-corrected vision (with glasses if applicable).
 - Examiner keeps his finger about 30 cm from the patient's eye.
 - Examiner moves his finger in the horizontal plane and then in vertical plane, for 10-20 seconds in each position.
 - Examiner should **NOT** move more than 30° from the midline.
 - Eyes are observed for nystagmus with Frenzel glasses then to fixate on the examiner's fingertip.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Jerk nystagmus.
 - Fixed direction.
 - Horizontal or torsional.
 - Enhanced with:
 - Frenzel glasses.
 - Gazing in the direction of fast phase.
 - Suppressed with:
 - Visual fixation.
 - Gazing in the direction of slow phase.
 - No Rebound nystagmus.
 - **CNS pathology:**
 - Jerk or pendular nystagmus.
 - Direction-changing.
 - Pure vertical or torsional.
 - No suppressed visual fixation.
 - Not affected by gazing in different directions.
 - Presence of Rebound nystagmus:
 - After holding the gaze for 30 seconds toward the direction of fast phase, reversed fast-phase direction rebound nystagmus occurs after eye is returned to primary position.
 - **Brun nystagmus:**
 - Combination of central and peripheral Gaze-Evoked Nystagmus.
 - Due to large CPA tumors compressing ipsilateral cerebellar flocculus.
 - If right CPA tumor:
 - Right-beating central nystagmus with right gaze (toward the lesion).
 - Left-beating vestibular nystagmus with left gaze (away from the lesion).

- **Saccades:**
- Allow rapid shifting of gaze from one object to another, maintaining the target image on the fovea.
- Abnormalities in saccadic eye movements may be difficult to detect in clinic examination and may require videonystagmography (VNG).
- Method:
 - Patient is asked to alternately fixate with the head still, on the examiner's nose and then on the finger held at 15° away from the primary position.
 - Repeat it in right and left horizontal plane and up and down in vertical plane.
 - Saccadic eye movement are observed for:
 - Latency of initiation.
 - Dysmetria (inaccurate saccades):
 - Hypometria (undershooting).
 - Hypermetria (overshooting).
- Interpretation:
 - **Peripheral vestibular pathology:**
 - Normal saccadic eye movement.
 - **CNS pathology:**
 - Abnormal saccadic eye movement.
- **Smooth Pursuit:**
- Visual ocular fixation and tracking of moving objects, maintaining the target image on the fovea.
- Requires intact central optic tracts.
- Method:
 - Patients is asked to track the examiner index finger that is initially positioned directly in front of the patient and moved smoothly 30° per second in horizontal then vertical planes.
 - Testing area is restricted 30° to left, right, up, and down from neutral position to avoid provoking normal end-gaze nystagmus.
 - Repeated 3-5 times in each plane.
 - Smooth pursuit eye movement are observed for:
 - Catch-up saccades.
- Interpretation:
 - **Peripheral vestibular pathology:**
 - Normal smooth pursuit.
 - **CNS pathology:**
 - Abnormal smooth pursuit.

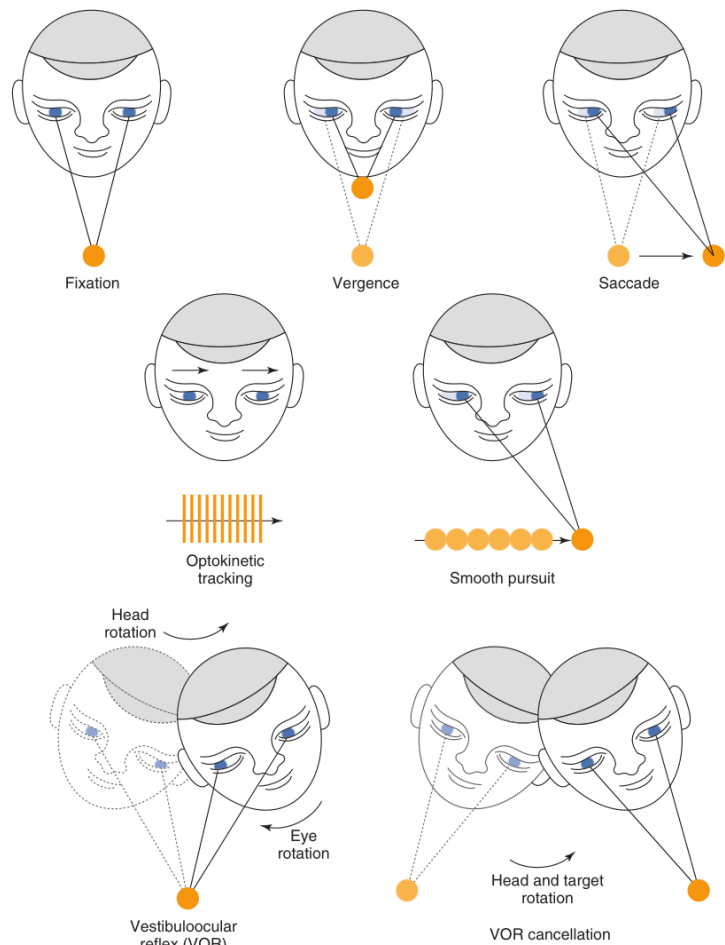


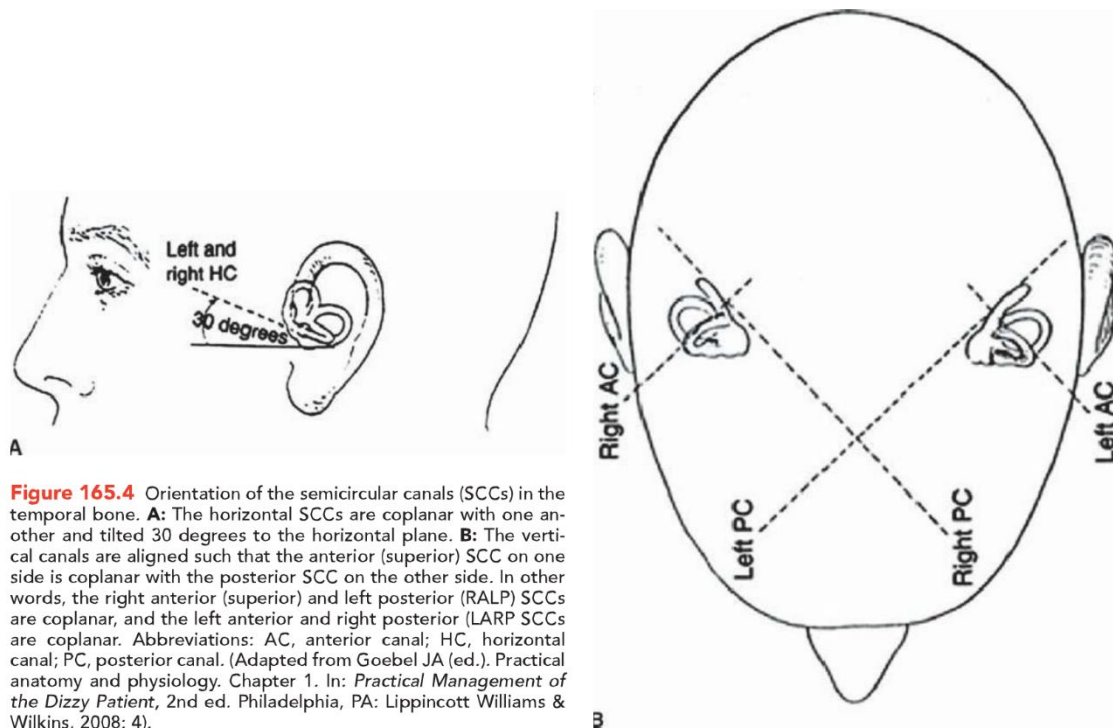
FIGURE 164-1. Schematic depiction of the functional classes of eye movements. Basic research in the neurophysiology of oculomotor control and clinical studies of eye movement disorders have been enhanced by the recognition of functionally distinct subsystems. Seven types of eye movements are shown.

TABLE 164-1. Functional Classes of Human Eye Movements	
Class of Eye Movement	Main Function
Visual fixation	Holds the image of a stationary object on the fovea
Vestibular	Holds images of the seen world steady on the retina during brief head rotations
Optokinetic	Holds images of the seen world steady on the retina during sustained head rotations
Smooth pursuit	Holds the image of a moving target on the fovea
Nystagmus	The repetition of a compensatory slow-phase and quick-phase resetting movement of the eyes; in quick phases, gaze directed toward the oncoming visual scene
Saccade	Brings images of objects of interest onto the fovea
Vergence	Moves the eyes in opposite directions so that images of a single object are placed simultaneously on both foveae

TABLE 164-3. Clinical Observations and Causes of Nystagmus

Observation	Cause and Treatment
Sinusoidal oscillation without fast phases, or <i>pendular nystagmus</i>	Multiple sclerosis, toluene intoxication, or brainstem infarction with inferior olivary hypertrophy (the syndrome of palatoclonus) Acquired pendular nystagmus: frequently dysconjugate and may even be horizontal in one eye and vertical in the other May be responsive to memantine
Purely torsional nystagmus	Intrinsic brainstem involvement within the vestibular nuclei suggestive of syringomyelia
Downbeat nystagmus	Arnold-Chiari deformity or degenerative lesions of the cerebellum Treated with 4-aminopyridine
Upbeat nystagmus	Lesions at the pontomedullary or pontomesencephalic junction or within the fourth ventricle
Horizontal jerk nystagmus that changes direction approximately every 2 minutes, or <i>periodic alternating nystagmus</i>	Lesions in the nodulus of the cerebellum Treated with baclofen
Nystagmus on attempted eccentric gaze and with slow phases that show a declining exponential time course	Side effect of certain medications—especially anticonvulsants, hypnotics, and tranquilizers—and in patients with disease of the vestibulocerebellum or its brainstem connections in the medial vestibular nucleus and nucleus prepositus hypoglossi
Accelerating slow phases accentuated by attempted fixation, decreased by convergence or active eyelid closure, associated with a head turn, and sometimes accompanied by “reversed” smooth pursuit, also known as <i>infantile</i> or <i>congenital nystagmus</i>	May be related to central or peripheral deficits May be associated with albinism, retinal diseases, and early visual deprivation May be responsive to neurontin or memantine
Gaze-evoked nystagmus that may dampen and actually change direction (“centripetal nystagmus”) and often is followed by rebound nystagmus (slow phases are directed toward the previous position of eccentric gaze) when the eyes return to the primary position	Peripheral vestibular lesions, drug intoxication, and cerebellar lesions Rebound nystagmus associated with many cerebellar lesions, including olivocerebellar atrophy
Slow phases with exponentially increasing waveforms	Congenital nystagmus and acquired lesions of the cerebellum
Nystagmus of greater intensity or present only in the abducting eye, or <i>dissociated nystagmus</i>	Commonly occurs in patients with internuclear ophthalmoplegia (INO) Mechanism unknown for abducting nystagmus in INO
One eye goes up and the other goes down, or <i>seesaw nystagmus</i>	Midbrain lesions May be related to an imbalance of activity in structures that receive projections from the labyrinthine otolith organs
Slow phases directed centrally and with retraction of the eye into the orbit, or <i>convergence-retraction nystagmus</i>	Midbrain lesions May be a failure of saccades or of vergence Usually coexists with upgaze paralysis (Parinaud syndrome)
Skew deviation and the ocular tilt reaction (one half-cycle of seesaw nystagmus)	Occurs with acute peripheral vestibular (otolith organ) lesions and with lesions in the medulla, pons, and midbrain Often associated with head tilt
Nystagmus that occurs in a plane other than that of vestibular stimulation, or <i>perverted nystagmus</i>	Central vestibular disorders; multiple sclerosis

- **Head Thrust (Impulse):**
- Used to detect asymmetries in vestibular gain from SSCs.
- Can be performed in all three canal pair orientations.
- In patients with unilateral vestibular loss:
 - VOR cannot produce signals to ocular muscles for eye stabilization during passive angular head rotations.
 - Brain will generate a re-fixation saccade toward the involved ear to acquire a visual target.
- **Method:**
 - Patient is asked to fixate on examiner's nose.
 - Patient's head is impulsively and unpredictably moved 30° from the midline in one direction.
 - **To test Horizontal SSC:**
 - Head is first tilted forward 30°.
 - Head is thrust to examined side in horizontal plane.
 - **To test Superior SSC:**
 - Head is first turned 45° away from examined side.
 - Then it is thrust downward.
 - **To test Posterior SSC:**
 - Head is first turned 45° toward the examined side.
 - Then it is thrust upward.
 - Eye movement are observed for:
 - Corrective saccades.



- Interpretation:

○ **Peripheral vestibular pathology:**

- Corrective saccade in opposite direction to the head movement.

○ **CNS pathology:**

- No corrective saccade.

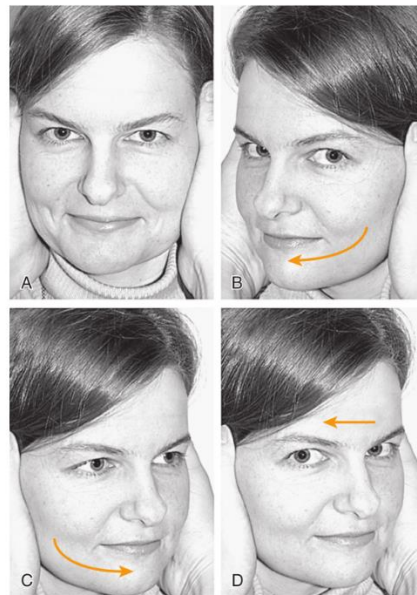
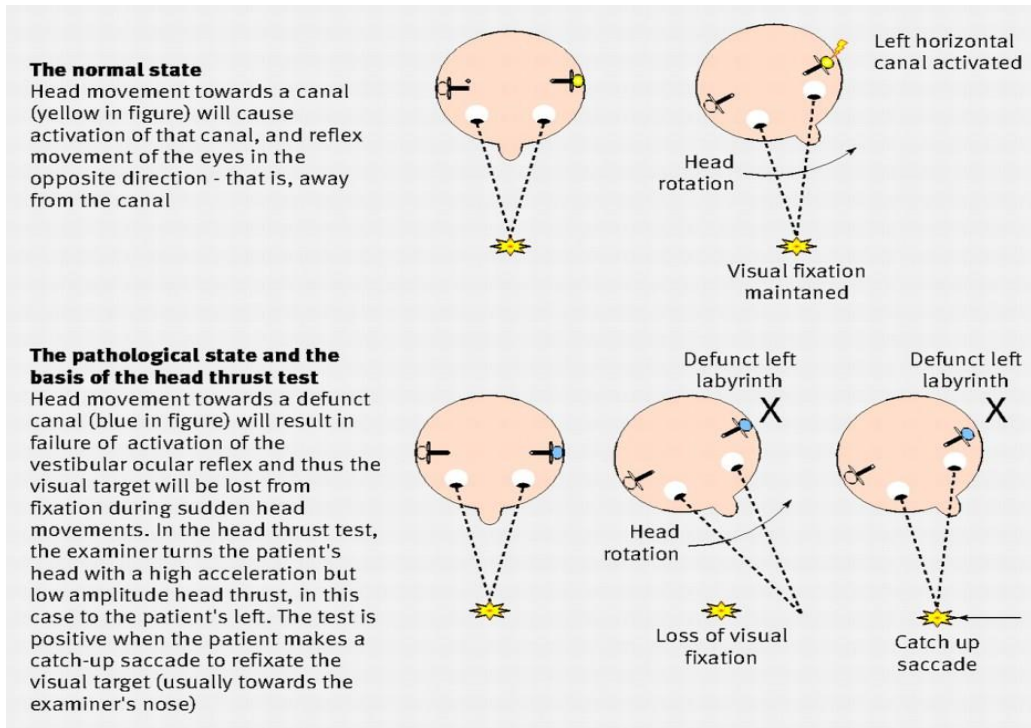


FIGURE 164-8. The head impulse sign in a patient with left unilateral horizontal canal hypofunction. Starting from a neutral position (A), a rapid head impulse to the right in the horizontal plane elicits compensatory eye movements to the left, and the patient's eyes remain stable on the examiner (B). With a similar movement to the left, a hypoactive labyrinth (C) results in a delayed catch-up saccade (D) to maintain gaze. The arrow in D shows the direction of the catch-up saccade. (From Hullar TE, Minor LB. The neurologic examination. In Jackler RK, Brackmann DE, editors: *Neurology*, ed 2, St Louis, 2004, Mosby, pp 215-227.)

- **Head Shake (Heave):**
- Used to detect utricular dysfunction by evaluating asymmetry in VOR.
- Method:
 - Patient is asked to fixate on examiner's nose with Frenzel glasses in place.
 - Patient's head is shake back and forth as quickly as possible in the horizontal plan for a period of 30 seconds.
 - Head shaking is stopped abruptly and the examiner looks for any nystagmus.



- Interpretation:
 - **Peripheral vestibular pathology:**
 - Horizontal nystagmus away from the affected side (due to unopposed stimulation of the intact labyrinth).
 - **CNS pathology:**
 - Vertical nystagmus.
 - Prolonged nystagmus.

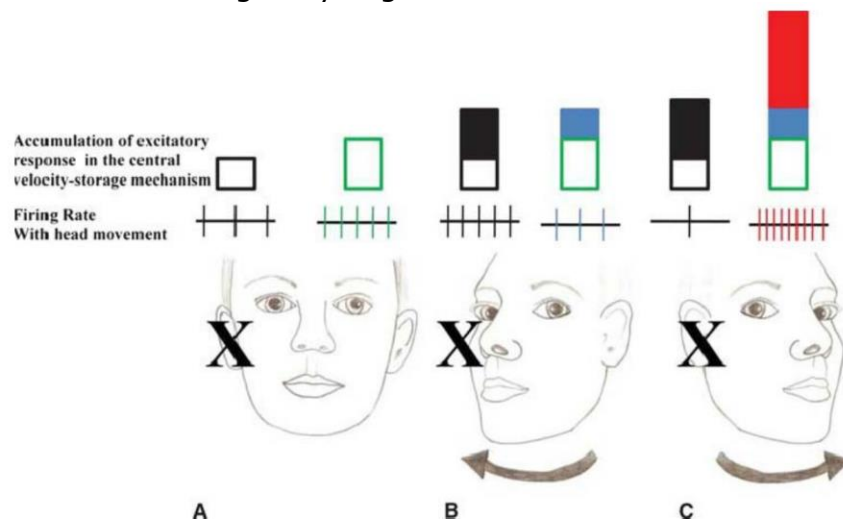


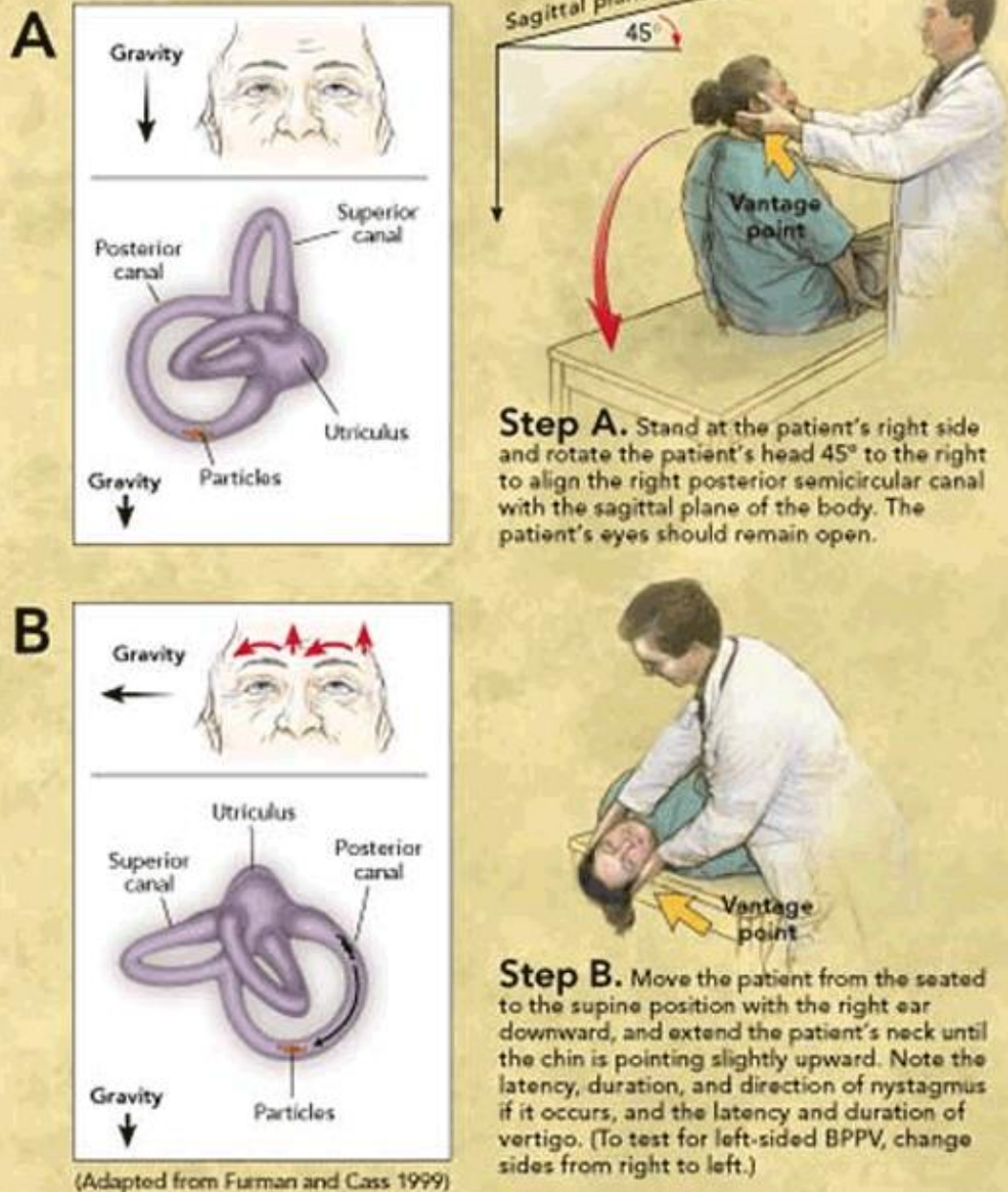
Figure 165.5 The postheadshake nystagmus test in a patient with right labyrinthine hypofunction. **A:** When the head is stationary, the asymmetry in basal firing rates between the healthy labyrinth (left, green) and the hypofunctioning labyrinth (right, black) is minimal. **B:** With head turns toward the hypofunctioning labyrinth (right, black), a weaker-than-normal excitatory response is elicited. The firing rate of the healthy labyrinth decreases (left, blue). **C:** Turning the head toward the healthy labyrinth (left, red) produces a normal excitatory response on the left. Each head rotation amplifies the asymmetry, and the activity accumulates in the central, velocity-storage mechanism. Following head shaking, stored activity is discharged from the velocity-storage center, and nystagmus results.

- **Dix-Hallpike Maneuver:**
- Standard clinical test for PC-BPPV.
- Allows maximal stimulation of posterior SCC.
- The test for BPPV can be made more sensitive by having the patient wear Frenzel glasses.
- **Method:**
 - Patient is seated on the exam table.
 - To test the Right Posterior SCC, examiner holds the patient's head, turns it 45° to the right and then places the patient in a supine position so that his head hangs 30° below the horizontal.
 - Patient's eyes are observed for nystagmus or complaint of vertigo.
 - The test is repeated with head turned to left and then again in straight head-hanging position.
- **Interpretation:**
 - **Peripheral vestibular pathology (PC-BPPV):**
 - Torsional nystagmus
 - Beating toward the stimulated ear (downward ear/geotropic)
 - Appears after latency period of 2-20 seconds.
 - Lasts 15-45 seconds (< 1 minute)
 - Fatigable (disappears with repeated testing).
 - Associated sometime with vertigo.
 - **CNS pathology:**
 - Direction-changing nystagmus
 - No latency
 - Prolonged lasting > 1 minute
 - Not fatigable

Table 6-2. Positional nystagmus in peripheral and central lesions of vestibular system. Positional nystagmus is elicited by Hallpike manoeuvre (vide infra)

	Peripheral	Central
Latency	2-20 seconds	No latency
Duration	Less than 1 minute	More than 1 minute
Direction of nystagmus	Direction fixed, towards the undermost ear	Direction changing
Fatiguability	Fatiguable	Non-fatiguable
Accompanying symptoms	Severe vertigo	None or slight

Fig. 1. The Dix-Hallpike test for right-sided benign paroxysmal positional vertigo

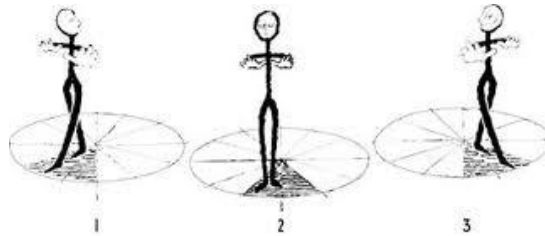


(Adapted from Furman and Cass 1999)

- **Limb Coordination:**
- Evaluate central lesions of the cerebellum and brainstem.
- **Method of Finger-Nose-Finger Test:**
 - Patient is asked to alternate touching the examiner's finger and his own nose.
 - As the patient touches his nose, the examiner quickly moves their finger to a new horizontal position.
 - Examiner observes for any dysmetria.
- **Method of Hand Rapid Alternating Movement Test:**
 - Patient is asked to alternate hand pronation and supination by tapping back of the hand on the thigh and then front of the hand on the thigh in a rapid succession.
 - Examiner observes for any dysdiadochokinesia.
- **Method of Heel-To-Shin Test:**
 - Patient is asked to fully extend their right leg and place their heel on the floor.
 - Then smoothly moves the heel of the left foot along the shin of extended right leg.
 - The test is repeated with the left leg extended.
 - Examiner observes for any dysmetria.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Normal tests.
 - **CNS pathology:**
 - Abnormal tests (dysmetria or dysdiadochokinesia) in cerebellar pathology.



- **Fukuda Stepping Test:**
- Used to evaluate the vestibulospinal function.
- Normal subjects deviate $<30^\circ$ to one side during the stepping test.
- **Method:**
 - Patient is asked to march in place with eyes closed and arms extended straight out at the level of shoulders for 1 minute.
 - Examiner evaluates degree of lateral rotation at the end of the maneuver.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Unilateral Non-compensated:
 - Deviation $>30^\circ$ to the affected side.
 - Unilateral Compensated:
 - Normal test.
 - **CNS pathology:**
 - Patient will be imbalanced even with eyes opened in cerebellar pathology.



- **Tandem Romberg Test:**
- Used to evaluate the somatosensation and proprioception function.
- More sensitive than Romberg test (standing with feet together) to detect vestibular pathology.
- With the eyes open, patient can still compensate the imbalance.
- With eyes closed, vestibular system is at more disadvantage.
- **Method:**
 - Patient is asked to stand with one heel in front of toes and arms extended by the sides with eyes first open and then closed.
 - Examiner observes the side of sway (if any) in both situations with and without vision.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Sway to the affected side with eye closed.
 - **CNS pathology:**
 - Patient will be imbalanced even with eyes opened in cerebellar pathology.



- **Tandem Gait:**
- Used to evaluate the somatosensation and proprioception function.
- **Method:**
 - Patient is asked to walk along a straight line to a fixed point in a heel to toe manner, first with eyes open and then closed.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Sway to the affected side with eye closed.
 - **CNS pathology:**
 - Patient will be imbalanced even with eyes opened in cerebellar pathology.



- **Hyperventilation Test:**
- Hyperventilation decreases serum PCO₂ levels leading to:
 - **Cerebrovascular vasoconstriction:**
 - Leads to lightheadedness and tingling of hands and lips.
 - Reproduce symptoms of hyperventilation syndrome or anxiety.
 - **Elevation of blood pH (Alkalosis):**
 - Leads to improved axonal conduction in partially demyelinated nerve fibers.
- **Method:**
 - With Frenzel glasses in place, patient is asked to hyperventilate for 90 seconds by taking deep breaths and then inhaling and exhaling in rapid succession.
 - Patient is evaluated for dizziness, light-headedness or nystagmus.
- **Interpretation:**
 - **Peripheral vestibular pathology:**
 - Demyelination of CN-VIII (Acoustic schwannoma):
 - Induce excitatory nystagmus to the affected side.
 - **CNS pathology:**
 - Psychiatric disorders (Anxiety and panic attacks):
 - Induce dizziness and light-headedness.
 - Central Demyelination (Multiple sclerosis):
 - Induce nystagmus.

Laboratory Test:

- Requested if the diagnosis is unclear after evaluation by history, physical, and the above mentioned screening tests.
- Includes:
 1. Electronystagmography (ENG)/ Videonystagmography (VNG)
 2. Rotational Chair Testing
 3. Posturography
 4. Vestibular Evoked Myogenic Potentials (VEMP)

Electronystagmography (ENG)/ Videonystagmography (VNG):

- Most widely used vestibular tests.
- Relies on the vestibulo-ocular reflex (VOR)
- Tests the peripheral vestibular function and its ability to generate efficient voluntary eye movements necessary for maintaining visual contact with the environment.
- Provides information regarding localization of lesion.
- Dependent on Anatomy of ear canal and temporal bone.
- Difficult to perform in children.
- Well-tolerated.
- Does not correlate with function.
- Consists of 3 Subtests:
 1. Oculomotor tests.
 2. Positional and positioning tests.
 3. Caloric tests.
- Two methods to measure eye movements:
 1. **Electronystagmography (ENG):**
 - ENG record eye movements by recording changes in the **corneoretinal potential** by placing electrodes around the eyes:
 - Cornea is (+ve) and Retina is (-ve).
 - Advantages:
 - Low cost; easy; noninvasive; no head restraint needed; lots of experience interpreting data.
 - Disadvantages:
 - Signal susceptible to changes in skin resistance due to perspiration; eye blink artifacts; poor signal:noise ratio.
 2. **Videonystagmography (VNG):**
 - VNG record eye movements with small cameras in goggle set.
 - Eyes illuminated with near-infrared light – camera sees the pt but the pt doesn't see the camera.
 - Advantages:
 - No artifact; no need to recalibrate; no need for impedance testing; vertical and torsional movements more easily assessed; disconjugate eye movements more easily seen; portable unit for testing off-site
 - Disadvantages of VOG:
 - must wear goggles; expensive

- **Oculomotor Testing**

- The common thread of these tests is they all test eye movements that originate in the cerebellum.
- Abnormalities in these tests suggest a [Central neurological origin](#).

1. Spontaneous nystagmus and fixation Tests:

- Spontaneous nystagmus may be observed with eyes centered and head upright and asking the patient to visually fixate on an object which is then followed by loss of fixation (ie. close eyes, turn lights off, or use Frenzel lenses).
- Nystagmus due to peripheral lesions usually decreases with fixation (Fixation suppression).

2. Gaze-evoked nystagmus:

- Induced by having the subject gaze 20-30 degrees to the left and right of center for 30 seconds each
- Maybe done with or without fixation.

3. Rebound nystagmus:

- Occur after prolonged gaze holding after the eye is returned to primary position.

4. Saccadic System:

- Saccades allow rapid shifting of gaze rapidly from one object to another, maintaining the target image on the fovea.
- Used to move a target from retinal periphery into fovea
- Tested by presenting targets at 10-20 degrees right and left of center gaze, Patient asked to rapidly shift gaze to each target.
- Evaluate Central components of the vestibular system; cerebellar or brainstem injury may cause ocular dysmetria (overshooting or undershooting of eye rotation).

5. Pursuit System:

- Allows ocular fixation on moving objects and maintains target image on the fovea.
- Tested with Sinusoidal-Tracking Tests by having the patient follow a spot moving in a sinusoidal pattern.
- At faster speeds the eyes may not be able to "keep up" causing saccadic eye jerks.
- Saccadic eye jerks at low velocity (rotational frequencies $<0.1-0.3$ Hz or target velocities $<30^\circ/\text{s}$) suggests a Central pathology.

6. Optokinetic System:

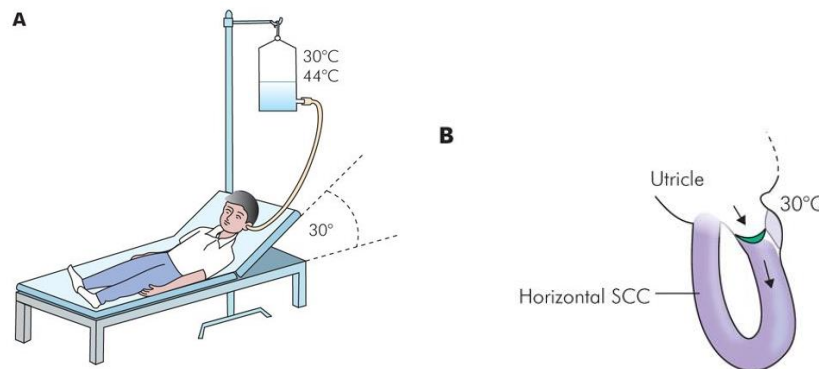
- Allows fixation on a moving field.
- Maintains image on the whole retina rather than specifically on the fovea as in saccades and pursuit systems
- Tested clinically by having the patient stay still and moving the environment (a series of black and white stripes on a moving field that encompasses the whole field of vision).
- Brainstem disease may cause bilateral reduced gain, cerebellar lesions may induce ataxia, peripheral lesions may demonstrate asymmetry.
- Not often used clinically

- **Positional/Positioning Nystagmus Testing**

- Abnormalities in these tests suggest a [Vestibular lesion](#).
- Determine whether the vestibular system responds normally and symmetrically to changes in head position.
- When placed in different positions, dizziness and nystagmus may occur as a result of incomplete compensation in that particular position.
- Patients with dizziness and NO nystagmus provoked by different positions must have non-vestibular etiologies considered.
- Vertical nystagmus or multidirectional nystagmus is highly suggestive of central disorders.
- Used to determine the presence of BPPV.
- [BPPV is most commonly the result of otoliths in the posterior SCC.](#)
- Stimulation using the Dix-Hallpike maneuver commonly causes torsional nystagmus which is difficult to measure if traditional ENG is used.
- It is important to note that patients may fatigue the response, and patients with true BPPV may test negative on the initial visit.
- It is therefore important in both ENG testing and clinical testing that patients with a strong history of BPPV be tested on multiple occasions using the Dix-Hallpike maneuver.
- [Positional Testing:](#)
 - Tests for nystagmus evoked by a new static head position (positional nystagmus is maintained as long as head remains in the evoked position).
- [Positioning Testing:](#)
 - Tests for nystagmus evoked by the action of motion of the head
 - Most common is **Dix-Hallpike**.

- **Caloric Testing:**

- Gold standard study for detecting unilateral vestibular loss.
 - Only test that evaluates vestibular function in each ear independently.
 - Determines right, left or bilateral vestibular weakness.
- The basis of this test is to induce nystagmus by thermal stimulation of the vestibular system.
- In case of TM perforation, Air should be used instead of water.
- **Method:**
 - In supine position, Head is elevated 30° to bring the horizontal SCC into a vertical position.
 - 250 cc of warm and cold water or air is then flushed into the external auditory canal at 7 degrees above (44°C) or below (30°C) body temperature for 40 seconds.
 - Nystagmus is recorded with eyes open and closed.
 - Time taken from the start of irrigation to the end point of nystagmus is recorded and charted on a calorigram.
 - If no nystagmus is elicited from any ear, test is repeated with water at 20°C for 4 minutes before labelling the labyrinth dead.



- Varying temperature causes a non-physiologic stimulation of one labyrinth that may evoke vertigo, nystagmus, and occasionally nausea and vomiting.
- **Cold water or air (30° C)** to **Right Ear** with patient in upright position causes flow in the Ampullifugal direction which Decreases the electrical activity of the ipsilateral vestibular nerve with a corresponding Increase in electrical activity of the opposite vestibular nerve resulting in **Left-Beating Nystagmus**.
- **Warm water or air (40° C)** to **Right Ear** with patient in upright position causes flow in the Ampullipetal direction which Increase the electrical activity of the ipsilateral vestibular nerve with a corresponding Decreases in electrical activity of the opposite vestibular nerve resulting in **Right-Beating Nystagmus**.

- **COWS:**
 - "Cool Opposite, Warm Same"
 - Represents the direction of nystagmus with warm and cool water.
 - Fast phase away from or toward the irrigated ear.
 - Cold irrigation is an inhibitory stimulus, and warm irrigation is excitatory.
 - Warm water causes the perilymph to rotate towards the ampula, resulting in stimulation of the ipsilateral labyrinth and a drift of the eyes away from the stimulated side. The eyes compensate with a saccade toward the stimulated side.
 - The opposite occurs with cold water stimulation.
- **Maximum Slow Phase Velocity:**
 - Determined by dividing the duration by the amplitude of the slow phase.
 - Standard measure of caloric response intensity.
- **Directional Preponderance:**
 - Denotes that the nystagmus response in a particular direction is weaker than the evoked response in the opposite direction.
 - Determined by comparing the duration or velocity of Right-beating nystagmus from both ears with Left-beating nystagmus from both ears.
- **Unilateral Caloric Weakness:**
 - Denotes that the response of one side to a stimulus is reduced in comparison to the other side.
 - Determined by comparing the duration or velocity response from left and right ears.
 - **> 20–25%** difference between sides suggest a unilateral peripheral weakness:
 - Seen in Meniere's disease, acoustic neuroma, post-labyrinthectomy or vestibular nerve section.
- **Bilateral Weakness:**
 - Suggested when the total caloric maximum slow phase velocity from each ear (all 4 irrigations) is Less than 12–24°/second.

Central v. Peripheral Findings on ENG

Findings suggestive of a central disorder

1. Spont or positional nystagmus with normal calorics
2. Direction-changing nystagmus independent of stimulus change
3. Failure of fixation suppression
4. Bilat reduced or absent calorics without history of labyrinthine or middle ear disease
5. Abnormal saccade/pursuit results (esp if calorics N)
6. Hyperactive caloric responses

Findings suggestive of a peripheral disorder

1. Unilateral caloric weakness
2. Bilat caloric weakness with a Hx of labyrinthine disease or ototoxic drug use
3. Fatiguing positional nystagmus
4. Intact fixation suppression response
5. Direction-fixed nystagmus

Rotational Chair Testing

- **Gold standard study for detecting bilateral vestibular loss**
- Sinusoidal (slow) Harmonic Acceleration Test.
- Test SCC under higher frequency conditions than caloric test.
- Evaluates the vestibulo-ocular reflex.
- **Method:**
 - o Patient is seated in the chair, eye movements are recorded while patient (chair) is rotated along the horizontal plane.
- **Vestibulo-ocular reflex measurements:**
 - 1. Gain:**
 - Ratio of slow phase eye velocity to chair (head) velocity
 - **Normal gain is 1.**
 - **Low gain:** Poor attention or testing conditions.
 - **Low gain in ideal conditions:** Bilateral loss.
 - **High gain:** Cerebellar lesion minimizing inhibition
 - 2. Phase:**
 - Compares peak responses of the slow component of the eye velocity with the maximum velocity of the chair (head).
 - Determines the timing relationship between stimulus and response.
 - 3. Symmetry:**
 - Compares eye movement during rightward chair rotation Vs. eye movement during leftward chair rotation.
 - Normally should be completely symmetric.

- Indications:
 1. Bilateral vestibular dysfunction. (Procedure of choice)
 2. Mild vestibular dysfunction undetected by traditional ENG testing.
 3. Identify residual labyrinthine function in those with no calorics.
 4. Monitor changes in vestibular function over time.
 5. Follow progress of vestibular compensation.
- Abnormalities are primarily seen at low frequencies (abnormal phase and gain reduction) and high frequencies (asymmetry)
- Normal vestibulo-ocular response results in similar slow eye phase velocity and chair velocity.
- Acute unilateral peripheral lesions typically reveal low frequency phase leads and high frequency asymmetry (the absence of asymmetry suggests vestibular compensation, eg, vestibular schwannoma)
- Bilateral vestibular disease typically demonstrates reduced gain at low frequencies but normal gain at high frequencies.



Posturography:

- ENG and rotational chair testing are designed to evaluate the horizontal VOR by stimulating the horizontal semicircular canals.
- Posturography evaluates other components of balance.
- Measures postural stability while variably changing the visual field references and support structures.
- Evaluates Spinal/proprioceptive, visual, and vestibular systems
- **Method:**
 - o Patients tries to maintain balance on a level platform with eyes open, eyes closed, and with stimulated moving visual environment.
 - o Test is then repeated on a physically swaying platform.
- Balance requires cerebellar integration of the information from vestibular, visual and somatosensory organs.
- Dysfunction of any of the necessary components of balance results in stronger reliance on other peripheral sensors for maintenance of balance.
- Posturography systematically takes away one or more sensory components to evaluate which component the patient is reliant upon for balance.
- [This is accomplished through one of six conditions:](#)
 - 1. Condition One:**
 - o Stable platform with eyes open in a stable visual environment (patient has full use of all information: visual, vestibular, and somatosensory).
 - 2. Condition Two:**
 - o Stable platform with eye closed (patient must rely on vestibular and somatosensory information)
 - 3. Condition Three:**
 - o Stable platform with moving visual surroundings (patient must suppress a false sense of visually induced movement and rely on vestibular and somatosensory inputs).
 - 4. Condition Four:**
 - o Unstable platform with eyes open in a stable visual environment (patient must rely on vestibular and visual inputs)
 - 5. Condition Five:**
 - o Unstable platform with eyes closed (patient must rely on vestibular input only because visual and somatosensory feedback have been eliminated)
 - 6. Condition Six:**
 - o Unstable platform and unstable visual environment (patient must rely on vestibular input alone and suppress a false sense of visually induced movement.
- **Controversial applications in diagnosis (Non-specific).**
- **Indications:**
 1. Confirm malingering.
 2. Monitor progress in vestibular rehabilitation.

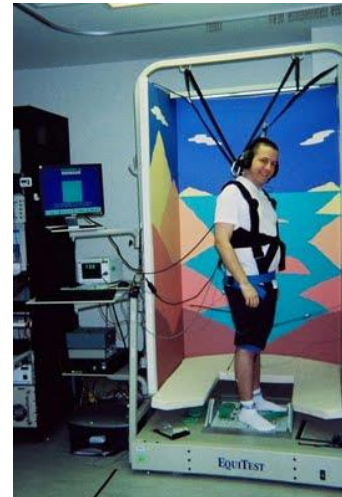
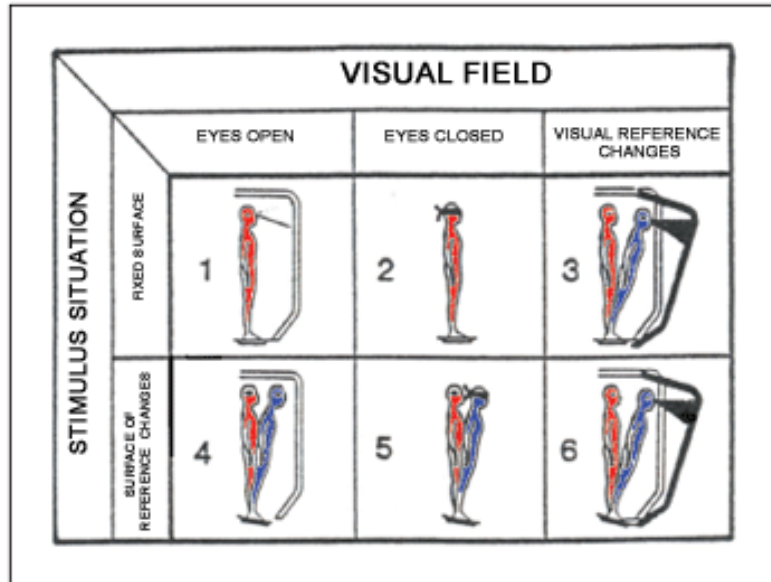


TABLE 131.4  **DIAGNOSIS**
CLASSIFICATION OF ABNORMAL
SENSORY PATTERNS

Pattern	Abnormal Condition (Test No.)
Vestibular dysfunction	5 and 6
Visual vestibular dysfunction	4, 5, and 6
Visual preference	3 and 6 or 6 alone
Visual preference and vestibular dysfunction	3, 5, and 6
Somatosensory and vestibular dysfunction	2, 3, 5, and 6
Severe dysfunction	Abnormal scores on four or more conditions not covered above
Aphysiologic or functional dysfunction	1, 2, 3, and 4 or any combination with normal 5 and 6

Vestibular Evoked Myogenic Potential (VEMP)

- **Cervical VEMP (cVEMP)**

- Provide 95 dB auditory click stimulation, gauge EMG response in ipsilateral sternocleidomastoid muscle.
- Measures sacculocolic reflex.
- Pathway:
 - Acoustic signal > saccule > inferior vestibular nerve > vestibular nucleus > vestibulospinal tract > sternocleidomastoid muscle action potential.
- Indications:
 - Ménière's disease (increase threshold)
 - Inferior vestibular nerve lesion (increase threshold)
 - SCDS (decrease threshold)

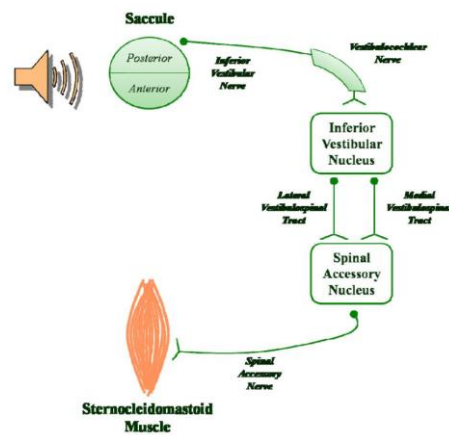


Figure 3. Diagram of the cVEMP neural pathway evoked by an air-conducted stimulus.

- **Ocular VEMP (oVEMP)**

- EMG response of contralateral ocular muscles in response to bone conducted vibration or air conducted sound.
- Air conduction elicits contralateral response, bone conduction elicits bilateral responses (represents vestibulo-ocular pathway)
- Pathway:
 - Acoustic signal > utricle > superior vestibular nerve > contralateral inferior oblique (for air conduction) or bilateral response (for bone vibration).
- Superior vestibular neuritis causes absent/reduced oVEMP but no effect on cVEMP.

	cVEMP	oVEMP
Muscle	SCM	Orbital (inferior oblique)
Response stimulation	- ve	+ ve
End-organ excitation	Saccule	Utricle
Nerve	Inf. Vest. N.	Sup. Vest. N.

Management Concepts of Dizzy Patient:

1. Safety:

- Avoid heights, ladders, driving, operating heavy machinery

2. Acute Vestibular Suppression:

- Indicated for intolerable symptoms.
- May delay central compensatory mechanisms in the long term.
- Common medical therapies:
- Betahistine (**Betaserc** 24,16,8 mg , Maximum 48mg/day).
- Phenothiazine
- Meclizine
- Diazepam
- Scopalmine
- Antiemetics
- Corticosteroid.

3. Vestibular Rehabilitation:

- Indicated for chronic complaints.
- Consists of a series of positional tasks, head movements, and oculomotor exercises to facilitate central compensation

4. Surgical Management:

- Indicated for specific diagnoses

Disorders of Vestibular System:

Disorders of vestibular system cause vertigo and are divided into:

- **Peripheral:**

- Responsible for 85% of all cases of vertigo.
- Involve vestibular end organs and their first order neurons.
 - Inner ear or the CN V-III nerve pathology.
- CNS compares the input that comes from each ear:
 - When inputs are symmetric, the system is balanced and no sense of movement is felt
 - When inputs are asymmetric, CNS interprets this as a head rotation and generates compensatory eye movements and postural adjustments leading to vertigo.
- In healthy individual, CNS compensation occurs after several days and results in near-normal clinical recovery, even if complete unilateral vestibular loss has occurred.

- **Central:**

- Involve central nervous system after the entrance of vestibular nerve in the brainstem and involve vestibulo-ocular, vestibulo-spinal and other central nervous system pathways.

Table 7-1. Vestibular disorders

Peripheral (Lesions of end organs vestibular nerve)	Central (Lesions of brainstem and central connections)
● Meniere's disease ● Benign paroxysmal positional vertigo ● Vestibular neuronitis ● Labyrinthitis ● Vestibulotoxic drugs ● Head trauma ● Perilymph fistula ● Syphilis ● Acoustic neuroma	● Vertebrobasilar insufficiency ● Posterior inferior cerebellar artery syndrome ● Basilar migraine ● Cerebellar disease ● Multiple sclerosis ● Tumours of brainstem and fourth ventricle ● Epilepsy ● Cervical vertigo

TABLE 8–6. General Characteristics of Peripheral and Central Causes of Vertigo

Characteristic	Peripheral	Central
Intensity	severe	mild
Fatigability	fatigues, adaptation	does not fatigue
Associated Symptoms	nausea, hearing loss, sweating	weakness, numbness, falls more likely
Eye Closure	symptoms worse with eyes closed	symptoms better with eyes closed
Nystagmus	horizontal, may be unilateral, rotary	vertical, bilateral
Ocular Fixation	suppresses nystagmus (may not suppress during acute phase)	no effect or enhances nystagmus

TABLE 166-1. Differentiation of Central and Peripheral Vertigo

Feature	Central Origin	Peripheral Origin
Imbalance	Severe	Mild to moderate
Neurologic symptoms	Frequent	Rare
Nystagmus	Changes direction on lateral gaze No change with fixation	Unidirectional Decreases with fixation
Hearing loss	Rare	Frequent
Nausea	Variable	Severe
Recovery	Slow	Rapid

TABLE 167.1**DISTINGUISHING CENTRAL FROM PERIPHERAL CAUSES OF DIZZINESS**

	Nausea and Vomiting	Ability to Walk	Hearing Loss	Focal Neurologic Signs and Symptoms	Associated Autonomic Symptoms (Sweating, Pallor)
Peripheral	Severe	Unsteadiness but able to walk	Common	Rare	Pronounced
Central	Moderate	Nearly incapable of walking	Rare except AICA infarct	Common	Less pronounced

TABLE 167.2**DISTINGUISHING CHARACTERISTICS OF PERIPHERAL VERSUS CENTRAL NYSTAGMUS**

	Type of Nystagmus	Effect of Fixation	Effect of Eye Position	Positional Nystagmus	Other Abnormalities on Testing
Peripheral: Labyrinth or vestibular nerve	Horizontal and torsional	Inhibited by fixation	Follows Alexander law	Latency, fatigues, torsional upbeat nystagmus, <1 min	May be associated with asymmetry of caloric response or of the VOR
Central: Brainstem or cerebellum or interconnections	Can be pure vertical, horizontal, or pure torsional	Unaffected by fixation	May be unidirectional, but if direction-changing is always central	No latency, does not fatigue, oftentimes downbeat or reversal of direction	Often associated with abnormalities of smooth pursuit, optokinetic nystagmus OKN, or saccades

**TABLE
166.1**

**BASIC PRESENTATIONS
OF PERIPHERAL VESTIBULAR
DYSFUNCTION**

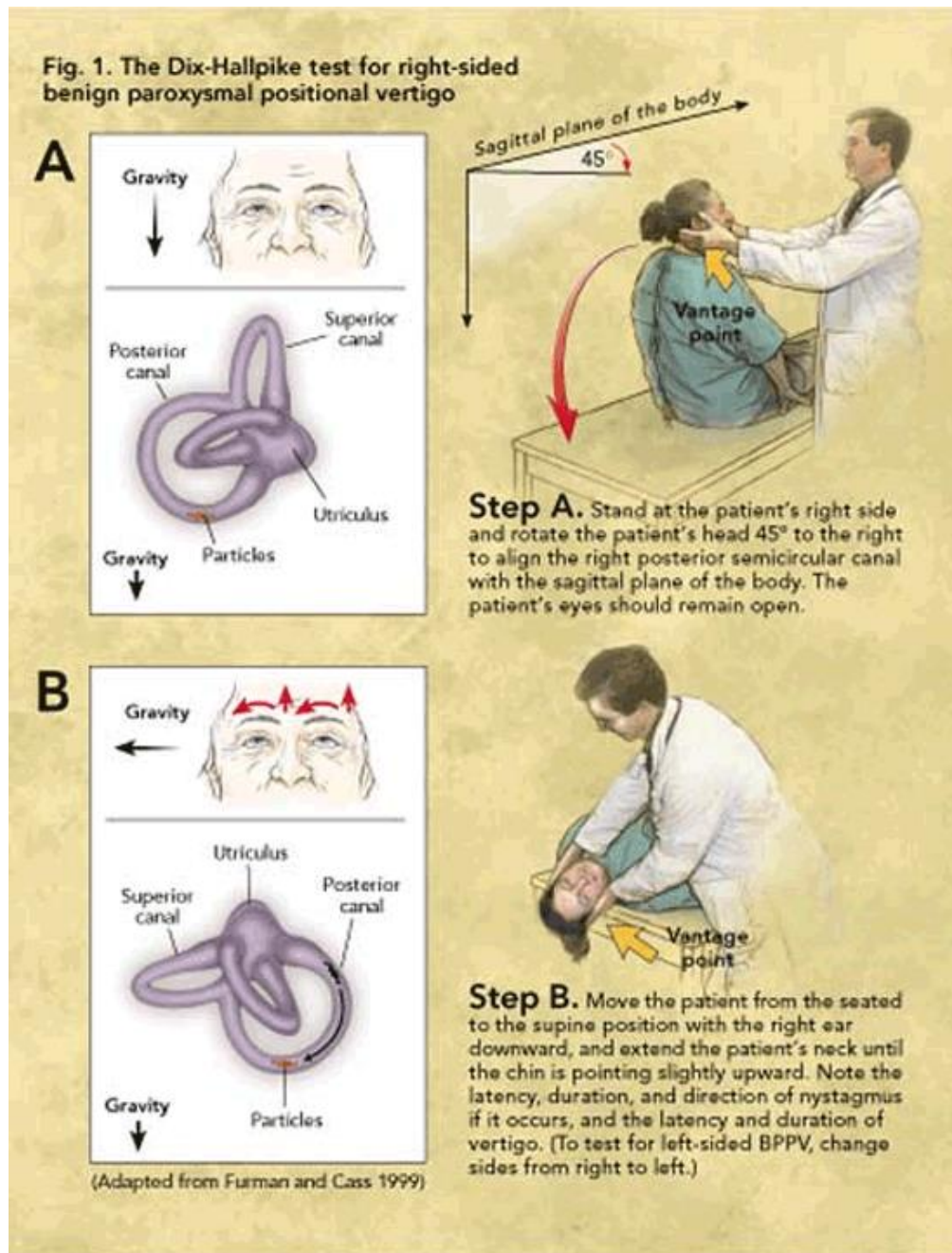
1. Episodic disruption of unilateral vestibular function
 - Lasting minutes-hours*
 - Ménière's disease
 - Perilymph fistula
 - Lasting >24 h*
 - Vestibular neuritis
 - Labyrinthitis
 - IMIED
 - Lasting variable periods of time*
 - Migrainous vertigo
 2. Brief/episodic excitation of unilateral vestibular function
 - BPPV
 - SCD syndrome
 - Perilymph fistula
 3. Chronically inadequate vestibular function
 - Unilateral*
 - Unilateral vestibular hypofunction following vestibular neuritis, trauma, etc.
 - Acoustic neuroma
 - Bilateral*
 - Aminoglycoside toxicity
 - Chemotherapy
 - Familial
-

Peripheral Vestibular Disorders:

Benign paroxysmal positional vertigo (BPPV):

- Most common cause of peripheral vertigo disorder.
 - o 40% of patients with peripheral vestibular vertigo.
- Characterized by brief spells of severe vertigo after specific movements of the head.
- Incidence increases with age with mean age of onset is 40-50s.
- About 1/3 of patients have a recurrence in the first year after treatment, and by five years, 50% of all patients have a recurrence.
- Pathophysiology:
 - 1. Canalithiasis theory:**
 - Most accepted theory.
 - Free-floating otoconia (calcium carbonate particles from utricle) is dislodged and migrate to the endolymph of SCC as a result of head movement in a specific position.
 - The inertial drag of the endolymph causes displacement of the cupula resulting in latent vertigo which resolves when the debris settles.
 - 2. Cupulolithiasis theory:**
 - Dislodged otoconia adheres to the cupula of the semicircular canal resulting in an ampulla that is gravity sensitive.
 - Objections to theory include no account for the transient nature of vertigo and the torsional nystagmus exhibited in BPPV.
- Types of BPPV:
 - o **Posterior canal BPPV (PC-BPPV): 90%**
 - Most common SCC affected because it is the lowest SCC when head is upright.
 - Vertigo occurs after:
 - Rolling over in bed
 - Extreme head extension while looking up
 - Rare to be bilateral (5%).
 - o **Lateral canal BPPV (LC-BPPV): 10%**
 - Most commonly induced by the repositioning maneuver for PC-BPPV (Epley maneuver).
 - Involved side is usually the same side of PC-BPPV.
 - Vertigo occurs after:
 - Turning the head to either side in bed.
 - Otoconia in lateral canal tend to fall out spontaneously with no treatment.
 - o **Superior canal BPPV (SC-BPPV): 2%**
 - Otoconia in superior canal tend to fall out spontaneously with no treatment.

- Causes of BPPV:
 - **Idiopathic (50%):**
 - Most common cause.
 - No known cause was identified.
 - **Head injury:**
 - Most common known cause.
 - **Viral infection:**
 - Vestibular neuronitis.
 - Labyrinthitis.
 - **Surgery:**
 - Stapedectomy.
 - **Prolonged bed rest:**
- Clinical features:
 - Typically self-limiting.
 - Recurrent episodes of:
 - Brief (lasting seconds).
 - Positional vertigo (turning over in bed, getting up, turning the head, bending over, looking up).
 - May be associated with nausea and prolonged light-headedness.
 - No associated hearing loss or other neurologic symptoms.
- Diagnosis of BPPV:
 - **Positioning Maneuvers:**
 - **Dix-Hallpike Maneuver:**
 - Standard clinical test for **PC-BPPV**.
 - Allows maximal stimulation of posterior SCC.
 - The test for BPPV can be made more sensitive by having the patient wear Frenzel goggles or a video goggle.
 - Method:
 - Patient is seated on the exam table.
 - To test the Right Posterior SCC, examiner holds the patient's head, turns it 45° to the right and then places the patient in a supine position so that his head hangs 30° below the horizontal.
 - Patient's eyes are observed for nystagmus or complaint of vertigo.
 - The test is repeated with head turned to left and then again in straight head-hanging position.



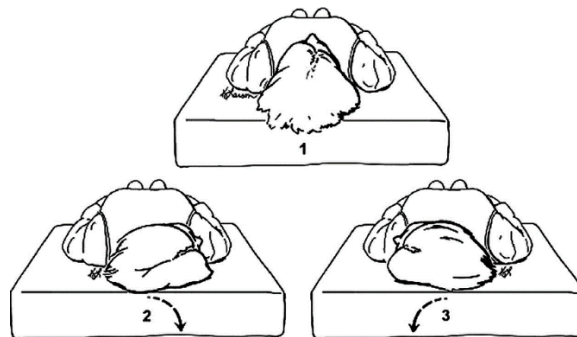
- Classic features of nystagmus in PC-BPPV:
 1. Torsional nystagmus
 2. Beating toward the stimulated ear (downward ear/geotropic)
 3. Appears after latency period of 2-20 seconds.
 4. Lasts 15-45 seconds (< 1 minute)
 5. Fatigable (disappears with repeated testing).
 6. Associated sometime with vertigo.

Table 6-2. Positional nystagmus in peripheral and central lesions of vestibular system. Positional nystagmus is elicited by Hallpike manoeuvre (vide infra)

	Peripheral	Central
Latency	2-20 seconds	No latency
Duration	Less than 1 minute	More than 1 minute
Direction of nystagmus	Direction fixed, towards the undermost ear	Direction changing
Fatiguability	Fatiguable	Non-fatiguable
Accompanying symptoms	Severe vertigo	None or slight

▪ **Supine Roll Maneuver:**

- Standard clinical test for **LC-BPPV**.
- Allows maximal stimulation of Lateral SCC.
- Method:
 - Patient is seated on the exam table.
 - Patient is placed in a supine position with head resting on exam table (not hyperextended).
 - Head is then turned rapidly to right so the patient's right ear rests on the table.
 - Eye movements are monitored with Frenzel glasses for 30 seconds.
 - Patient's head is then returned back to supine position (looking upward).
 - Head is then turned rapidly to left so the patient's left ear rests on the table.
 - Eye movements are monitored again.
 - Which one is the bad ear?
 - **The side that when placed down, elicits greater symptomatic complaints and nystagmus amplitude.**



- Classic features of nystagmus in LC-BPPV:

1. Horizontal nystagmus
2. Geotropic (most commonly) or ageotropic.
3. Shorter latency period than in PC-BPPV.
4. Less fatigable than in PC-BPPV.

- **Rotatory chair test:**

- Indications:

- Diagnose and treat patients with spine abnormalities who can't tolerate manual manipulation.
 - Diagnose and treat patients with more difficult cases of BPPV (Bilateral or lateral BPPV).



- Treatment of BPPV:

- **In-Office Re-Positioning Maneuvers:**

- Based on repositioning free floating particle (otoconial debris) from involved SCC back into the utricle.

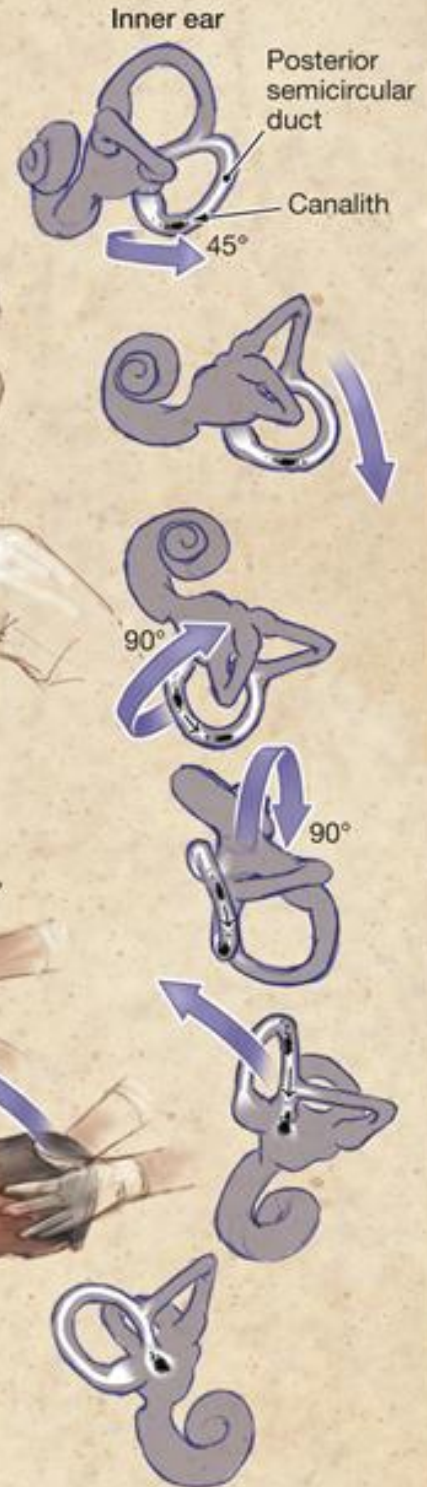
- **Maneuvers for treating PC-BPPV:**

- 1. Epley's Maneuver:**

- Simple and effective to treat PC-BPPV.
 - 80% of patients will be cured by a single maneuver.
 - If the patient remains symptomatic, the maneuver can be repeated.
 - Method:
 - Starting with Dix-Hallpike position to the affected side.
 - Wait till vertigo and nystagmus subside.
 - Head is turned 90 degree and wait till vertigo and nystagmus subside.
 - Head and body are turned 90 degree more in lateral recumbent position so that affected ear is up.
 - Patient is then brought to upright sitting position.

Canalith Repositioning Procedure for Left-Sided Benign Paroxysmal Positional Vertigo

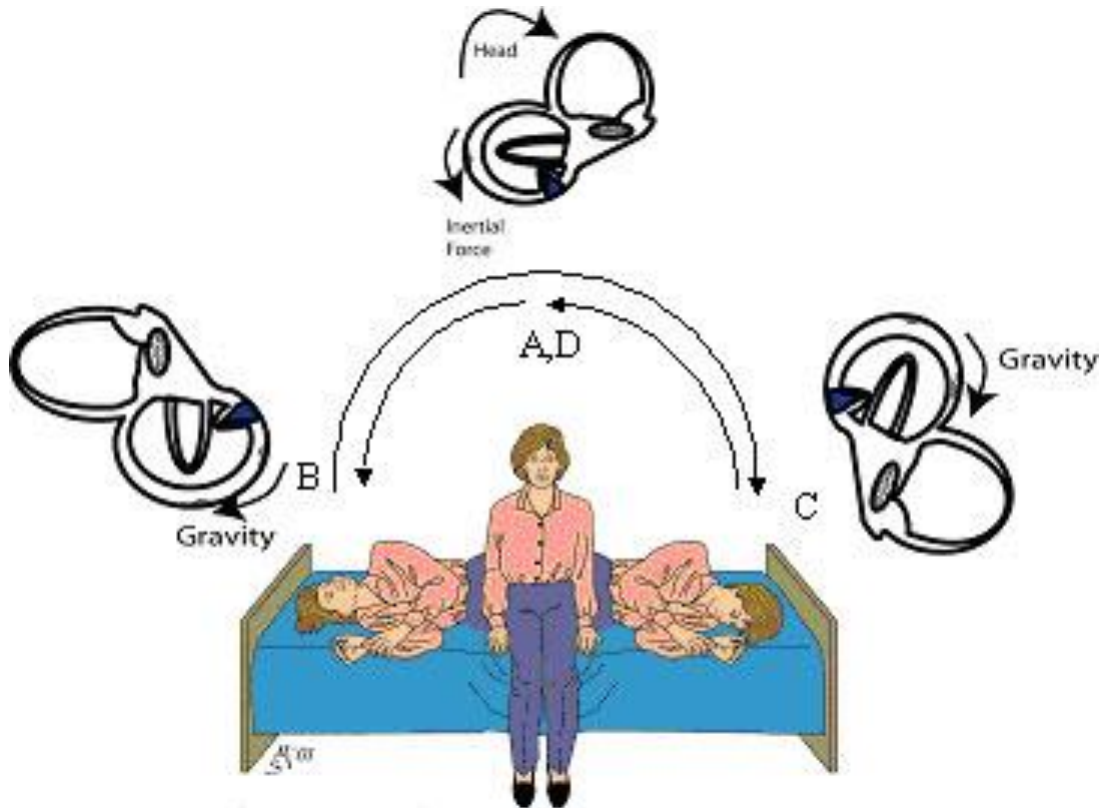
- 1 With patient seated on table, turn head 45° toward the affected side while extending the neck.
- 2 Lay patient down keeping head rotated and extend the neck 10° to 20° depending on patient's ability and comfort. Hold this position for 20 to 30 seconds or until nystagmus or vertigo ceases.
- 3 Turn head 90° toward the unaffected side. Hold this position for 20 to 30 seconds or until nystagmus or vertigo ceases.
- 4 Turn head another 90° rolling body toward the unaffected side. Hold this position for 20 to 30 seconds or until nystagmus or vertigo ceases.
- 5 Return patient to upright, seated position with neck flexed for 20 to 30 seconds.



C. Lynn

2. Semont Maneuver:

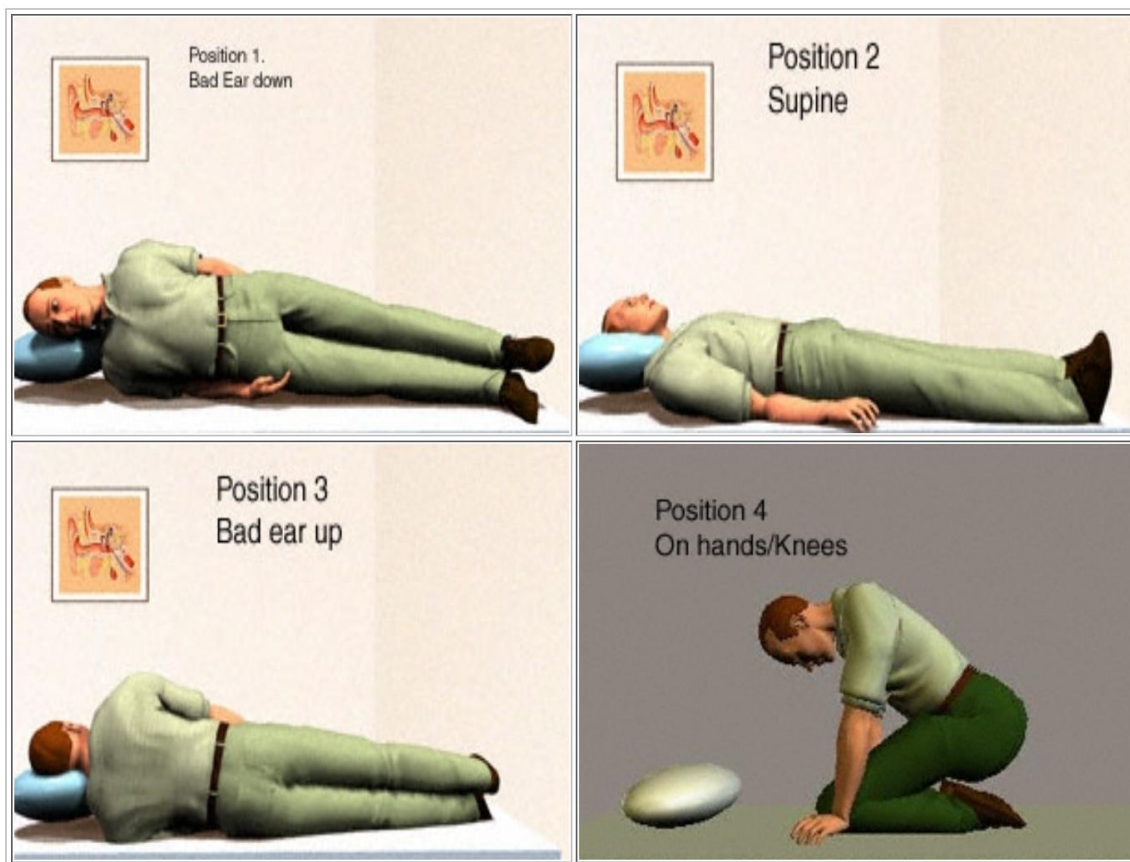
- Effective to treat PC-BPPV.
- More difficult compared to Epley maneuver.
- Method for treating Left PC-BPPV:
 - Patient is setting in upright position with head is turned 45 degree to the affected side (left).
 - Patient is then turned into right lateral recumbent position while maintaining this head position (looking upward) and remains for 4 minutes.
 - Patient is then rapidly moved 180 degree into left lateral recumbent position while maintaining this head position (looking downward) and remains for 4 minutes.
 - Patient is then sit patient up slowly.



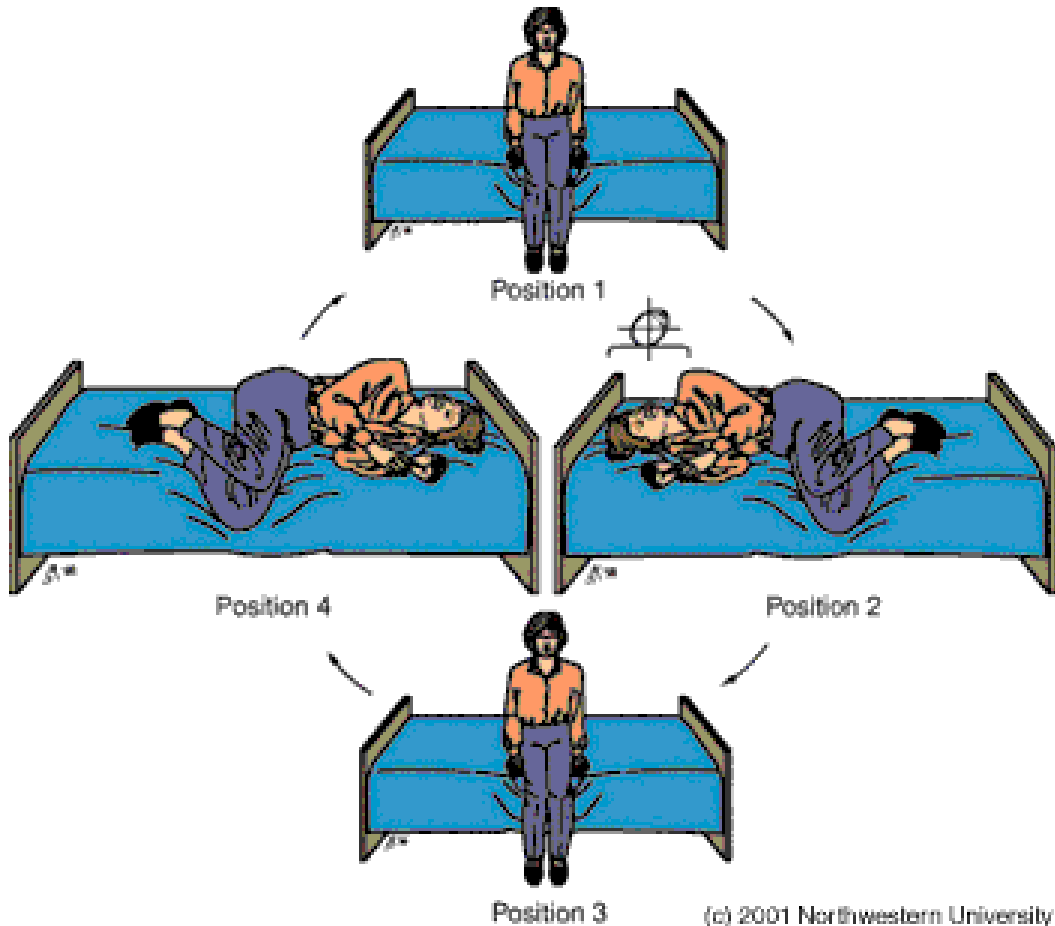
- **Maneuvers for treating LC-BPPV:**

- 1. Log Roll / Barbecue Maneuver:**

- Effective to treat LC-BPPV.
 - Method:
 - Patient is lying in lateral recumbent position with affected ear is down for 1 minute.
 - Patient body is turned 90 degree into supine position for 1 minute.
 - Patient body is turned 90 degree into lateral recumbent position with affected ear is up for 1 minute.
 - Patient body is turned 90 degree into prone position with face looking down for 1 minute.
 - Patient is then setting in upright position.



- Instructions post repositioning maneuvers:
 - Remain upright for 48 hours.
 - For at least one week, avoid provoking head positions that might bring BPPV:
 - Use 2 pillows when you sleep.
 - Avoid sleeping on the "bad" side.
 - Don't turn your head far up or far down.
- Causes of failure of repositioning maneuvers:
 - **Maneuver didn't work:**
 - Should keep treating for 4 attempts.
 - **Canal conversion (5%):**
 - From PC-BPPV to ipsilateral LC-BPPV during Epley maneuver.
 - Change of nystagmus from upbeating/torsional nystagmus into horizontal nystagmus.
 - Treated by log roll / barbecue maneuver.
 - **Another problem in addition to BPPV:**
 - Should change treatment.
- **Home Exercises:**
 - Used to treat PC-BPPV at home when the affected side is not clear.
 - **Brandt-Daroff Exercises:**
 - Succeed in 95% of cases.
 - Performed in **3 sets/day for 2 weeks.**
 - **In each set, maneuver is performed for 5 times.**
 - In most persons, complete relief from symptoms is obtained after 30 sets, or about 10 days.
 - Method (1 set):
 - Start sitting upright.
 - Then move into the side-lying position with the head angled upward about halfway for 30 seconds, or until the dizziness subsides.
 - Then go back to the sitting position and stay for 30 seconds.
 - Then go to the opposite side and follow the same routine.



○ **Surgical treatment of BPPV:**

- Rarely indicated for retractable disabling disease.
- Options:
 - **Posterior SCC Occlusion:**
 - Blocks the canal lumen to prevent movement of endolymph so it becomes unresponsive to angular acceleration.
 - Performed through transmastoid approach.
 - **Singular Neurectomy:**
 - Transection of the inferior vestibular nerve (singular) supplying posterior SCC.
 - Performed by accessing the singular nerve deep to the round window niche via a transcanal approach.
 - Technically difficult.
 - Risk of hearing loss (40%).

Ménière's Disease (Idiopathic Endolymphatic hydrops):

- Idiopathic disorder of inner ear where the endolymphatic system is distended with endolymph.
- Incidence of about 1 in 500.
- Peak age of onset is in the 40-50s.
- **Bilateral in 20% of patients and occurs years after the unilateral symptoms.**
- Symptomatic Triad:
 - 1. Episodic Vertigo**
 - 2. Tinnitus**
 - 3. Fluctuating SNHL**
 - 4. Aural fullness (+/-).**
- Classic Ménière's disease:
 - Presents with ALL of the above symptoms.
- Variants of Ménière's disease:
 - Presents with any combination of the above symptoms
 - Types:
 - 1. Cochlear Hydrops (25%):**
 - Isolated cochlear variant.
 - After several years, vertigo may occur due to block at the level of ductus reuniens.
 - Characterized by:
 - Hearing loss
 - Tinnitus
 - **NO vertigo.**
 - 2. Vestibular Hydrops (20%):**
 - Isolated vestibular variant.
 - Characterized by:
 - Episodic vertigo.
 - **NO tinnitus.**
 - **NO hearing loss.**
 - 3. Lermoyez Syndrome:**
 - Endolymphatic hydrops and membrane ruptures isolated to the basal turns of cochlea and the saccule.
 - Symptoms of Meniere's disease are seen in reverse order.
 - Progressive SNHL
 - Tinnitus
 - Vertigo (at which the hearing loss and tinnitus resolves).

4. Otolithic Crisis of Tumarkin (Drop Attack):

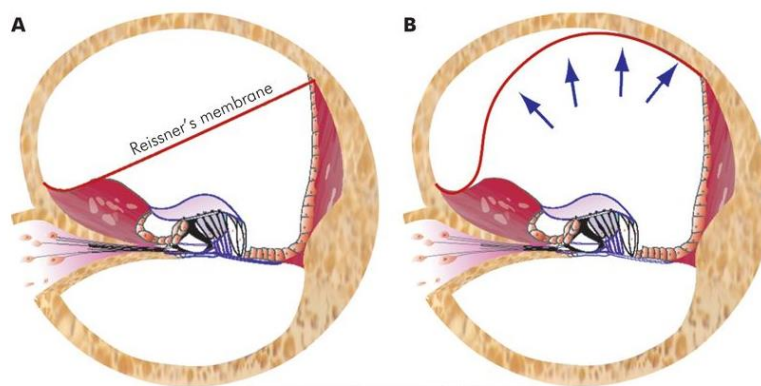
- Acute utriculo-saccular dysfunction.
- Reported in 5% of patients with Ménière's disease.
- Sudden drop attack due to loss of extensor function followed by complete recovery.
- Characterized by:
 - Sudden unexplained falls.
 - NO loss of consciousness.
 - NO vertigo
 - NO fluctuations in hearing loss.

5. Delayed Endolymphatic Hydrops:

- Loss of hearing later followed by typical Ménière's symptoms.

- Pathophysiology of Ménière's disease (Controversial):

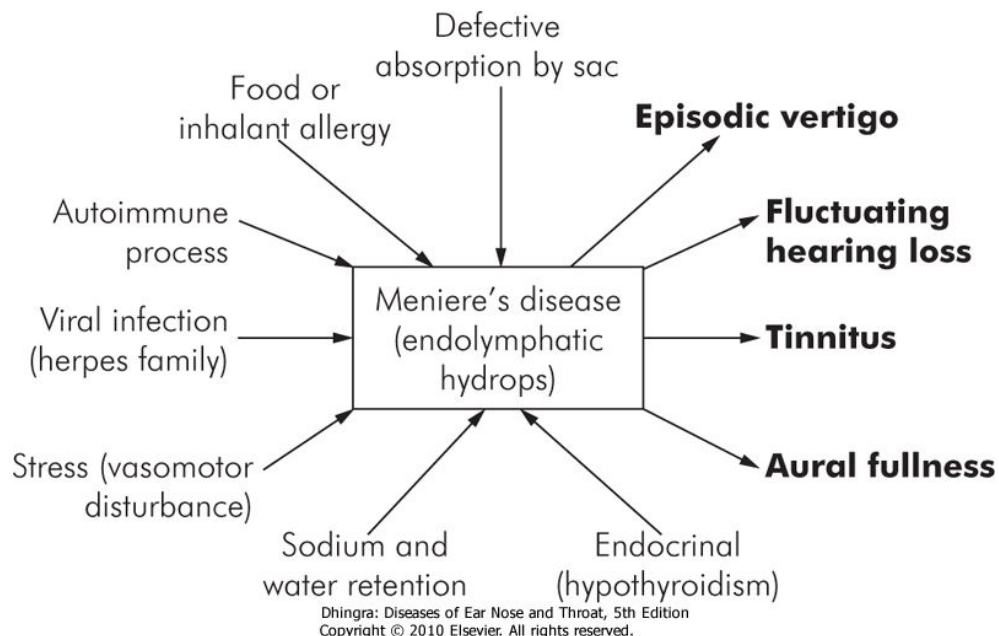
- Normally, endolymph is secreted by stria vascularis, fills the membranous labyrinth and is absorbed through the endolymphatic sac.
- Main pathology in Ménière's disease is distension of endolymphatic system (endolymphatic hydrops) as a result of either excessively synthesized or inadequately resorbed endolymphatic fluid.
- Mainly affecting the cochlear duct (scala media) and the saccule leading to:
 - Dilatation of cochlear duct and bulging of Reissner's membrane.
 - Distention of saccule which may come to lie against the stapes footplate.
- Membranous labyrinth distention may cause micro-tears (rupture) which mixes endolymph and perilymph resulting in some permanent damage to hair cells and instant vertigo.
- The tears then spontaneously seal
- After 2–3 hours the inner ear fluid re-equilibrates with resolution of the vertigo.
- Repeated ruptures may cause progression of the SNHL



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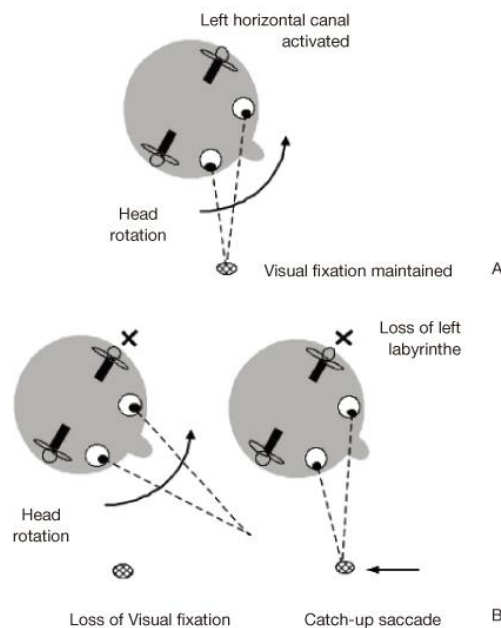
Figure 15.1 (A) Normal cochlear duct. (B) Cochlear duct is distended with endolymph pushing the Reissner's membrane into scala vestibuli.

- Etiology:
 - **Idiopathic endolymphatic hydrops (Ménière's Disease):**
 - Idiopathic with no known cause.
 - **Secondary endolymphatic hydrops (Ménière's Syndrome):**
 - Endolymphatic hydrops secondary to a suggested cause:
 - Allergy:
 - 50% of patients with Meniere's disease have concomitant inhalant and/or food allergy.
 - Inner ear acts as the "shock organ" producing excess of endolymph.
 - Respond to allergic desensitization.
 - Autoimmune (Familial):
 - 10-20% of cases.
 - Autosomal-dominant inheritance.
 - Associated strongly with:
 - Migraine
 - Gluten sensitivity
 - Hypothyroidism:
 - 3% of cases of Meniere's disease are due to hypothyroidism.
 - May benefit from replacement therapy.
 - Vascular/ischemic insult:
 - Sympathetic over-activity resulting in spasm of internal auditory artery branches.
 - Interfering with the function of cochlear or vestibular sensory neuroepithelium.
 - Viral infection
 - Trauma
 - Excessive sodium and water retention



- Clinical presentation:
 - Episodic attacks lasting for minutes to hours with periods of spontaneous remission lasting for weeks, months or years.
 - Consist of:
 - **Vertigo (96%):**
 - Onset is sudden.
 - Feeling of rotation of himself or his environment.
 - Exacerbated with any head movement.
 - Accompanied by nausea and vomiting with ataxia and nystagmus.
 - Severe attacks may be accompanied by other symptoms of vagal disturbances such as abdominal cramps, diarrhea, cold sweats, pallor and bradycardia.
 - **Tinnitus (91%):**
 - Ipsilateral to the side of the hearing loss.
 - Low-pitched non-pulsatile (whistling or roaring).
 - Continuous or intermittent.
 - Sometimes precede the attack and may persist during periods of remission.
 - Change in intensity and pitch of tinnitus may be the warning symptom of attack.
 - **Hearing loss (88%):**
 - Ipsilateral to the side of tinnitus.
 - Fluctuating and progressive in nature.
 - Hearing improves after the attack and may be normal during the periods of remission.
 - With recurrent attacks, recovery between episodes may be incomplete resulting in a **progressive SNHL initially at lower frequencies**.
 - Over time, hearing loss flattens and involve higher frequencies.
 - 1-2% of patients progress to profound deafness.
 - Additional features:
 - **Recruitment (56%):**
 - Intolerance to loud sounds.
 - Poor candidates for hearing aids.
 - **Diplacusis (44%):**
 - Difference in the perception of pitch between the ears.
 - **Aural fullness:**
 - Fluctuates.
 - Accompany or precede an attack of vertigo.

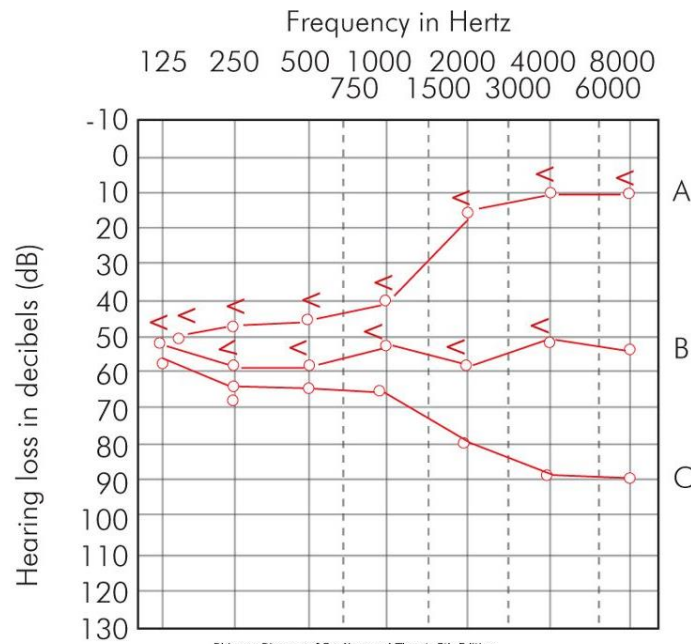
- Physical examination:
 - **Otoscopy:**
 - No abnormality is seen in the tympanic membrane.
 - **Hennebert sign:**
 - False positive fistula test.
 - Pressure induced vertigo and nystagmus in absence of labyrinthine fistula.
 - Formation of fibrous bands between stapes footplate and utricle.
 - Present in:
 - 25% of patients with Meneire's disease.
 - Congenital syphilis.
 - **Tuning fork test:**
 - Indicate SNHL.
 - Rinne test is positive.
 - Weber is lateralized to the better ear.
 - **Nystagmus:**
 - Horizontal nystagmus.
 - Seen only during acute attack.
 - Quick component of nystagmus is towards unaffected ear (paralytic).
 - **Head-Thrust Test:**
 - Assesses integrity of angular vestibuloocular reflex (AVOR).
 - Normal subjects are able to maintain visual fixation on a target during rapid head movement.
 - Asymmetry is only present in 30% of patients with Meniere disease.



- **Investigation:**

○ **PTA:**

- In early stages, lower frequency SNHL and the curve is of rising type.
- As the disease progresses, middle and higher frequencies get involved and audiogram becomes flat type.



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Figure 15.3 (A) Audiogram in early Meniere's disease. Note: Hearing loss is sensorineural and more in lower frequencies-the rising curve. As the disease progresses, middle and higher frequencies get involved and audiogram becomes flat or falling type (B & C).

○ **Speech Audiometry:**

- Discrimination score 55-85% between the attacks.
- Discrimination ability is impaired during and immediately following an attack.

○ **Special audiometry tests:**

- Indicate the cochlear nature of disease.
 - Recruitment test:
 - Positive.
 - SISI (short increment sensitivity index):
 - SISI score is better than 70% in two-thirds of the patients
 - SISI score in normal hearing is 15%.
 - Tone decay test:
 - Decay of less than 25 dB.

Table 15-1. Results of various tests to differentiate a cochlear from a retrocochlear lesion

	Normal	Cochlear lesion	Retrocochlear lesion
• Pure tone audiogram	Normal	Sensorineural hearing loss	Sensorineural hearing loss
• Speech discrimination score	90-100%	Below 90%	Very poor
• Roll over phenomenon	Absent	Absent	Present
• Recruitment	Absent	Present	Absent
• SISI score	0-15%	Over 70%	0-20%
• Threshold tone decay test	0-15 dB	Less than 25 dB	Above 25 dB
• Stapedial reflex	Present	Present	Absent
• Stapedial reflex decay (page 109)	Normal	Normal	Abnormal
• E.R.A	Normal interval between wave I & V	Normal interval between wave I & V	Wave V delayed or absent

- **Cervical Vestibular Evoked Myopotentials (cVEMP):**
 - Reflect otolith function.
 - Standard test in the workup of a patient with Meniere's disease.
 - Measure the disease severity
 - Able to detect bilateral disease.
 - Saccule is second most common site affected by hydrops.
 - cVEMP is generated by playing loud clicks in the ear which moves stapes footplate and stimulates the saccule.
 - This stimulation passes through inferior vestibular nerve and vestibular nuclei and sends inhibitory impulses that relax the sternocleidomastoid muscle.
 - Patients with Meniere's disease will have increase threshold or absent response to sounds.

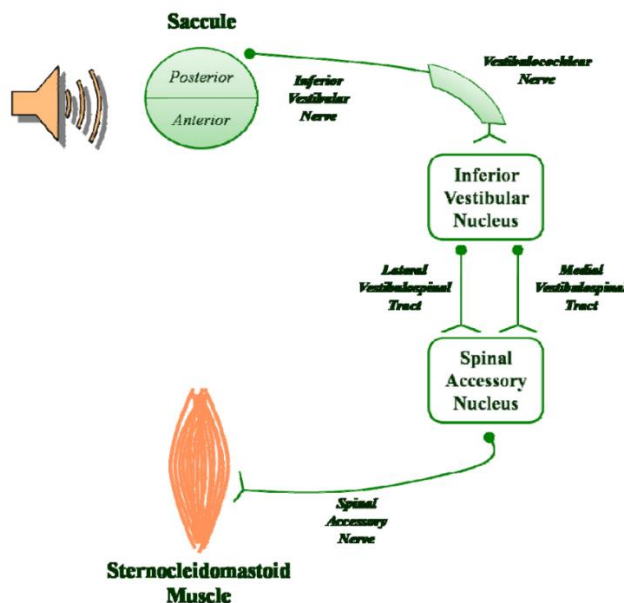


Figure 3. Diagram of the cVEMP neural pathway evoked by an air-conducted stimulus.

- **Electrocochleography (ECoG):**
 - Distension of basilar membrane into scala tympani (hydrops) causes increase in the normal asymmetry of basilar membrane vibration.
 - Summating potential (SP) in Meniere patients is larger and more negative.
 - SP/AP Ratio:
 - Most commonly used value.
 - Ratio of SP amplitude and CN-8th action potential (AP).
 - Sensitivity of 50-70%.
 - **Normally:**
 - SP/AP ratio is 20-30%.
 - **In Meniere's disease:**
 - SP/AP ratio is greater than 50%.

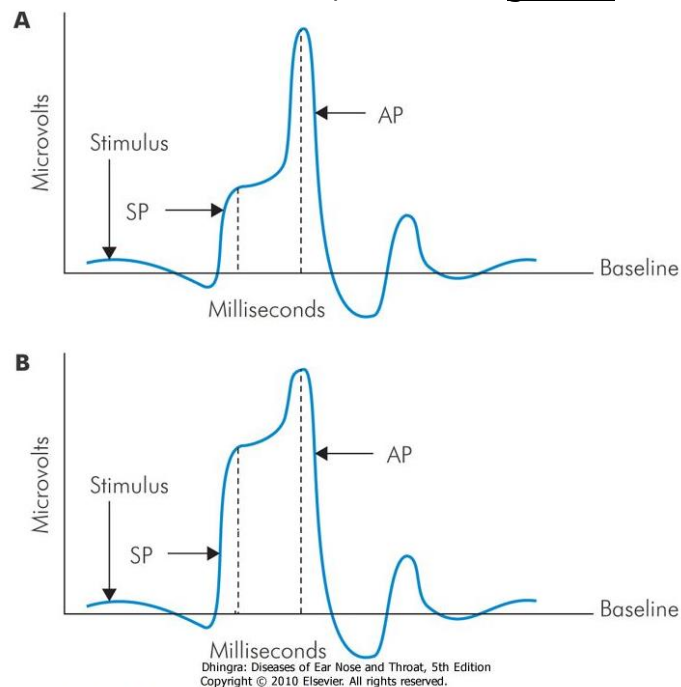


Figure 15.4 Electrocochleography. (A) Normal ear; (B) Ear with Meniere's disease. Voltage of summating potential (SP) is compared with that of action potential (AP). Normally SP is 30% of AP. This ratio is enhanced in Meniere's disease.

- **Videonystagmography (VNG):**
 - Reveal unilateral weakness on affected side.
 - Caloric testing:
 - Caloric asymmetry of $\geq 20\%$ is indicative of unilateral peripheral vestibular hypofunction.
 - Caloric testing is showing reduced response on the affected side in 75% of cases.

- **Dehydration test:**
 - Administration of dehydrating agents (urea, glycerol, and furosemide) to reduce the increased endolymph volume in the inner ear and produce a measurable change in response.
 - 60% sensitivity in cases of known Meniere disease.
 - Improvement is measured by:
 - Audiometrics
 - ECoG (reduction in SP negativity)
 - Change in the gain of vestibuloocular response to rotational stimulation.
- Diagnosis:
 - Based on clinical history, physical examination, and Audiological findings, with exclusion of other causes of hearing loss and vertigo is adequate for diagnosis and initiating empirical therapy.
- **AAOHNS classification of the diagnosis of Meniere's disease:**
- **Certain:**
 - Definite Meniere's disease confirmed by histopathology.
- **Definite:**
 1. Two or more definitive spontaneous episodes of vertigo lasting 20 minutes or longer.
 2. Audiometrically documented hearing loss on at least one occasion.
 3. Tinnitus or aural fullness in the affected ear.
 4. All other causes excluded.
- **Probable:**
 1. One definitive episode of vertigo.
 2. Audiometrically documented hearing loss on at least one occasion.
 3. Tinnitus or aural fullness in the treated ear.
 4. Other causes excluded.
- **Possible:**
 1. Episodic vertigo of Meniere's type without documented hearing loss **or**
 2. Sensorineural hearing loss, fluctuating or fixed, with disequilibrium but without definitive episodes.
 3. Other causes excluded.

Box 165-1. AMERICAN ACADEMY OF OTOLARYNGOLOGY–HEAD AND NECK SURGERY CRITERIA FOR MENIERE DISEASE DIAGNOSIS^{1,20}

Major Symptoms

Vertigo

- Recurrent, well-defined episodes of spinning or rotation
- Duration from 20 minutes to 24 hours
- Nystagmus associated with attacks
- Nausea and vomiting during vertigo spells (common)
- No neurologic symptoms with vertigo

Deafness

- Fluctuating hearing deficits
- Sensorineural hearing loss
- Progressive hearing loss, usually unilateral

Tinnitus

- Variable, often low pitched and louder during attacks
- Usually unilateral
- Subjective

Diagnosis of Meniere Disease

Possible Meniere Disease

- Episodic vertigo without hearing loss or
- Sensorineural hearing loss, fluctuating or fixed, with dysequilibrium but without definite episodes
- Other causes excluded

Probable Meniere Disease

- One definitive episode of vertigo
- Hearing loss documented by audiogram at least once
- Tinnitus or aural fullness in the suspected ear
- Other causes excluded

Definite Meniere Disease

- Two or more definitive spontaneous episodes of vertigo lasting at least 20 minutes
- Audiometrically documented hearing loss on at least one occasion
- Tinnitus or aural fullness in the suspected ear
- Other causes excluded

Certain Meniere Disease

- Definite Meniere disease plus histopathologic confirmation

**Box 165-2. AMERICAN ACADEMY OF
OTOLARYNGOLOGY–HEAD AND NECK SURGERY
CRITERIA FOR MENIERE DISEASE SEVERITY^{120,126}**

In 1996, the Committee on Hearing and Equilibrium reaffirmed and clarified the 1985 guidelines, adding initial staging and reporting guidelines.

Vertigo

- a. Any treatment should be evaluated no sooner than 24 months.
- b. Formula to obtain numeric value for vertigo: ratio of average number of definitive spells per month after therapy divided by definitive spells per month before therapy (averaged over a 24-month period) $\times 100 =$ numeric value
- c. Numeric value scale
 - 0: Class A: Complete control of definitive spells
 - 41 to 80: Class B: Limited control of definitive spells
 - 81 to 120: Class C: Insignificant control of definitive spells
 - > 120: Class D
 - Class E: Secondary treatment initiated

Disability

- a. No disability
- b. Mild disability: intermittent or continuous dizziness/unsteadiness that precludes working in a hazardous environment
- c. Moderate disability: intermittent or continuous dizziness that results in a sedentary occupation
- d. Severe disability: symptoms so severe as to exclude gainful employment

Hearing

- a. Hearing is measured by a four-frequency pure tone average (PTA) of 500 Hz and 1, 2, and 3 kHz
- b. Pretreatment hearing level: worst hearing level during 6 months prior to surgery
- c. Posttreatment hearing level: poorest hearing level measured 18 to 24 months after institution of therapy
- d. Hearing classification:
 - i. Unchanged = ≤ 10 -dB PTA improvement or worsening or $\leq 15\%$ speech discrimination improvement or worsening
 - ii. Improved > 10 -dB PTA improvement or $> 15\%$ speech discrimination improvement
 - iii. Worse > 10 -dB PTA worsening or $> 15\%$ speech discrimination worsening

In 1996, the Committee on Hearing and Equilibrium reaffirmed and clarified the guidelines, adding initial staging and reporting guidelines.

Initial Hearing Level

Four-Tone Average (dB)

Stage 1: ≤ 25

Stage 2: 26-40

Stage 3: 41-70

Stage 4: > 70

Functional Level Scale

Regarding my current state of overall function, not just during attacks:

1. My dizziness has no effect on my activities at all.
2. When I am dizzy, I have to stop for a while, but it soon passes, and I can resume my activities. I continue to work, drive, and engage in any activity I choose without restriction. I have not changed any plans or activities to accommodate my dizziness.
3. When I am dizzy, I have to stop what I am doing for a while, but it does pass, and I can resume activities. I continue to work, drive, and engage in most activities I choose, but I have had to change some plans and make some allowance for my dizziness.
4. I am able to work, drive, travel, and take care of a family or engage in most activities, but I must exert a great deal of effort to do so. I must constantly make adjustments in my activities and budget my energies. I am barely making it.
5. I am unable to work, drive, or take care of a family. I am unable to do most of the active things that I used to do. Even essential activities must be limited. I am disabled.
6. I have been disabled for 1 year or longer and/or I receive compensation because of my dizziness or balance problem.

- **Staging of Meniere's Disease:**
- Done in certain and definite cases of Meniere's disease.
- Based on average of the pure tone thresholds at 0.5, 1, 2, 3 kHz (rounded to nearest whole) of the worst audiogram during interval of 6 months before treatment.

Table 15-2. Staging of Meniere's disease

Stage	Pure tone average in dB in previous 6 months
1	≤ 25
2	26-40
3	41-70
4	> 70

- Management:
 - All proven therapy is directed at relieving vertigo which is the most distressing symptom.
 - 80% of patients can be effectively managed by medical therapy alone.
 - Acute management:
 - **Reassurance:**
 - Patient's anxiety can be relieved by reassurance and by explaining the true nature of disease.
 - **Bed rest:**
 - Head supported on pillows to prevent excessive movements.
 - **Precautions:**
 - Avoid activities requiring good body balance during the attacks.
 - **Vestibular sedatives:**
 - Relieve vertigo.
 - Administered intramuscularly or intravenously if vomiting prevent oral administration.
 - Drugs useful in acute attack:
 - Dimenhydrinate (Dramamine).
 - Prochlorperazine (Stemetil).
 - Promethazine theoclate (Avomine)
 - Diazepam (Valium).
 - **Vasodilators:**
 - Improves labyrinthine circulation.
 - Betahistine (Betaserc).
 - **Corticosteroids:**
 - May be considered for acute exacerbations.

- Chronic Management:
 - **Precautions:**
 - As the attack of Meniere's disease is abrupt, sometimes with no warning symptom, professions such as flying, under-water diving or working at great heights should be avoided.
 - **Behavioral:**
 - Salt Restriction:
 - **First-line therapy.**
 - Reduce endolymph volume by fluid removal.
 - Avoid fluid shifts by restricting salt (<1.5g/day).
 - Lack of hard evidence of its efficacy.
 - Stress Reduction:
 - Symptoms are exacerbated by stress.
 - Mental relaxation exercises and yoga helpful to decrease stress.
 - Cessation of smoking:
 - Nicotine causes vasospasm.
 - Smoking should be completely stopped.
 - Elimination of allergen:
 - Sometimes, a food or inhalant allergen is responsible for such attacks.
 - **Medical:**
 - Diuretics:
 - **First-line therapy.**
 - Reduce endolymph volume by reduce the production.
 - Encourages constant renal output
 - Must avoid dehydration which would exacerbate symptoms.
 - Furosemide or acetazolamide taken on alternate days helps to control recurrent attacks.
 - Lack of hard evidence of its efficacy.
 - Vasodilators:
 - Improves labyrinthine circulation.
 - Betahistine (Betaserc) is H1-histamine receptor agonist.
 - Reduce frequency and severity of vertigo episodes.

- **Surgical:**

- Indicated after failure of medical therapy.
- Needed in 10–15% of patients.
- **Hearing preservation (Serviceable hearing):**

- 1. Trans-tympanic dexamethasone injection:**

- Non-destructive intervention to improve hearing and vertigo attacks.
- Allows to avoid ablative therapies.
- Single injection (**12 mg/ml**) to be repeated after 6-8 weeks if vertigo recurs.
- Complete resolution of vertigo in 80%.

- 2. Trans-tympanic Gentamicin injection (Chemical labyrinthectomy):**

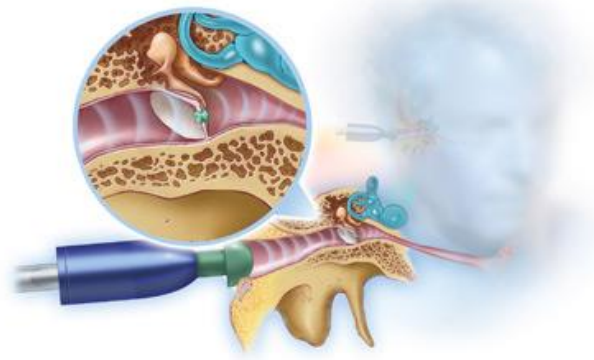
- Gentamicin is mainly vestibulotoxic.
- Destructive intervention to ablate the vestibular function while preserving the hearing.
- Has advantage of avoiding morbidity of surgical labyrinthectomy.
- Used as weekly injections (**40mg/ml**) for a total of 4-6 weeks titrated to the patient's response.
- 10% risk of profound SNHL.
- Patient should be followed up closely with hearing tests to detect early HL.
- Superior to dexamethasone for vertigo control in a RCT.
- Complete resolution of vertigo in 90%.

- 3. Endolymphatic Sac Surgery:**

- Includes:
 - Endolymphatic sac decompression.
 - Endolymphatic shunt to the subarachnoid space or mastoid cavity.
- Non-destructive surgery to preserves auditory and vestibular function.
- Low morbidity and less successful than ablative procedures.
- Efficacy of these procedures remains controversial.
- Rate of success 60–80%.
- Can be done in bilateral Meniere's.

4. Meniett Device Therapy:

- Non-destructive intervention.
- Intermittent positive pressure waves therapy.
- Delivered from external device in the EAC through ventilation tube to reach round window and inner ear fluids.
- Pressure waves pass through the perilymph and cause reduction in endolymph pressure by redistributing it through various communication channels such as the endolymphatic sac or the blood vessels.
- Improves symptoms of Meniere's disease.
- Patient can self-administer the treatment at home.
- It may require a few months before complete remission of disease is obtained.
- Meniett device therapy has been recommended for patients who have failed medical treatment and the surgical options are being considered.



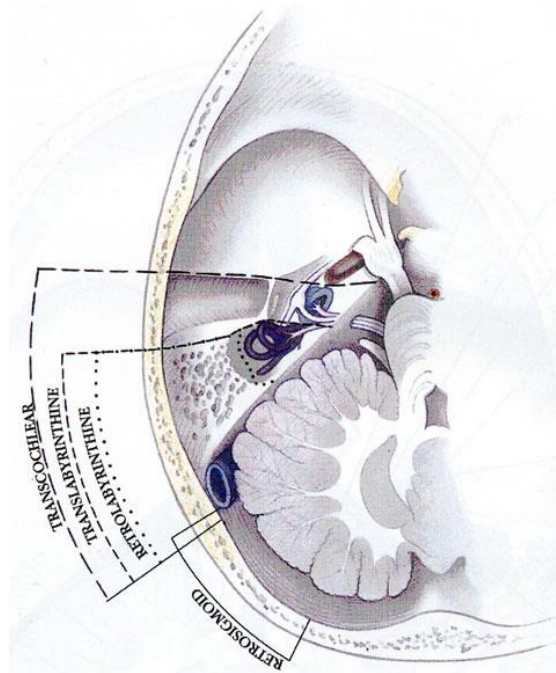
5. Selective Vestibular Nerve Section:

- Approach:
 - Retro-sigmoid.
 - Middle cranial fossa.
- Destructive surgery.
- Achieve vertigo control in 90% of patients.
- Risk of hearing worsening in 30% of patients.

- **NON Hearing preservation (Non-Serviceable hearing):**

1. Labyrinthectomy:

- Approaches:
 - Trans-mastoid.
 - Trans-canal.
- Destructive surgery that totally destroy cochlear and vestibular function.
- Considered for non-serviceable hearing (>50 – 60 dB HL or $<50\%$ word recognition).
- High rate of success (90%).
- Must take into account risk of development of Ménière's disease in the contralateral ear later on.



- Cochlear Implantation is a viable option for hearing restoration in patients Meniere's disease and SNHL.
- In patient with bilateral Meniere's disease, destructive surgeries to the vestibular system should be limited to one ear to avoid development of oscillopsia.

Vestibular Neuritis:

- Vestibular dysfunction due to infection of vestibular nerve.
- Characterized by:
 - o **Severe vertigo:**
 - Sudden onset.
 - Lasting several hours to days.
 - Gradual improvement throughout the course.
 - May associate with URTI.
 - o **NO Hearing loss.**
 - o **NO neurologic impairment**
- Pathophysiology:
 - o Vestibular nerve degeneration secondary to reactivation of a latent herpes simplex virus infection in the vestibular ganglia.
 - In most cases, responsible virus is never identified.
 - o **Superior vestibular nerve** is more commonly involved as it is longer and pass through a narrower bony canal that makes it more susceptible to compressive swelling.
 - o 1/3 of patients will develop PC-BPPV months to years after resolution due to otolithic debris released from the damaged utricle.
- Diagnosis:
 - o Clinical history and physical exam.
 - o Vestibular lab:
 - Unilateral decreased caloric response on VNG.
 - o No associated HL on audiogram
- Management:
 - o **Steroids:**
 - Mainstay of treatment.
 - Improve caloric function.
 - Recommended to be started within 3 days of onset of symptoms.
 - o **Supportive:**
 - Anti-vertiginous:
 - For first 72 hours.
 - Long-term anti-verginous medications should be avoided as it tend to decrease vestibular habituation.
 - Anti-emetics.
 - o **Vestibular rehabilitation.**
 - Should be initiated as early as possible to promote central compensation.
 - Full recovery may requires weeks to months.

Labyrinthitis:

- Vestibular and auditory dysfunction due to infection of labyrinth.
- Pathophysiology:
 - o Spread of infection from middle ear into inner ear through:
 - Round window
 - Annular ligament
 - Pre-existing labyrinthine fistula
 - Stapedotomy
- Types:
 1. Serous labyrinthitis
 2. Suppurative labyrinthitis
- **Serous Labyrinthitis:**
- Diffuse intra-labyrinthine inflammation secondary to viral infection without pus formation.
- Reversible condition if treated early.
- Clinical features
 - o **Sudden severe vertigo**
 - o **Sudden SNHL**
 - o **Nystagmus towards affected ear (irritative)**
- Management:
 - o **Steroids**
 - o **Supportive**
- **Suppurative Labyrinthitis:**
- Diffuse bacterial invasion of the labyrinth with permanent loss of vestibular and cochlear functions.
- Clinical features
 - o **Sudden severe vertigo**
 - o **Sudden SNHL**
 - o **Nystagmus away from affected ear (paralytic)**
 - o **Meningitis**
 - o **Fever**
- Management:
 - o **Steroids**
 - o **IV antibiotic**
 - o **Surgical management of middle ear infection (if indicated)**

Labyrinthine Fistula:

- Defect in the bony capsule of labyrinth.
- Part of membranous labyrinth is exposed and becomes sensitive to pressure changes.
- Locations:
 - **Otic capsule**
 - **Oval window**
 - **Round window**
- Causes:
 - **Cholesteatoma:**
 - Most common locations:
 - Horizontal SCC (75%)
 - Oval window
 - Promontory
 - **Temporal bone fracture**
 - **Stapes surgery**
 - **Barotrauma**
 - **Acoustic trauma**
 - **Mondini malformation**
- Clinical features:
 - **SNHL (70%):**
 - No specific audiometric pattern.
 - SNHL vary from isolated high-frequency loss to low-frequency or flat one.
 - **Transient Vertigo (60%):**
 - Induced by:
 - Valsalva maneuver.
 - Pressure-induced (**Fistula test**)
 - Noise-induced (**Tullio phenomenon**).
 - **Nystagmus:**
 - Quick component towards the affected ear (irratative).
 - **CSF otorrhea**
- Diagnosis (No definitive tests):
 - **Fistula test:**
 - **50% sensitivity to detect labyrinthine fistula.**
 - Increases air pressure in the ear canal (tragal pressure or pneumatic otoscopy) stimulates the labyrinth.
 - Causes vertigo and nystagmus.
 - **CT temporal bone:**
 - **60% sensitivity to detect labyrinthine fistula.**
 - Signs:
 - Otic capsule bony defect
 - Presence of pneumolabyrinth.

- Treatment

○ **Surgical exploration for possible cases:**

▪ Positive exploration:

- Diagnosed intra-op by:
 - Presence of bony defect
 - Perilymph leak
 - Staging:
 - **Type I:** Bony erosion but intact endosteum
 - **Type IIa:** Violated endosteum but intact perilymphatic space.
 - **Type IIb:** Violated perilymphatic space.
 - **Type III:** Violated membranous labyrinth
 - Treated by:
 - Patching with tissue graft (bone pate and fascia) can achieve hearing preservation and relief of vertigo.
 - If caused by cholesteatoma:
 - CWD mastoidectomy
 - Complete removal of cholesteatoma matrix over the fistula with patching of bony defect
- EXCEPT the following (High risk of SNHL):**
- **Firmly adherent**
 - **Infection**
 - **Size >2mm**
 - **Tympanium (promontory)**
 - **Only hearing ear**
 - **LA**

▪ Negative exploration:

- Reasons:
 - No fistula exists
 - Intermittent fistula
 - Too small fistula to be detected.

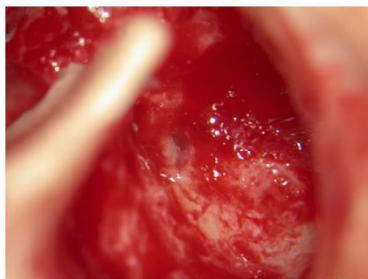


Fig. 2. Intraoperative picture of a horizontal canal fistula.

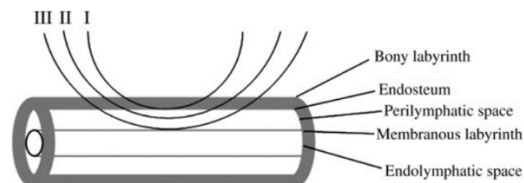
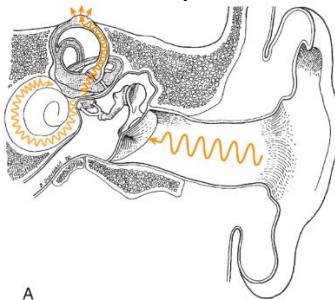


Fig. 3. A fistula staging system. (*Adapted from Dornhoff JL, Milewski C. Management of the open labyrinth. Otolaryngol Head Neck Surg 1995;112(3):410-4.*)

- **Medical care:**
 - Bed rest with head elevation.
 - Avoidance of any straining (sneezing with mouth open, stool softeners) for 5–10 days
 - Systemic antibiotic:
 - Prevent spread of infection into the labyrinth.
 - Systemic steroid:
 - Protective effect on hearing.

Superior Canal Dehiscence Syndrome (SCDS/ Minor's Syndrome):

- Absence of bone over the superior SCC.
- Mean age range at time of diagnosis is 40s.
- Causes:
 - Congenital dehiscence
 - Increased CSF pressure
 - Post-traumatic
- Pathophysiology:
 - Creates "**Third-window effect**" into the inner ear.
 - Under normal circumstances sound pressure enters inner ear through stapes footplate in the oval window and passing around the cochlea to exit through round window.
 - Presence of dehiscence in superior SCC allows abnormal movement of endolymph during presentation of loud sounds, tragal compression or Valsalva maneuver.
 - Lead to:
 - Decreased sensitivity to air conducted sound
 - Increased sensitivity to bone conducted sound



A



FIGURE 163-19. **A,** In superior canal dehiscence syndrome, sound waves can excite the superior canal, because the "third mobile window" created by the dehiscence allows some sound pressure to be dissipated along a route through the superior canal in addition to the conventional route through the cochlea. **B,** Computed tomography scan demonstrates dehiscence (arrows) of the superior canal. (**A,** Courtesy Dr. B. Dunham.)

- Clinical features:
 - **Transient Vertigo (97%):**
 - Induced by:
 - Valsalva maneuver.
 - Pressure-induced (**Fistula test**)
 - Noise-induced (**Tullio phenomenon**).
 - **CHL:**
 - Not due to middle ear pathology.
 - Caused by the third-window effect that causes dissipation of acoustic energy transmitted through AC mechanisms.
 - **Nystagmus:**
 - **Quick component Away from affected ear (paralytic).**
 - **Autophony:**
 - Resulted from increased sensitivity to bone-conducted sounds, due to the low impedance for bone-conducted sound to enter the inner ear at the dehiscence.
 - Sensation of increased loudness of patient's own voice
 - Hearing eye movements or blinking.
 - Hearing footsteps loudly.
 - Differentiated from Autophony resulted from Patulous Eustachian Tube by:
 - **NOT hearing own breathing sounds.**
 - **Continuous from the time of onset.**
 - **TM not moving during breathing.**
 - **Pulsatile tinnitus:**
 - Sensation of hearing patient's own pulse (Autophony).
- Diagnosis:
 - **Otoscope:**
 - Normal examination.
 - NO movement of TM synchronous with nasal breathing as seen with patulous eustachian tube.
 - **Fistula Test:**
 - **Nystagmus with quick component Away from affected ear (paralytic).**

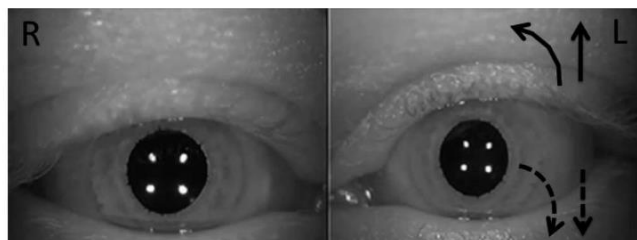
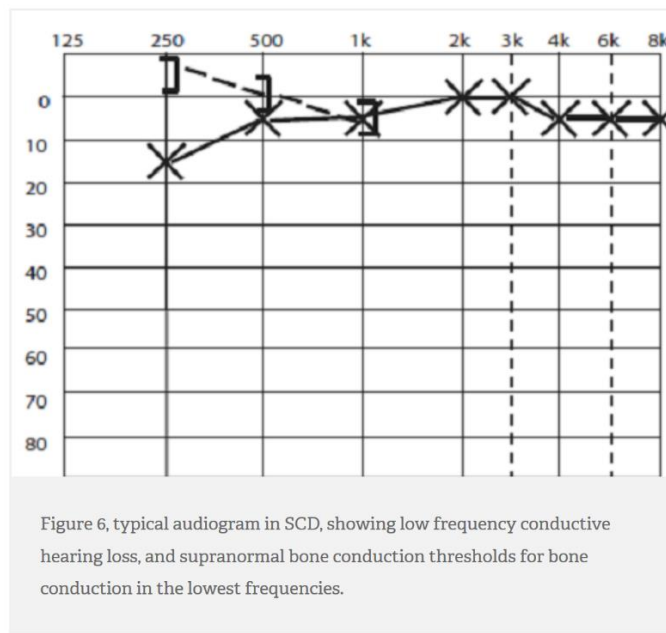


Fig. 1. Hennebert's sign associated with left superior semicircular canal dehiscence syndrome as seen under video-oculography goggles. The video with audio is available online. Positive pressure in the left ear causes a conjugate vertical-torsional ocular deviation where the eyes rotate up and away from the left ear (solid arrows). This reverses with negative pressure (dotted arrows). These eye movements align in the plane of the left superior semicircular canal. R = right; L = left.

- **Tuning fork:**
 - Weber tuning fork test lateralizes to affected ear.
 - Patients may hear a tuning fork placed on the lateral malleolus of the foot.
- **PTA:**
 - Bone-conduction thresholds can be below 0 dB (supra-normal BC threshold).
 - ABG can exist even when air conduction thresholds are normal (ALWAYS request for masked BC when suspecting SCDS).
 - A low Frequency CHL (250 to 1,000 Hz).
 - Moderate CHL (mimicking otosclerosis).
 - Normal discrimination



- **Tympanometry:**
 - Should be normal in SCDS.
 - Important to differentiate it from CHL due to otosclerosis or ossicular fixation, middle ear fluid,
- **Stapedial reflex:**
 - Should be normal in SCDS.
 - Important to differentiate it from CHL due to otosclerosis where reflexes are absent.
 - Patients with intact stapedial reflex and an ABG on PTA should undergo high resolution temporal CT scan before proceeding with surgical exploration of middle ear.

- **CT temporal bone:**
 - Diagnosis should never be based on CT scan alone due to false positive results.
 - Specificity and positive predictive value are improved with high resolution thin-cuts CT scan (0.5 mm) on the following planes:
 - Poschl:
 - Parallel to superior SCC.
 - Stenver's:
 - Perpendicular to superior SCC.

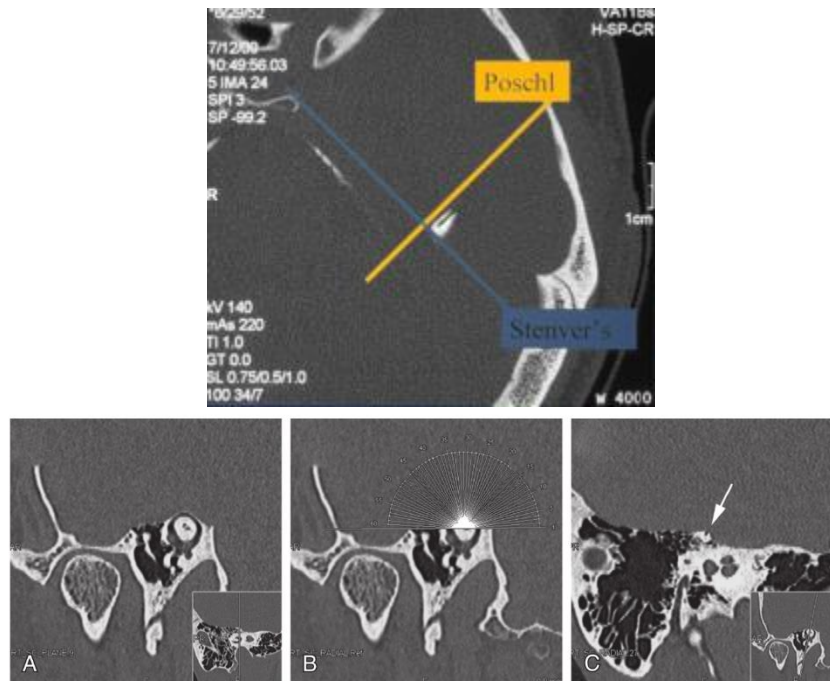


FIGURE 165-8. Computed tomography (CT) of the right superior semicircular canal demonstrates dehiscence. **A**, CT is reconstructed in the plane of the torus of the right superior canal (SC). **B**, The center of the SC is identified, and radial orthogonal reconstructions are performed at 3-degree intervals. **C**, The radial reconstructions are sequentially examined along the course of the SC, and dehiscence in the canal is identified if bone does not appear to completely cover the canal (white arrow). Inset images in **A** and **C** show the perpendicular view.

- **Cervical Vestibular Evoked Myopotentials (cVEMP):**
 - The "third mobile window" associated with dehiscence creates low impedance pathway that increases sensitivity of vestibular receptors to sound and pressure stimuli.
 - Patients with SCDS will have low threshold response to sounds.

- Treatment

- **Observation:**
 - For patients with mild symptoms.
 - Avoidance of the stimuli that evoke these effects.
- **Ventilation tubes :**
 - May be beneficial in patients with pressure-induced symptoms.
- **Surgical Repair:**
 - Indicated for patients with severe symptoms.
 - Types:
 - Reinforcement of oval and round window:
 - Trans-canal approach.
 - Minimally invasive procedure.
 - Using fascia to reinforce and dampening of oval and round window.
 - May alleviate symptoms in some patients.
 - Risk of recurrent symptoms.
 - Resurfacing of the bony defect:
 - Middle cranial fossa approach.
 - Eliminate the third mobile inner ear window.
 - Using bone cement and fascia to resurface the bony plate of middle fossa.
 - Preserve the physiological function of SCC.
 - Risk of recurrent symptoms from slippage of fascia or bone graft out of the place.
 - Less effective than plugging.
 - Plugging of superior canal:
 - Middle cranial fossa approach.
 - Eliminate the third mobile inner ear window.
 - Using fascia and bone graft to obliterate the superior SCC.
 - More affective and reliable.
 - Vestibular function will be lost.
 - Risk of SNHL.
 - Combined plugging and resurfacing:
 - Most effective approach.
 - Effective in alleviating vestibular symptoms and closing the ABG and normalization of cVEMP.
 - Low risk of recurrence.
 - Trans-mastoid approach:
 - Alternative approach for SCDS, provided that the skull base / tegmen is not too low and that no evidence for large skull base defects or brain sagging into the ear.

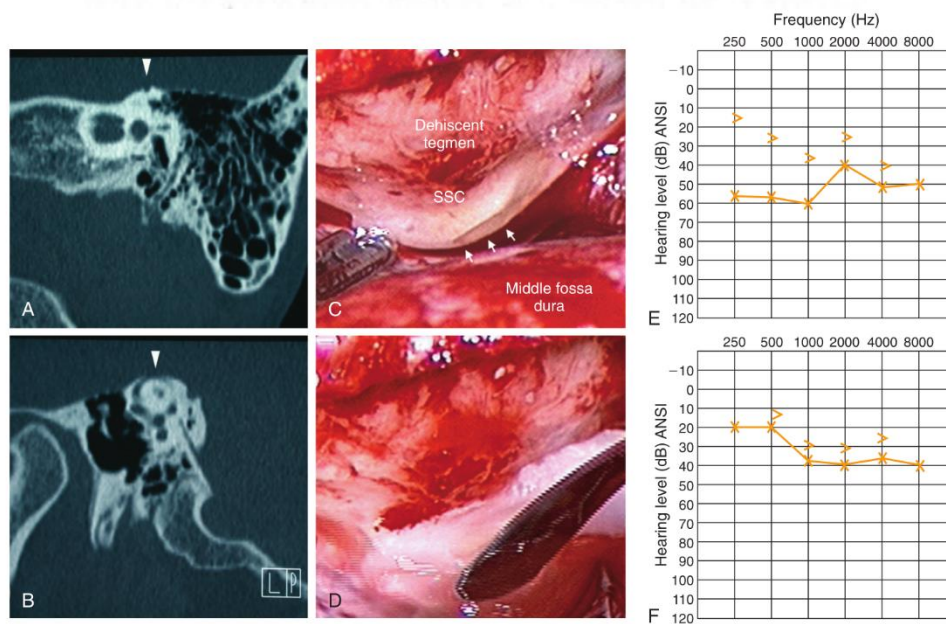
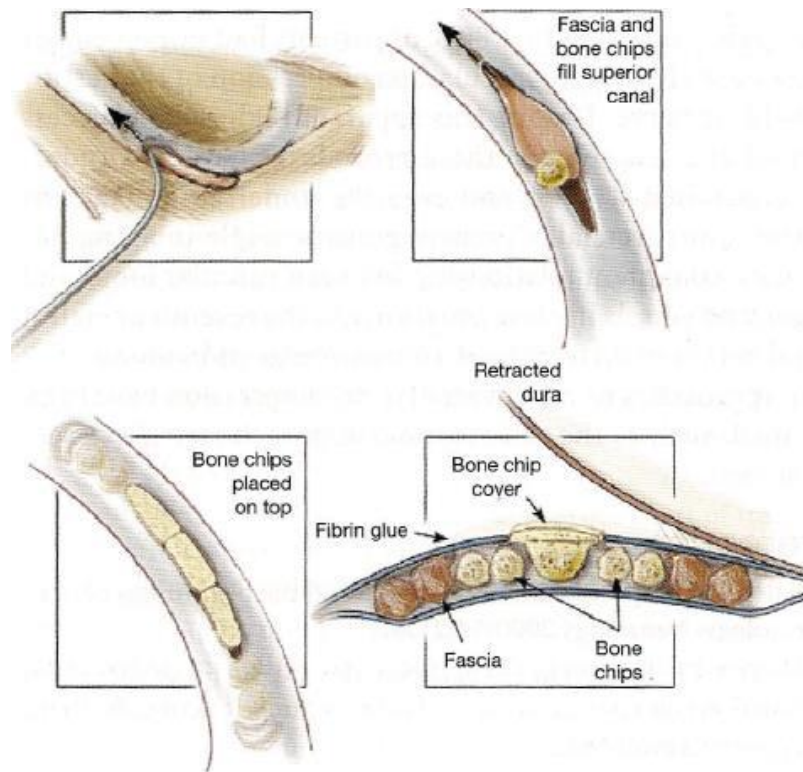


FIGURE 129-7. Inner ear impedance changes because of superior canal dehiscence are associated with an air-bone gap on audiometric testing. **A** through **F**, Clinical images from a 28-year-old woman with autophony, hearing loss, and sound- and pressure-induced oscillopsia and dizziness. Left-sided superior canal dehiscence (SCD) syndrome was diagnosed. **A** and **B**, Temporal bone computed tomography scans reformatted to Stenver (perpendicular to the superior canal, **A**) and Pöschl (parallel to the superior canal, **B**) views of the left ear show a 4-mm dehiscence (arrowheads). **C** and **D**, Intraoperative photos of left SCD during middle fossa craniotomy and dural retraction. Note the exposed membranous labyrinth of the superior canal and surrounding dehiscence tegmen in **C** that correlates with radiology in **A** and **B**. Plugging of the left SCD with bone wax is shown in **D**. Temporalis fascia and a split-calvarial bone graft were also used to reconstruct the dehiscence middle fossa floor. Preoperative audiogram (**E**) and post-SCD plugging audiogram (**F**) demonstrate closure of air-bone gap associated with SCD. SSC, superior semicircular canal. ANSI, American National Standards Institute. (Modified from Watters KF, Rosowski JJ, Sauter T, Le DJ. Superior semicircular canal dehiscence presenting as postpartum vertigo. *Otol Neurotol* 2006;27[6]:756-768.)

Trauma:

- Trauma can cause peripheral vertigo by:
 - Labyrinthine concussion
 - Temporal bone fracture
 - Penetrating trauma
 - Blast trauma
 - Barotrauma
- **Labyrinthine Concussion:**
- Post-traumatic disorders of inner ear function.
- NO violation of otic capsule or intra-labyrinthine membranes.
- Short lived symptoms that gradually subside over days to weeks.
- May result in residual BPPV-like symptoms.
- Clinical features:
 - Self-limiting acute vertigo.
 - Imbalance.
 - Nystagmus to affected side.
 - Recoverable SNHL
 - Tinnitus.
 - Normal otoscopic findings
 - Normal radiologic findings
- Management:
 - Reassurance.
 - Observation
 - Mild sedatives
- **Blast trauma:**
- Explosive blasts can produce pressure waves > 200 dB sound-pressure level which cause injury to the inner ear.
- Includes:
 - Open-handed slap to the ear
 - Explosions.
- Lead to:
 - TM Perforation
 - Ossicular disruption
 - Inner ear damage
- SNHL loss is common and commonly recovers spontaneously.
- Dizziness present in 15% of patients, and it is thought that the otoliths may be especially vulnerable.

- **Barotrauma:**
- Inner ear damage secondary to pressure changes.
- Common injury in divers.
- Types:
 - **Alternobaric trauma:**
 - Produced by elevated or asymmetric middle ear pressure.
 - Seen in:
 - Divers while in ascent to the surface.
 - Fliers during ascent of the aircraft.
 - Secondary to ET dysfunction.
 - URTI is a predisposing factor (affect ET patency)
 - Symptoms (self-limiting for 10-15 minutes):
 - Hearing loss
 - Tinnitus
 - Vertigo
 - Prevented by:
 - Frequent equilibration of ME pressure.
 - Use of topical decongestants.
 - Avoidance of diving or flying during URTI.
 - **Atmospheric barotrauma:**
 - Produced by either:
 - Extremes of pressure
 - Abrupt change in air pressure.
 - Symptoms (hours to days):
 - Hearing loss (HF-SNHL)
 - Tinnitus
 - Vertigo (35%)
 - Treatment:
 - Conservative with bed rest, head elevation and close monitoring of symptoms.
 - Symptoms spontaneously resolve in hours to days.
 - **Inner ear decompression sickness (IEDS):**
 - Produced by bubble formation within labyrinth or its blood supply as consequence of "oxyhelium" use for deep-water diving.
 - The most common decompression injury experienced by individuals diving to depths > 100m.
 - Symptoms (Permanent if treatment is delayed):
 - Vertigo (Mainly)
 - Hearing loss
 - Tinnitus
 - Treatment:
 - Hyperbaric oxygen within 24 hours after the injury.

Vestibulotoxic Drugs:

- Several drugs cause ototoxicity by damaging hair cells of inner ear.
- Examples:
 - **Aminoglycoside:**
 - Mainly streptomycin, gentamicin, kanamycin.
 - Cisplatin.
 - Antihypertensives
 - Labyrinthine sedatives
 - Estrogen preparations
 - Diuretics
 - Antimicrobials (nalidixic acid, metronidazole)
 - Antimalarials

Cogan Syndrome:

- Autoimmune process causing hydrops similar to Ménière's Disease.
- Disease progresses over months
- Clinical features (Triad):
 - **Interstitial keratitis:**
 - Blurriness
 - Rapidly progresses to blindness)
 - **Episodic vertigo.**
 - **Bilateral fluctuating SNHL**
- Diagnosis:
 - Clinical history
 - Physical exam
 - Elevated ESR and CRP.
 - Non-reactive syphilis tests.
- Management:
 - High dose oral corticosteroids 1mg/kg daily (usually resolves hearing and vestibular dysfunction)
 - May consider cyclophosphamide and methotrexate.

Vogt-Koyanagi-Harada Syndrome

- Similar to Cogan syndrome (SNHL, vertigo).
- Associated with:
 - Granulomatous Uveitis.
 - Depigmentation of hair and skin
 - Aseptic meningitis
 - Loss of eyelashes.

Otosyphilis

- Syphilis is sexually transmitted disease that can disseminated and affect any organ system.
- Types:
 - **Congenital:**
 - 30% will have inner ear involvement.
 - **Acquired:**
 - 80% will have inner ear involvement.
- Timing of onset:
 - **Early syphilis:**
 - Symptoms occur within 2 years of exposure
 - Vestibular symptoms are less frequent.
 - Most common otosyphilis manifestation is sudden hearing loss.
 - **Late syphilis:**
 - Symptoms occur after 2 years of exposure.
 - Otosyphilis presentation is similar to Meniere disease with episodes of vertigo combined with progressive hearing loss and tinnitus that is often unilateral.
 - Interstitial keratitis is found in 90% of patients with late-onset otologic syphilis.
 - Hutchinson's triad in late congenital syphilis:
 - Interstitial keratitis
 - SNHL
 - Notched incisors
- Diagnosis:
 - Hennebert's sign
 - Lab tests:
 - Non-treponemal tests (70% sensitivity for otosyphilis):
 - VDRL
 - Rapid plasma regain test
 - Treponemal tests (95% sensitivity for otosyphilis):
 - Fluorescent treponemal antigen absorption test [FTA-ABS].
 - Micro-hemagglutination assay for Treponema pallidum antibodies [MHA-TP].
- Management:
 - Penicillin IM
 - Prednisone

Acoustic neuroma:

- Classified in peripheral vestibular disorders as it arises from CN VIII within internal acoustic meatus.
- Causes only unsteadiness or vague sensation of motion.
- Severe episodic vertigo is usually missing.
- Other tumors of temporal bone (e.g. glomus tumour, carcinoma of external or middle ear and secondaries), destroy the labyrinth directly and cause vertigo.

**TABLE
166.1**

BASIC PRESENTATIONS OF PERIPHERAL VESTIBULAR DYSFUNCTION

1. Episodic disruption of unilateral vestibular function
 - Lasting minutes-hours*
 - Ménière's disease
 - Perilymph fistula
 - Lasting >24 h*
 - Vestibular neuritis
 - Labyrinthitis
 - IMIED
 - Lasting variable periods of time*
 - Migrainous vertigo
2. Brief/episodic excitation of unilateral vestibular function
 - BPPV
 - SCD syndrome
 - Perilymph fistula
3. Chronically inadequate vestibular function
 - Unilateral*
 - Unilateral vestibular hypofunction following vestibular neuritis, trauma, etc.
 - Acoustic neuroma
 - Bilateral*
 - Aminoglycoside toxicity
 - Chemotherapy
 - Familial

Central Vestibular Disorders:

Vestibular Migraine/ Migraine-Associated Vertigo:

- Migraine is characterized by recurrent headaches associated with:
 - o Nausea and vomiting
 - o Hypersensitivity to light, sound and smell.
- Migraine affects 25% of women, 15% of men and 5% of children.
- Typically begins in young adulthood.
- Vertigo is extremely common in patients with migraine **(25%)**.
- Pathophysiology of vestibular migraine remains unclear.
- Patients often report a prior history of migraine headaches that seem to have resolved.

Box 166-1. DIAGNOSTIC CRITERIA FOR MIGRAINE

Migraine Without Aura

- A. At least five attacks that fulfill B through D
- B. Headache lasting 4 to 72 hours when untreated
- C. Headache has at least two of the following characteristics:
 - 1. Unilateral location
 - 2. Pulsating quality
 - 3. Moderate or severe intensity
 - 4. Aggravation by physical activity
- D. During the headache, either nausea and vomiting or photophobia
- E. Not attributed to another disorder

Typical Aura with Migraine Headache

- A. At least two attacks that fulfill B through D
- B. Fully reversible visual, sensory, or aphasic aura symptoms with no motor weakness
- C. At least two of the following characteristics are present:
 - 1. Homonymous positive features or unilateral sensory symptoms
 - 2. At least one symptom that develops gradually over more than 5 minutes
 - 3. Symptom duration of 5 to 60 minutes
- D. Headache follows aura with a headache-free interval of less than 60 minutes
- E. Not attributed to another disorder

**Box 166-2. CRITERIA FOR DIAGNOSIS
OF VESTIBULAR MIGRAINE**

Vestibular Migraine*

- A. At least five episodes of vestibular symptoms of moderate or severe intensity lasting 5 minutes to 72 hours
- B. Current or prior history of migraine according to the International Classification of Headache Disorders (ICHD)
- C. One or more migraine features with at least 50% of the vestibular episodes; may include migraine headache, photophobia or phonophobia, and visual aura
- D. Not better accounted for by another vestibular or ICHD diagnosis

Probable Vestibular Migraine

- A. At least five episodes of vestibular symptoms of moderate or severe intensity occur that last 5 minutes to 72 hours
- B. Only one of criteria B and C above is fulfilled (i.e., migraine history or migraine features during the episode)
- C. Symptoms are not better accounted for by another diagnosis

*Vestibular symptoms rated as "moderate" mean they interfere with but do not prohibit daily activities. They are rated as "severe" if daily activity cannot be continued.

- Clinical picture:

- Vertigo does not usually present as an aura immediately preceding the headache, as a prodrome.
- Dizziness is described as vertigo (spinning, rocking, swaying) or simply disequilibrium.
- Variable in duration:
 - Lasting minutes to days in episodic cases, or constant disequilibrium lasting months.
- **When to suspect Migraine-Associated Vertigo:**
 - Dizziness associated with photophobia, menstrual association, and nasal stuffiness at time of the attack.
 - No history of hearing fluctuation or positional component.
 - Spells of vertigo or disequilibrium last more than a day.
 - Positive family history of migraines.

- Treatment:

- **Dietary and lifestyle modifications:**
 - Regular sleep and exercise.
 - Avoid trigger factors:
 - Stress
 - Weather changes
 - Food:
 - Red wine, aged cheese, yogurt, caffeine and chocolate.

- **Medical management:**
 - Prophylactic medications:
 - Goal is to reduce symptom frequency and severity by 50-70%.
 - Benefits are seen after 6-8 weeks of therapy.
 - **Beta blockers:**
 - Propranolol
 - **Calcium channel blockers:**
 - Nifedipine or Verapamil
 - **Antidepressants:**
 - Nortriptyline or venlafaxine
 - **Anticonvulsants:**
 - Sodium valproate or gabapentin
 - Abortive medications:
 - Should not be used > 6-8 times per month due to their known rebound effects
 - **Sumatriptan**

Treatment of Migraine-Associated Vertigo

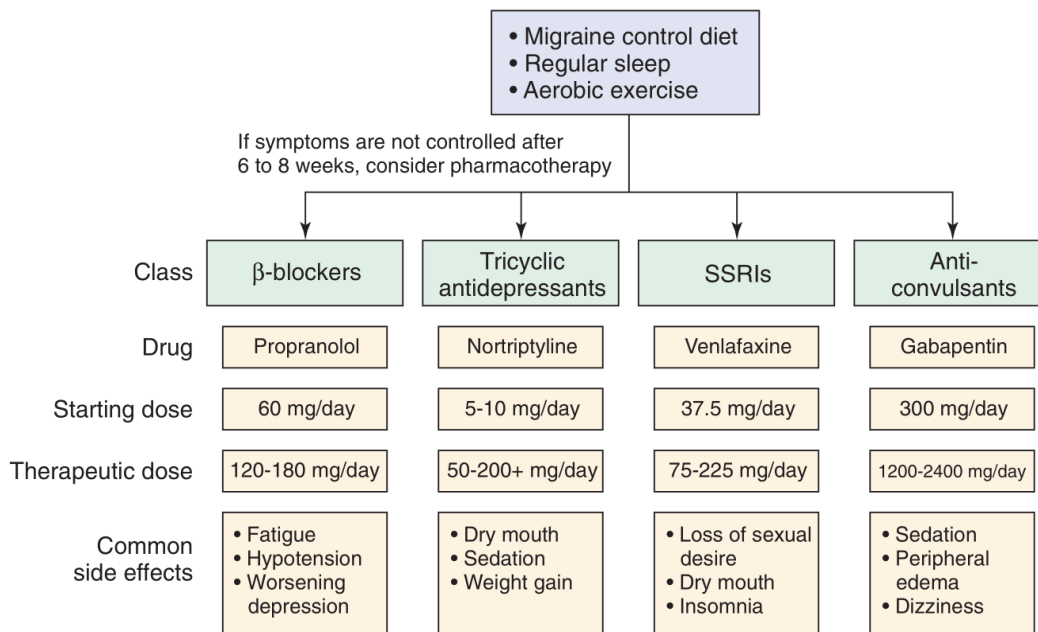


FIGURE 166-1. Flowchart of suggested migraine treatment. Lifestyle modification is usually a good first option, because it is not associated with side effects and will provide symptomatic improvement in most patients. Pharmacotherapy should be considered if more conservative measures fail. SSRI, selective serotonin reuptake inhibitor.

Vertebrobasilar insufficiency (VBI):

- Common cause of central vertigo in patients over age of 50 years.
- Pathophysiology:
 - o Transient decrease in cerebral blood flow due to Atherosclerosis.
 - o Compression of vertebral artery compromises flow to the posterior (PICA) and Anterior Inferior cerebellar Arteries (AICA).
- Precipitated by:
 - o Hypotension
 - o Neck Hyperextension or excessive rotation.
- Clinical picture:
 - o **Abrupt onset of (lasting for minutes):**
 - Vertigo
 - Nausea and vomiting
 - Headache
 - Diplopia
 - Ataxia
 - Numbness
 - Weakness
 - Dysphagia
- Subclavian Steal Syndrome:
 - o Occlusion or stenosis of the subclavian artery proximal to the vertebral artery.
 - o Results in reverse flow of the vertebral artery in favor of the ipsilateral arm.
 - o Symptoms are precipitated by exercise of upper extremities.
- Diagnosis:
 - o Clinical history and physical exam
 - o Radiography (CT and MRI brain).
- Management:
 - o Anticoagulation (antiplatelet medication)

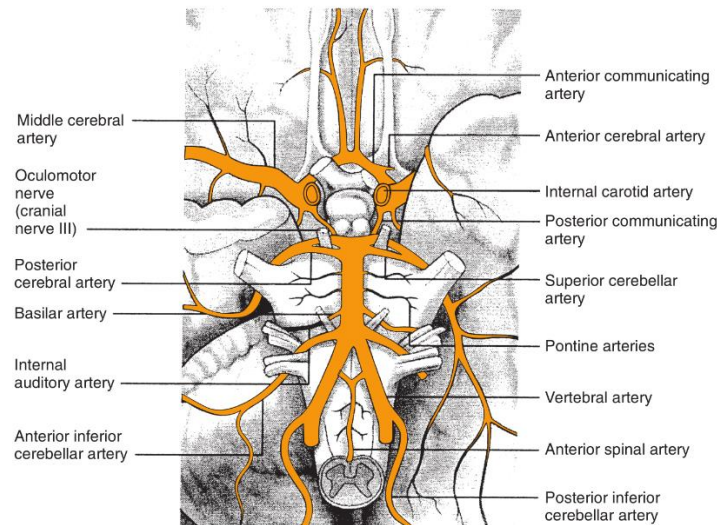
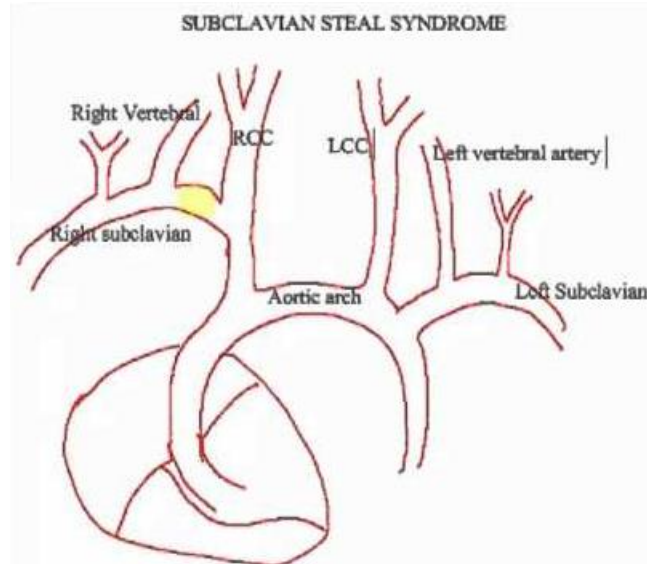


FIGURE 166-2. Blood vessels that supply the brain. The brainstem is supplied by branches of the vertebral and basilar arteries. Note that although this figure shows the internal auditory artery arising directly from the basilar artery, it is commonly a branch off of the anterior inferior cerebellar artery instead. (From Kandel ER, Schwartz JH, Jessell TM: *Principles of neural science*, New York, 2000, McGraw-Hill, p 1303.)



Lateral Medullary Syndrome (Wallenberg's syndrome):

- Posterior Inferior Cerebellar Artery (PICA) Syndrome.
- Pathophysiology:
 - Embolic event or thrombosis of ipsilateral vertebral or Posterior inferior cerebellar artery (PICA).
 - Results in an infarction of lateral medullary region of brain stem (serving CN V-X, Cerebellum, and sympathetic ganglion), spares cochlear nucleus.
- Clinical features:
 - 1. Ipsilateral loss of facial sensation:** (CN-V)
 - 2. Ipsilateral loss of lateral rectus:** (CN-VI)
 - 3. Ipsilateral loss of facial musculature:** (CN-VII)
 - 4. Acute vertigo and nystagmus:** (Vestibular CN-VIII).
 - 5. Ipsilateral palatal paresis (dysphagia):** (Pharyngeal plexus, Nucleus Ambiguus, CN-IX, CN-X).
 - 6. Ipsilateral vocal fold paralysis (Dysphonia):** (Pharyngeal plexus, Nucleus Ambiguus, CN-IX, CN-X).
 - 7. Ataxia:** Incoordination of ipsilateral limbs (falls toward lesion).
 - 8. Ipsilateral horner syndrome:** (Anhydrosis, Ptosis, Miosis) from damage to Preganglionic sympathetic fibers.
 - 9. Contralateral loss of pain and temperature:** Injury to the crossed spino-thalamic fibers
- Diagnosis:
 - Cerebral Angiogram, CT Brain (wedge-shaped infarct)
- Management:
 - As per cerebrovascular accident protocols.

Cerebellar disease:

- Affected by:
 - Hemorrhage (hypertension)
 - Infarction (occlusion of arterial supply)
 - Infection (otogenic cerebellar abscess)
 - Tumours (glioma, teratoma or haemangioma).
- Clinical features:
 - Severe vertigo
 - Vomiting
 - Ataxia simulating an acute peripheral labyrinthine disorder.
 - Incoordination
 - Past-pointing
 - Adiadokokinesia
 - Rebound phenomenon
 - Wide-based gait.

Multiple sclerosis

- Demyelinating disease of CNS.
- Affecting young adults.
- Result in "plaques" within the central vestibular system.
- Clinical features:
 - Vertigo and dizziness:
 - Blurring or loss of vision
 - Diplopia
 - Dysarthria
 - Paraesthesia
 - Ataxia.
 - Spontaneous nystagmus may be seen.
 - Acquired pendular nystagmus, dissociated nystagmus and vertical upbeat nystagmus are important features in diagnosis.

Tumors of brainstem and floor of IVth ventricle:

- Gliomas, astrocytomas may arise from pons and midbrain.
- Medulloblastoma, epidermoid cysts or teratomas may arise from floor of IVth ventricle.
- Cause vertigo and dizziness and other neurological signs and symptoms.
- CT scan and magnetic resonance imaging are useful in diagnosis.

Epilepsy

- Vertigo may occur as an aura in temporal lobe epilepsy.
- History of seizure and/or unconsciousness following the aura may help in the diagnosis.
- E.E.G. may show abnormalities during the attack.

Cervical Vertigo:

- Dizziness caused by disorders of neck and cervical spine is poorly understood and relatively uncommon.
- It is known that neck afferents have a role in the coordination of eye, head, and body spatial orientation.
- Perception of head rotation can be driven by vestibular, proprioceptive, or visual inputs.
- Cervical vertigo must be by definition proprioceptive in nature.
- Unilateral local anesthesia of cervical roots can cause ataxia without nystagmus in humans.
- Patients with chronic cervicobrachial neck pain have worse results on posturography tests.
- In clinical practice, it is necessary to exclude neurologic, vestibular, and psychosomatic disorders before a disorder of the craniovertebral junction can be given serious consideration.

Approach to Tinnitus:

- **Tinnitus:**
- Ringing sound or noise in the ear.
- Origin of this sound is within the patient.
- Usually unilateral but may also affect both ears.
- Vary in pitch and loudness.
- Described by the patient as roaring, hissing, swishing, rustling or clicking type of noise.
- Most prevalent from 40-70 yrs.
- Men > Women.

Table 1. Abbreviations and Definitions of Common Terms.

Term	Definition
Tinnitus	The perception of sound when there is no external source of the sound
Primary tinnitus	Tinnitus that is idiopathic ^a and may or may not be associated with sensorineural hearing loss
Secondary tinnitus	Tinnitus that is associated with a specific underlying cause (other than sensorineural hearing loss) or an identifiable organic condition
Recent onset tinnitus	Less than 6 months in duration (as reported by the patient)
Persistent tinnitus	6 months or longer in duration
Bothersome tinnitus	Distressed patient, affected quality of life ^b and/or functional health status; patient is seeking active therapy and management strategies to alleviate tinnitus
Nonbothersome tinnitus	Tinnitus that does not have a significant effect on a patient's quality of life but may result in curiosity of the cause or concern about the natural history and how it might progress or change

^aThe word *idiopathic* is used here to indicate that a cause other than sensorineural hearing loss is not identifiable.

^b*Quality of life* is the degree to which persons perceive themselves as able to function physically, emotionally, mentally, and/or socially.

1. Subjective Tinnitus:

- Perception of sound in the absence of any stimulation (Acoustic, Electrical, or External stimulation).
- Not audible to another person.
- Most common type.
- Typically associated with a high frequency HL (3000–5000 Hz)
- Pitch of tinnitus may correlate with the frequency of hearing loss.
- Pathophysiology is largely unknown.
- 25% improve spontaneously.
- **Otologic Causes:**
 - Presbycusis: very common (75%)
 - NISNHL: High freq SNHL most consistent factor with tinnitus
 - Meniere's: Almost all have tinnitus
 - Prolonged otitis
 - Recurrent labyrinthitis.
 - Otosclerosis
- **Drugs:**
 - ASA: Most common drug.
 - Mercury, Arsenic and Lead.
 - Caffeine.

TABLE 6–3. Differential Diagnosis of Subjective Tinnitus

Hearing Loss and Otologic Disorders

- Hearing Loss (Presbycusis, Autoimmune Hearing Loss)
- Retrocochlear Lesions (Acoustic Neuromas)
- Ménière's Disease

Medications: (see Table 6–1)

- Aspirin/NSAIDs
- Hypertensive Agents
- Aminoglycosides
- numerous medications have been reported to have the potential to cause tinnitus

Trauma

- Head Injuries (Whiplash)
- Loud Noise Exposure
- Barotrauma

Systemic Diseases

- Hypertension
- Depression and Anxiety
- Neurological Disease (Multiple Sclerosis, Brainstem Stroke)

Metabolic

Hypo- Hyper Thyroidism

Hyperlipidemia

Vit. deficiency (A,B, Zinc)

2. Objective Tinnitus:

- Perception of sound caused by an internal body sound or vibration.
- Audible to another person.
- Pulsating in character (Asking the patient to perform light physical activity may confirm the pulsatile nature).
- Exacerbated with a CHL.
- **Vascular Causes:**
 - Soft, rushing, pulsatile sound.
 - Synchronous with heartbeat.
 - Increase with exercise
 - < 10% of tinnitus.
 - Small percentage have bruit
 - Benign Intracranial Hypertension Syndrome, Atherosclerotic carotid artery disease and Glomus tumors compose two thirds of definable causes.

1. Pseudotumor Cerebri (Benign Intracranial Hypertension Syndrome):

- Most common cause of pulsatile tinnitus.
- Overweight females, 20–30 years old
- Increased ICP without focal neurological signs.
- Caused by systolic pulsations of CSF transmitted to dural venous sinuses.
- Diagnosed by MRI and LP to measure CSF pressure.
- May be associated with a mild SNHL.
- Rx: weight loss and diuretics.

2. Vascular Tumors:

- Most common is **Glomus Tympanicum or Jugulare.**
- Reddish/blue mass behind TM.
- Decreased hearing.
- Diagnosed with CT with contrast to look for erosion of carotico-jugular spine or lesion in ME space.

3. Venous Hum:

- Soft and low-pitched.
- Caused by turbulent blood flow through the jugular bulb or transverse sinus.
- Found commonly in patients with:
 - High jugular bulb.
 - HTN
- Tinnitus typically disappears in:
 - Upright position:
 - Pressure over ipsilateral IJV.

4. Arterial Bruits:

- Whooshing sound synchronous with heartbeat.
- Transmitted sounds.
- Caused by:
 1. Apparent course of ICA in the middle ear.
 2. Atherosclerotic carotid artery disease.
 3. Vascular loops in internal auditory meatus pressing on CN-VIII.
- Diagnosed by CTA, MRA or Duplex U/S of carotids.

5. AV malformations:

- Intracranial or Preauricular.
- Connections between Occipital artery and Transverse sinus, ICA and vertebral vessels, or middle meningeal artery and GSPA.
- Diagnosed by MRA.
- Rarely need Tx.

- Mechanical Causes:

1. Patulous Eustachian Tube:

- Abnormally patent Eustachian tube.
- Ocean roar in ear synchronous with Nasal Respiration.
- Absent on lying flat.
- Autophony (Hyperacusis to one's own speech and bodily sounds).
- Hypermobility TM moves with respiration.
- Causes:
 - Post-radiation patients
 - After significant weight loss
 - Stroke.
 - Injury to CN-V
 - Iatrogenic injury to Tensor veli palatini in Cleft palate surgery.
- Rx: Mucosal irritants, Eustachian diathermy probe (often requires a pressure equalization tube), may consider Teflon paste injection into the Torus tubaris (less effective).

2. Palatal Myoclonus:

- Rapid clicking sound caused by the contraction of Palatal muscles and ET.
- May be evaluated by Nasopharyngoscopy in an awake patient.
- Treat with Botox injections, Antiepileptics or muscle relaxants (Clonazepam, Diazepam).

3. Tensor Tympani/ Stapedius Syndrome:

- Spasm or myoclonus of Tensor tympani or stapedius muscle.
- Fluttering, low frequency tinnitus.
- Accentuated by External sound.
- Dx: Tinnitus synchronous with TM movement.
- Rx: Avoidance of Stimulants, Reassurance, rarely requires section of the tensor tympani muscle.

4. Spontaneous Otoacoustic Emissions:

- Rare cause of objective tinnitus

- Evaluation of pulsatile tinnitus:

○ **Retro-tympanic mass on otoscope:**

- Request contrast CT to evaluate for :
 - Glomus tumors
 - Aberrant carotid artery
 - Jugular bulb abnormalities

○ **No Retro-tympanic mass on otoscope:**

- If tinnitus improves with upright position or ipsilateral IJV pressure:
 - Venous hum.
 - Doesn't require further evaluation.
- If tinnitus doesn't improve with upright position or ipsilateral IJV pressure:
 - Request MRI/MRA to evaluate for:
 - BIH.
 - Vascular loops.
 - AV malformations.
 - Request Carotid Duplex US to evaluate for:
 - Atherosclerotic carotid artery disease
 - Carotid duplex ultrasonography.

Approach to Tinnitus:

- Similar approach to Hearing loss with special attention to:
 - Character of Tinnitus:
 - Pulsatile or Non-pulsatile.
 - Unilateral or Bilateral.
 - High-pitched (Ringing, Hissing) or Low-pitched (Roaring, Buzzing).
 - Progression and Frequency
 - Level of discomfort (Difficulty with sleeping)
 - Complete H&N examination including stethoscope to auscultate for objective tinnitus.
- Work-up:
 - **Audiology evaluation:**
 - Diagnostic testing should include PTA, speech discrimination testing, and tympanometry.
 - Indications:
 - Recommended indications:
 - 1. Unilateral tinnitus.**
 - 2. Associated with hearing difficulties.**
 - 3. Persistent tinnitus (≥ 6 months).**
 - Optional indications:
 - Any tinnitus.
 - **Imaging:**
 - Indications:
 - 1. Unilateral tinnitus:**
 - MRI with contrast to rule out VS.
 - 2. Asymmetric hearing loss:**
 - MRI with contrast to rule out VS.
 - 3. Pulsatile tinnitus:**
 - CT/CTA to evaluate vascular abnormalities (Glomus, apparent carotid in ME).
 - MRI/MRA to evaluate AVM or vascular loops.
 - 4. Focal neurological abnormalities:**
 - MRI/CT brain.

Table 6. Key Details of Medical History in the Tinnitus Patient.^a

Key Issue	Significance	Implication
Unilateral tinnitus	Concern for focal auditory lesions, some serious, such as VS or vascular tumor	Referral for comprehensive audiologic assessment and an otologic evaluation; additional testing such as imaging where indicated
Pulsatile tinnitus	Concern for vascular lesion, systemic cardiovascular illness	Consider cardiovascular and general physical examination (hypertension, heart murmurs, carotid bruits, venous hums); examination of the head and neck for signs of vascular tumors or other lesions; comprehensive audiology; imaging and other testing where indicated
Hearing loss	Tinnitus is frequently associated with hearing loss, particularly SNHL; differentiate between conductive and SNHL, unilateral and bilateral; establish severity of hearing loss	Referral for comprehensive audiology; otologic evaluation to look for the wide range of pathologies that could cause hearing loss associated with tinnitus; consider hearing aid evaluation when indicated
Sudden onset of hearing loss with tinnitus	Sudden hearing loss requires prompt treatment to stabilize or improve hearing.	See sudden SNHL guideline ⁴⁴
New onset tinnitus	Tinnitus perception may diminish or disappear, and/or tinnitus reactions may be reduced.	Evaluation and treatment is based on severity, and presence and absence of other symptoms
Noise exposure	Tinnitus may be associated with prolonged noise exposure from occupational or recreational activities.	Counseling and education related to potential damaging effect of noise, acoustic trauma, and pertinent environmental exposures; referral for comprehensive audiologic assessment
Medications and potential ototoxic exposures	Some medications such as salicylates are associated with tinnitus; ototoxins can cause hearing loss and tinnitus. Interactions between medications have unknown effects and can exacerbate tinnitus symptoms.	Counseling regarding medication use, etiology of tinnitus is facilitated; patients can be provided list of known ototoxic medications as part of counseling; comprehensive audiologic assessment
Unilateral or asymmetric hearing loss	Possible presentation of serious lesion such as VS	Audiologic and otologic assessment; imaging where indicated
Vertigo or other balance malfunction	Possible cochlear, retrocochlear, or other central nervous system disorder (Ménière's disease, superior canal dehiscence, VS, other)	Audiologic, otologic, vestibular assessment; imaging and referral where indicated
Symptoms of depression and/or anxiety	Tinnitus is often accompanied by symptoms of depression and anxiety. The presence and severity of such symptoms will dictate the pace of evaluation and treatment as well as the need for referral to treat these issues.	Referral to mental health professionals for assessment and treatment of depression and/or anxiety; urgent referral for suicidal patients
Apparent cognitive impairments	Elderly patients at risk for tinnitus are also at risk for cognitive decline from dementia.	The presence of dementia will affect the results of tinnitus and audiologic assessments.

Abbreviations: SNHL, sensorineural hearing loss; VS, vestibular schwannoma.

^aA definition of *comprehensive audiologic assessment* can be found in **Table 8**.

Table 7. Key Details of Physical Examination in the Tinnitus Patient.

Key Issue	Significance	Implication
Objective tinnitus	Rarely, tinnitus can be heard by the clinician as well as the patient.	Objective tinnitus may be caused by identifiable diseases, such as vascular abnormalities and myoclonus.
Heart murmurs, carotid bruits, or vascular sounds	Cardiovascular disease and vascular lesions may cause tinnitus.	Treatment of the underlying disease may help tinnitus symptoms. Cardiovascular disease (carotid stenosis, heart murmurs, hypertension) can have morbidities more substantial than tinnitus and requires appropriate evaluation and treatment.
Focal neurologic signs	Tinnitus patients should undergo neurologic assessment. Any focal neurologic deficits will dictate additional evaluation and treatment.	Referral to appropriate specialists (neurologists, otologists/neurotologists, head and neck surgeons, etc) and for appropriate workup, which may include imaging of the central nervous system
Otorrhea	Sign of middle ear infection or otitis externa	Treatment of otitis media/externa may improve tinnitus as well as associated hearing difficulties.
Signs of other external or middle ear disease on examination and/or otoscopy	Simple problems such as cerumen impaction or otitis media can be detected. Cholesteatoma, glomus tumors, and other uncommon middle ear disorders can be detected by otoscopy.	Appropriate referral can be made for diagnosis and treatment of external auditory canal issues such as cerumen, and middle ear disease such as otitis media or middle ear masses. Imaging can be performed when indicated.
Head and neck masses	A head and neck mass associated with ipsilateral tinnitus requires prompt investigation.	Referral to appropriate specialists; imaging when indicated

Table 10. Patient Education Discussion Points for Bothersome Tinnitus.

1. Definition of tinnitus	Tinnitus is sound that is created in the ears or in the head. It is a symptom and not a disease. People with chronic tinnitus usually hear it all or most of the time. For some people, tinnitus is intermittent.
2. Distinguishing tinnitus from transient ear noise (brief spontaneous tinnitus)	Transient ear noise is a sudden whistling sound accompanied by the perception of hearing loss. The event is unilateral and seems to occur completely at random without anything precipitating the sudden onset of symptoms. Often, the ear feels blocked during the episode. The symptoms generally dissipate within a period of about a minute. Transient ear noise, sometimes also called brief spontaneous tinnitus, is normal.
3. Assessment of tinnitus and associated hearing loss	Patients with tinnitus commonly attribute hearing problems to tinnitus. The clinician should determine how much of a patient's complaint is due to a hearing problem and how much is due specifically to the tinnitus. Such assessment may require an audiologic examination and appropriate questionnaires.
4. Tinnitus can be temporary	Exposure to loud noise can cause temporary threshold shift as well as temporary tinnitus. Tinnitus induced in this fashion will likely resolve within a few days following the insult. Repeated episodes of noise exposure increase the likelihood that the tinnitus will become permanent.
5. Drugs and tinnitus	Tinnitus can be induced by a number of medications and drug interactions. Such tinnitus is usually temporary (typically lasting 1 to 2 weeks postexposure) but can be permanent—especially with the use of aminoglycoside antibiotics or the cancer chemotherapeutic drug cisplatin. Aspirin is well known to cause temporary tinnitus, although the dosage generally has to be rather high to induce tinnitus. Other medications that can cause temporary tinnitus include nonsteroidal anti-inflammatory drugs, loop diuretics, and quinine. Drugs used to treat mental health and sleep conditions also may trigger or exacerbate tinnitus.
6. No cure for primary tinnitus	A cure for primary tinnitus does not yet exist, and despite claims to the contrary, no method has been proven to provide long-term suppression of tinnitus. We can help patients by relieving the functional effects of tinnitus, such as sleep disturbance, difficulty concentrating, problems with hearing, and difficulty relaxing. Patients need to be informed that although tinnitus cannot be cured, they can learn to manage their reactions to it, thereby improving their QOL. Health care professionals should be compassionate regarding patients' concerns and fears about tinnitus. A brief overview of the evidence-based interventions discussed later in this guideline can be presented.
7. Current theory on the pathophysiology of tinnitus	Research suggests that tinnitus results from the compensatory adaptation of the central auditory system to hearing loss. Clinical observations establish the near universal association of tinnitus with hearing loss. Hearing loss associated with tinnitus can range in severity from minimal to profound, and most people with hearing loss do not experience tinnitus. Changes in inhibitory and excitatory neurotransmitters occur throughout the auditory pathway in association with tinnitus.

Management:

1. Treat Reversible causes:

- Medications Review (ASA/NSAIDs)

2. Behavioral:

- Lifestyle modifications:
 - Smoking cessation
 - Avoidance of caffeine, chocolate, tea
 - Avoid loud noise
- Home-masking techniques:
 - White Noise at night.
 - Placing Radio between stations.
- Cognitive behavioural therapy (CPT):
 - Counseling/psychological therapy.
 - Biofeedback for stress reduction
 - Habituation:

3. Medical:

- Strong recommendations against its routine use.
- No medications approved by FDA for treatment of tinnitus.
- No medications or dietary supplements showed improvement in tinnitus perception.
- Examples:
 - Intra-tympanic steroid injections.
 - Antidepressants (Tricyclic antidepressants, SSRI).
 - Anxiolytics (Benzodiazepines).
 - Ginkgo Biloba
 - Melatonin
 - Zinc

4. Hearing Aids:

- Indicated for subjective tinnitus with HL.
- Reduces Tinnitus by amplifying ambient sound to mask Tinnitus.
- Simplest method of "Direct Masking".
- 25% improvement with severe tinnitus.

5. Masking Devices:

- Masking devices utilize a band of White Noise centered around the Tinnitus Frequency (Pitch matched).
- May be combined with Hearing Aids.
- Indicated for patients with persistent, bothersome tinnitus.

6. Habituation:

- Tinnitus retraining therapy.
- Stimulation with Broad-band noise up to 16 hours/day.
- Level is increased until it is audible but not mask the tinnitus
- Over months, Patients symptom improves

7. Surgery:

- Correct otologic conditions (Otosclerosis).
- CI

Table 5. Summary of Guideline Action Statements.

Statement	Action	Strength
1. History and physical exam	Clinicians should perform a targeted history and physical examination at the initial evaluation of a patient with presumed primary tinnitus to identify conditions that if promptly identified and managed may relieve tinnitus.	Recommendation
2A. Prompt audiologic examination	Clinicians should obtain a prompt, comprehensive audiologic examination in patients with tinnitus that is unilateral, persistent (≥ 6 months), or associated with hearing difficulties.	Recommendation
2B. Routine audiologic examination	Clinicians may obtain an initial comprehensive audiologic examination in patients who present with tinnitus (regardless of laterality, duration, or perceived hearing status).	Option
3. Imaging studies	Clinicians should not obtain imaging studies of the head and neck in patients with tinnitus, specifically to evaluate the tinnitus, unless they have 1 or more of the following: tinnitus that localizes to 1 ear, pulsatile tinnitus, focal neurological abnormalities, or asymmetric hearing loss.	Strong recommendation against
4. Bothersome tinnitus	Clinicians must distinguish patients with bothersome tinnitus from patients with nonbothersome tinnitus.	Strong recommendation
5. Persistent tinnitus	Clinicians should distinguish patients with bothersome tinnitus of recent onset from those with persistent symptoms (≥ 6 months) to prioritize intervention and facilitate discussions about natural history and follow-up care.	Recommendation
6. Education and counseling	Clinicians should educate patients with persistent, bothersome tinnitus about management strategies.	Recommendation
7. Hearing aid evaluation	Clinicians should recommend a hearing aid evaluation for patients with hearing loss and persistent, bothersome tinnitus.	Recommendation
8. Sound therapy	Clinicians may recommend sound therapy to patients with persistent, bothersome tinnitus.	Option
9. Cognitive behavioral therapy	Clinicians should recommend cognitive behavioral therapy to patients with persistent, bothersome tinnitus.	Recommendation
10. Medical therapy	Clinicians should not routinely ^a recommend antidepressants, anticonvulsants, anxiolytics, or intratympanic medications for a primary indication of treating persistent, bothersome tinnitus.	Recommendation against
11. Dietary supplements	Clinicians should not recommend Ginkgo biloba, melatonin, zinc, or other dietary supplements for treating patients with persistent, bothersome tinnitus.	Recommendation against
12. Acupuncture	No recommendation can be made regarding the effect of acupuncture in patients with persistent bothersome tinnitus.	No recommendation
13. Transcranial magnetic stimulation	Clinicians should not recommend transcranial magnetic stimulation for the routine ^a treatment of patients with persistent, bothersome tinnitus.	Recommendation against

^aThe words *routine* and *routinely* are used to avoid setting a legal precedent and to acknowledge that there may be individual circumstances for which clinicians and patients may wish to deviate from the prescribed action in the statement.

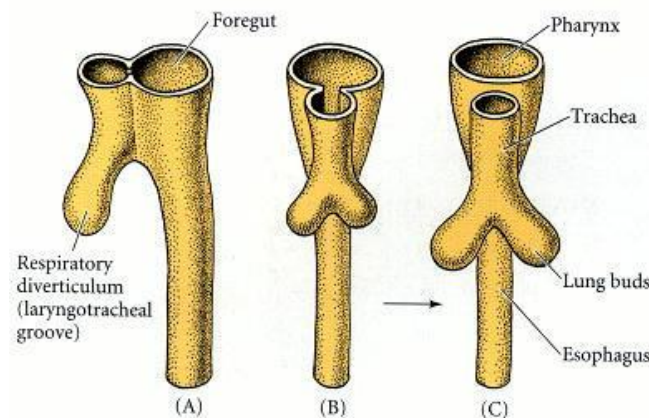
- **Hyperacusis:**
- Increase sensitivity to sound that wouldn't normally trouble normal individual.
- Painful at 40-50 dB.
- 25-40% association with tinnitus.
- Different from Recruitment – painful with loud sounds.
- May develop Overprotection (Overzealous earplug use) and Phonophobia.
- Therapy to wean off earplugs/earmuffs
- Tinnitus Retraining Therapy (TRT)
- High association with Williams Syndrome:
 - Rare Neurodevelopmental disorder.
 - Caused by a deletion of about 26 genes from Long arm of chromosome 7.
 - Characterized by a distinctive "Elfin" facial appearance, along with a low nasal bridge.
 - Idiopathic infantile hypercalcemia.
 - Mental retardation.
 - Cardiovascular anomalies:
 - Supravalvular aortic stenosis
 - Peripheral pulmonary artery stenosis

Laryngology:

- 1- Laryngotracheal Embryology, Anatomy and Physiology**
- 2- Voice Production and Dysphonia**
- 3- Vocal Folds Paralysis**
- 4- Benign Laryngeal Lesions and Tumors**
- 5- Phonomicrosurgery and Laser Surgery**
- 6- Endoscopic Laser Cordectomy Classification**
- 7- Tracheostomy**
- 8- Tracheostomy Sizing Chart**
- 9- Laryngospasm**
- 10- Laryngitis**
- 11- Summary of Laryngeal Granulomatous Diseases**
- 12- Laryngopharyngeal Reflux**
- 13- Laryngeal Trauma**

Laryngotracheal Embryology:

- Occurs during **4th** week of gestation.
- Starts as **Respiratory Diverticulum**:
 - Laryngotracheal groove develops along ventral wall of Foregut Endoderm near level of last pharyngeal arch.
 - Groove gradually deepens.
 - Its edges fuse to form a Tracheoesophageal septum.
 - Tracheoesophageal septum separates Laryngotracheal tube from Pharynx and Esophagus.
 - Fusion starts caudally and extend cranially.
 - Formation of a blind outgrowth (Diverticulum).
 - Elongates in Caudal direction to form **Laryngotracheal Tube**:
 - **Proximal End** → Larynx.
 - **Middle Portion** → Trachea.
 - **Distal End** → Bifurcates into 2 Lung buds → Grow into Right and Left bronchi and Lungs.



- Epithelial proliferation obliterates the laryngeal lumen.
- Recanalization occurs by **10th week**.
- Failure of Recanalization results in stenosis.
 - **Laryngotracheal Tube Endoderm** gives rise to:
 - Epithelial lining of Larynx, Trachea, Bronchi and Alveoli.
 - **4th and 6th Pharyngeal Arch Mesoderm** give rise to
 - Cartilaginous and Muscular components of Larynx.
 - **Splanchnic Mesoderm** gives rise to:
 - Cartilaginous and Muscular components of Trachea and Lungs.
- Any congenital malformation of Pharynx and Esophagus is always associated with certain degree of malformation of Larynx and Trachea.

- **Embryology of Larynx:**

- Develops from:

1. Endoderm of Cranial end of Laryngotracheal tube:

- Epithelial lining of Larynx.

2. Mesoderm of 4th Pharyngeal Arch:

- Cartilages:

1. Epiglottis.
2. Thyroid.

- Muscle:

1. Cricothyroid Muscle (**Tensor**).

- Nerve:

1. Superior Laryngeal Nerve.

3. Mesoderm of 6th Pharyngeal Arch:

- Cartilages:

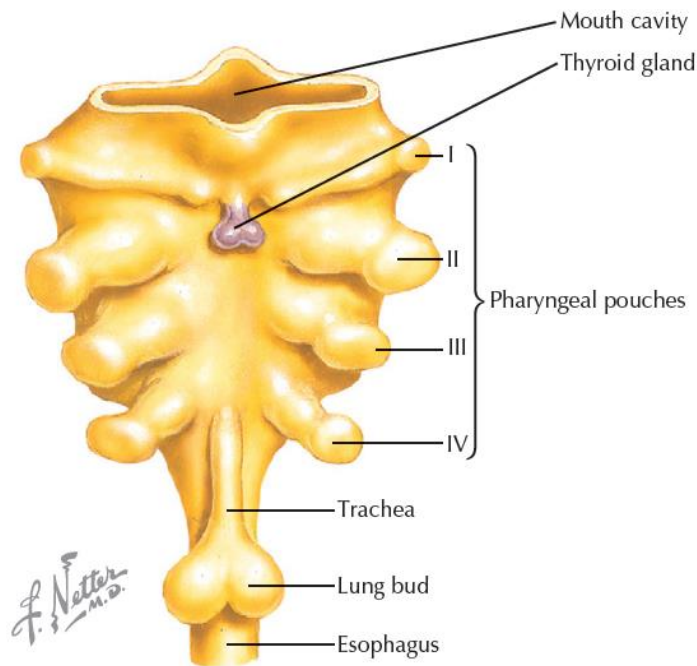
1. Arytenoid.
2. Corniculate.
3. Cuneiform.
4. Cricoid.

- Muscles:

1. Posterior Cricothyroid (**Abductor**)
2. Lateral Cricothyroid. (**Adductor**)
3. Interarytenoid (**Adductor**)
4. Thyroarytenoid (**Adductor**)
5. Vocalis (**Tensor**)

- Nerve:

- Recurrent Laryngeal Nerve.



- **Embryology of Trachea:**

- Develops from:

- 1. Endoderm of Middle portion of Laryngotracheal tube Middle**

- Epithelial lining and glands of Trachea.

- 2. Splanchnic Mesoderm:**

- Surround Tracheal tube
 - Form Tracheal cartilages, connective tissue, and smooth muscles of its walls

- By week 8:

- Mesenchymal rudiments of the 16-20 tracheal cartilages are seen.

- By week 10:

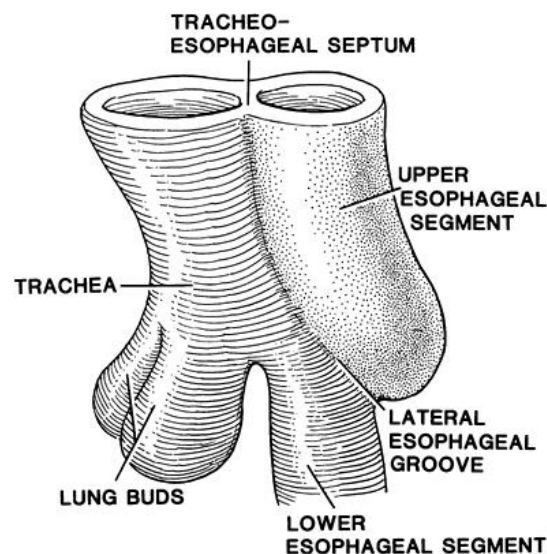
- Cilia appear.
 - Cartilages start to appear cranially and extending caudally.
 - Fibroelastic tissue of the tracheal wall arises from mesenchyme between the cartilage and, posteriorly, between the ends of the embryonic rings smooth muscle (the trachealis) arises.

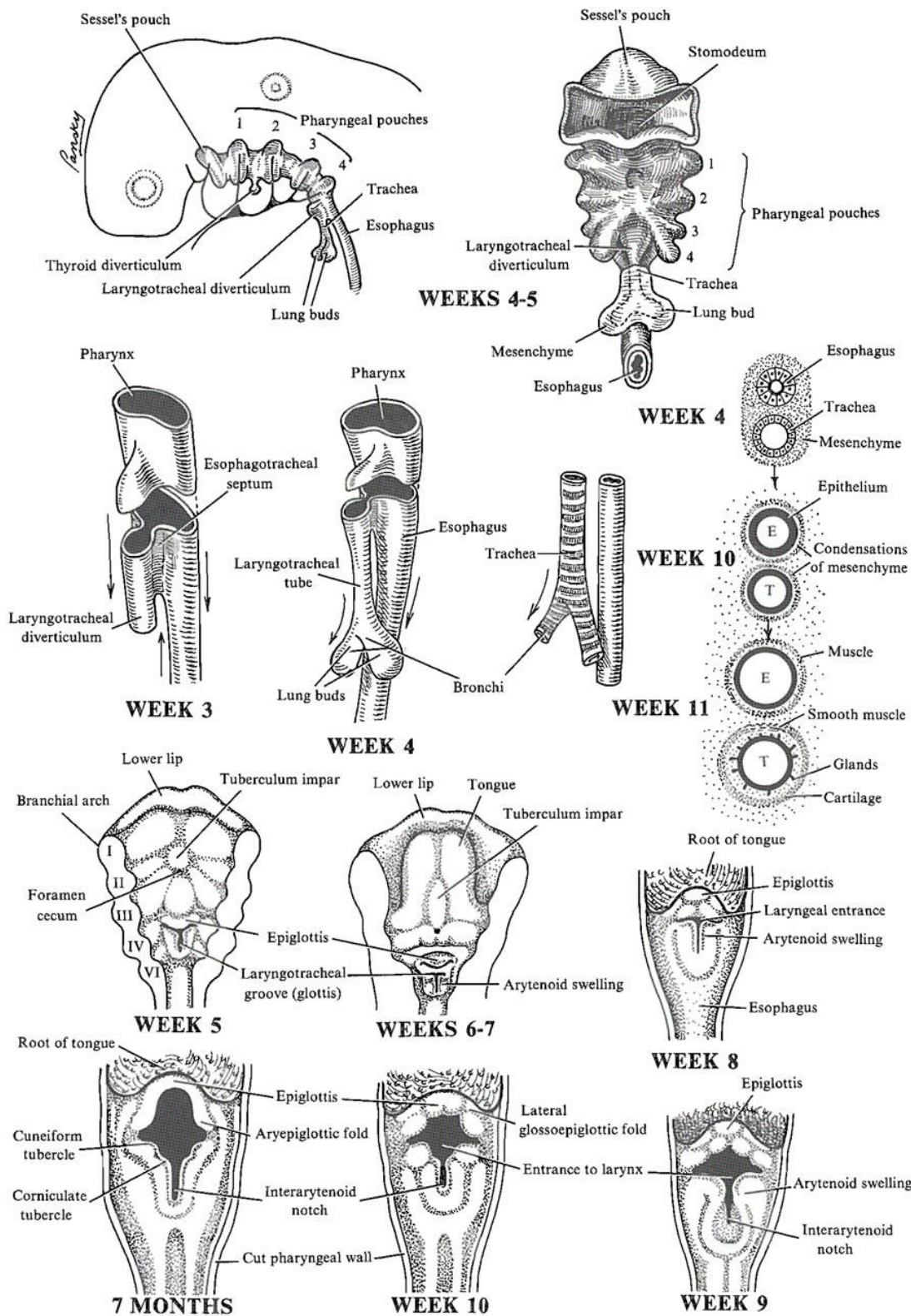
- By week 12:

- Mucosal glands are seen
 - Develop in a craniocaudal direction

- By the end of week 20:

- All major microscopic features of Trachea are visible
 - It is Short and narrow while the larynx is relatively long
 - This relationship remains until after birth when the trachea outgrows the larynx to reach its final form





Laryngeal Anatomy:

- Lies anterior to Hypopharynx:
 - o Opposite to **C3-C6** vertebrae.
- Extend from upper border of Epiglottis to lower border of Cricoid.
 - o Upper end opens into Hypopharynx by Laryngeal inlet.
 - o Its lower end is continuous with Trachea at the level of C6.
- Larynx moves vertically and in anteroposterior direction during swallowing and phonation.
- It can be passively moved from side to side producing a characteristic grating sensation called **Laryngeal crepitus**.

Relations of Larynx:

Anterolateral:

- o Infrahyoid muscles & Platysma.

Lateral:

- o Lobes of thyroid gland & carotid sheath.

Posterior:

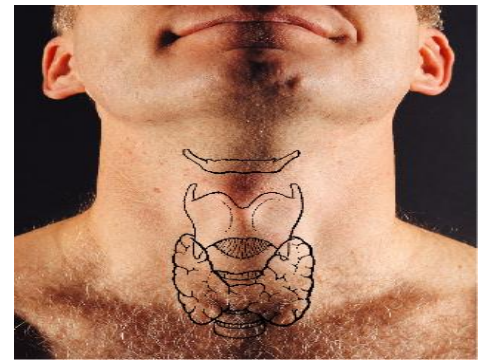
- o It forms anterior wall of Hypopharynx.

Superior:

- o Base of Tongue & vallecula.

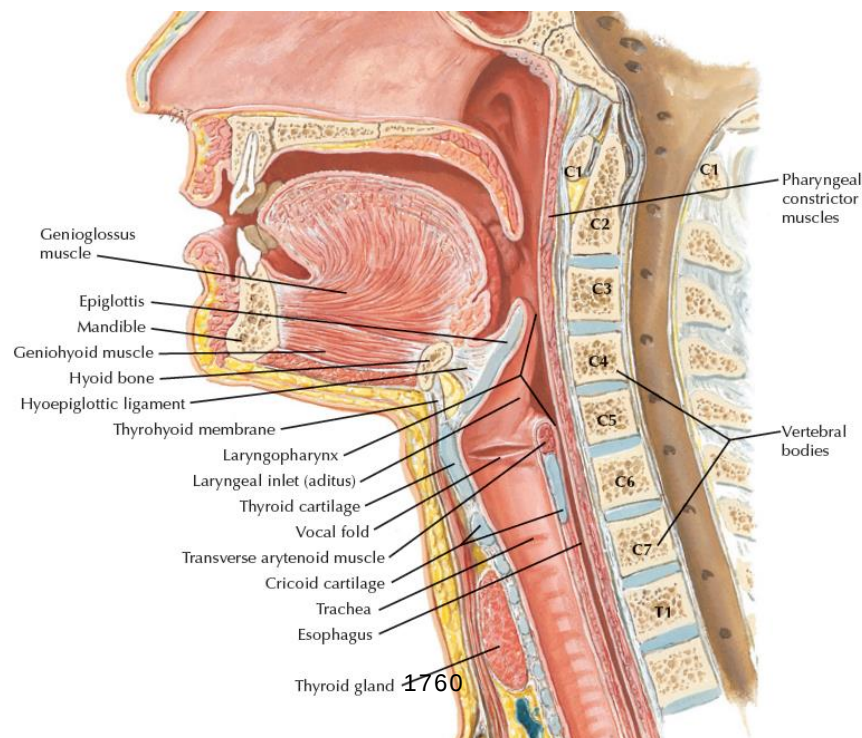
Inferior:

- o Trachea.

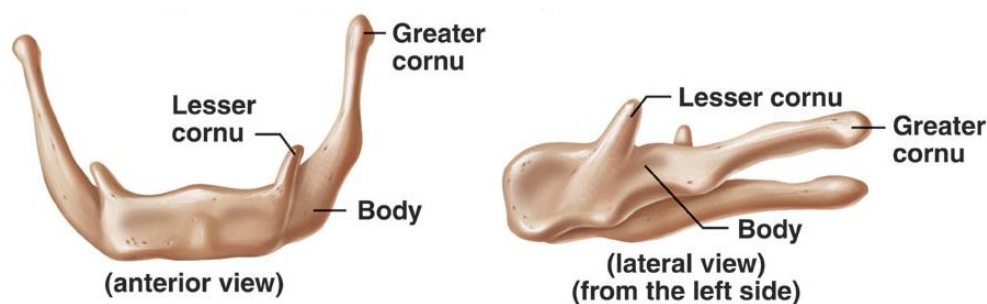


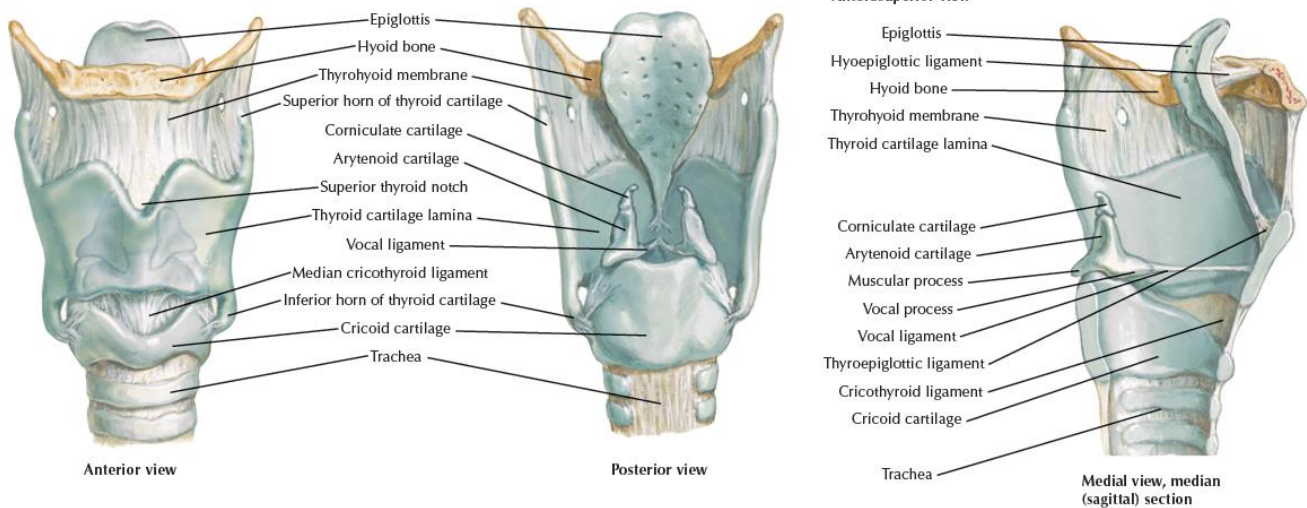
Laryngeal skeleton is made up from:

1. Hyoid Bone (Not main part of Larynx)
2. Laryngeal Cartilages.
3. Laryngeal Joints.
4. Laryngeal Ligaments.
5. Laryngeal Membranes.
6. Laryngeal Muscles.



- **Hyoid Bone:**
- Horseshoe shaped bone.
- Situated in anterior midline of neck between chin and thyroid cartilage.
- Lies at the level of C3 at rest.
- Suspended from the skull by **Stylohyoid ligaments**.
- It is only distantly articulated to other bones by muscles or ligaments.
- Due to its position, Hyoid bone is not susceptible to easy fracture.
 - o In a suspected case of murder, a fractured hyoid strongly indicates throttling or strangulation.
- Hyoid supports the larynx, stabilizes the Hypopharynx and aids in swallowing and tongue movement.
- **Consists of:**
 - o **Body:**
 - Anterior surface is convex and posterior surface is concave.
 - Posterior surface is attached to Epiglottis by **HyoEpiglottic ligament**.
 - Inferior surface of body and Greater Cornua is attached by **ThyroHyoid membrane** to upper border of Thyroid Cartilage:
 - **Median ThyroHyoid ligament** is median thickening of ThyroHyoid membrane and it attaches inferior surface of body of hyoid to Superior Horn of Thyroid Cartilage.
 - o **Greater Cornua:**
 - Project backward from lateral borders of the body.
 - Inferior surface of body and Greater Cornua is attached by **ThyroHyoid membrane** to upper border of Thyroid Cartilage:
 - **Lateral ThyroHyoid ligament** is lateral thickening of ThyroHyoid membrane and it attaches greater cornua end to Superior Horn of Thyroid Cartilage.
 - o **Lesser Cornua:**
 - Small conical eminences attached to superior surface of the body.
 - Gives attachment to **StyloHyoid ligament**.





- Hyoid gives Muscular attachments to:

○ **Supra-Hyoid Muscles:**

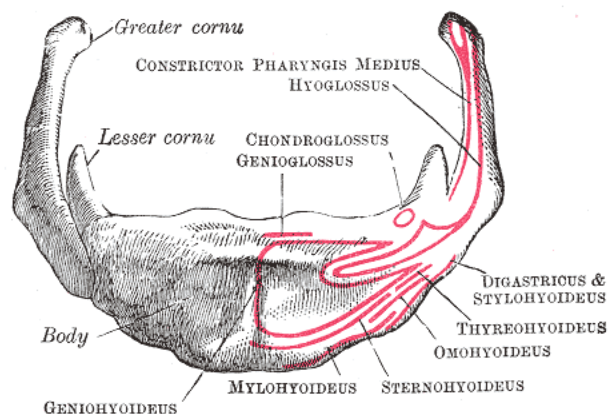
- Assist in elevating hyoid bone and widening the esophagus during swallowing.

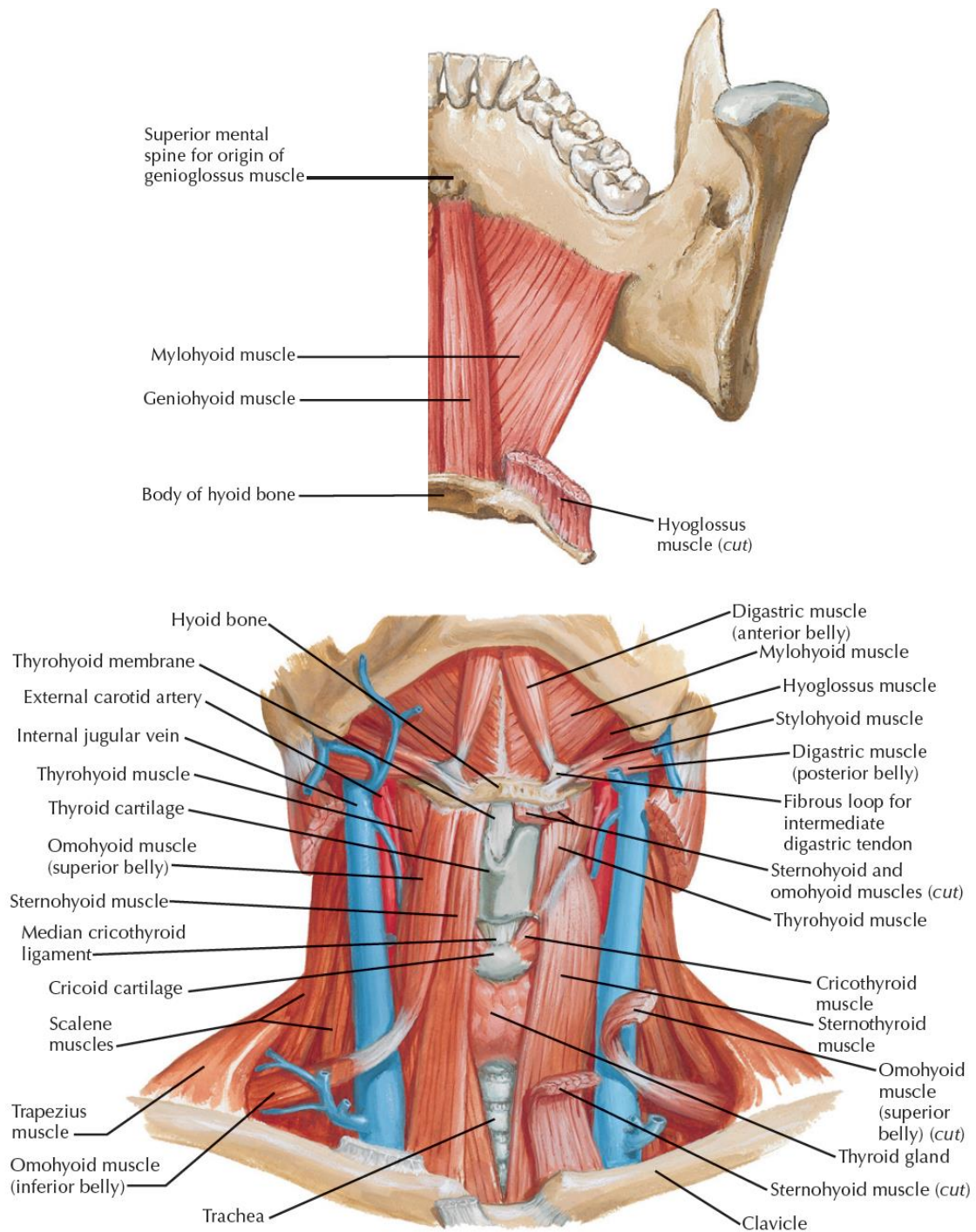
1. GenioHyoid
2. MyloHyoid
3. Digastric
4. StyloHyoid
5. HyoGlossus
6. Middle pharyngeal constrictor

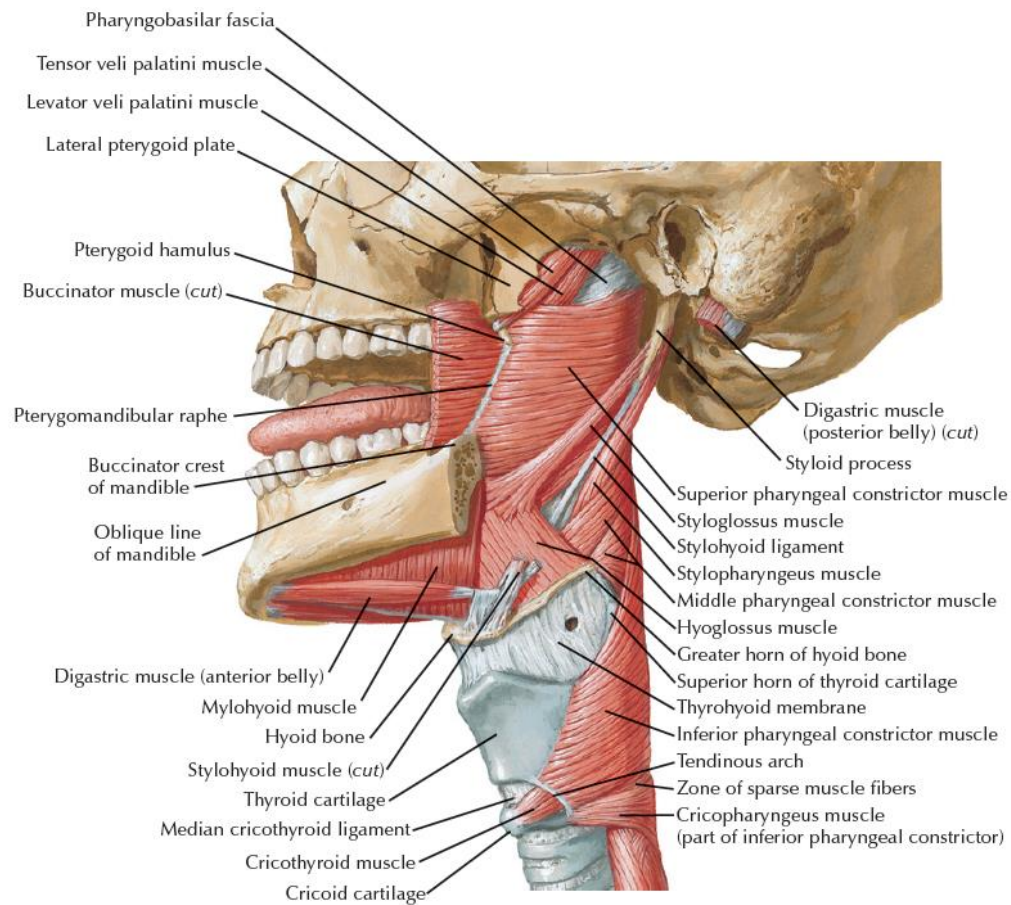
○ **Infra-Hyoid Muscles (Strap):**

- Assist in depressing hyoid bone and larynx during swallowing and speech.

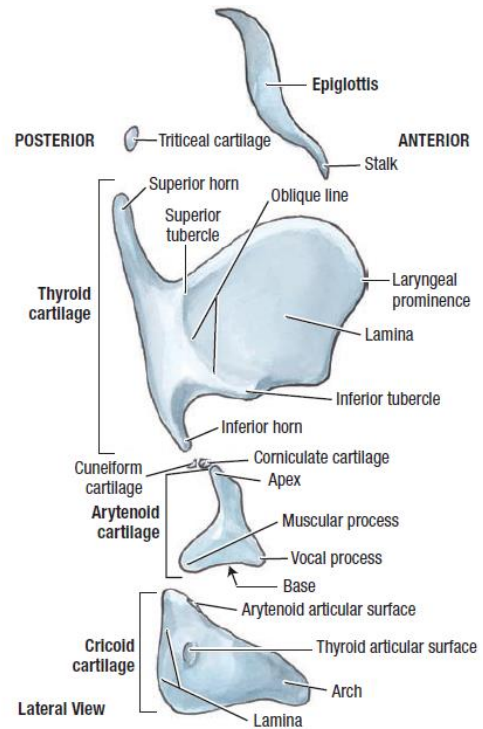
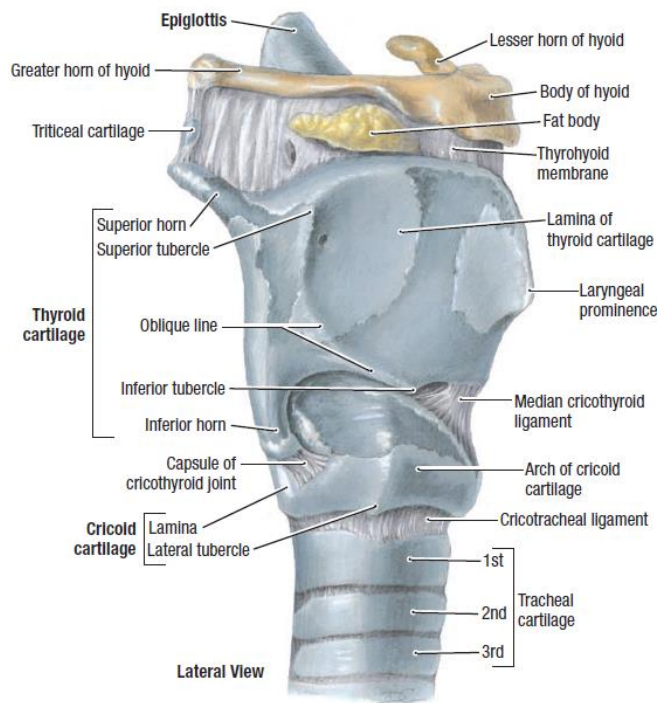
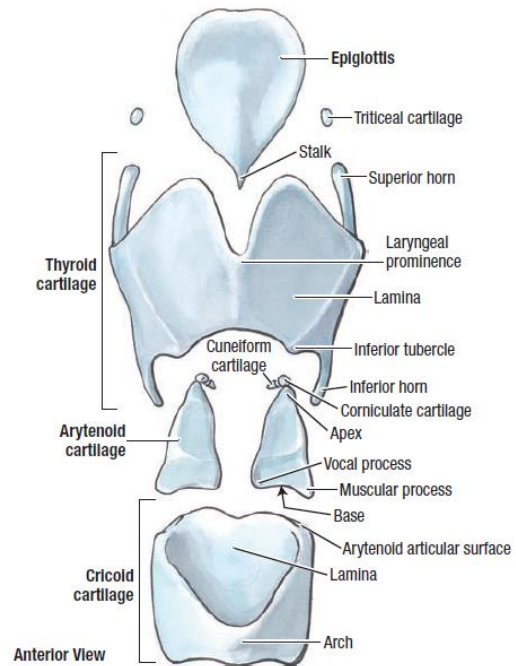
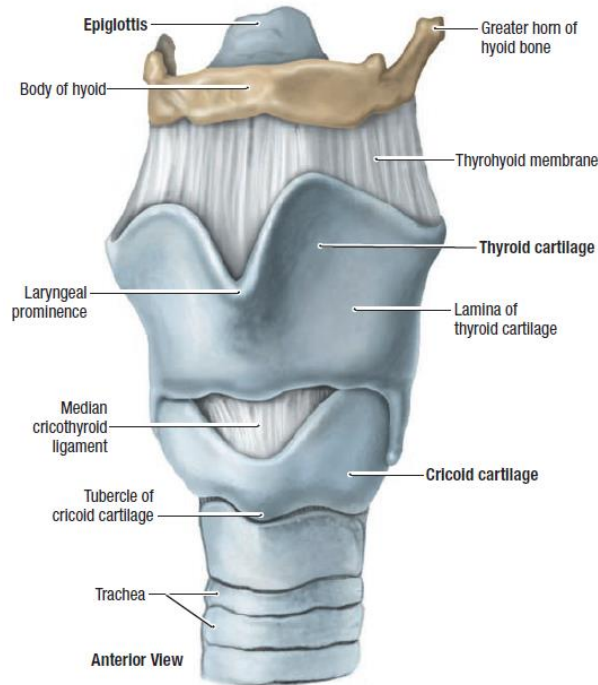
1. OmoHyoid
2. SternoHyoid
3. SternoThyroid
4. ThyroHyoid

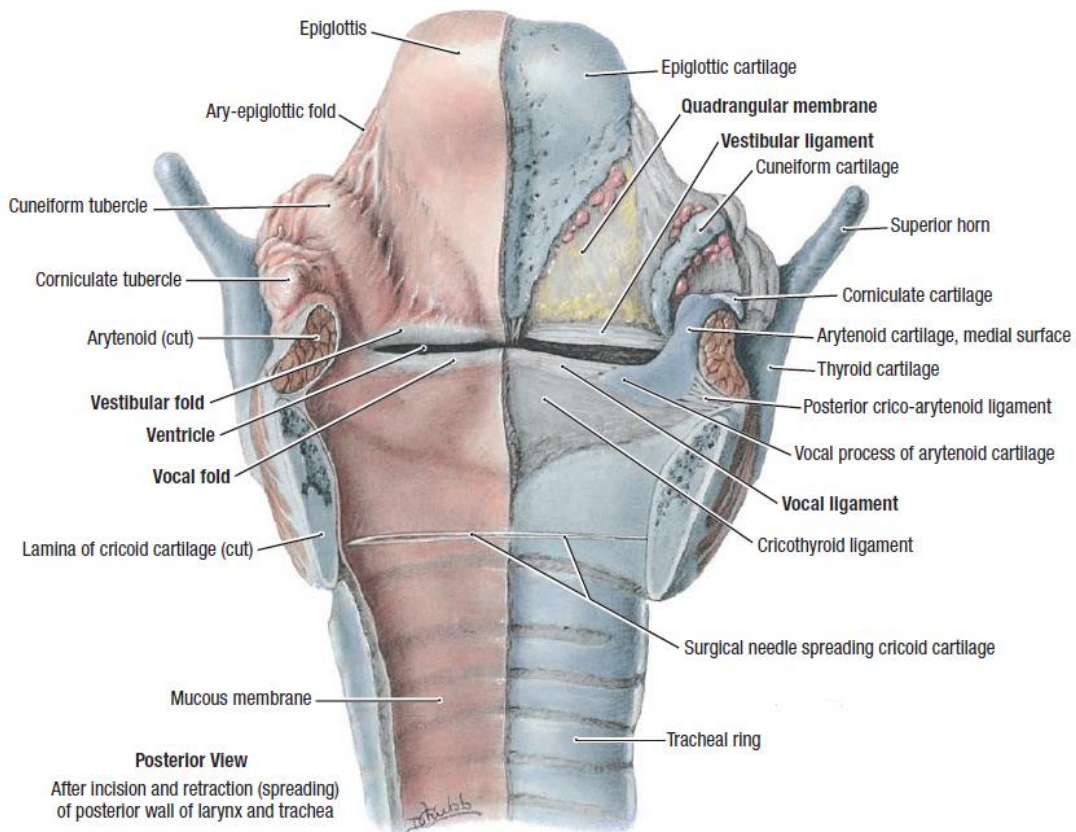
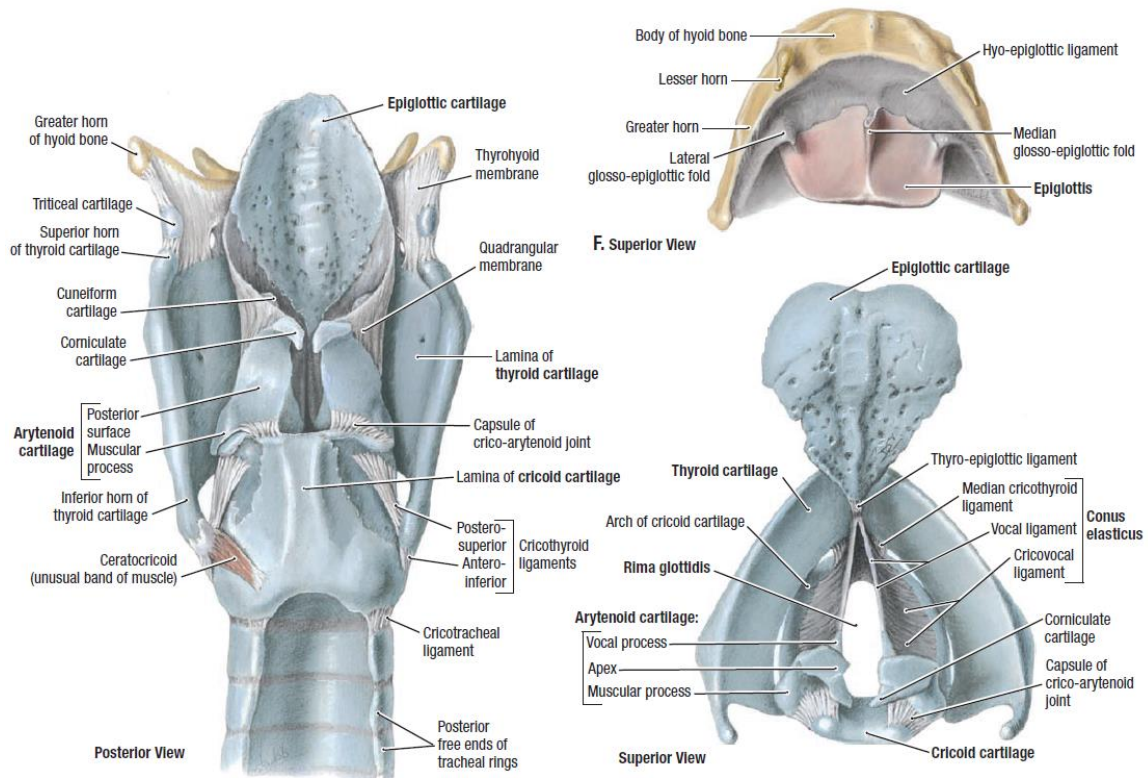






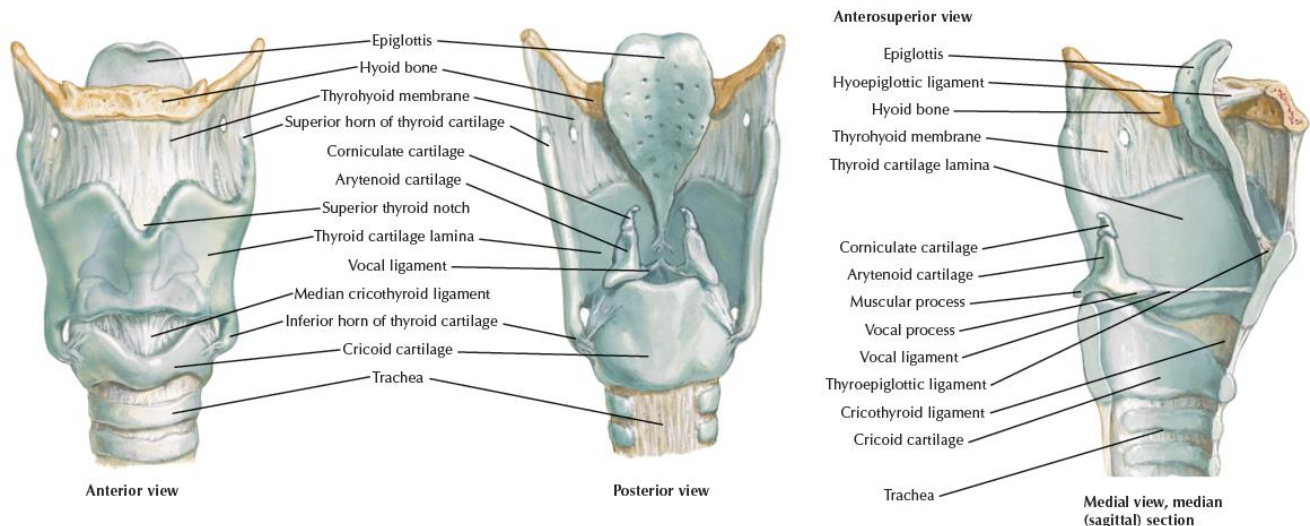
- **Laryngeal Cartilages:**
- **9 Cartilages:**
 - **3 Unpaired cartilages:**
 - Epiglottis
 - Thyroid
 - Cricoid
 - **3 Paired cartilages:**
 - Arytenoid
 - Corniculate
 - Cuneiform

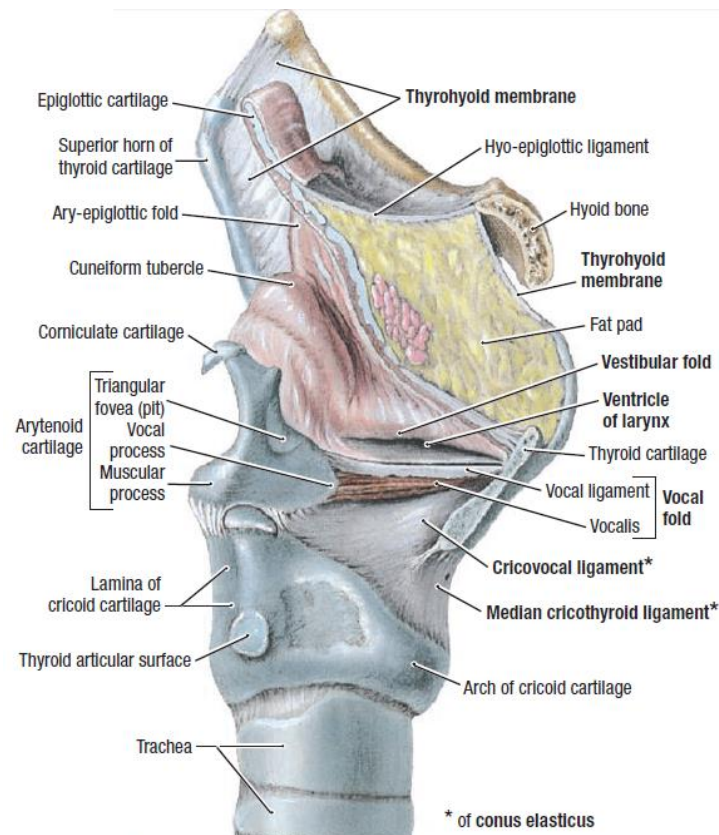




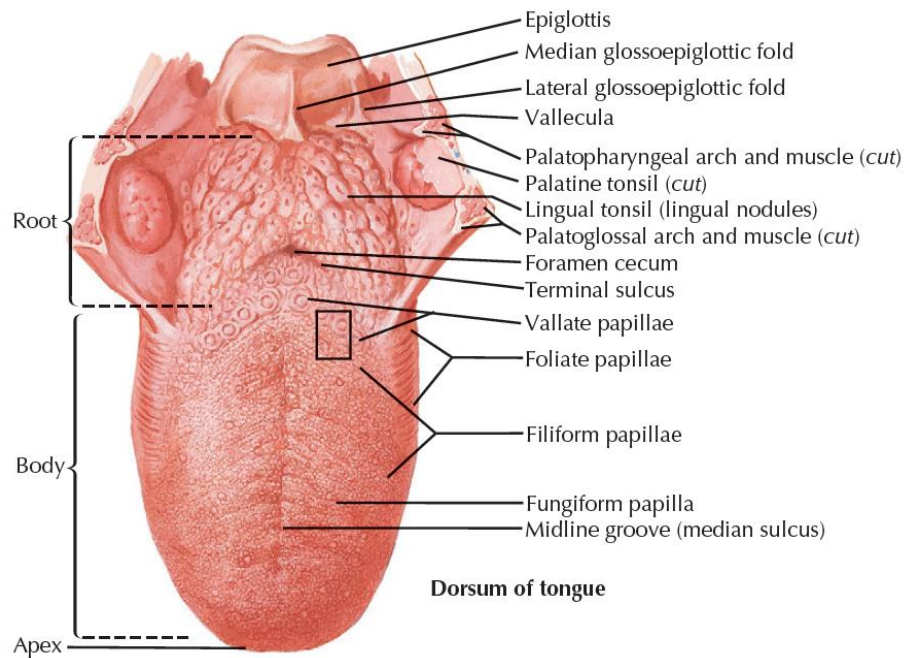
1. Epiglottis:

- Fibroelastic cartilage.
- Leaf-like shape.
 - Has upper broad free end and a lower tapering end.
- Projects upwards behind Tongue and Hyoid bone
- Forms Anterior wall of laryngeal inlet.
- Connected Anteriorly to:
 - **Base of Tongue:**
 - By Median and Lateral GlossoEpiglottic folds.
 - The depressions between Epiglottis and root of Tongue, on either side of the median fold, are named **Valleculae.**
 - **Body of Hyoid:**
 - By HyoEpiglottic ligament.
 - Divides epiglottic to suprahyoid and infrahyoid epiglottis .
 - **Inner surface of Thyroid cartilage:**
 - By ThyroEpiglottic ligament.
 - **Patiole:** is stalk-like process of epiglottis attached to ThyroEpiglottic ligament.
- AryEpiglottic (A-E) Folds:
 - Mucosal folds extend between lateral margin of Epiglottis and Arytenoid cartilage on each side.
 - Constitute the lateral borders of the laryngeal inlet.
- Pre-Epiglottic space:
 - Potential space filled with fat.
 - Separates anterior surface of epiglottis from ThyroHyoid membrane and upper part of Thyroid cartilage.
 - Invaded in supraglottic or base of tongue Ca.



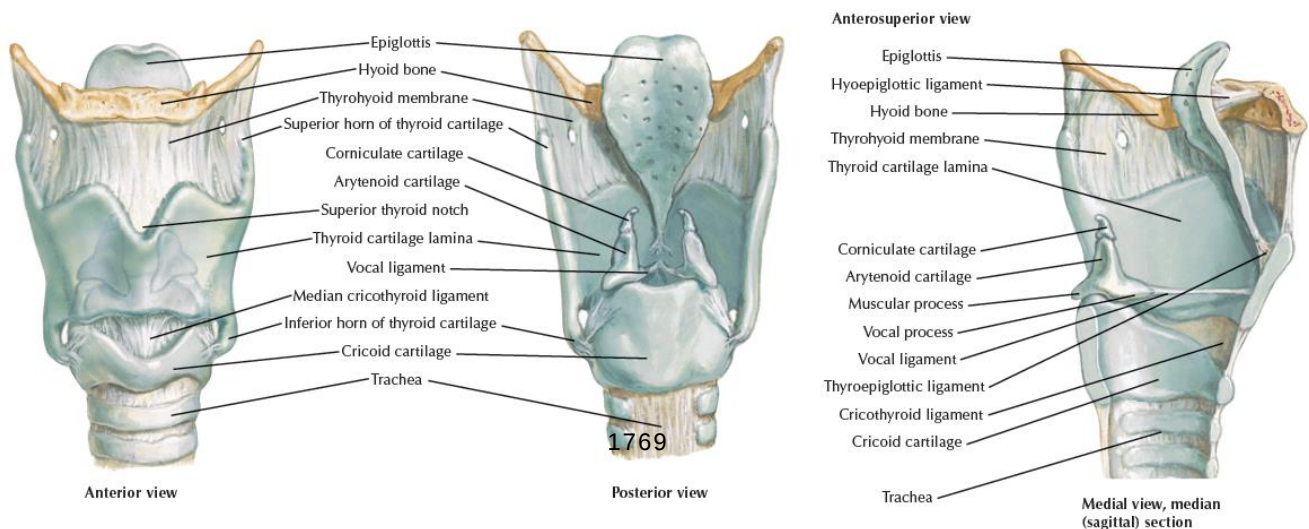


B. Lateral View After Removal of the Right Thyroid Cartilage



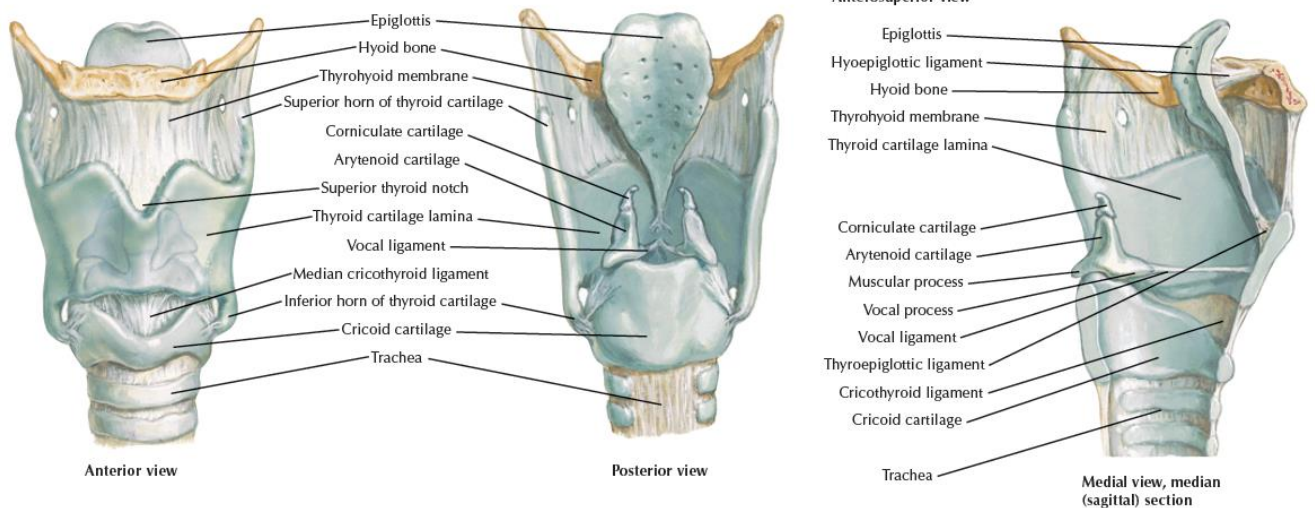
2. Thyroid Cartilage:

- Hyaline cartilage.
 - Start to ossifies around age of 20 years.
- Largest laryngeal cartilage.
- Lies between C4 and C6.
- Formed of 2 Laminae (Ala):
 - **Fused Anteriorly:**
 - Form Laryngeal prominence (Adams Apple):
 - Angle 90° in males
 - Angle 120° in Females.
 - Superior Thyroid Notch:
 - V-shaped notch.
 - Lies in superior portion of laryngeal prominence.
 - Connected to Epiglottis at Thyroid angle by ThyroEpiglottic ligament.
 - Inferior surface of Thyroid cartilage is attached to upper border of Cricoid Cartilage by CricoThyroid Membrane:
 - Median CricoThyroid ligament is median thickening of CricoThyroid membrane.
 - Vocal cord are attached to middle of thyroid angle in the medial surface .
 - **Separated Posteriorly:**
 - Posterior borders has 2 horns :
 - Superior Horn:
 - Attached to tip of greater horn of hyoid bone by Lateral ThyroHyoid ligament.
 - Inferior Horn:
 - Articulates with Cricoid cartilage to form the CricoThyroid joint.
- Oblique Line:
 - Oblique ridge on outer surface of each lamina.
 - Extends between Superior and Inferior tubercles.
 - Gives attachment for:
 1. Sternothyroid muscle.
 2. Thyrohyoid muscle.
 3. Inferior constrictor muscle.



3. **Cricoid Cartilage:**

- Hyaline cartilage.
- Smaller, but thicker than Thyroid cartilage.
- Lies below and behind Thyroid cartilage (level of C6).
- Both intrinsic & extrinsic laryngeal muscles attach to Cricoid .
- Only cartilage forming a complete ring.
- Signet ring in shape:
 - **Anteriorly:**
 - Form a narrow Arch.
 - **Posteriorly:**
 - Expand to form Lamina.
 - **Superiorly:**
 - Sloping border.
 - Attached to inferior surface of Thyroid cartilage by CricoThyroid Membrane:
 - Median CricoThyroid ligament is median thickening of CricoThyroid membrane.
 - Articulates with Arytenoid cartilage to form CricoArytenoid joint .
 - **Inferiorly:**
 - Horizontal border.
 - Articulates with inferior horn of Thyroid cartilage to form CricoThyroid joint .
 - Connected to 1st Tracheal ring by CricoTracheal ligament.



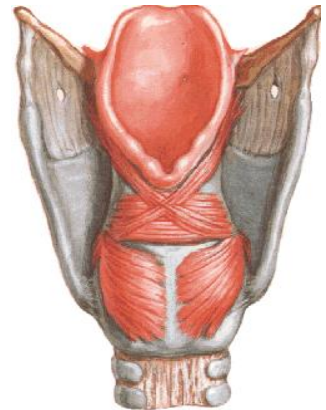
4. **Arytenoid Cartilage:**

- Paired Hyaline cartilage.
- The chief moving parts of Larynx.
- Pyramidal in shape:
 - **Apex:**
 - Supports Corniculate cartilage.
 - **Base:**
 - Articulates with Cricoid cartilage.
 - **Muscular process:**
 - Directed laterally.
 - Give attachment to intrinsic laryngeal muscles.
 - **Vocal process:**
 - Directed Anteriorly.
 - Gives attachment to vocal cord.
- Arytenoids have 3 surfaces:
 - Posterior surface .
 - Anterolateral surface.
 - Medial surface .



5. **Corniculate Cartilage:**

- Cartilages of Santorini.
- Paired Fibroelastic cartilages.
- Articulates with Apex of Arytenoid cartilage.
- Located posteriorly within AryEpiglottic (A-E) folds forming Corniculate Tubercle.
- Provides rigidity to AryEpiglottic (A-E) folds.



6. **Cuneiform Cartilage:**

- Cartilages of Wrisberg.
- Paired Fibroelastic cartilages.
- Located anterior to Corniculate cartilage within AryEpiglottic (A-E) folds forming Cuneiform Tubercle.
- Provides rigidity to AryEpiglottic (A-E) folds.

• **Triticeous Cartilage:**

- Sometimes found in Lateral ThyroHyoid ligament.
- May be mistaken on x-ray as a foreign body when calcified.



- **Laryngeal Joints:**

- **3 joints:**

1. CricoThyroid Joint:

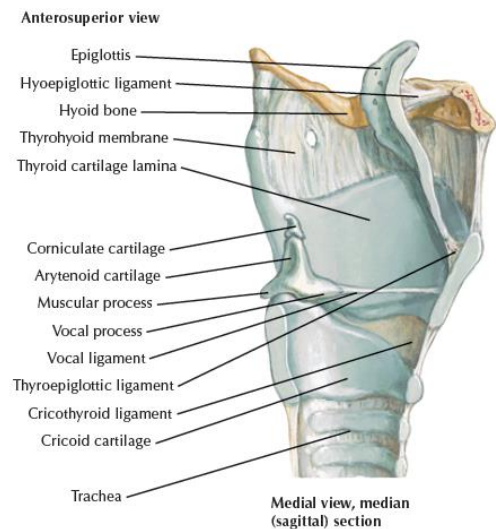
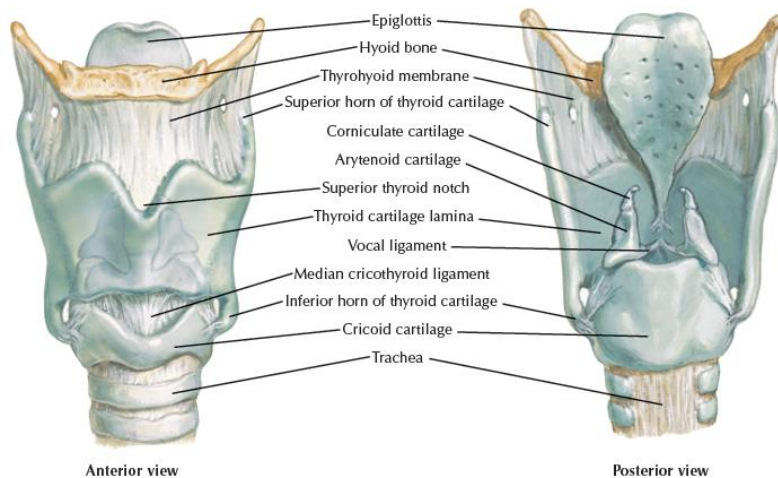
- Synovial joint.
- Formed by Inferior cornua of thyroid cartilage with a facet on Cricoid cartilage.

2. CricoArytenoid Joint:

- Synovial joint.
- Formed by base of Arytenoid and with a facet on upper border of Cricoid lamina.
- Two types of movements:
 - **Rotatory movement:**
 - Arytenoid cartilage moves around a vertical axis.
 - Abducting or Adducting the vocal cord
 - **Gliding movement:**
 - One Arytenoid glides towards the other cartilage or away from it.
 - Closing or opening the posterior part of glottis.

3. AryCorniculate Joint:

- One on each side.
- A joint between the apex of Arytenoid cartilage and Corniculate cartilage.



- **Laryngeal Membranes:**
- **3 Extrinsic Membranes:**

1. ThyroHyoid Membrane:

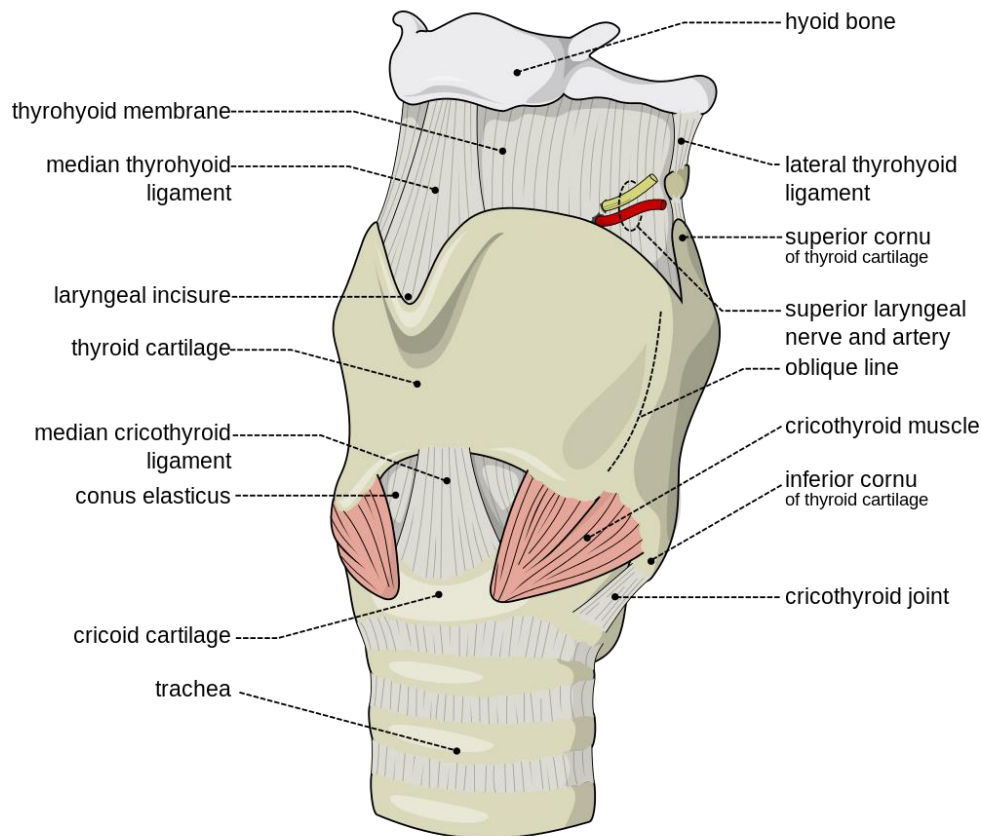
- Broad, Fibroelastic layer connects Thyroid cartilage to Hyoid bone.
- Pierced by:
 1. Superior laryngeal vessels.
 2. Internal branch of Superior laryngeal nerve.
- Median part of the membrane is thickened to form **Median ThyroHyoid Ligament**
- Posterior border of the membrane is thickened to form **Lateral ThyroHyoid Ligament**.
 - Contains Triticeous Cartilage.

2. CricoThyroid Membrane:

- Connects Thyroid cartilage to Cricoid cartilage.
- Avascular structure.
- Pierced for emergency Cricothyrotomy.

3. CricoTracheal Membrane:

- Connects Cricoid cartilage to 1st Tracheal ring.
- Resembles the fibrous membrane which connects the cartilaginous rings of trachea to each other.



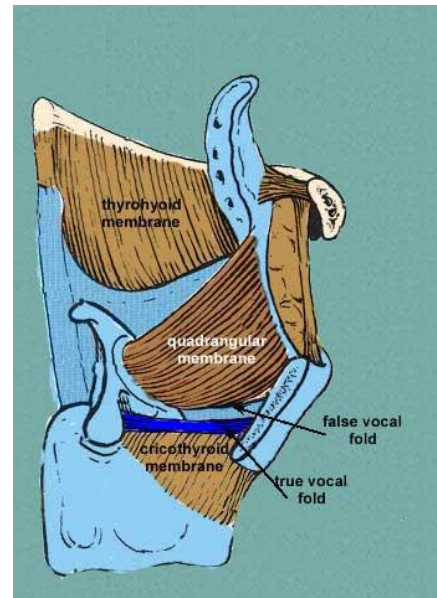
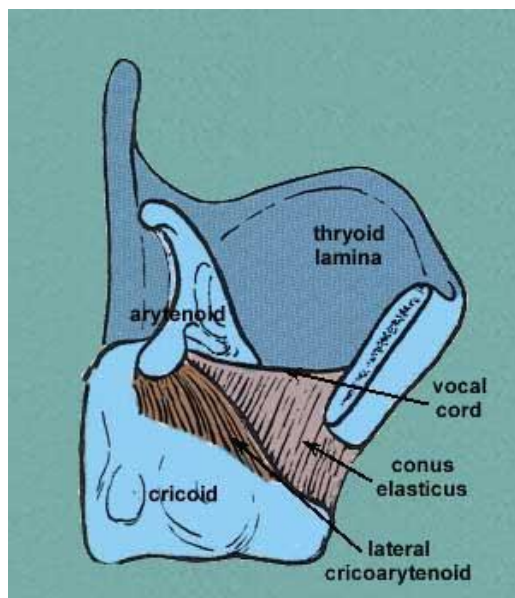
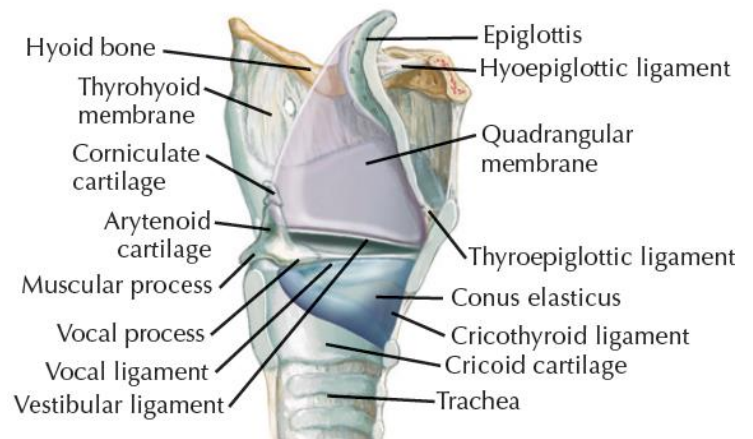
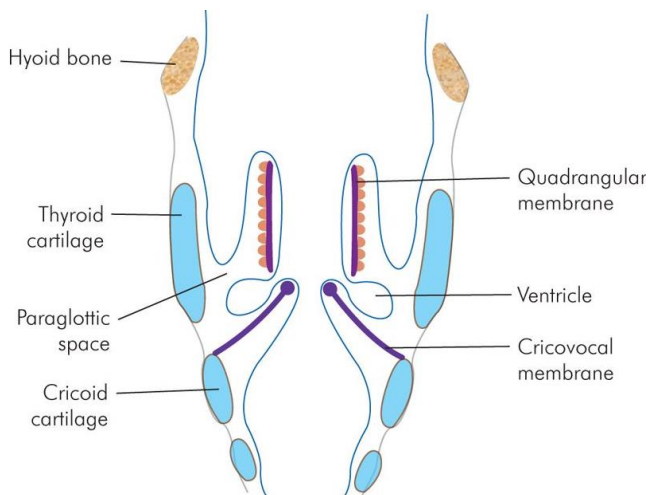
- 2 Intrinsic Membranes:

1. Quadrangular Membrane:

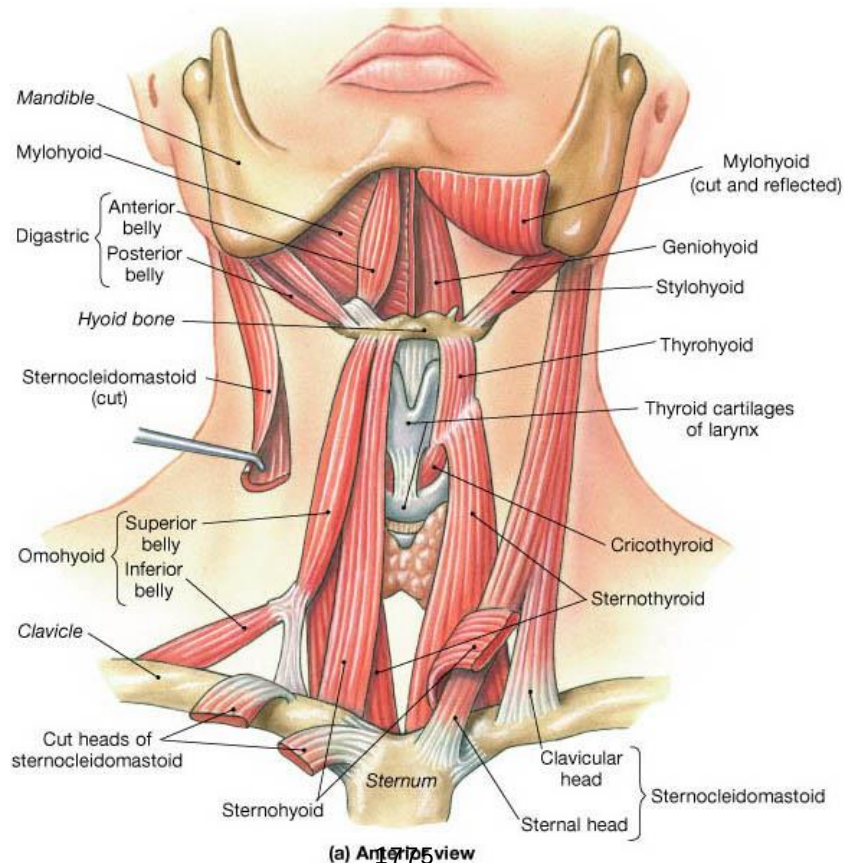
- Extends between lateral aspect of Epiglottis and Aterolateral surface of Arytenoid cartilage on each side .
- Its free lower border forms Vestibular ligament which forms Vestibular folds (False vocal folds) once covered by mucosa.

2. Conus Elasticus (CricoVocal Membrane):

- Triangular fibroelastic membrane.
- Extends superiorly from Anterior Arch Cricoid and attached to Thyroid cartilage anteriorly and Vocal processes of Arytenoid cartilages posteriorly.
- Its free upper border forms Vocal ligament, which forms True vocal folds once covered by mucosa.



- **Laryngeal Muscles:**
- **Extrinsic Muscles:**
 - Attach Larynx to adjacent structures.
 - **Elevators:**
 - Primary Elevators:
 - Act directly as they are attached to Thyroid cartilage:
 1. ThyroHyoid (**C1**)
 2. StyloPharyngeus (**CN-IX**)
 3. PalatoPharyngeus (**CN-X**)
 4. SalpingoPharyngeus (**CN-X**)
 - Secondary Elevators:
 - Act indirectly as they are attached to Hyoid bone:
 1. GenioHyoid (**C1**)
 2. Mylohyoid (**CN-V3**)
 3. Digastric:
 - Ant. belly (**CN-V3**)
 - Post. belly (**CN-VII**)
 4. StyloHyoid (**CN-VII**)
 - **Depressors:**
 - Infra-Hyoid Muscles (Strap):
 1. ThyroHyoid (**C1**)
 2. OmoHyoid (**C1-C3**)
 3. SternoThyroid (**C1-C3**)
 4. SternoHyoid (**C1-C3**)



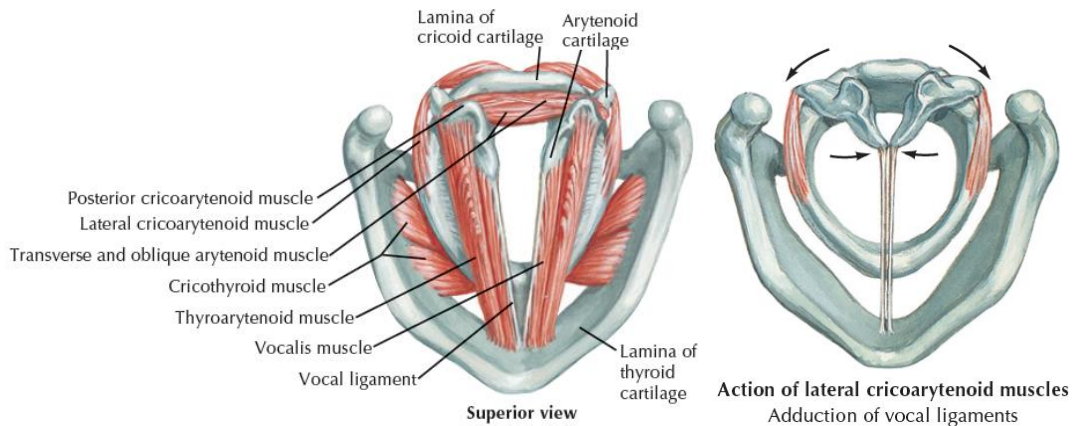
- Intrinsic Muscles:

- Attach laryngeal cartilages to each other.
- Act on vocal cords.

▪ **ADDuctors (Close Rima Glottidis):**

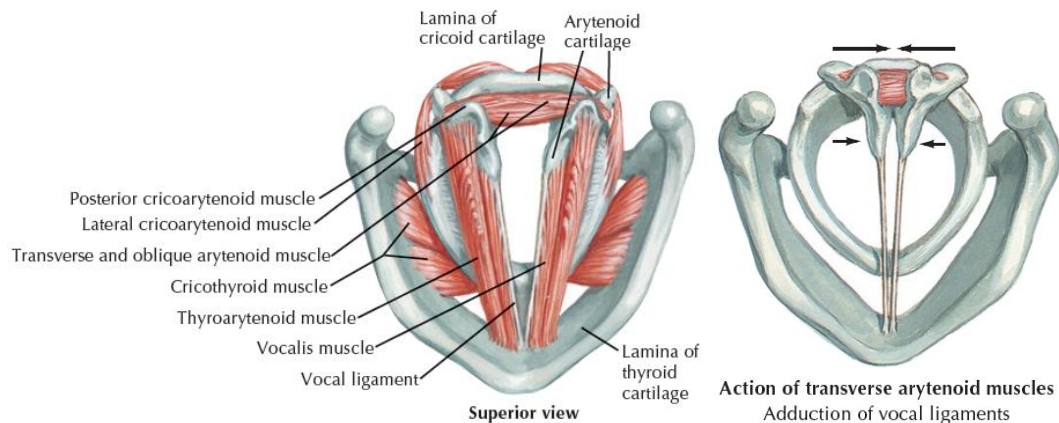
1. Lateral CricoArytenoid:

- Origin → Lateral Cricoid Arch
- Insertion → Muscular process of Arytenoid
- Nerve Supply → **(RLN)**



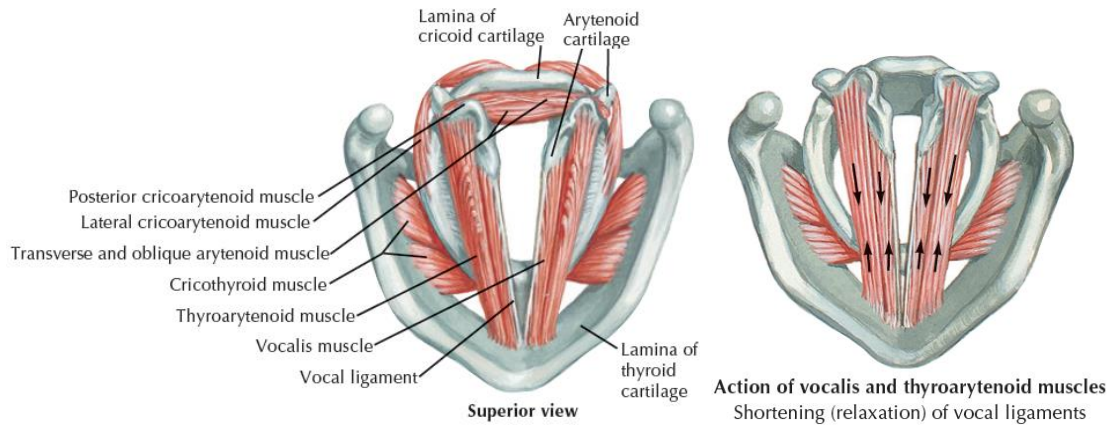
2. InterArytenoid:

- Only single intrinsic laryngeal muscle.
- Only muscle receive bilateral nerve supply.
- Origin → Muscular process of Arytenoid
- Insertion → Muscular process of other Arytenoid
- Nerve Supply → **(Bilateral RLN)**



3. ThyroArytenoid (External Part):

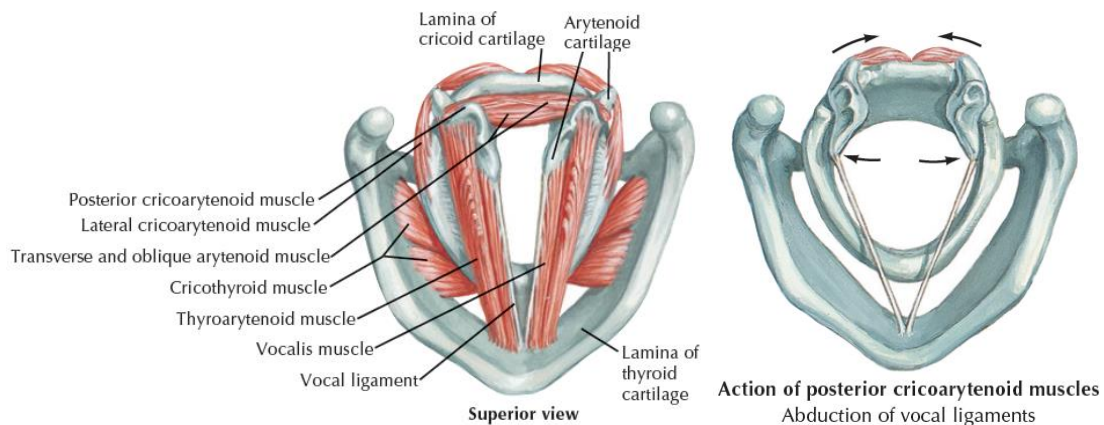
- Origin → Inner surface of Thyroid angle.
- Insertion → Vocal process of Arytenoid
- Nerve Supply → **(RLN)**



▪ **ABDuctor (Open Rima Glottidis):**

1. Posterior CricoArytenoid:

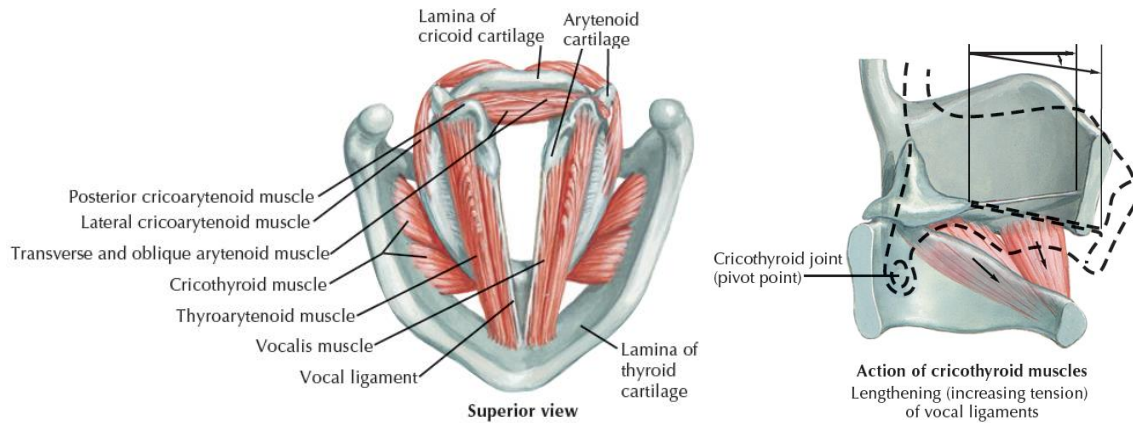
- Only ABDuctor muscle.
- Origin → Posterior Cricoid Lamina
- Insertion → Muscular process of Arytenoid
- Nerve Supply → **(RLN)**



▪ **Tensors:**

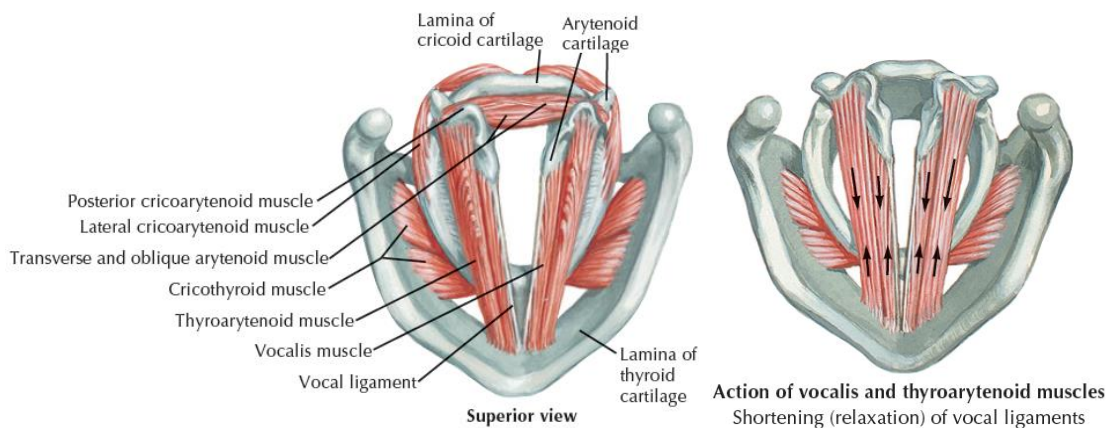
1. Cricothyroid:

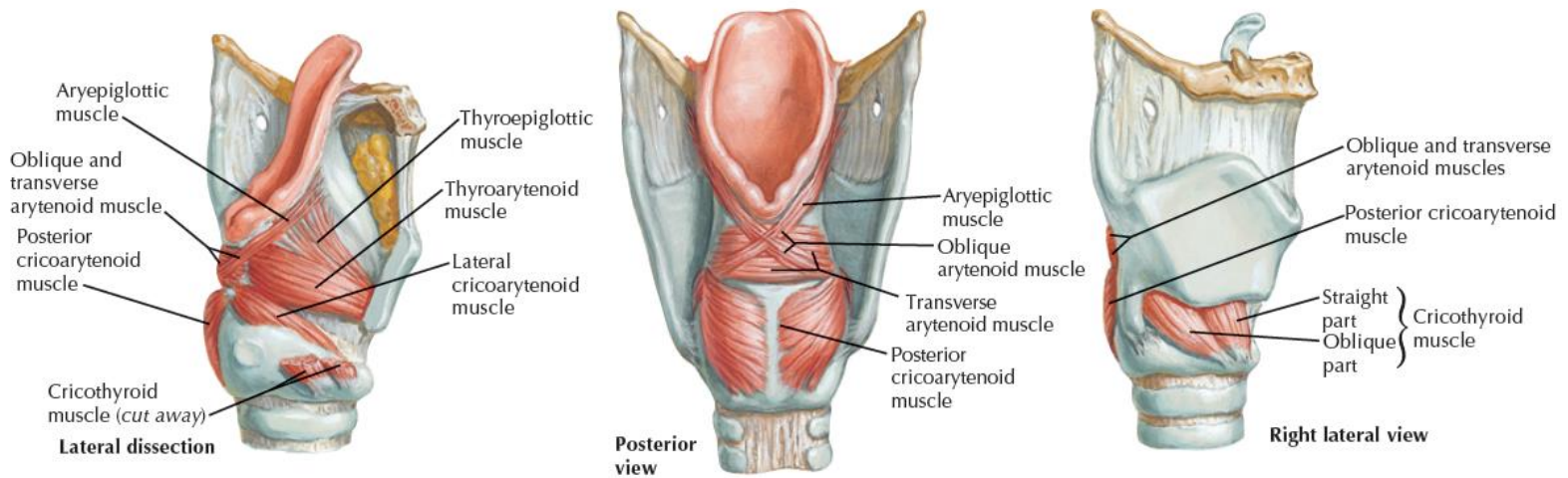
- Chief pitch-changing muscle.
- Only muscle seen on outside of Larynx.
- Only muscle supplied by External SLN.
- Origin → Arch of Cricoid
- Insertion → Inferior horn and Lamina of Thyroid cartilage.
- Nerve Supply → **(External SLN)**



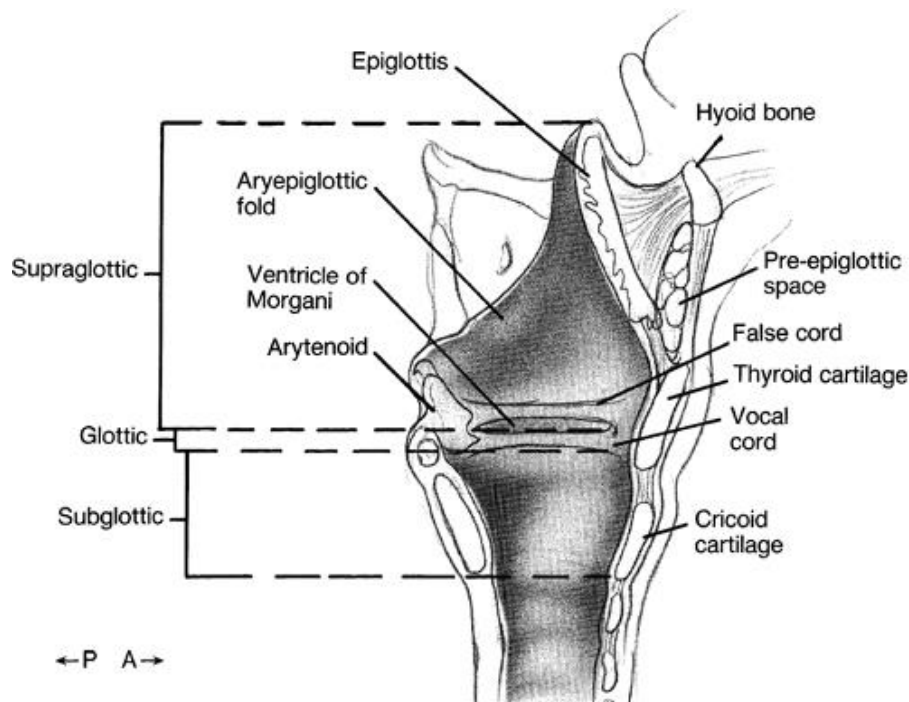
2. Vocalis (Internal Part of ThyroArytenoid):

- Origin → Inner surface of Thyroid angle.
- Insertion → Vocal process of Arytenoid
- Nerve Supply → **(RLN)**

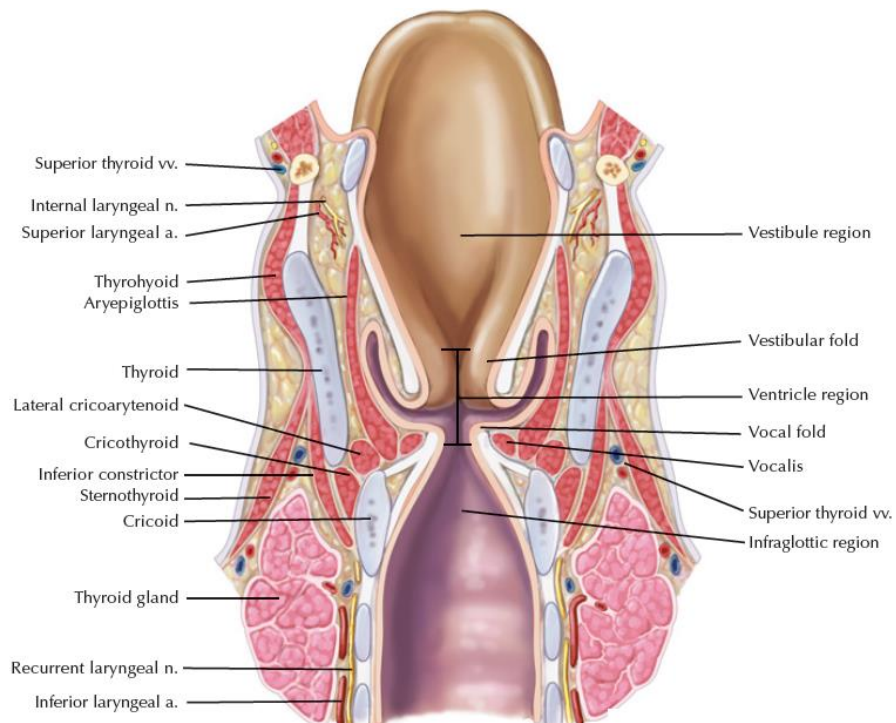




- **Cavity of Larynx:**
- Starts at Laryngeal inlet where it communicates with Hypopharynx and ends at lower border of Cricoid where it is continuous Trachea.
- **Laryngeal Sites:**
 - **Supraglottis:**
 - Extends from Epiglottis to junction of Ventricle and True vocal folds.
 - **Glottis:**
 - Extends from superior surface of True vocal folds to 1cm below True vocal folds.
 - **Subglottis:**
 - Extends from 1 cm below True vocal folds to inferior Cricoid cartilage.

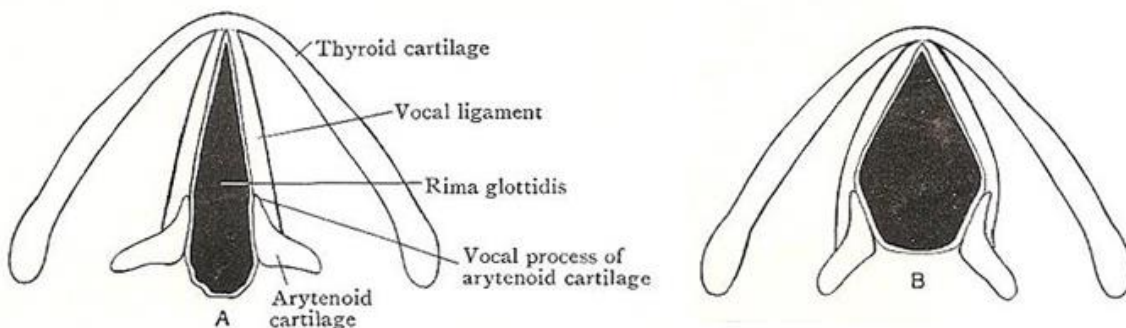


- **Supraglottis:**
 - Extends from Epiglottis to junction of Ventricle and True vocal folds.
 - **Laryngeal inlet:**
 - Oblique opening bounded by:
 - Anteriorly: Epiglottis
 - Posteriorly: Apex of Arytenoid
 - Laterally: AryEpiglottic folds
 - **Vestibule:**
 - Space between laryngeal inlet and False vocal folds (vestibular folds).
 - **Vestibular Folds (False vocal folds):**
 - Two thick folds of mucous membrane enclosing a narrow band of fibrous tissue (ventricular ligament).
 - Attached anteriorly to Thyroid angle and posteriorly to antero-lateral surface of Arytenoid cartilage.
 - **Ventricle (Sinus of Larynx):**
 - Deep elliptical space between False and True vocal folds.
 - Ventricle of larynx opens into **Saccule**:
 - Blind-ended pouch ascends forwards from ventricle between vestibular fold and thyroid cartilage.
 - 60-70 mucous glands open onto its luminal surface help to lubricate vocal cords which lack mucous glands.



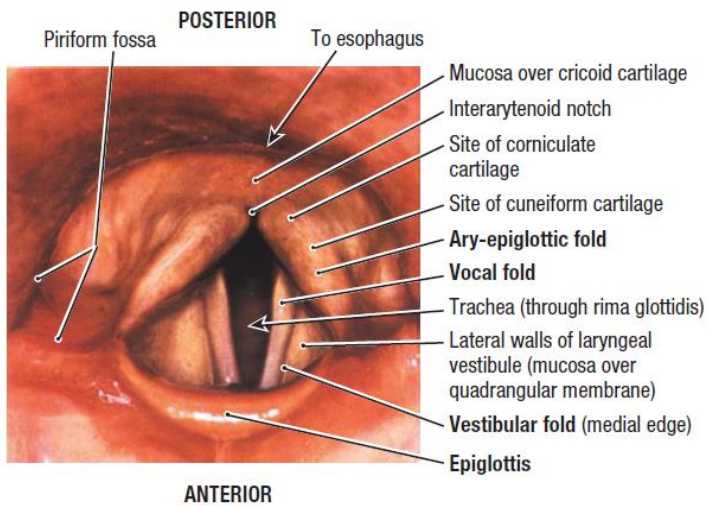
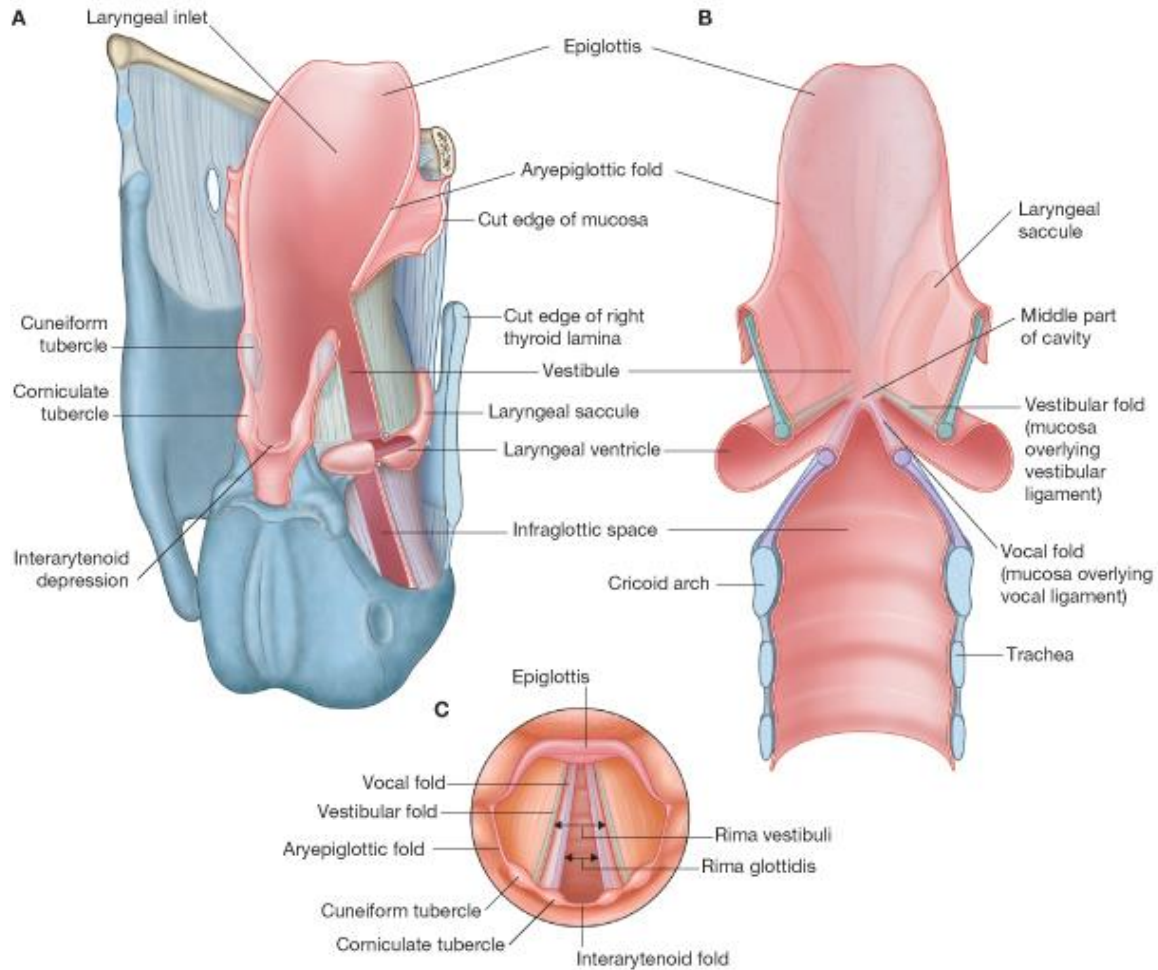
○ **Glottis:**

- Extends from superior surface of True vocal folds to 1cm below True vocal folds.
- **Vocal folds (True vocal folds):**
 - Two pearly-white strong bands extending from Thyroid angle to Vocal processes of Arytenoids.
 - Consists of a vocal ligament which is upper edge of Cricovocal membrane (Conus Elasticus) covered by closely bound mucous membrane with scanty subepithelial connective tissue.
 - Anterior commissure does not have an inner perichondrium which allows for Thyroid cartilage invasion in cases of Laryngeal ca.
- **Glottis (Rima Glottidis):**
 - Elongated space between vocal folds anteriorly, and vocal processes and base of Arytenoids posteriorly.
 - Narrowest part of laryngeal cavity in Adults.
 - **Anteroposteriorly:**
 - 24 mm in men
 - 16 mm in women.
 - **Anterior 2/3 of glottis:**
 - Formed by membranous cords
 - **Posterior 1/3 of glottis:**
 - Formed by Vocal processes of Arytenoids.

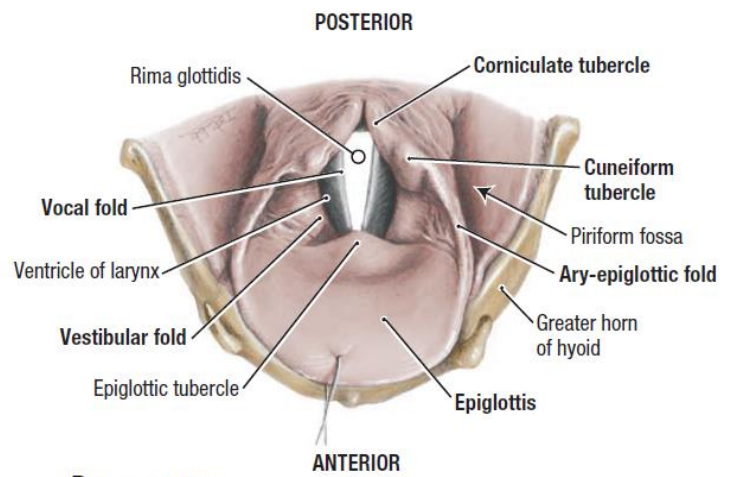


○ **Subglottis:**

- Extends from 1 cm below True vocal folds to inferior Cricoid cartilage.
- Narrowest part of laryngeal airway in pediatrics.



A. Laryngoscopic Examination



B. Superior View

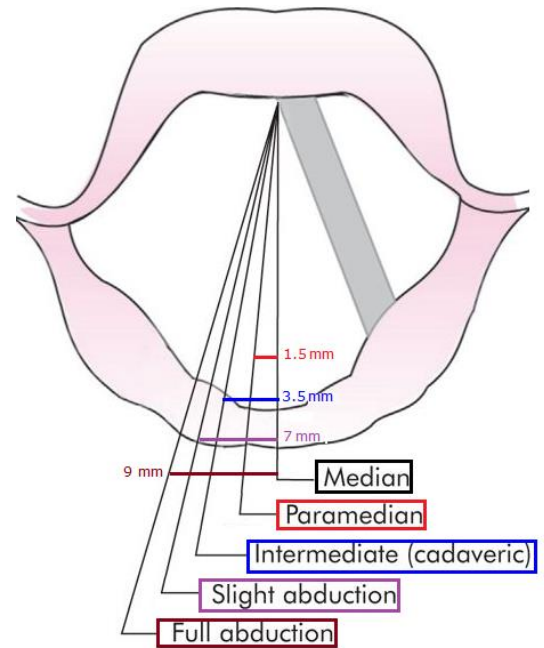
- **Positions of Vocal folds:**
- Does not necessarily predict site of the lesion.
- Caused mainly by degree of Reinnervation and Synkinesis.

1. Median Position:

- In the Midline.
- Normally occurs during:
 - Phonation.

2. ParaMedian Position:

- 1.5 mm from Midline.
- Normally occurs during:
 - Strong whispering.
- Abnormally occurs during:
 - Isolated RLN Paralysis.
 - Partial ADduction is preserved from:
 1. CricoThyroid (Intact SLN).
 2. InterArytenoid (Receives Bilateral innervation)



3. Cadaveric (Intermediate) Position:

- 3.5 mm from Midline.
- Neutral position of Cricoarytenoid joint.
- ABduction and ADduction take place from this position
- Abnormally occurs during:
 - Complete Paralysis of both RLN and SLN.

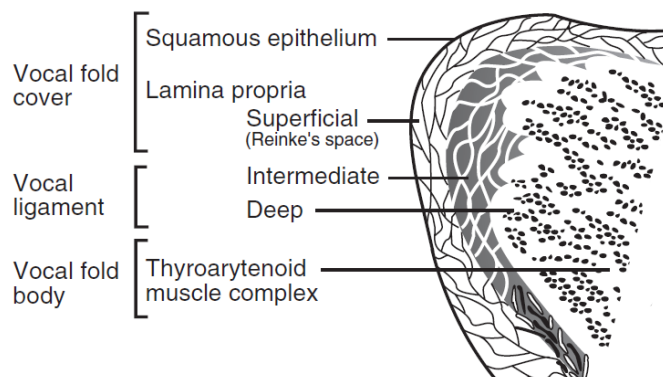
4. Gentle ABduction Position:

- 7 mm from Midline.
- Normally occurs during:
 - Quiet Respiration.

5. Full ABduction Position:

- 9.5 mm from Midline.
- Normally occurs during:
 - Deep Respiration.

- **Mucous Membrane of Larynx:**
 - Lined mainly with respiratory epithelium:
 - Ciliated pseudostratified columnar epithelium with goblet cells.
 - **EXCEPT:**
 - Lingual surface of Epiglottis
 - True Vocal folds
 - Both lines with Stratified squamous epithelium.
 - Mucous glands are distributed all over the mucous lining and are particularly numerous on:
 - Posterior surface of Epiglottis
 - Posterior part of Aryepiglottic folds
 - Sacculi.
 - **NO mucous glands in the vocal folds.**
-
- **Vocal Fold Layers:**
 - **Vocal Fold Cover:**
 - 1. **Stratified Squamous Epithelium:**
 - Non-keratinizing.
 - 2. **Superficial Lamina Propria (Reinke's Space):**
 - Loose fibrous matrix (few fibroblasts).
 - Gelatinous consistency permits fluency of vocal fold vibration (mucosal wave).
 - Lacks of lymphatics and blood vessels which permits rapid tumor extension.
 - Edema of this space causes fusiform swelling of the membranous cords (Reinke's edema).
 - **Vocal Ligament:**
 - 1. **Intermediate Lamina Propria:**
 - Elastin (some fibroblasts).
 - 2. **Deep Lamina Propria:**
 - Fibroblasts and collagen (dense).
 - **Vocal Fold Body:**
 - 1. **ThyroArytenoid Muscle Complex:**
 - Thyroarytenoid and Vocalis muscle.



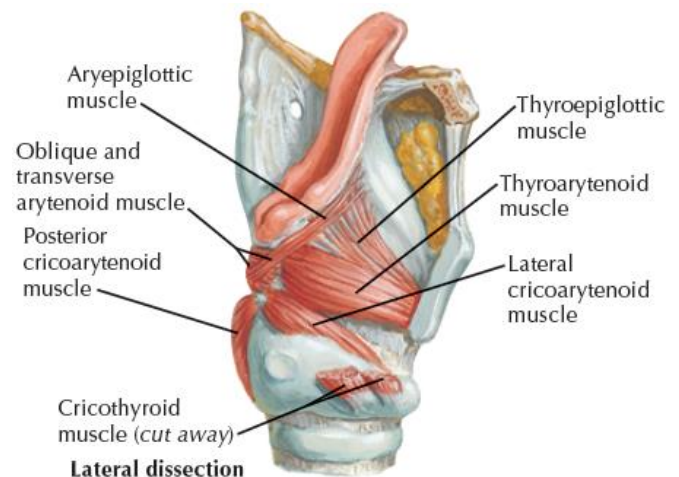
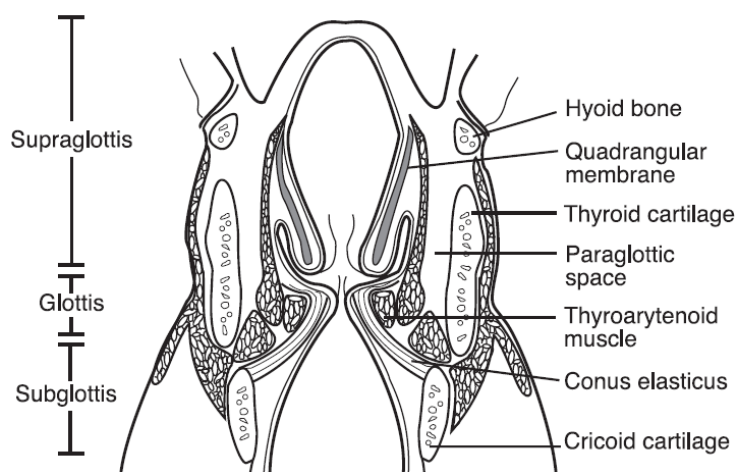
- **Spaces of Larynx:**

○ **Pre-Epiglottic space:**

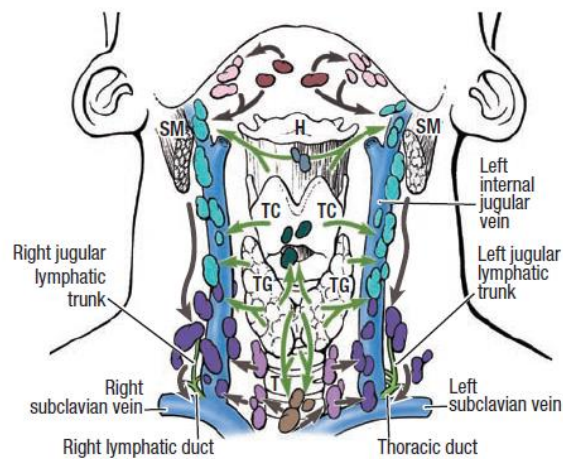
- Midline fibrofatty-filled space.
- Continuous laterally with Paraglottic space.
- Bounded by:
 - Anteriorly → Hyoid bone + Thyroid cartilage + ThyroHyoid membrane.
 - Posteriorly → Epiglottis + Quadrangular membrane.
 - Superiorly → HyoEpiglottic ligament
- Tumor may enter into Pre-Epiglottic space from Anterior commissure or supraglottic extension.

○ **Para-Glottic space:**

- Fibrofatty-filled space.
- Located outside of Conus Elasticus and Quadrangular membrane.
- Continuous with Pre-Epiglottic space.
- Allows transglottic extension of Tumors.
- Bounded by:
 - Superomedially → Quadrangular membrane.
 - Mid-Medially → Ventricles.
 - Inferomedially → Conus Elasticus.
 - Laterally → Thyroid cartilage + CricoThyroid membrane.
 - Posteriorly → Pyriform fossa mucosa.
 - Inferior → Space between Thyroid and Cricoid cartilage.
- Tumors invade this space can present in the neck through CricoThyroid space.



- **Lymphatic Drainage of Larynx:**
- **Supraglottis:**
 - Lymphatics pierce ThyroHyoid membrane.
 - Drains into levels II, III, IV LN.
- **Glottis:**
 - No lymphatics in vocal folds.
 - Glottic Ca rarely shows lymphatic metastases.
 - Drains into levels VI (Pretracheal), II, III, IV LN.
- **Infraglottis:**
 - Lymphatics pierce CricoThyroid membrane
 - Drains into levels VI (Pretracheal), III, IV LN.

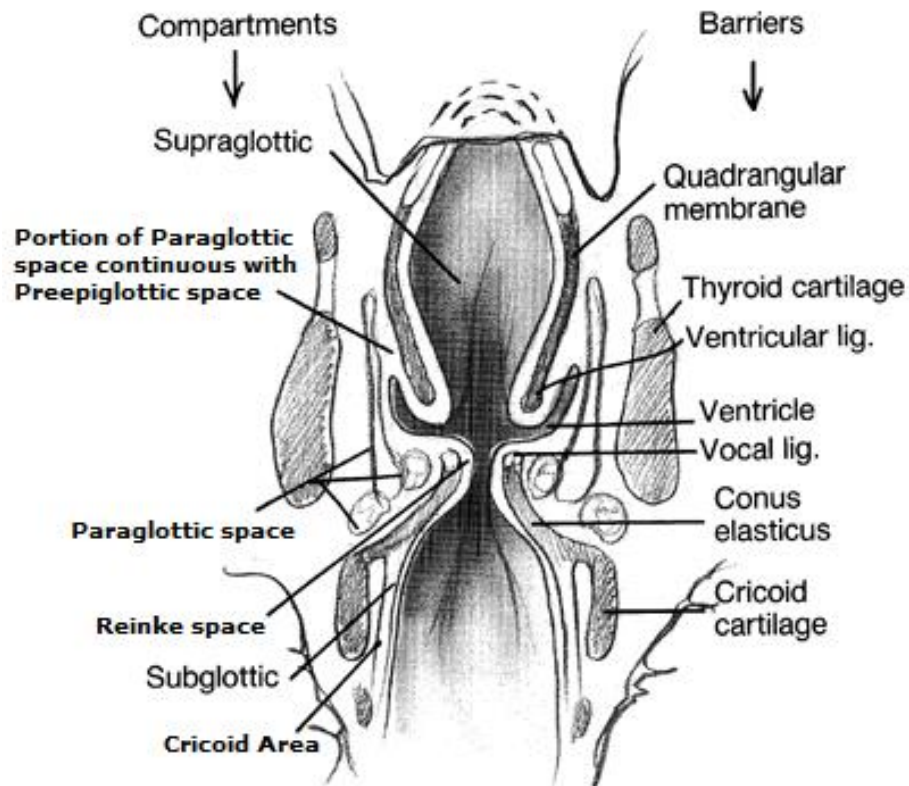


B. Anterior View

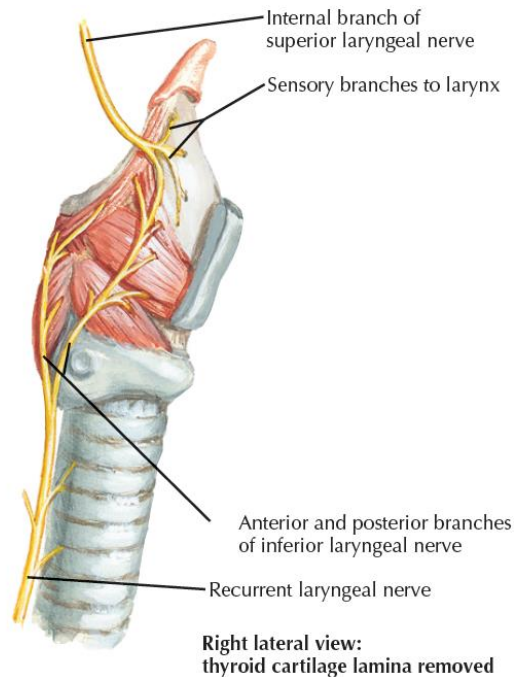
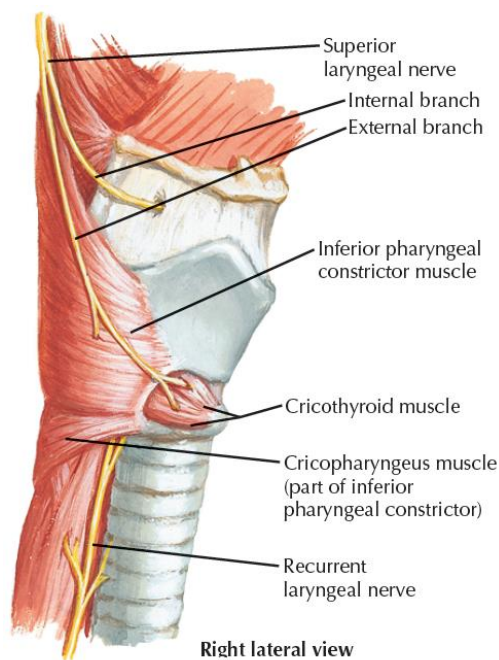
Lymph nodes:			
 Buccinator	 Paratracheal	 Superficial cervical	SM Sternocleidomastoid
 Inferior deep cervical	 Parotid	 Superior deep cervical	T Trachea
 Infrahyoid	 Prelaryngeal	Structures:	
 Jugulodigastric	 Pretracheal	→ Initial drainage	TG Thyroid gland
 Jugulo-omohyoid	 Retropharyngeal	→ Secondary (subsequent) drainage	P Palatine tonsil
 Mastoid (retro-auricular)	 Submandibular	H Hyoid	PG Parotid gland
 Occipital	 Submental		Ph Pharyngeal tonsil

- **Connective tissue barriers to Laryngeal Cancer spread:**

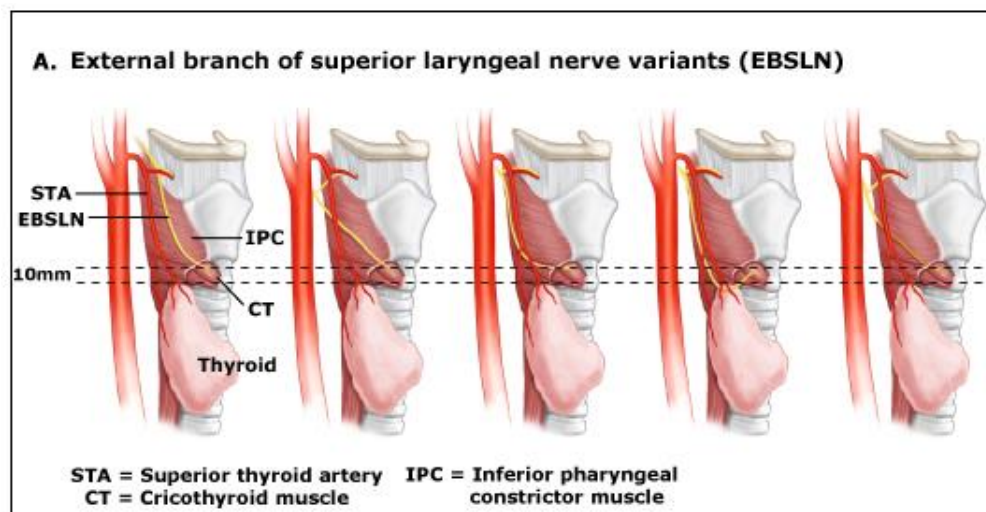
1. Thyroid Cartilage Perichondrium.
2. Cricoid Cartilage Perichondrium.
3. Conus Elasticus.
4. Quadrangular membrane
5. Ventricles
6. ThyroHyoid membrane
7. HyoEpiglottic ligament
8. Ventricular ligament

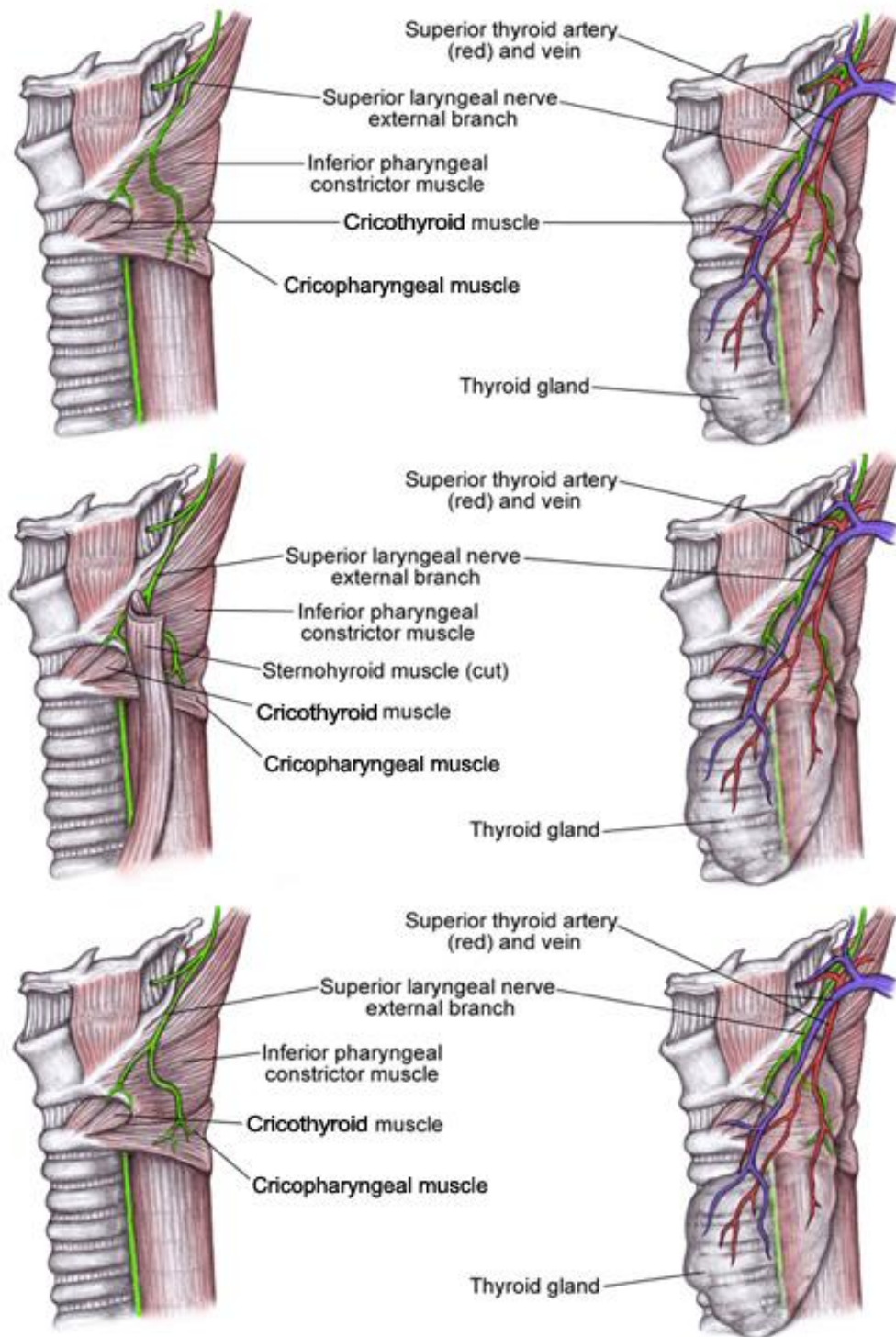


- **Motor Supply of Larynx:**
- **ALL** intrinsic muscles of larynx which move vocal folds are supplied by Recurrent Laryngeal Nerve (**RLN**):
 - Posteriore CricoArytenoid (**ABdcutor**)
 - Lateral CricoArytenoid. (**ADdcutor**)
 - InterArytenoid (**ADdcutor**)
 - ThyroArytenoid (**ADdcutor**)
 - Vocalis (**Tensor**)
- **EXCEPT:**
 - CricoThyroid (**Main Tensor + ADdcutor**)
 - External branch of Superior Laryngeal Nerve (**SLN**).
- **Sensory Supply of Larynx:**
- Laryngeal mucosa Above Vocal folds:
 - Internal branch of Superior Laryngeal Nerve (**SLN**).
- Laryngeal mucosa Below Vocal folds:
 - Recurrent Laryngeal Nerve (**RLN**).

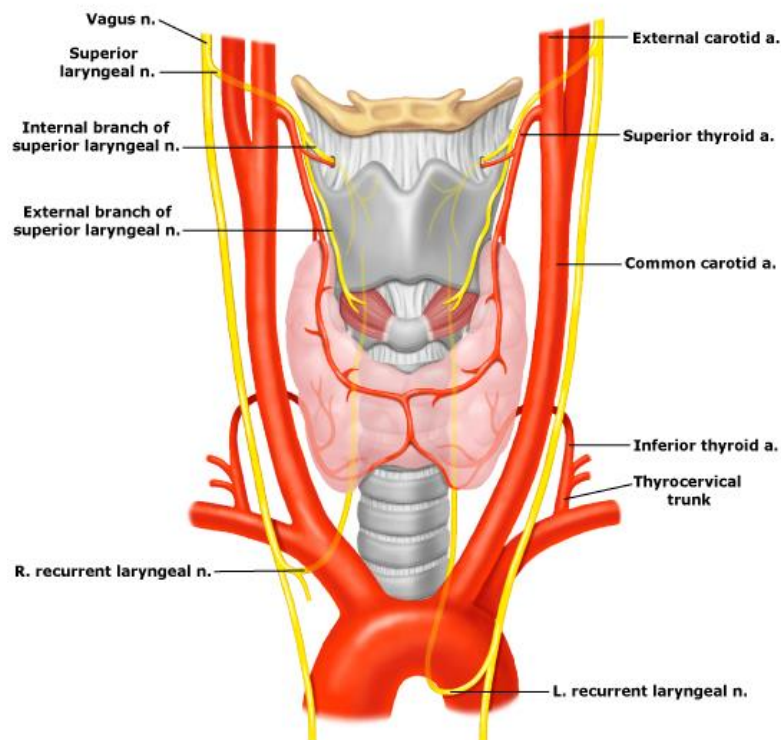


- **Superior Laryngeal Nerves (SLN):**
 - Originate from vagus nerve as it exits base of the skull.
 - Runs transversally behind the carotid artery.
 - Courses with Superior thyroid artery until approximately 1 cm before the artery enters the capsule of Superior pole of the thyroid.
 - Two primary branches:
 - **External branch:**
 - Primarily motor:
 1. Inferior constrictor muscle.
 2. Cricothyroid muscle.
 - Travels with Superior thyroid artery until approximately 1 cm before the artery enters superior thyroid pole.
 - Divides into branches that enter Lateral inferior pharyngeal constrictor muscle and Cricothyroid muscle.
 - **Internal branch:**
 - Sensory to Supraglottis and Glottis.
 - Afferent branch of cough reflex arc.
 - Enters the larynx through Thyrohyoid membrane superior to the external branch.





- **Recurrent Laryngeal Nerve (RLN):**
 - Provides both sensory and motor function to Larynx.
 - Sensory to Subglottis and Trachea.
 - Innervates all muscles to the larynx Except Cricothyroid muscle.
 - **Course of Left RLN:**
 - Originates from Left Vagus nerve below Arch of Aorta in the thorax.
 - Loops posterior to Arch of Aorta
 - **Course of Right RLN:**
 - Originates from Right Vagus nerve at level of Subclavian Artery.
 - Loops posterior to Subclavian Artery.
 - **Common Course:**
 - Associated with Inferior thyroid artery at junction of lower and middle thirds of thyroid gland.
 - Posterior to Inferior Thyroid Artery (**40%**).
 - Between branches of the artery (35%)
 - Anterior to Inferior Thyroid Artery (20%)
 - Ascends to the side of Trachea at Tracheoesophageal groove.
 - Terminal portion of RLN accompanies Laryngeal branch of Inferior thyroid artery to enter the larynx behind Cricothyroid joint.
 - In 90% of cases, RLN divides into two to three branches just before entering the larynx deep to inferior constrictor muscle.



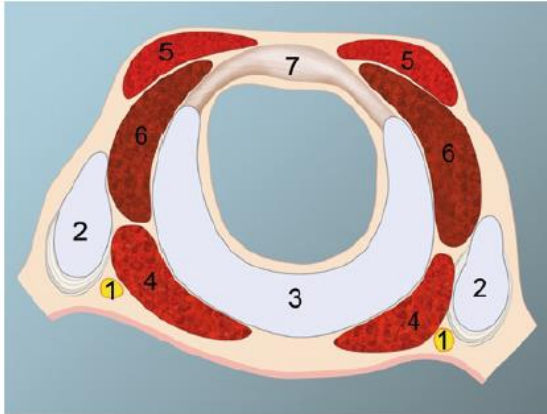
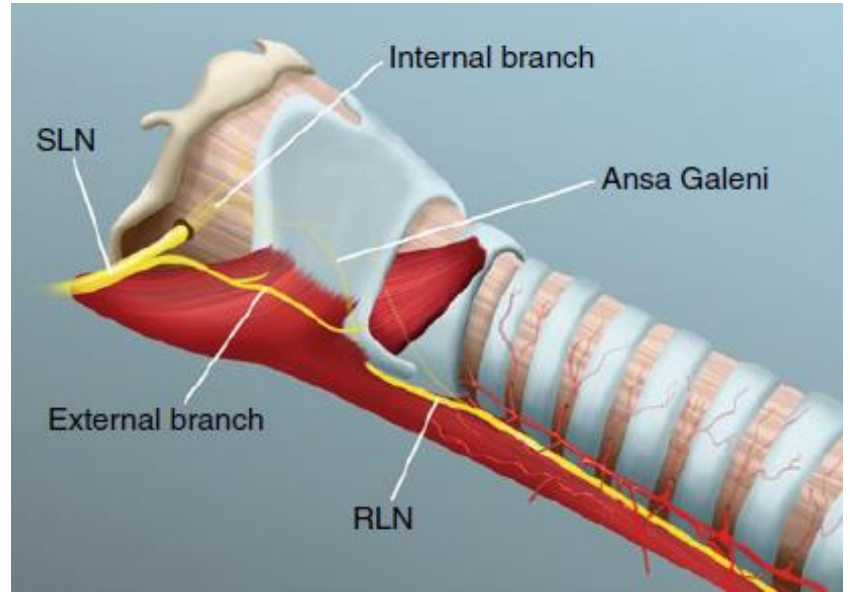
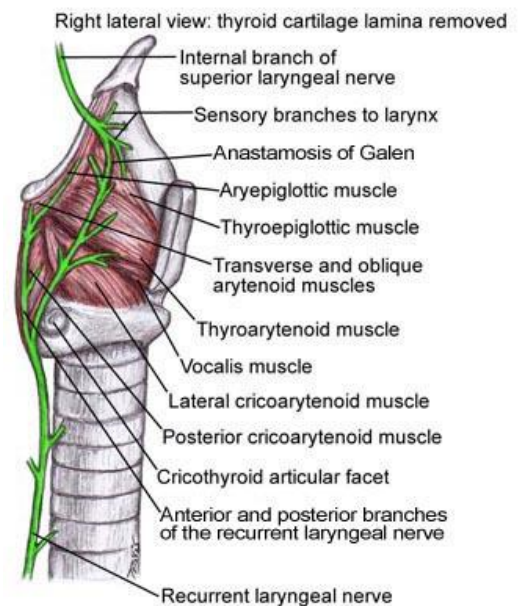


Fig. 2.10 Relationship of the RLNs with the cricothyroid joint: horizontal section at the level of the cricothyroid membrane (diagram): (1) RLN, (2) cricothyroid joint, (3) cricoid plate, (4) posterior cricoarytenoid muscle, (5) cricothyroid muscle, (6) lateral cricoarytenoid muscle and (7) cricothyroid membrane. The RLNs are located immediately behind the cricothyroid joints



- **Galen Anastomosis (Ansa of Galen):**
- Connection between RLN and internal branch of SLN.
- Provide purely sensory and autonomic innervation.
- May also contain motor fibers.
- Present as a single nerve, single trunk, several branches, or a plexus.



- **Blood Supply:**

1. Superior Thyroid Artery:

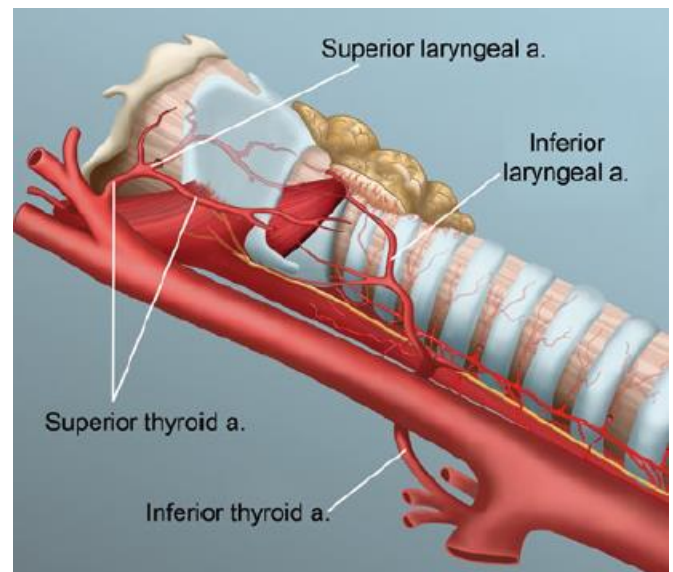
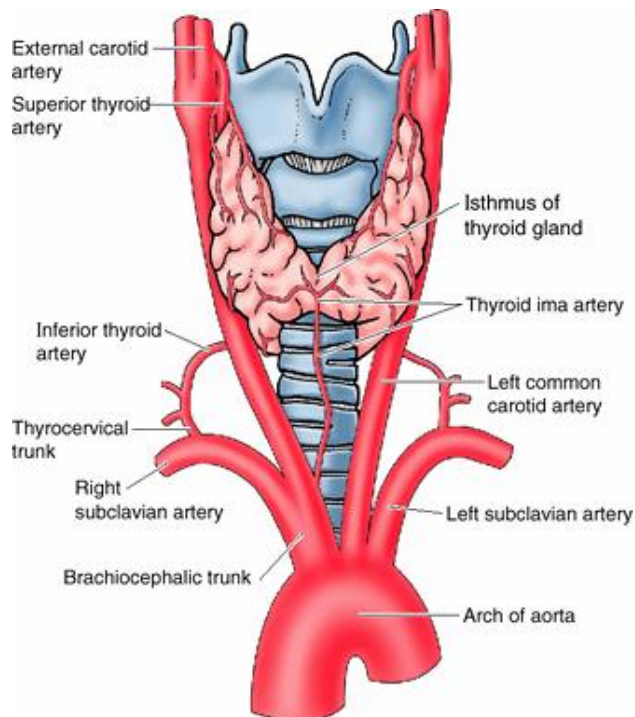
- 1st branch of External Carotid Artery.
- Arises just below Greater cornu of Hyoid bone.
- Descends along Inferior constrictor muscle to reach upper pole of Thyroid gland.
- Cephalic to Upper pole, External branch of Superior Laryngeal Nerve runs with Superior Thyroid Artery before turning medially to supply Cricothyroid muscle.
- Provides blood supply to Supraglottis and Glottis.
- Gives rise to:

1. Superior Laryngeal Artery:

- Prices Thyrohyoid membrane to supply the larynx.
- Accompanied with Internal branch of Superior laryngeal nerve.

2. Cricothyroid Artery:

- Runs cephalic to upper pole and runs toward the midline on the cricothyroid ligament.
- Lacerated during emergent cricothyroidotomy.



2. Inferior Thyroid Artery:

- Branch of Thyrocervical trunk of Subclavian Artery.
- Ascends vertically along Medial border of Anterior Scalene Muscle to enter Tracheoesophageal groove.
- Penetrates posterior aspect of Lower lobes of Thyroid.
- Provides the blood supply to Glottis , Subglottis and Cervical Trachea.
- Gives rise to:

1. Inferior Laryngeal Artery:

- Reaches posterior border of larynx at level of Cricothyroid joint.
- Lies immediately deep to Inferior constrictor muscle.
- Travels beside Recurrent laryngeal nerve.

- Venous Drainage:

1. Superior Thyroid Vein:

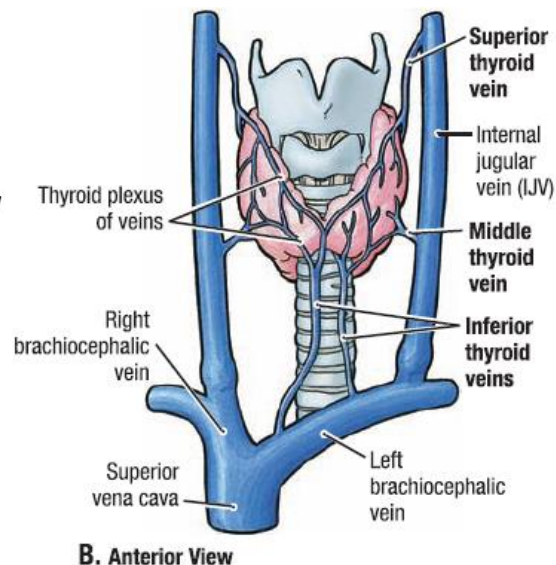
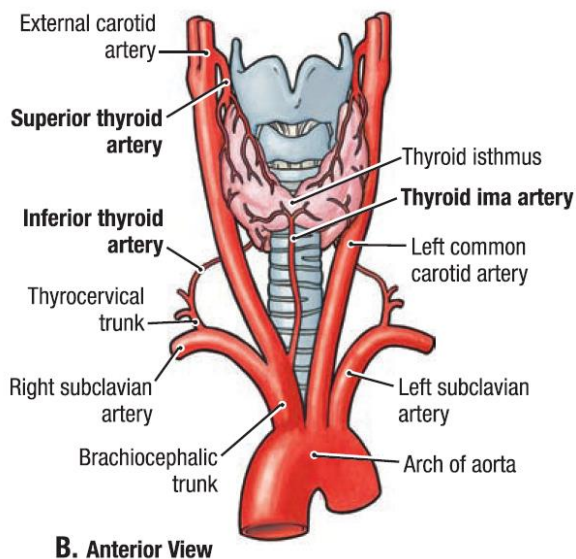
- Ascends along Superior Thyroid Artery.
- Drains into Internal Jugular Vein.

2. Middle Thyroid Vein:

- Follows a direct course laterally to drain into Internal Jugular Vein.

3. Inferior Thyroid Vein:

- Drains into Left Brachiocephalic Vein.
- Occasionally, both inferior veins form a common trunk called Thyroid ima vein, which empties into the left brachiocephalic vein.



- **Physiology of Larynx:**

- The larynx performs the following important functions:
 1. Protection of lower airways.
 2. Phonation.
 3. Respiration.
 4. Fixation of the chest.

1. Protection of Lower Airways:

- Earliest function to develop.
- Mechanism:

1. Sphincteric closure of Laryngeal Airway:

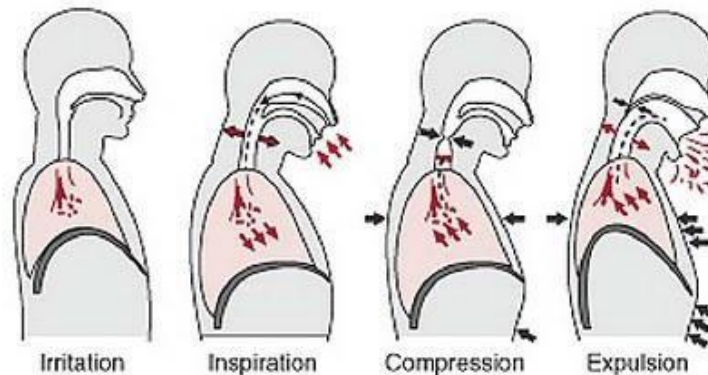
1. Laryngeal inlet.
2. False vocal cords.
3. True vocal cords (**Most Significant**)

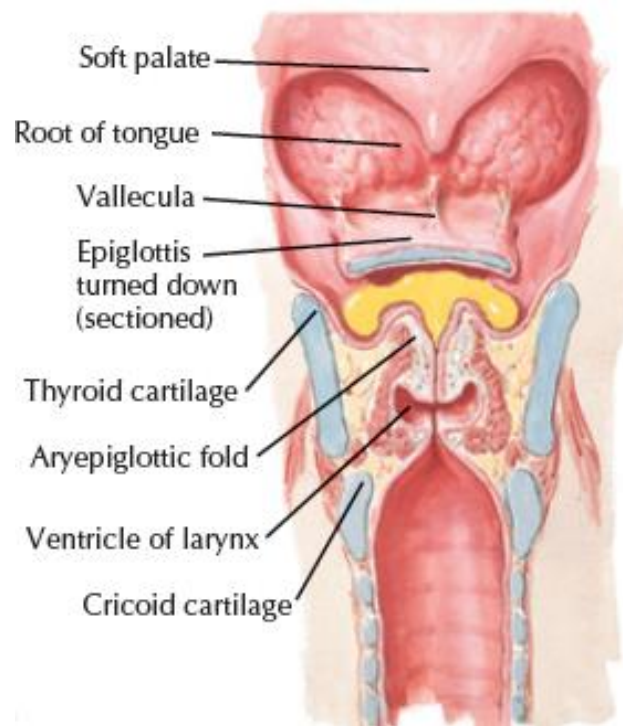
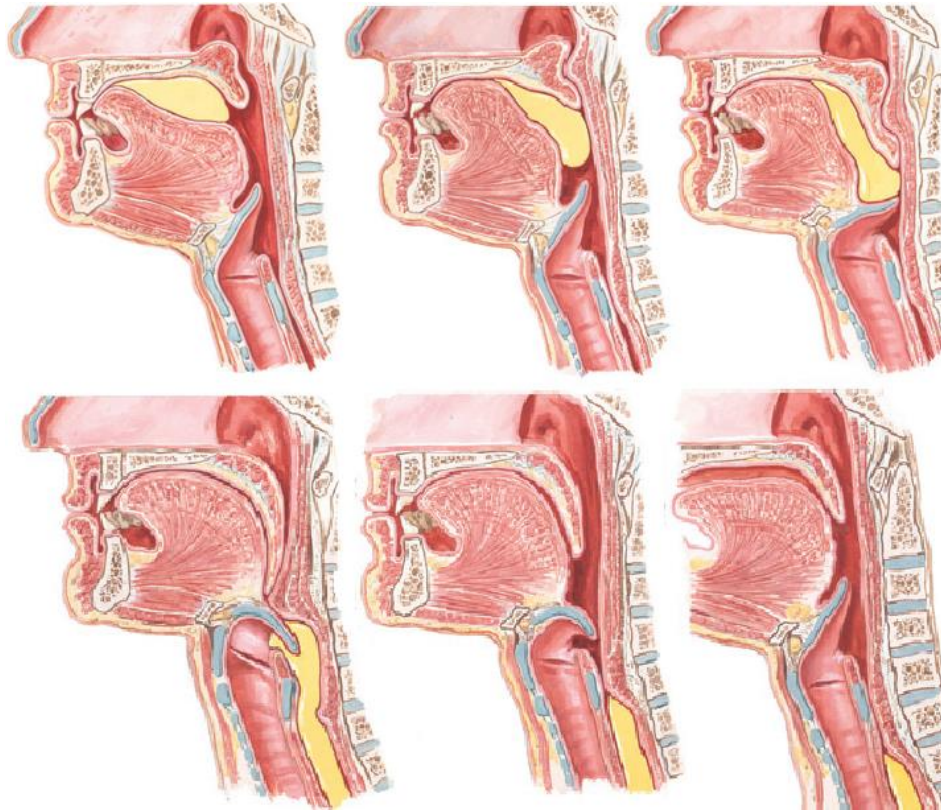
2. Cessation of respiration:

- Reflex generated by afferent fibers of **CN-IX**, when food comes in contact with posterior pharyngeal wall or the base of tongue.

3. Cough Reflex:

- Important mechanism to expel a foreign particle when it comes into contact with respiratory mucosa.
- Reflex generated by afferent fibers of **SLN**.
- Three Phases:
 - Larynx opens very widely to permit rapid and deep inspiration.
 - Tight closure of the glottis and strong activation of expiratory muscles.
 - Larynx suddenly opens widely, with a sudden outflow of air.

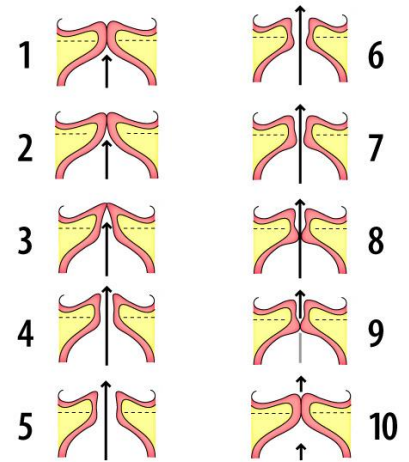




- **Laryngeal Reflexes:**
- Elicited by mechanical stimulation of the larynx.
- Because larynx is in a very protected position, such responses do not usually occur naturally.
 - 1. **Laryngeal ADductor Reflex:**
 - Rapid brief closure of True vocal folds in response to stimulation of mucosa innervated by SLN.
 - 2. **Laryngospasm:**
 - Exaggerated prolonged ADduction reflex due to laryngeal irritation caused by:
 - **Intubation or Extubation:**
 - When the patient is well oxygenated and under light anesthesia.
 - **URTI**
 - **Reflux**
 - **Foreign bodies**
 - **Mucus.**
 - Mediated by fibers of **SLN**.
 - 3. **Apnea:**
 - Prominent response to laryngeal stimulation.
 - Prevents aspiration of the stimulating material into the lower airway.
 - Caused by:
 - **Ammonia**
 - **Cigarette smoke.**
 - **Water:**
 - Typical response to water in normal conscious adult is vigorous coughing.
 - Apnea occurs only in the followings:
 - If the patient is under GA.
 - During URTI.
 - In infants.
 - 4. **Cardiovascular Reflex:**
 - Stimulation of larynx can produce changes in HR and BP.
 - Occurs mainly during:
 - **Endotracheal intubation:**
 - Direct result of laryngeal stimulation is hypertension.
 - If laryngeal stimulation produces significant Bradycardia , Indirect result can be Hypotension.
 - **Obstructive sleep apnea:**
 - Narrowing of upper airway results in increase in negative airway pressure which stimulates receptors in the larynx causing Cardiac arrhythmias.

2. Phonation:

- Larynx is like a wind instrument.
- Myoelastic-Aerodynamic theory of voice production:
 - True vocal folds are Adducted and tensed.
 - → Increase subglottic pressure by exhaled air from the lungs due to contraction of thoracic and abdominal muscles.
 - → Air force open the cords and is released as small puffs which vibrate the vocal cords and produce sound.
 - → Glottis opens from an inferior to superior direction
 - → Glottis closes from an inferior to superior direction



- This sound is converted into speech by the modulatory action of lips, tongue, palate, pharynx, and teeth.
- Intensity of sound depends on the air pressure produced by the lungs.
- Pitch of sound depends on the frequency with which the vocal cords vibrate.

3. Regulation of Airflow:

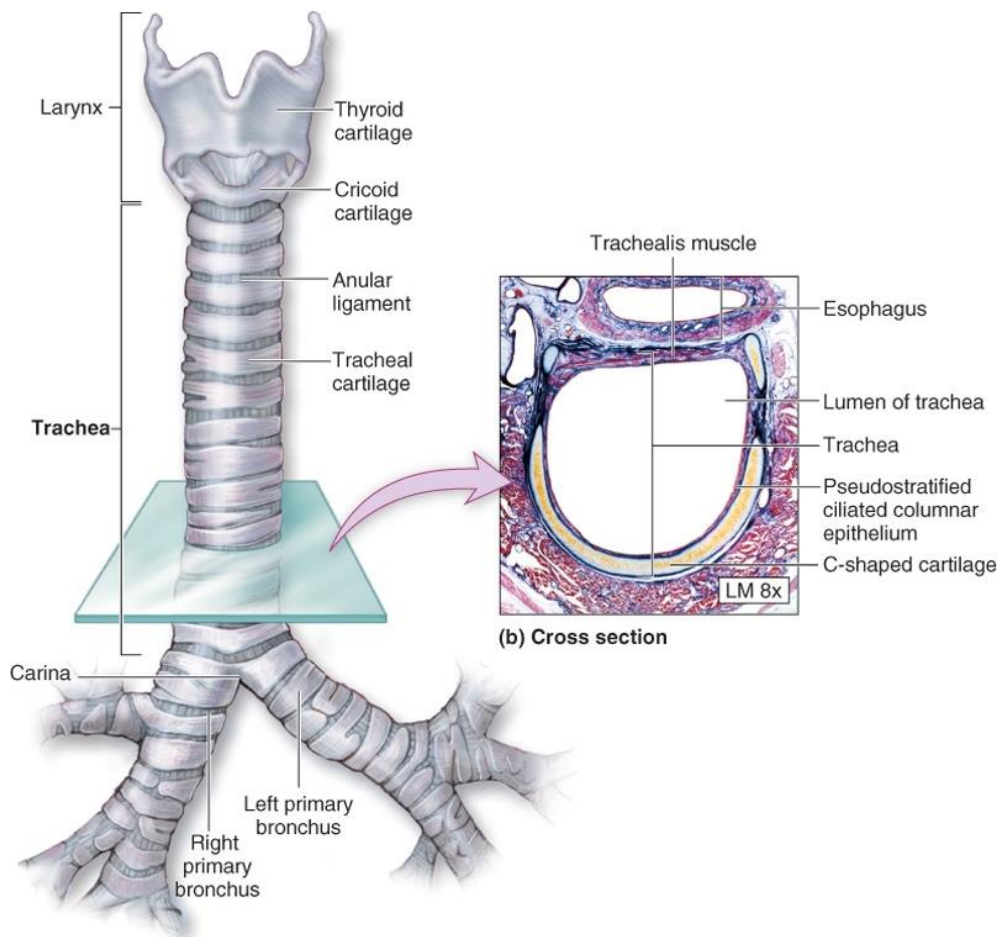
- Larynx regulates airflow into the lungs.
- During Respiration at Rest:
 - Laryngeal glottis is opened passively.
 - Air comes in and out without active participation of the larynx.
- During High-volume Respiration:
 - Laryngeal glottis is widely open by Active ABduction of PTC.

4. Fixation of the Chest (Valsalva Maneuver):

- Forced expiration against a tightly closed glottis.
- When larynx is closed, chest wall gets fixed and various thoracic and abdominal muscles can then act best.
- This function is important in digging, pulling and climbing.
- Coughing, vomiting, defaecation, micturition and childbirth also require a fixed thoracic cage against a closed glottis.

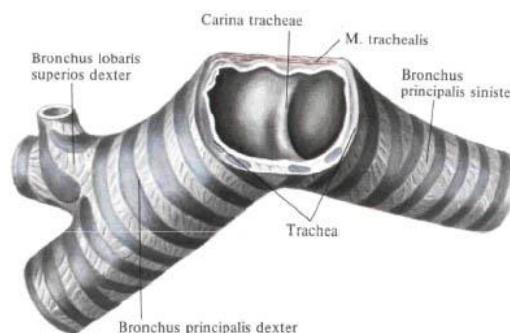
Tracheal Anatomy:

- Trachea is nearly cylindrical, flattened posteriorly.
- D-shaped in cross section:
 - o Anteriorly and Laterally: Incomplete cartilaginous rings.
 - o Posteriorly: Straight membranous wall.
- Starts from inferior border of Cricoid cartilage of larynx in the neck, opposite to C6.
- Ends at the Carina in the thorax where it divides into the right and left bronchi, opposite to T4-5.
- Trachea measures 11 cm in length.
- Its inner diameter is 25 mm.
- Mucous membrane is continuous with larynx above and intrapulmonary bronchi below.
 - o Pseudostratified ciliated columnar epithelium with numerous goblet cells.



- **Tracheal Cartilages:**

- 15-20 U-shaped rings.
- Hyaline cartilages.
 - Highly elastic but may become calcified in advanced life.
- Responsible for the lateral rigidity of the Trachea.
 - Hold the airway open and give it flexibility.
 - Prevent collapse of the conducting pathway.
- Each of the cartilages is enclosed in perichondrium.
- Tracheal cartilages are placed horizontally above each other.
 - Separated by narrow intervals.
 - Tracheal annular ligaments join the tracheal rings together.
- Outer surfaces are flattened in a vertical direction.
- The internal surfaces are convex.
- Tracheal cartilages are thicker in middle than at the margins.
- 1st Tracheal Cartilage:
 - Broader than the rest.
 - Connected with inferior border of cricoid cartilage by CricoTracheal ligament.
- Last Tracheal Cartilage:
 - Thick and broad in the middle.
 - Its lower border is prolonged into a triangular hook-shaped process, which curves downward and backward between the 2 bronchi.
 - It ends on each side in an imperfect ring, which encloses the commencement of the bronchus. The cartilage above the last is somewhat broader than the others at its center.
- Carina:
 - At the bottom of the trachea.
 - Keel-like partition.
 - Separating the 2 bronchi.
 - Right bronchus is larger diameter in and more direct continuation of the trachea than the left.
 - During intubation, if ETT is pushed beyond the carina, it will enter the right side.
 - Also foreign bodies in the trachea fall more frequently into the right bronchus.



- **Membranous wall (Pars Membranacea):**

- Tracheal cartilage rings are deficient posteriorly.
- Composed of bands of fibroelastic tissue and **Trachealis muscle**:
 - Non-striated smooth muscle.
 - Connects the ends of the incomplete rings posteriorly:
 - Allows for esophageal dilatation during passage of food or liquid.
 - Contracts during coughing to reduce the size of the lumen of the trachea aiming to increase the rate of air flow.
 - Supplied by Vagus nerve (CN-X) and upper thoracic spinal nerves.

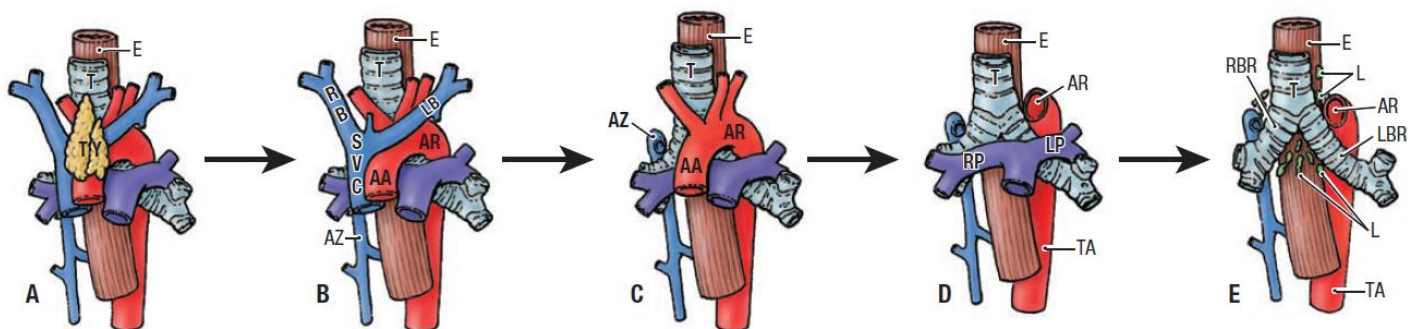
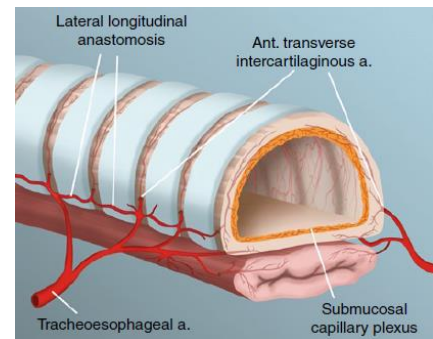
- **Blood supply of Trachea:**

- **Cervical segment:**

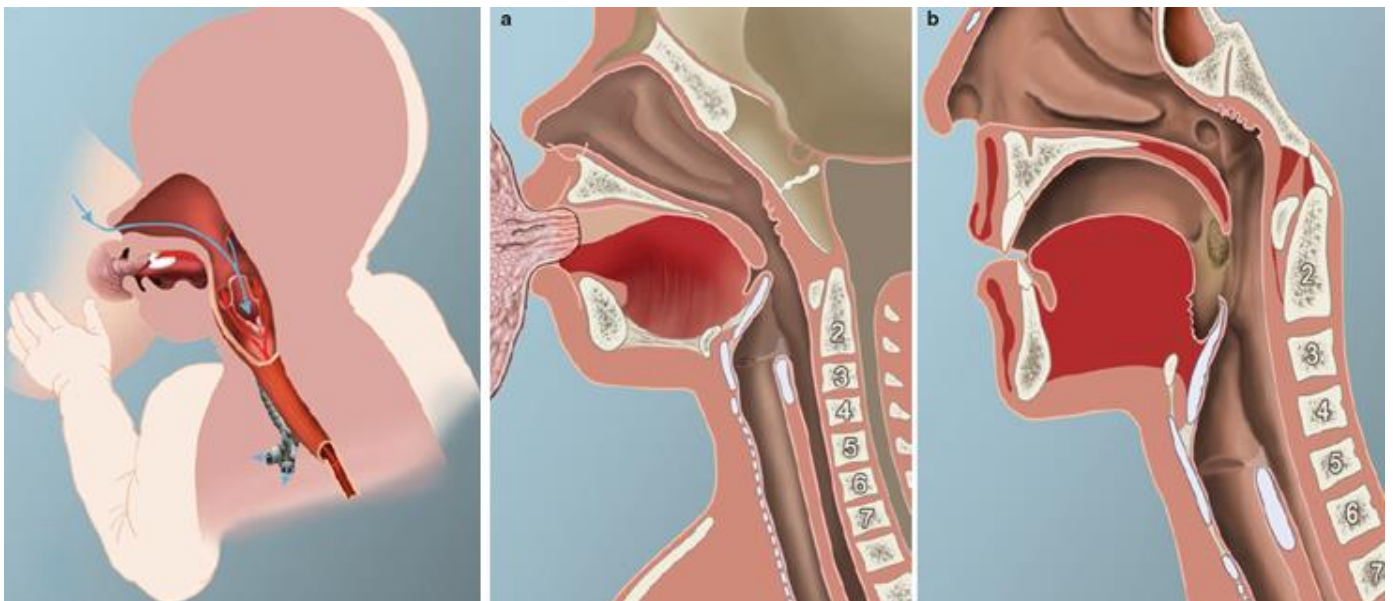
- Inferior thyroid artery.

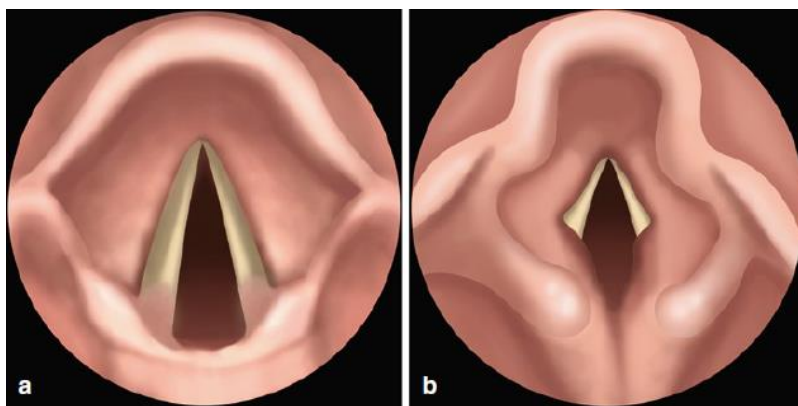
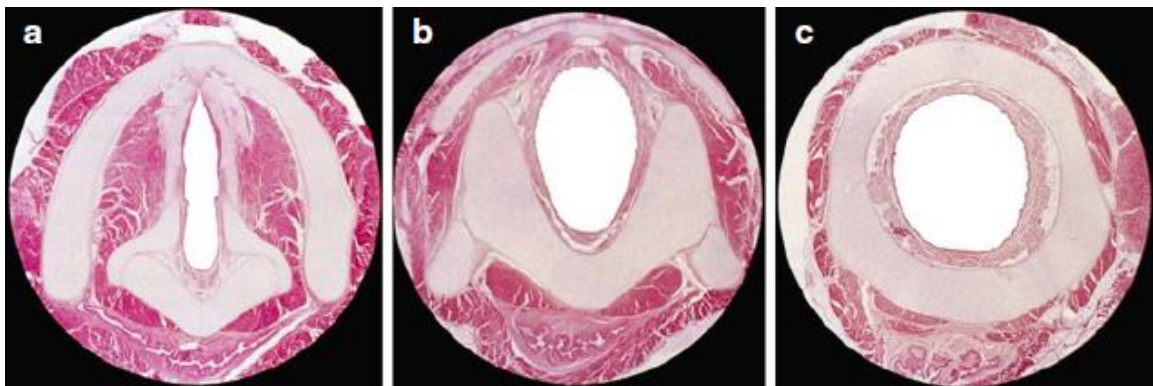
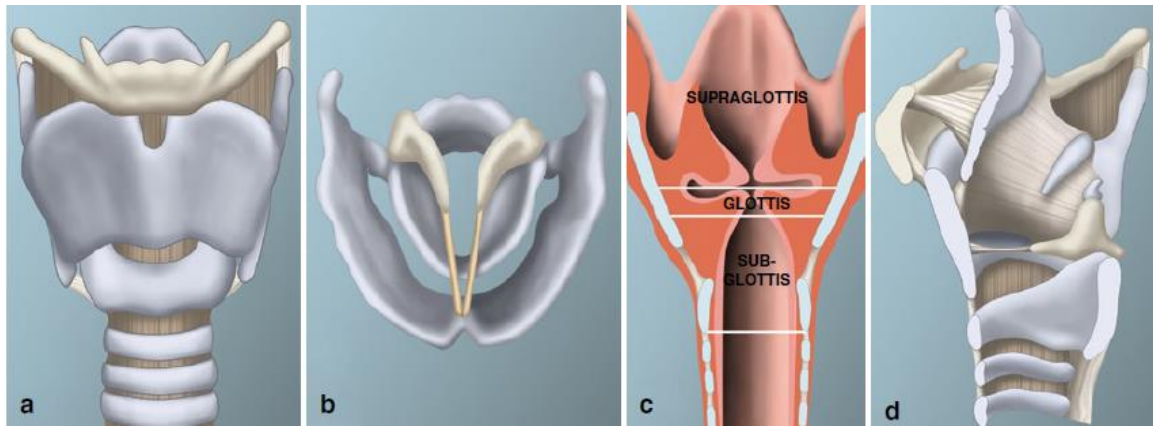
- **Thoracic segment:**

- Innominate-subclavian system.
- Bronchial arteries.



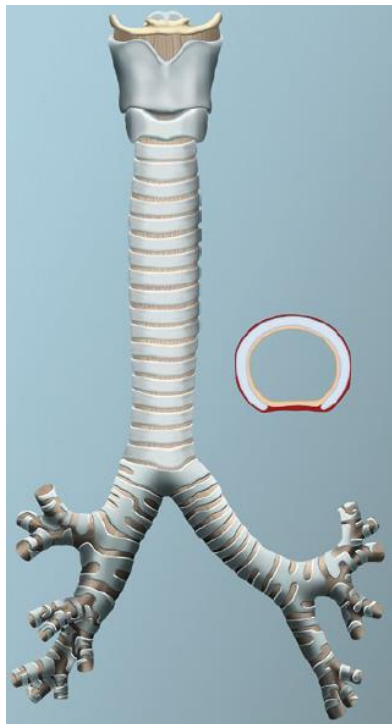
- **Pediatric Airway:**
- Infant larynx is different from the adult larynx by the following:
 1. Larynx is small and conical:
 - 1/3 size of the adult larynx.
 2. Positioned high in the neck:
 - Opposite to C3-C4 at rest
 - Reaches C1-C2 during swallowing.
 - Allows Epiglottis to meet soft palate and make a nasopharyngeal channel for nasal breathing during suckling.
 - Milk feed passes separately over the dorsum of tongue and the side of epiglottis, thus allowing breathing and feeding to go on simultaneously.
 3. Laryngeal cartilages are soft and collapse easily.
 4. Epiglottis is omega-shaped.
 5. Aryepiglottic folds are shorter.
 6. Thyroid cartilage is more rounded.
 - Overlaps cricoid cartilage and hyoid bone.
 - CricoThyroid and ThyroHyoid membranes are narrow.
 7. Arytenoid cartilages are larger.
 - Comprising > 50% of glottic length until 3 years of age.
 8. Glottis length in a full-term newborn is 7 mm
 9. Posterior glottis width in a full-term newborn is 3–4 mm.
 10. Cricoid cartilage is smaller than size of glottis:
 - **Subglottis is the narrowest part.**
 - Upper half of infant cricoid is V-shaped (Elliptical) and becomes rounded at its lower level.
 - Normal subglottic diameter:
 - 4.5 mm in Full-term neonate.
 - 3.5 mm in Premature neonate.
 11. Laryngeal mucosa are lax and more prone to edema when inflamed or injured.



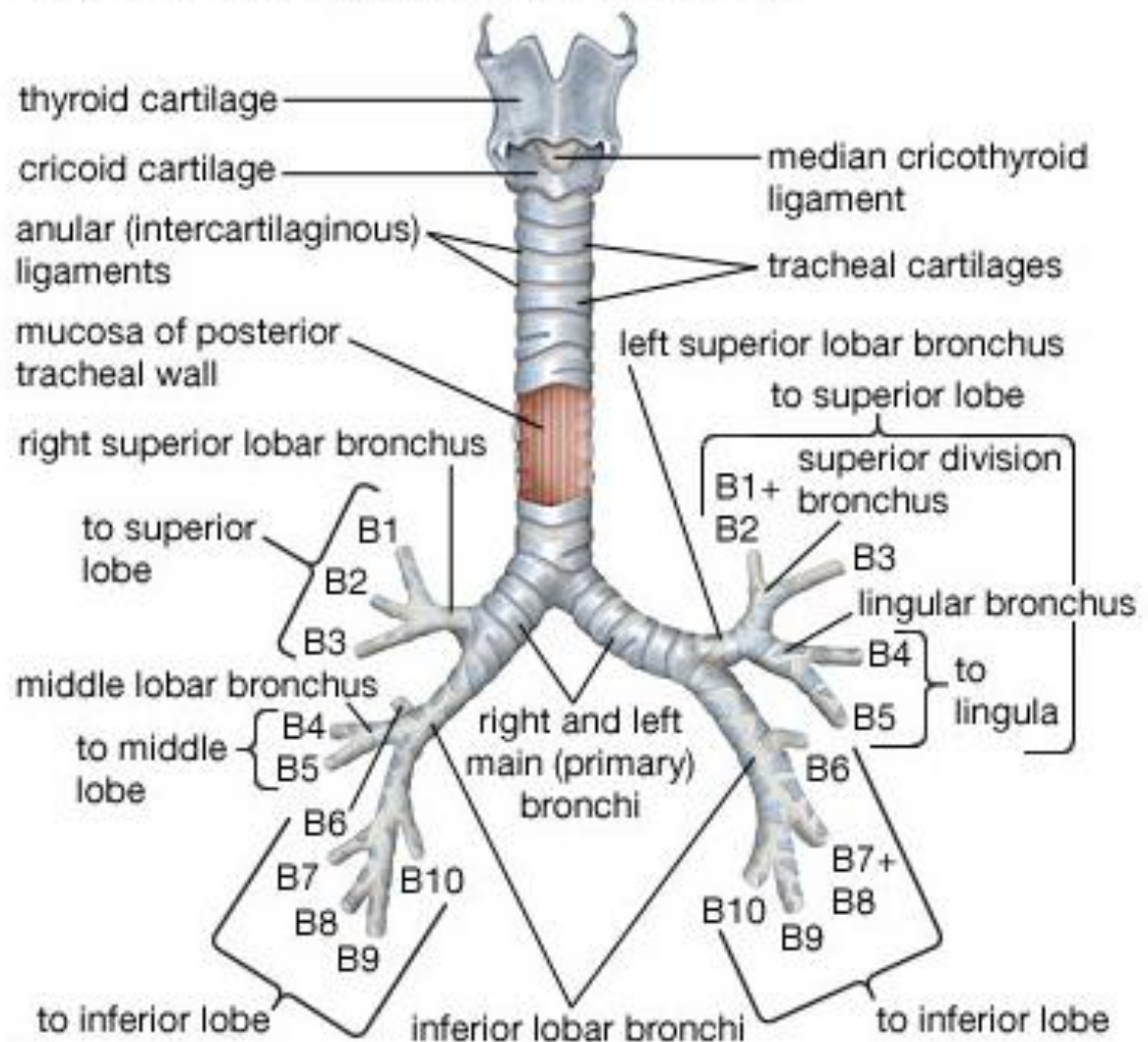


- Infant's larynx shows two spurts in growth.
 - **1st 3 years of life:**
 - Larynx grows in width and length.
 - Obviates the need for any airway surgery in certain congenital anomalies.
 - **Adolescence:**
 - Thyroid angle develops.
 - Length of vocal cords increases.
 - In the female its increase after puberty is only slight.
 - In the male it undergoes considerable increase:
 - All the cartilages are enlarged.
 - Thyroid cartilage becomes prominent in the middle line of the neck.
 - Length of Rima glottidis is nearly doubled.
 - Voice changes to one of lower pitch.
 - With growth of the neck, larynx gradually descends to adult; the vocal cords lying opposite C5.

- Pediatric Trachea has the same overall configuration as the normal adult trachea (15–20 horseshoeshaped tracheal rings), **EXCEPT** for its size:
 - More than doubles in length
 - Triples in diameter
 - Increases by 6-fold in cross-sectional area



Trachea and major bronchi of the lungs



Voice Production:

- Normal speech consists of 3 processes:

1. Phonation.
2. Resonance.
3. Articulation.

- **Phonation:**

- Generation of sound by vibration of vocal folds.
- Sound is produced when Expiratory airflow induces vibration of free edges of vocal folds.
- Vocal fold position is a critical factor in phonation.

- Requirements for Sound Production:

- **Source of energy:**
 - Airflow from lungs.
- **Source of vibration:**
 - True vocal folds
 - Neoglottis
 - Articulator constrictions

- Requirements for Normal Phonation:

1. Normal VC shape.
2. Normal vibratory characteristics.
3. Sufficient subglottic pressure.
4. Proper approximation of VC
5. Appropriate control of VC tension and length.

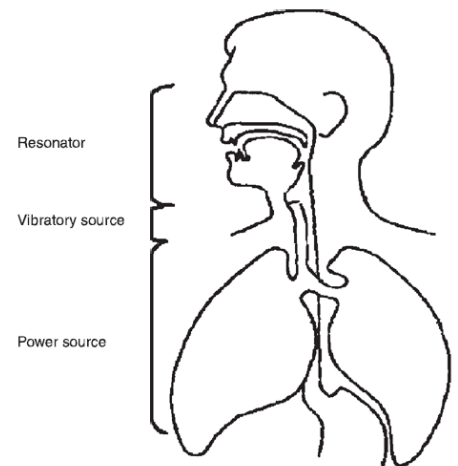


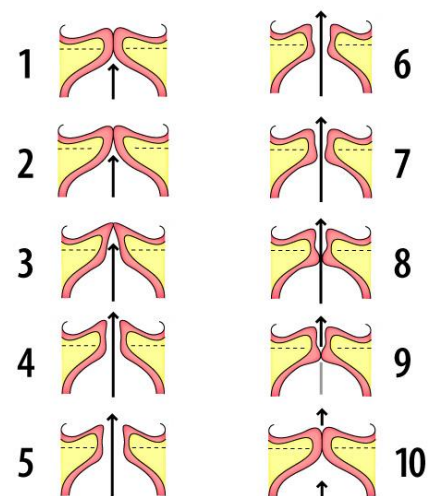
Figure 72.4 Vocal tract.

- Bernoulli's Effect:

- Forced air across a constricted zone produces negative pressure.
- Allows true vocal folds to be "sucked" back together.

- Myoelastic-Aerodynamic Theory:

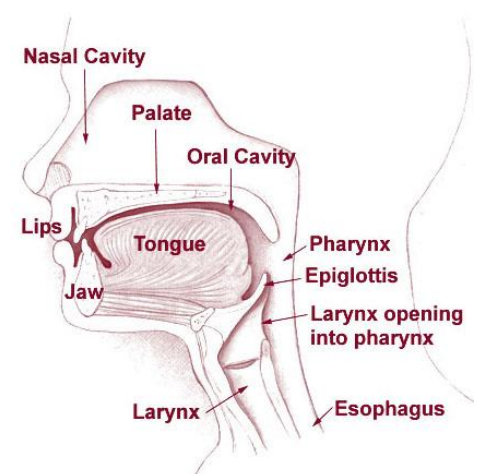
- True vocal folds are Adducted and tensed.
- → Exhalation cause increase subglottic pressure.
- → Vocal folds are pushed apart and Glottis opens from an inferior to superior direction.
- → Vocal folds then returned back to midline and Glottis closes from an inferior to superior direction as a result of:
 - Sudden decrease in subglottic pressure.
 - Elastic forces in vocal fold
 - Bernoulli effect.
- → Subglottic pressure builds once more and the cycle is repeated causing vocal folds vibration.



- Normal vocal fold vibration occurs in two direction:
 - Horizontally from medial to lateral along superior surface of vocal fold (Seen with Stroboscopy).
 - Vertically from inferior to superior (Not seen with Stroboscopy).
- During Normal conversational speech:
 - Driving force for phonation is Passive exhalation due to release of energy stored in the rib cage and diaphragm.
- During Loud and prolonged speech (singing or shouting):
 - Require deeper inspiration and contraction of abdominal and intercostal to permit louder and longer phonation.
- When vocal folds are loosely approximated:
 - Gap exists between the vocal folds.
 - Efficiency is markedly reduced.
 - Higher airflow is required to induce vibration.
 - Resulting voice is breathy and duration of phonation is shortened.
 - With a wider gap:
 - Airflow requirement exceeds the expiratory capacity.
 - Voice may dwindle to a whisper.
- Phonation Types:
 - **Modal phonation:**
 - Vocal folds are fully approximated.
 - Moderate longitudinal tension.
 - Phonation is regular, of moderate pitch, has rapid closures and a long closed phase duration.
 - **Whisper phonation:**
 - Vocal folds are tensed and rigid.
 - Held slightly apart.
 - Rigidity prevents the folds from vibrating, while the partially opened glottis forms a narrow opening which causes turbulence.
 - **Breathy phonation:**
 - Vocal folds are tensed appropriately for vibration but not fully approximated so that complete closures do not occur.
 - Air flow continues throughout the cycle which can lead to turbulence at the glottis.
 - Glottic closures is less sharp.
 - Vocal folds remain open for a longer portion of the cycle.

- **Creaky phonation:**
 - Vocal folds are lax but tightly approximated and this can lead to cycles which are closed for a longer proportion of the cycle and which are irregular in duration.
 - Creaky voice is commonly found at the bottom of a speaker's pitch range when the folds are slack anyway.
 - A common form of creaky voice is called Diplophonia, where long and short cycles alternate.
- **Falsetto phonation:**
 - Vocal folds are extremely tense and are held in such a way as that only the internal edges of the vocal folds are able to vibrate.
 - This means that the amplitude of phonation is small and of high fundamental frequency.
- **Resonance:**
- Phonatory output is modulated by resonance.
- Amplification and filtering of sound.
 - Induction of vibration in the chest, pharynx and head with selective amplification of certain component frequencies.
- Controlled by:
 - Altering shape and volume of pharynx (**Most significant**).
 - Raising or lowering the larynx.
 - Moving tongue or jaw position
 - Varying the amount of sound transmission through Nasopharynx and Nose.
- In English, ALL phonemes are produced with oral airflow that requires Velopharyngeal closure to exclude nasopharyngeal airflow.
- Exception of /**m**/, /**n**/, and /**ng**/ which require an open nasopharyngeal valve and nasal airflow to produce these sounds.

- **Articulation:**
- Formation of consonants and vowels.
 - Consonants are sounds where there is airflow obstruction caused by articulators.
 - Vowels are sounds where there is no airflow obstruction caused by articulators (**A, E, I, O, U**).
- Controlled by:
 - Lips
 - Jaw
 - Tongue
 - Velopharynx.



- Voice Parameters:

- **Loudness (dB):**
 - Amplitude of the pressure wave is perceived as loudness or sound intensity.
 - Simplest means of increasing loudness is to raise the subglottic air pressure by using forced or active exhalation.
 - Determined by:
 - Subglottic pressure
 - Glottal resistance
 - Airflow rate
 - Amplitude of VC vibration
 - Force of vocal contact
- **Pitch (Hz):**
 - Frequency of vocal folds vibration.
 - Determined by:
 - Vocal folds Length.
 - Vocal folds Tension.
 - Vocal folds speed of vibration.
- **Fundamental Frequency (Hz):**
 - Lowest frequency of all the individual waves that add up to create voice source spectrum.
 - Reflects rate at which vocal folds vibrate.
 - Fundamental frequency in males:
 - **128 Hz**
 - Long thick vocal folds.
 - Fundamental frequency in females:
 - **256 Hz**
 - Short thin vocal folds.
- **Quality (Timbre):**
 - Determined by:
 - Synchronicity of vocal folds vibration and glottal competence.

Evaluation of Dysphonic Patient:

- Dysphonia is change of voice from patient's habitual.
- Hoarseness is roughness & harshness of voice.
- Symptom not a diagnosis.

History:

- Character of Dysphonia:

- o Onset
- o Duration
- o Time course (Acute vs. Chronic).
- o Periodicity:
 - Morning hoarseness with GERD.
 - Evening hoarseness with vocal abuse.

- Associated Symptoms:

- o Stridor
- o Choking
- o Aspiration
- o Dysphagia
- o Odynophagia
- o Cough
- o FB sensation in throat
- o Frequent throat clearing
- o Postnasal drip
- o Heartburn
- o Neck mass
- o Weight loss
- o Hearing loss

- Contributing Factors:

- o Recent URTI
- o Voice Abuse
- o Patient's job
- o Smoking
- o Alcohol
- o GERD
- o Medications:
 - Steroid inhalers.
 - Warfarin.
- o Hypothyroidism.
- o Psychological stressors.
- o Laryngeal trauma.
- o History of Surgery (Cardiothoracic, Thyroid)
- o History of Intubation
- o History of Airway manipulation

- [Voice Handicap Index \(VHI\)](#):
 - 10-question questionnaire to quantify functional, physical and emotional impacts of a voice disorder on quality of life.
 - Good indicator for severity of voice pathology.

Table 62-1. Causes of hoarseness

1. Inflammations	
Acute	Acute laryngitis usually following cold, influenza, exanthematous fever, laryngo-tracheo-bronchitis, diphtheria
Chronic	(i) <i>Specific</i> . Tuberculosis, syphilis, scleroma, fungal infections
	(ii) <i>Non-specific</i> . Chronic laryngitis, atrophic laryngitis
2. Tumours	
Benign	Papilloma (solitary and multiple), haemangioma, chondroma, fibroma, leukoplakia
Malignant	Carcinoma
Tumour-like masses	Vocal nodule, vocal polyp, angiofibroma, amyloid tumour, contact ulcer, cysts, laryngocele
3. Trauma	Submucosal haemorrhage, laryngeal trauma (blunt and sharp), foreign bodies, intubation
4. Paralysis	Paralysis of recurrent, superior laryngeal or both nerves
5. Fixation of cords	Arthritis or fixation of cricoarytenoid joints
6. Congenital	Laryngeal web, cyst, laryngocele
7. Miscellaneous	Dysphonia plica ventricularis, myxoedema, gout
8. Functional	Hysterical aphonia

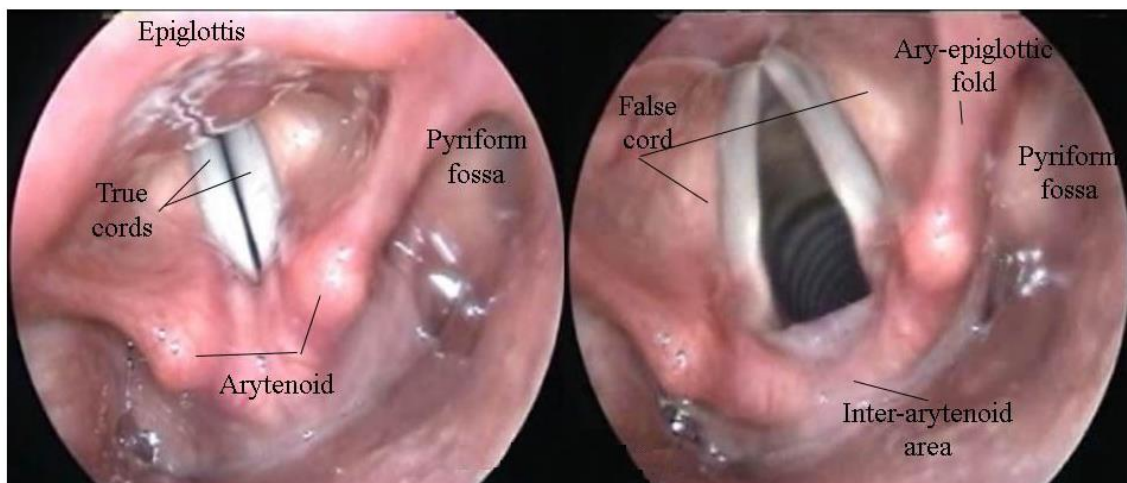
TABLE 3–1. Differential Diagnosis of Dysphonia: KITTENS Method

(K) Congenital	Infectious & Idiopathic	Toxins & Trauma	Tumor (Neoplasia)	Endocrine	Neurologic	Systemic
Congenital webs	Laryngitis (viral, bacterial, and fungal)	Laryngeal cysts, nodules, polyps, and ulcers	Recurrent laryngeal papillomatosis	Hypothyroidism (laryngeal myxedema)	Cerebral palsy	GERD
Underdeveloped larynx	Vocal fold paralysis	Voice abuse	Laryngeal cancer	Adrenal, pituitary, and gonadic disorders	Multiple sclerosis	Connective tissue disorders (rheumatoid arthritis, SLE)
	Adductor spasmodic dysphonia	Reinke's edema	Benign laryngeal neoplasms (hemangiomas, cystic hygromas)	Pubescence	Extrapyramidal lesions (Parkinson's)	Psychogenic
	Muscle-tension disorders	Arytenoid dislocation			Stroke	
		Vocal fold granulomas			Guillain Barré	
		Caustic inhalation injuries			Myasthenia gravis	
					Other neurological disorders	

Physical Exam:

- Maximum Phonation Time (MPT):
 - Measures the time an individual can sustain a sung tone, after having filled the lungs maximally.
 - The best of three attempts at sustaining a vowel (E) is used as Maximum Phonation Time
 - Normal MPT in male: **32 seconds**
 - Normal MPT in Female: **26 seconds**
- Complete ENT H&N Exam:
 - Neck masses
 - Thyroid masses
 - Complete cranial nerve exam
- Endoscopic Laryngoscopy:
 - Uses Halogen light.
 - **Flexible scope:**
 - Advantages:
 - Examine Nasal cavity in addition to larynx.
 - Easy to tolerate by the patient.
 - Examine the larynx in the normal posture.
 - Allows for connected speech.
 - Disadvantages:
 - Low resolution (Available now HD flexible scopes)
 - **Rigid scope (90 or 70 degree):**
 - Advantages:
 - Excellent Resolution.
 - Disadvantages:
 - Difficult to tolerate by the patient (gag reflex).
 - Needs tongue protrusion which distorts laryngeal posture.
 - Doesn't allow for connected speech.
 - Used to evaluate the following:
 1. Gross laryngeal appearance.
 2. Laryngeal lesions.
 3. Supraglottic hyperfunction.
 4. Vocal Folds:
 - Mobility.
 - Lesions.
 5. Glottic competence:
 - Gap: Glottic space during ADduction (Phonation)
 - Chink: Glottic space during ABduction (Respiration)

- **AAO-HNS guidelines for indications of Endoscopic Laryngoscopy:**
 1. Hoarseness does not resolve after 3 months of onset.
 2. If serious underlying cause is suspected:
 - Tobacco or alcohol use
 - Concomitant neck mass
 - Trauma
 - Hemoptysis
 - Dysphagia
 - Odynophagia
 - Otalgia
 - Airway compromise
 - Accompanying neurologic symptoms
 - Unexplained weight loss
 - Worsening hoarseness
 - Immunocompromise
 - Possible aspiration of foreign body
 - Neonatal hoarseness
 - After intubation or neck surgery



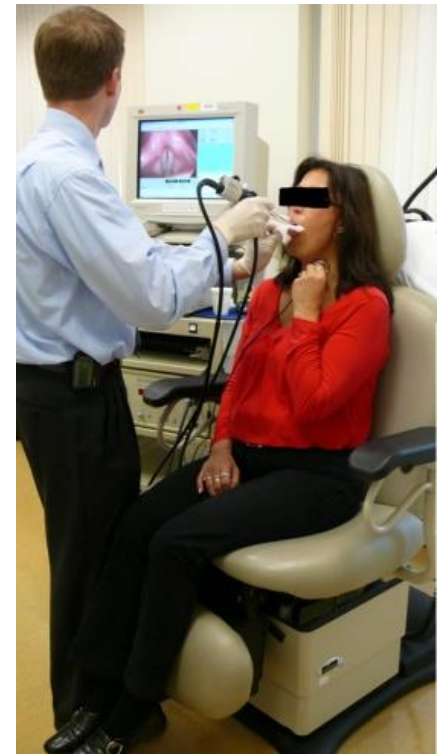
VC Adduction (Phonation)

VC Abduction (Deep inspiration)

Investigations:

- Stroboscopy:

- Special method used to visualize vocal fold vibration.
- Flashing light source which gives an "illusion" of continuous, slow-motion of vocal folds mucosal vibration.
- Light flashes at a frequency just slightly different from that of Glottal cycle, generating a series of still images that are fused to generate a continuous sequence seen by the examiner.
- Stroboscopy does not demonstrate fine detail of each individual vibratory cycle.
- It shows a pattern averaged over many successive nonidentical cycles.
- It is a less-than-perfect illustration of true vibratory nature.



○ Talbot's law:

- Images on the human retina linger for 0.2 seconds after exposure (persistence of vision).
 - Eye can perceive no more than 5 images/second.
 - Sequential images produced at intervals less than 0.2 seconds produce the illusion of a continuous image.
 - True vocal folds vibrate at rates of 75 to 1000 cycles/second, so the vibratory patterns cannot be visualized without assistance.
- Vocal folds vibration is too rapid to be perceived by human eye.
 - Strobe light flashes on the vibrating vocal folds.
 - Each light pulse illuminates a point of the vibratory cycle.
 - Illuminated points are visually fused providing an averaged vibratory pattern over successive cycles.
- When strobe flashes are emitted at the same frequency as phonation:
 - They optically occur at the same phase point in the successive cycles and the images appear frozen.
 - When strobe flashes are slightly slower (2 Hz) than the frequency of phonation:
 - They optically occur at different phase points in successive cycles yielding a simulated slow-motion effect of the vocal fold vibration.

○ Importance of Stroboscopy:

1. Assessment of vibratory patterns of true vocal folds.
2. Enhances clinician's diagnosis ability.
3. Early and accurate detection of laryngeal pathologies.
4. Documents laryngeal condition to computer.
5. Allows pre-/post-intervention follow-up of vocal folds.
6. Used in patient education and biofeedback.
7. Voice therapy.

○ Limitations of Stroboscopy:

1. Severe dysphonia or Aphonía.
2. Supraglottic hyperfunction (constriction).
3. Mostly subjective.
4. Patient's cooperation is needed.
5. "Illusion" of slow motion (Not a real motion)
6. Not capable of recording onset and offset of phonation.



○ Stroboscopy Examination:

1. Mucosal wave propagation:

- Longitudinal flexibility of fold, seen as traveling wave on vibration.
- Reflects pliability of phonatory mucosa.

2. Amplitude of vibration:

- Extent of lateral displacement from midline.
- Normally 1/3 width of VC.

3. Phase symmetry:

- Shape-changing relationship.

4. Presence of Adynamic segment.

5. Closure pattern:

- Musculomembranous portion of vocal folds completely closes during vibratory cycle.
- Posterior cartilaginous glottis may remain partially open (posterior glottic chink) in some healthy people.

6. Phase closure:

- Ratio of open to closed phase.
- 50:50

7. Periodicity:

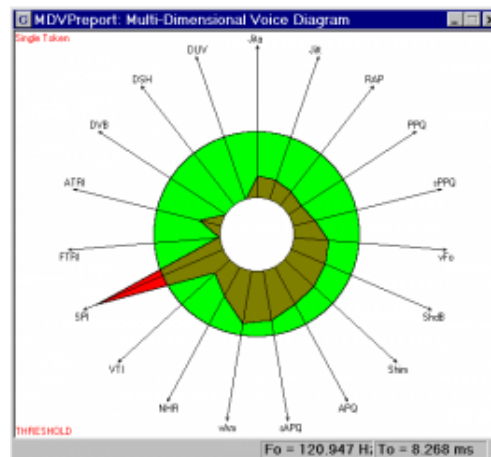
- Regularity of successive apparent cycles of vibration.

8. Fundamentals frequency:

- Measured by using strobe unit and is used to set the frequency of light flashes.
- Strobe light is typically produced at a frequency several hertz slower than vocal frequency to produce the illusion of a slow-motion vibratory cycle.
- An identical frequency is emitted in the locked mode that produces a still image of a single portion of the vibratory cycle.

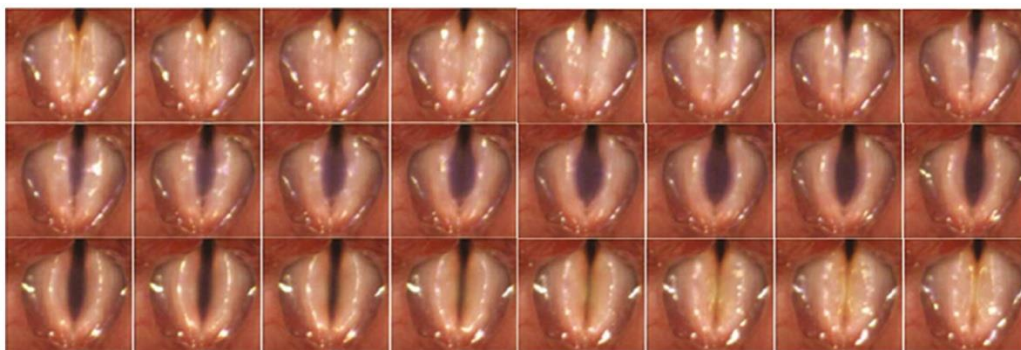
- **Multidimensional Voice Program (MDVP):**

- Gold standard software tool for quantitative acoustic assessment of voice quality.
- Calculates more than 22 parameters on a single vocalization.
- **Jitter:**
 - Peak to peak fluctuation in **Frequency** of sound wave.
- **Shimmer:**
 - Peak to peak fluctuation in **Amplitude** of sound wave.
- **Fundamental Frequency:**
 - Lowest frequency of a periodic waveform.



- **High Speed Camera:**

- Allows for images to be recorded at a speed of up to 4,000 frames per second (fps).
- Video then played in slow motion to examine mucosal wave.
- Newer and more expensive than stroboscopy
- Gives a true picture of vocal folds (Not illusion).
- **Advantages:**
 1. Records details of vocal fold vibration in real time.
 2. Records onset and offset of phonation, phonatory breaks, and laryngospasm.
 3. Better with Spasmodic dysphonia.



- **CT Scan with Contrast:**

○ Indications:

1. Laryngeal Tumors:

- CT Neck with contrast.
- Evaluate the extent of the tumor.

2. Idiopathic vocal fold paralysis:

- CT with contrast from Skull base to chest.
- Evaluate lesions along course of Vagus Nerve.

- **Laryngeal Electromyography (LEMG):**

- Evaluates integrity of laryngeal neuromuscular system in by recording action potentials generated in laryngeal muscles during voluntary and involuntary contraction.
- Most effective between 1-6 months post onset of paralysis.

○ Tests 3 laryngeal muscles:

1. ThyroArtyenoid (TA):

- Evaluation of RLN.
- Needle placement is confirmed by vocalisation.

2. Posterior CricoArytenoid (PCA):

- Evaluation of RLN.
- Needle placement is confirmed by sniffing.

3. CricoThyroid (CT):

- Evaluation of SLN.
- Needle placement is confirmed by glide from one pitch to another.

○ Indications of LEMG:

1. Neuropathy vs. Mechanical Fixation in immobile VF:

•Vocal fold Paralysis:

- EMG findings of Neuropathy.

•CricoArytenoid Fixation:

- Normal EMG findings.

2. Localization of lesion (SLN vs RLN):

- CricoThyroid for SLN Paralysis
- ThyroArtyenoid for RLN Paralysis

3. Prognosis of Vocal fold Paralysis:

•Good Prognosis:

- Presence of Motor-Unit Potentials.

•Poor Prognosis:

- Absence of Motor-Unit Potentials.
- Fibrillation Potentials

4. Detects Synkinesis:

- When ThyroArtyenoid activated by Sniffing or Posterior CricoArytenoid activated by vocalisation.

5. Confirm placement of needle for Botox injections.

6. Intra-op nerve monitoring.

7. Biofeedback in speech and swallowing disorders.



○ Interpretation of LEMG:

1. Normal LEMG:

- Bi or Triphasic Motor-Unit Potentials.
- Seen in patients with CricoArytenoid Fixation.

2. Neuropathy:

- Fibrillation Potentials:
 - Abnormal spontaneous activity at rest.
 - Due to ongoing axonal degeneration.
 - Starts after 3 weeks of injury.
 - Persist until Re-innervation occurs or until the motor endplates of the muscle degenerate (Myopathic injury).
- Sharp waves:
 - Lower Frequency
 - Normal Amplitude

3. Muscle Re-innervation after Neuropathic injury:

- Polyphasic Potentials
 - > 4 phases.
 - Due to Not well synchronized muscle fibers.

4. Myopathy:

- Decreased functioning muscle fibers.
 - Polyphasic.
 - Low Amplitude.
 - Low Duration.



TABLE 61.3

CLASSIFICATION OF LARYNGEAL ELECTROMYOGRAPHY FINDINGS IN LARYNGEAL PARALYSIS

Class	Spontaneous Activity	Recruitment	Motor Unit Morphology	Interpretation
I	Absent	Normal	Normal	Normal
II	Absent	Reduced	Low-amplitude polyphasics	Reinnervation
III	Absent	Reduced	Giant polyphasic units	Old injury
IV	Present	Reduced	Polyphasic units	Equivocal
V	Present	None	Fibrillations, positive sharp waves	Denervation

- **Labs:**

- CBC with differential
- Trepemonal studies (FTA-ABS)
- Lyme titers
- Thyroid function tests
- Toxin screen (lead and arsenic levels)
- Fasting blood sugar.

- **Microalaryngoscopy:**

- Direct detailed Microscopic examination of larynx under GA.
- Surgical excision and biopsy of the lesions.
- Assessment of Cricoarytenoid joint mobility.



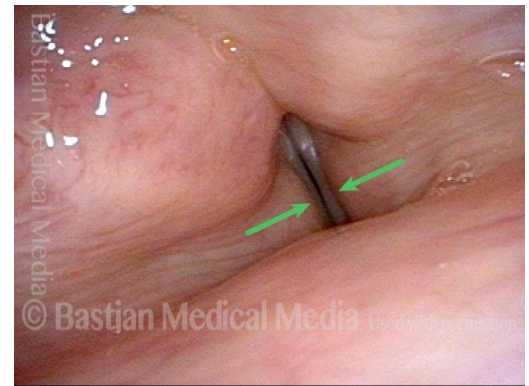
- **AAO-HNS Guideline Statements for Treatment of Hoarseness:**
 - Treat the underlying cause when discovered.
 - Recommend against prescribing antireflux meds without signs or symptoms of acid reflux.
 - Option of providing antireflux meds for hoarseness with signs of chronic laryngitis.
 - Recommend against routinely prescribing oral corticosteroids.
 - Strongly recommend against routinely prescribing antibiotics.
 - Recommend endoscopic laryngoscopy before prescribing voice therapy and document/communicate results to speech pathology.
 - Strongly recommend voice therapy for reduced voice-related QOL.
 - Recommend surgery for:
 - Suspected laryngeal malignancy
 - Benign laryngeal soft tissue lesions
 - Glottic insufficiency
 - Option of education/counseling about control/preventive measures

- **Speech Disorders:**
- **Organic disorders:**
 - **Structural**
 - Caused by laryngeal lesions causing physical abnormality.
 - Examples:
 - Contact Ulcers
 - Cysts
 - Granuloma
 - Hemorrhage
 - Hyperkeratosis
 - Laryngitis
 - Leukoplakia
 - Nodules
 - Papilloma
 - Polyps
 - Trauma
 - Miscellaneous growths
 - **Neurogenic:**
 - Caused by problems in the nervous system as it interacts with the larynx.
 - Examples:
 - Paralysis/Paresis
 - Spasmodic Dysphonia (Laryngeal Dystonia)
 - Essential Tremors
 - Tic Disorders
 - Oculopalatolaryngopharyngeal Myoclonus
 - Dysphonia caused by other neurological disorder.
- **Functional disorders (Muscle tension dysphonia):**
 - Caused by poor muscle functioning.
 - Examples:
 - Ventricular phonation
 - Pharyngeal constriction
 - HyperABduction
 - HyperADduction
- **Psychogenic disorders:**
 - No structural reason for the voice disorder, and there may or may not be some pattern of muscle tension.
 - Examples:
 - Conversion Dysphonia or Aphonia
 - Puberphonia (Mutational Falsetto)
 - Paradoxical Vocal Fold Motion Disorder

Neurogenic Speech Disorders:

- **Spasmodic Dysphonia (SD):**
 - Focal task-specific laryngeal dystonia with spasmodic contractions of internal laryngeal musculature.
 - Adult onset disorder with a female predominance.
 - Unknown pathophysiology.
 - Site of pathology involves the basal ganglia.
 - 25% of patients with SD report family history of dystonia or other movement disorders.
 - Associated with:
 - Essential tremor (30%)
 - Blepharospasm (15%)
 - Writer's cramp (15%)
 - **SD affects the speaking voice only.**
 - Spares other vocal tasks such as singing or yelling.
 - Spares non-speech laryngeal functions such as breathing, swallowing, or coughing.
 - Symptoms exacerbated by:
 - Anxiety.
 - Stress.
 - Public speaking.
 - Symptoms suppressed by:
 - Alcohol
 - Sedatives
 - Sensory trick (Geste antagonist) such as:
 - Voicing while chewing.
 - Voicing while biting the tongue.
 - Holding a finger in the corner of the mouth.
 - Insertion of a flexible scope.
 - Sometimes it is difficult to distinguish organic spasmodic dysphonia from psychogenic voice disorders or muscle tension dysphonia.
 - **Types of Spasmodic Dysphonia:**
 1. ADducted SD
 2. ABducted SD
 3. Mixed SD (Rare)

- **ADductor Spasmodic Dysphonia (AddSD):**
 - Most common type of SD (**85%**).
 - Causes vocal fold hyper-ADduction:
 - Phonation against a closed glottis.
 - Sudden and strong contraction of ThyroArytenoid muscle.
 - Voice Profile:
 - Harsh strained voice.
 - Strangled quality.
 - Voice breaks in connected speech.
 - Prominent with words begin with vowels.
 - Counting from 80 to 90.
 - "We eat eggs every day"
 - Diagnosis:
 - Flexible Nasolaryngoscopy during connected speech:
 - Hyper-ADduction of vocal folds with hyperfunction of Supraglottis.
 - Stroboscopy.
 - High Speed Camera:
 - Best tool to diagnose SD.
 - Treatment:
 - Voice therapy.
 - **Botulinum toxin (Botox) injections:**
 - Treatment of choice for SD.
 - 90% improvement.
 - Injection of ThyroArytenoid (**TA**) and Lateral CricoArytenoid (**LCA**) muscles.
 - Done transcutaneously under LEMG guidance.
 - 0.05-10 U of Botox (average of 1 U) is injected to bilateral TA muscles.
 - Effect lasts for 3 months.
 - Side Effects:
 - Excessive glottal weakness and breathiness.
 - Liquid dysphagia if toxin diffused to the adjacent constrictors.
 - Surgical therapy:
 - Destruction of RLN branches.
 - Resection dystonic musculature.
 - Type II Thyroplasty
 - Reinnervation of dystonic musculature with a branch of Ansa cervicalis.



- **ABductor Spasmodic Dysphonia (AbdSD):**
 - Less common than AddSD (**15%**).
 - Causes vocal fold hyper-ABduction:
 - Phonation against a opened glottis.
 - Sudden and strong contraction of Posterior CricoArytenoid muscle.
 - Voice Profile:
 - Abnormal whispered or sustained breathiness with breathy voice breaks during phonation.
 - Especially during voice onset
 - Prominent in vowels following a voiceless consonant:
 - Counting from 60 to 70
 - "The puppy bit the tape"
 - Diagnosis:
 - Flexible Nasolaryngoscopy during connected speech:
 - Inappropriate vocal fold ABduction during connected speech.
 - Stroboscopy.
 - High Speed Camera:
 - Best tool to diagnose SD.
 - Treatment:
 - Voice therapy.
 - **Botulinum toxin (Botox) injections:**
 - Treatment of choice for SD.
 - 90% improvement.
 - Injection of Posterior CricoArytenoid (**PCA**) muscle.
 - Done transcutaneously under LEMG guidance.
 - 2-5 U of Botox is injected to unilateral PCA muscle.
 - Lasts for 3 months.
 - Side Effects:
 - Stridor with airway compromise.
 - Liquid dysphagia if toxin diffused to the adjacent constrictors



- **Essential Tremor:**

- Common, benign and inherited movement disorder.
- Female predominance.
- Most common presentation:
 - Shaking of the hands and rhythmic head titubation.
- **30% of patients will have symptomatic vocal involvement.**
 - Tremor in muscles of larynx, pharynx, soft palate and the strap muscles of the neck.
- Clinical picture:
 - Steady shaking voice.
 - Ranging from gentle and continuous to a staccato, almost hiccuping sound.
 - Tremor is rhythmic and steady at 5-7 cycles per second.
 - Occurs in all speech contexts.
 - Symptoms exacerbated by:
 - Anxiety.
 - Stress.
 - Public speaking.
 - Symptoms suppressed by:
 - Alcohol
 - Sedatives
- Essential tremor differs from spasmodic dysphonia by:
 - Its rhythmicity.
 - Present across all speech tasks.
- Treatment:
 - Voice therapy.
 - Medical Management:
 - Beta-blocker (Propranolol)
 - Anti-epileptic (Primidone)
 - Botulinum toxin (Botox) injections:
 - Useful to "dampen" the tremor by weakening the affected musculature, but oscillatory movements will persist at diminished amplitude.
 - Will not eliminate the tremor.
 - Botox is injected into ThyroArytenoid muscle within the vocal fold.
 - TA is responsible for the strength of the staccato, hiccuping effect of the tremor.

- **Tic Disorders:**

- Tics are sudden, recurrent, quick, abnormal movements or vocalizations that abruptly interrupt normal activity.
- There is a compulsion to perform a movement, which is relieved once the movement is completed.
- Mainly affects children with male predominance.
 - Up to 30% of children may have transient tics.
- Many adults have unrecognized simple tics including:
 - Throat clearing
 - Snoring
 - Squeals
 - Coughs
 - Belching noises.
- Symptoms exacerbated by:
 - Anxiety.
 - Stress.
 - Public speaking.
- Symptoms suppressed by:
 - Alcohol
 - Sedatives
- Treatment:
 - Voice therapy.
 - Medical Management:
 - Tetrabenazine
 - Clonazepam
 - Botulinum toxin (Botox) injections.

- **Oculopalatolaryngopharyngeal Myoclonus:**

- Uncommon disorder consisting of twitch-like, rhythmic contractions of soft palate, pharynx and larynx.
- Occurs at a rate of 1-2 contractions per second.
- May affect only the palate, or all of the laryngopharynx.
- Causes:
 - Part of a seizure disorder
 - Posttraumatic
 - Viral, toxic, or metabolic encephalopathy.
- Clinical picture:
 - Choppy speech
 - Intermittent hypernasality from palatal dysfunction.
 - Persistent tinnitus (clicking) in the ear.
 - ET dysfunction.
- Treatment:
 - Anti-epileptic medications.
 - Botulinum toxin (Botox) injections.
 - Injections of palate and vocal folds to decrease severity of the contractions
 - Will not improve ET dysfunction and velopharyngeal insufficiency.

- **Dysphonia caused by other neurological disorder:**

- Examples:
 - Stroke
 - Multiple Sclerosis
 - Bulbar Palsy
 - Poliomyelitis
 - Guillain-Barre Syndrome
 - Myasthenia Gravis
 - Amyotrophic Lateral Sclerosis
 - Friedreich Ataxia
 - Arnold-Chiari Malformations
 - Parkinson's disease

**TABLE
70.3**

NEUROLARYNGEAL DISORDERS, BY LARYNGEAL PRESENTATION

Presentation	Spastic	Hypokinetic	Hyperkinetic	Ataxic	Paresis/Paralysis
Vocal Fold Appearance	Variable	Rigid/bowing on phonation	Hyperabducted	Uncoordinated, decreased mobility	Atrophied, flaccid, fasciculating
Anatomical Structures Involved	UMN/corticobulbar tracts	Basal ganglia	Basal ganglia	Cerebellum	Brainstem—nucleus ambiguus; peripheral nerve; neuromuscular junction; muscle
Example Disorders	PLS, ALS, stroke, MS	Parkinson disease, MSA	Dystonia, tremor, myoclonus	Friedreich ataxia, stroke, MS	ALS, brainstem stroke, GBS, superior laryngeal nerve injury, MG, polymyositis
Treatment	Botulinum toxin, voice and swallowing therapy, vocal fold medialization	Voice and swallowing therapy, vocal fold medialization	Botulinum toxin, voice and swallowing therapy	Voice and swallowing therapy, vocal fold medialization	Tracheostomy(severe), vocal fold medialization or lateralization, voice and swallowing therapy

UMN, Upper Motor Neuron; PLS, Primary Lateral Sclerosis; ALS, Amyotrophic Lateral Sclerosis; MS, Multiple Sclerosis; MSA, Multiple Systems Atrophy; GBS, Guillain-Barre Syndrome; LSVT, Lee Silverman Vocal Therapy.

Functional Speech disorders (Muscle Tension Dysphonia):

- Voice disturbance without structural or neurologic laryngeal pathology.
- Excessive tension in intrinsic and/or extrinsic laryngeal muscles.

- Examples:

- **Ventricular Phonation (Plica ventricularis):**
 - Voice is produced by ventricular folds (false vocal folds) which have taken over the function of true vocal folds.
 - Diagnosis is made on Endoscopic laryngoscopy:
 - False vocal folds are seen to approximate partially or completely and obscure the view of true cords on phonation.
- **Pharyngeal Constriction:**
 - Pharyngeal muscles contract excessively while talking, leaving the pharynx very constricted.
- **Hyper Abduction.**
- **Hyper Adduction.**

- Causes:

- **Primary (Normal larynx):**
 - Prolonged voice overuse:
 - Teachers
 - Singers and actors
 - People talking on the telephone all day.
 - Learned adaptations after URTI.
 - Increased pharyngolaryngeal tone secondary to LPR.
 - Psychologic.
- **Secondary (Abnormal Larynx):**
 - Extreme compensation for minor glottic insufficiency and/or underlying mucosal disease.

- Clinical picture:

- Breathy or harsh voice with use through the day and recovers with rest.
- Vocal fatigue and strain.
- Organic changes in vocal cords may occur secondary to such faulty use or overloading.

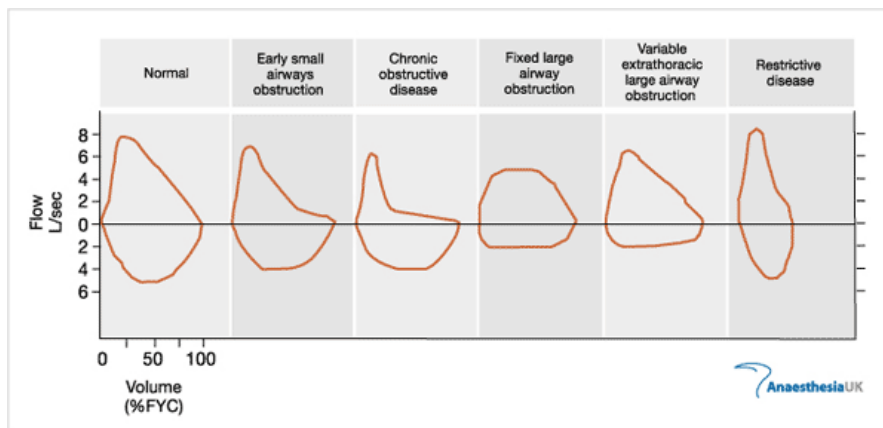
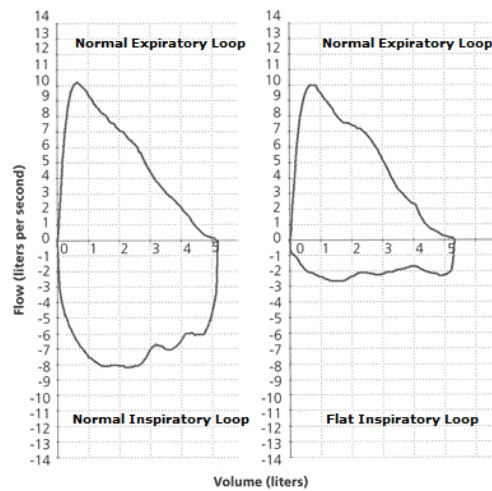
- Management:

- Voice therapy.
- Psychological counseling if needed.

Psychogenic Speech Disorders:

- **Conversion Dysphonia/Aphonia:**
 - Functional disorder mostly seen in emotionally labile females in the age group of 15-30.
 - Exists when there is psychological trauma or conflict that is manifested physically.
 - Accident
 - Death
 - Psychologically damaging event
 - Clinical picture:
 - Breathy–hoarse dysphonia with a normal laryngeal exam.
 - Aphonia is usually sudden and unaccompanied by other laryngeal symptoms.
 - Patient communicates with whisper.
 - On examination of patient with Conversion aphonia:
 - Vocal folds are seen in ABducted position.
 - Fail to ADduct on phonation.
 - Normal cough voice and with normal vocal folds ADduction during coughing.
 - Management:
 - Voice therapy.
 - Psychological counseling.
- **Puberphonia (Mutational Falsetto Voice):**
 - Persistence of childhood high-pitched voice.
 - More common in adolescent males, less common in females.
 - Occurs during puberty in boys who are emotionally immature, feel insecure and show excessive fixation to their mother.
 - Psychologically, they shun to assume male responsibilities though their physical and sexual development is normal.
 - Pathophysiology:
 - Normally, childhood voice has a higher pitch.
 - When the larynx matures at puberty, vocal folds lengthen, and voice changes to lower pitch.
 - This is a feature exclusive to males.
 - Clinical picture:
 - Recurrent “cracked,” shrill, high-pitched voice.
 - Gutzmann's pressure test:
 - Pressing thyroid prominence in a backward and downward direction relaxes the overstretched folds and low tone voice can be produced.
 - Management:
 - Voice therapy.
 - Psychological counseling.

- **Paradoxical Vocal Fold Motion Disorder (Vocal fold Dysfunction):**
 - Attacks of stridor in which true vocal folds paradoxically ADduct during inspiration.
 - Seen in young females.
 - Very common among asthmatics.
 - Causes:
 - **Organic (Neurogenic):**
 - Stroke
 - Multiple Sclerosis
 - Myasthenia Gravis
 - Amyotrophic Lateral Sclerosis
 - **Psychogenic (Conversion or Somatization):**
 - History of stressful periods (unconsciously and without intentional gain).
 - Clinical picture:
 - Easily misdiagnosed.
 - Frequent, episodic attacks of:
 - Cough
 - Inspiratory/expiratory wheeze
 - Dyspnea with/without exertion
 - Stridor
 - Hoarseness
 - Diagnosis:
 - **Laryngoscopic examination:**
 - Paradoxical vocal folds motion (inspiratory anterior vocal cord closure with posterior chinking).
 - **Pulmonary Function Test (PFT):**
 - PFT with a flow-volume loop is used to confirm vocal fold dysfunction and differentiate it from Asthma.
 - Flow-volume loop include forced inspiratory and expiratory maneuvers.
 - Extra-thoracic upper airway obstruction:
 - Normal expiratory loop.
 - Flat inspiratory loop.
 - Asthma:
 - Both expiratory and inspiratory loop are diminished but with predominant prolongation of expiratory loop.



- Management:
- Acute Management:
 - **Heliox:**
 - Gaseous mixture of oxygen and helium.
 - Found in ratios of 20/80 and 30/70.
 - Less dense than air.
 - Inhalation reduces turbulence in the airway and eliminates respiratory noise.
 - Reduces anxiety that is the predisposing factor to many attacks.
 - **Intermittent or continuous positive airway pressure:**
 - Widen Rima glottidis.
 - **Benzodiazepines:**
 - Reduce anxiety.
 - **Intralaryngeal injection of Botox:**
 - Recommended in severe cases of PVCM.
- Chronic Management:
 - **Voice therapy.**
 - **Psychological counseling.**

- **Hypernasality (Rhinolalia Aperta):**
 - It is seen when certain words which have little nasal resonance are resonated through nose.
- **Hyponasality (Rhinolalia Clausa):**
 - It is lack of nasal resonance for words which are resonated in the nasal cavity (m, n, ng).

Table 62-2. Causes of hyponasality and hypernasality

Hyponasality	Hypernasality
Common cold	Velopharyngeal insufficiency
Nasal allergy	Congenitally short soft palate
Nasal polypi	Submucous palate
Nasal growth	Large nasopharynx
Adenoids	Cleft of soft palate
Nasopharyngeal mass	Paralysis of soft palate
Familial speech pattern	Post-adenoidectomy
Habitual	Oronasal fistula
	Familial speech pattern
	Habitual speech pattern

**TABLE
72.5**

LARYNGEAL HYGIENE MEASURES

Smoking cessation
 Avoid excessively dry environments
 Eliminate extraneous and loud voice use
 Avoid eating prior to bed
 Avoid foods high in fat content
 Avoid foods containing natural diuretics
 Increase free water intake

**TABLE
71.1**

COMPARISONS OF WHICH VOICE DISORDERS ARE MORE OR LESS AMENABLE TO VOICE THERAPY

Voice Disorders Amenable to Voice Therapy (Infrequent Surgical Management)

Nodules
 Primary muscle tension dysphonia
 Functional aphonia (muscle tension aphonia)
 Mild vocal fold atrophy
 Paradoxical vocal fold motion disorder
 Parkinson disease

Voice Disorders Less Amenable to Voice Therapy (Frequent Surgical Management Required)*

Cysts, polyps, fibrous masses, pseudocyst
 Scar/sulcus
 Papilloma
 Moderate-to-severe atrophy
 Reinke edema
 Paralysis/dense paresis
 Spasmodic dysphonia/vocal essential tremor

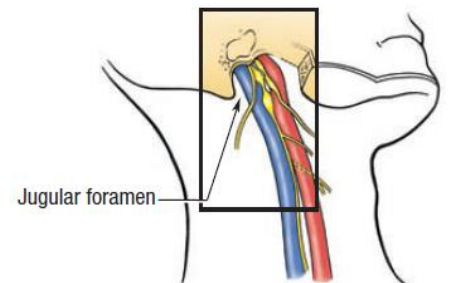
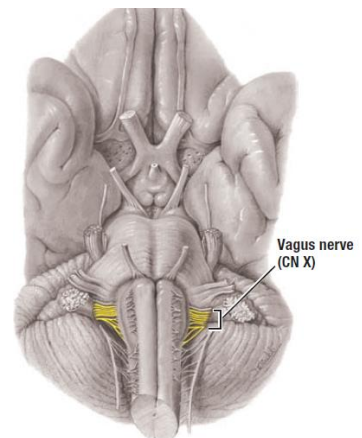
*Although patients may require a brief session of voice therapy, to be educated about their current laryngeal mechanism and engaged in indirect voice therapy, possibly reduce edema around the lesions for better surgical success, and to be instructed in ways to reduce compensatory extralaryngeal tensions, voice therapy most likely will not resolve the problem.

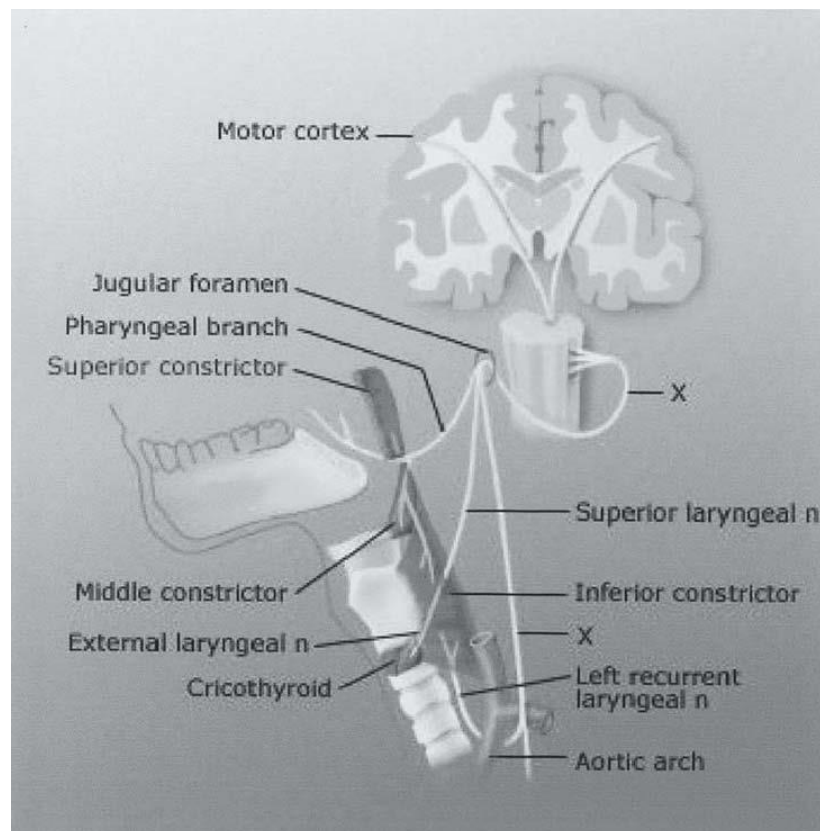
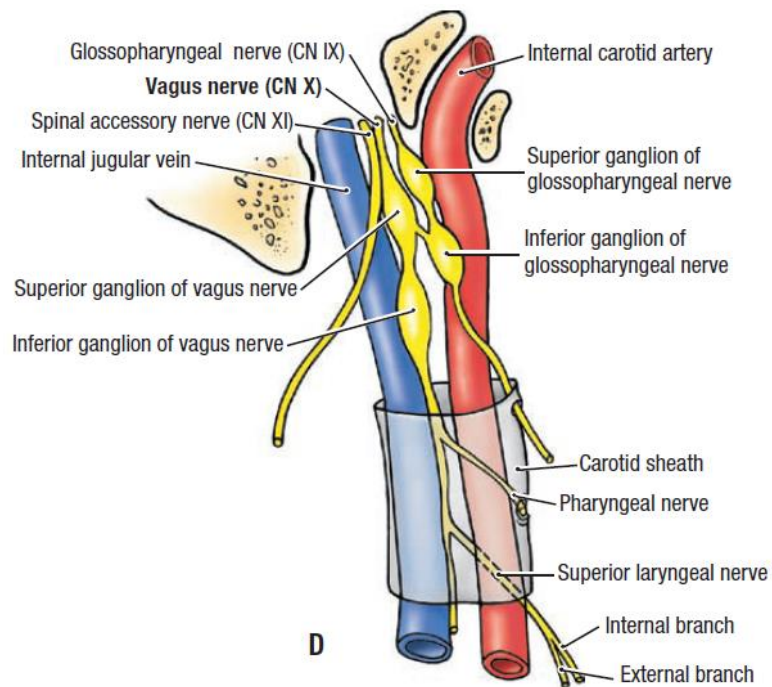
Vocal Folds Paralysis (VFP):

- Terminology:
 - **Immobility:**
 - Non-movement of vocal fold from any cause (Neurologic or Mechanical).
 - **Hypomobility:**
 - Partial non-movement of vocal fold from any cause (Neurologic or Mechanical).
 - **Paralysis:**
 - Non-movement of vocal fold from neurologic cause (RLN injury).
 - **Paresis:**
 - Partial non-movement of vocal fold from neurologic cause (RLN injury).
- **VFP is a physical finding and not a diagnosis, and the cause of the immobility must be determined.**

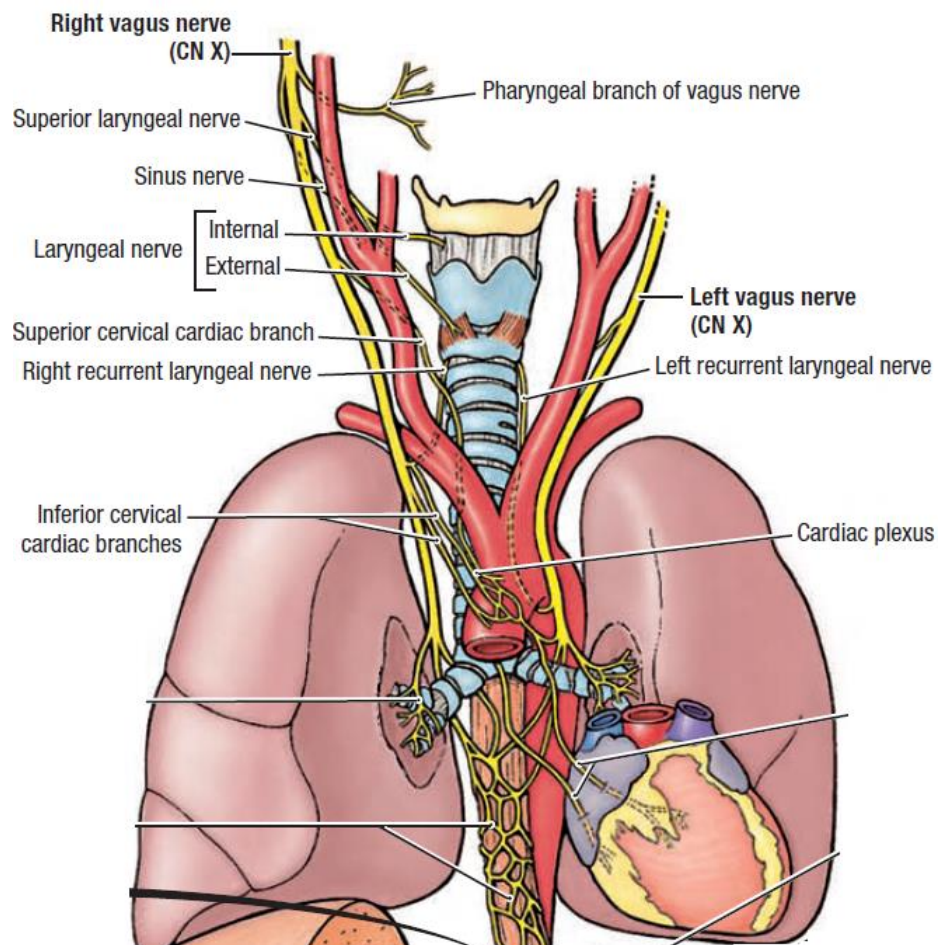
- Anatomy of Vagus Nerve (CN-X):

- Corticobulbar fibers from the cerebral cortex descend through the internal capsule and synapse on the motor neurons in Nucleus Ambiguus.
- Nucleus Ambiguus is the motor nucleus of CN-X at Medulla oblongata.
- CN-X fibers emerge from Anterior surface of Medulla oblongata between Olive and Inferior cerebellar peduncle.
- Passes in the posterior cranial fossa.
- Leaves the skull through Jugular Foramen.
- Descends through the neck Between the Carotid arteries and Internal Jugular vein within Carotid Sheath.
- Vagus Nerve has 2 Ganglionic swellings, which are the sensory ganglia of the nerve:
 - Superior (Jugular) Ganglion.
 - Inferior (Nodosum) Ganglion.
- Then it passes through the mediastinum of thorax, pierces the diaphragm with esophagus and terminates within Abdomen.
- Longest and most extensive distribution of all Cranial nerves.
- Mixed Motor and Sensory Nerve.

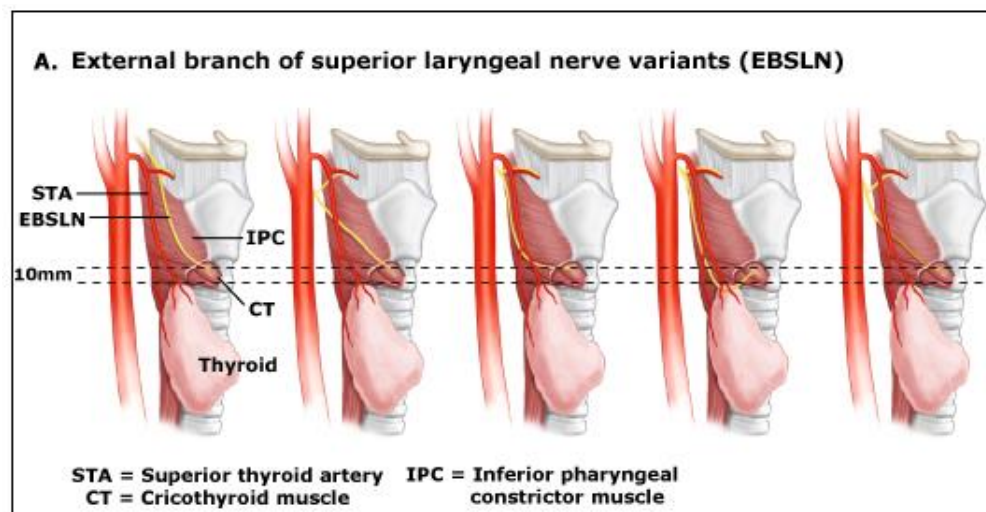




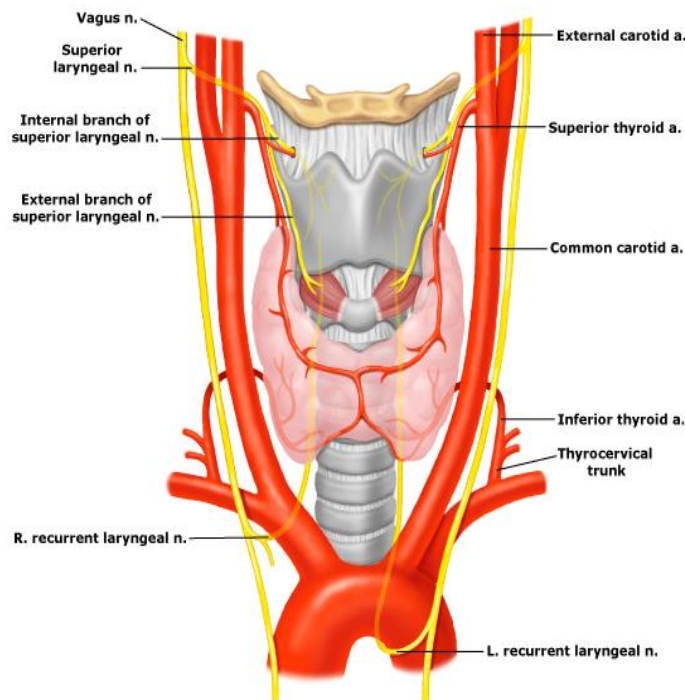
- **Right Vagus Nerve:**
 - Crosses in front of 1st part of Subclavian Artery.
 - Travels behind the innominate vessels.
 - Reaches the thorax on the right side of the trachea.
- **Left Vagus Nerve:**
 - Crosses in front of Left subclavian artery.
 - Enter the thorax between the left common carotid and subclavian arteries.
 - Descends on left side of the aortic arch.



- **Superior Laryngeal Nerves (SLN):**
 - Originate from vagus nerve as it exits base of the skull.
 - Runs transversally behind the carotid artery.
 - Courses with Superior thyroid artery until approximately 1 cm before the artery enters the capsule of Superior pole of the thyroid.
 - Two primary branches:
 - **External branch:**
 - Primarily motor:
 1. Inferior constrictor muscle.
 2. Cricothyroid muscle.
 - Travels with Superior thyroid artery until approximately 1 cm before the artery enters superior thyroid pole.
 - Divides into branches that enter Lateral inferior pharyngeal constrictor muscle and Cricothyroid muscle.
 - **Internal branch:**
 - Sensory to Supraglottis and Glottis.
 - Afferent branch of cough reflex arc.
 - Enters the larynx through Thyrohyoid membrane superior to the external branch.

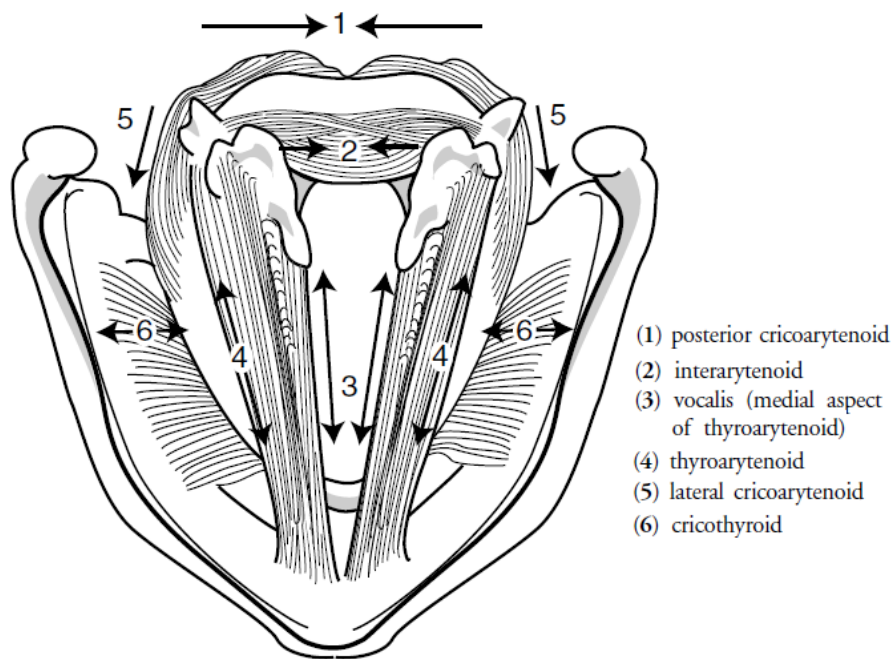


- **Recurrent Laryngeal Nerve (RLN):**
 - Provides both sensory and motor function to Larynx.
 - Sensory to Subglottis and Trachea.
 - Innervates all muscles to the larynx Except Cricothyroid muscle.
 - **Course of Left RLN:**
 - Originates from Left Vagus nerve below Arch of Aorta in the thorax.
 - Loops posterior to Arch of Aorta
 - **Course of Right RLN:**
 - Originates from Right Vagus nerve at level of Subclavian Artery.
 - Loops posterior to Subclavian Artery.
 - **Common Course:**
 - Associated with Inferior thyroid artery at junction of lower and middle thirds of thyroid gland.
 - Posterior to Inferior Thyroid Artery **(40%)**.
 - Between branches of the artery (35%)
 - Anterior to Inferior Thyroid Artery (20%)
 - Ascends to the side of Trachea at Tracheoesophageal groove.
 - Terminal portion of RLN accompanies Laryngeal branch of Inferior thyroid artery to enter the larynx behind Cricothyroid joint.
 - In 90% of cases, RLN divides into two to three branches just before entering the larynx deep to inferior constrictor muscle.



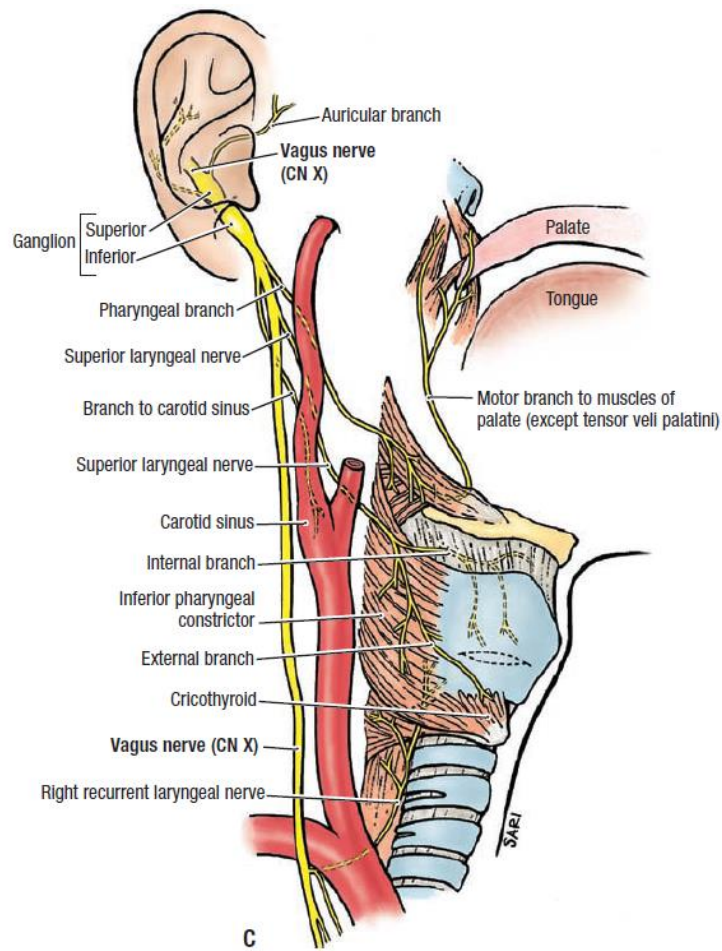
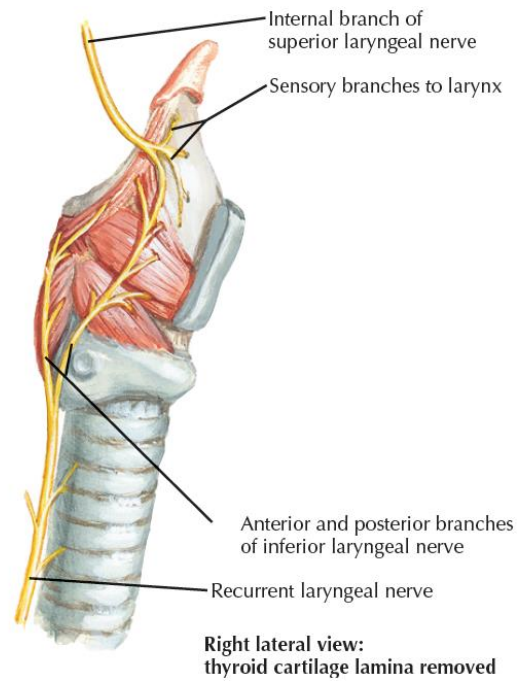
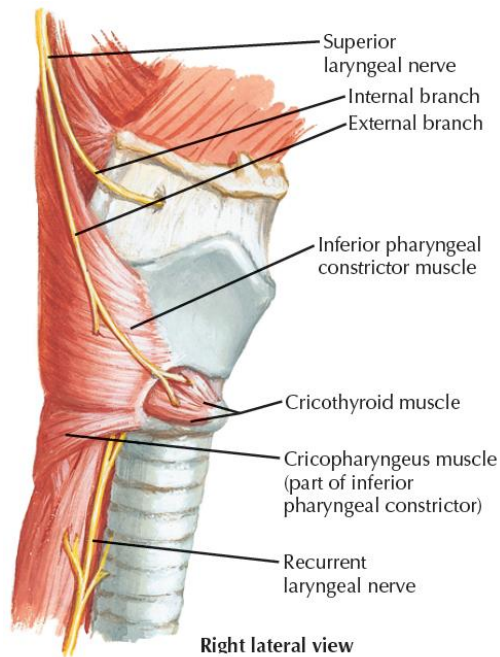
- **Motor Supply of Larynx:**

- **ALL** intrinsic muscles of larynx which move vocal folds are supplied by Recurrent Laryngeal Nerve (**RLN**):
 - Posteriior CricoArytenoid (**ABdcutor**)
 - Lateral CricoArytenoid. (**ADdcutor**)
 - InterArytenoid (**ADdcutor**)
 - ThyroArytenoid (**ADdcutor**)
 - Vocalis (**Tensor**)
- **EXCEPT:**
 - CricoThyroid (**Main Tensor + ADdcutor**)
 - External branch of Superior Laryngeal Nerve (**SLN**).



- **Sensory Supply of Larynx:**

- Laryngeal mucosa of Glottis and Supraglottis:
 - Internal branch of Superior Laryngeal Nerve (**SLN**).
- Laryngeal mucosa of Subglottis:
 - Recurrent Laryngeal Nerve (**RLN**).



- **Classification of VFP:**
- Unilateral or Bilateral.
- May involve:
 1. Recurrent Laryngeal Nerve.
 2. Superior Laryngeal Nerve.
 3. Both RLN and SLN (Complete/Combined).

- **Causes of VFP:**
- In topographical manner:
 - **SupraNuclear Lesions:**
 - Cerebral lesions.
 - Cause bilateral VFP and other CN involvement.
 - **Brainstem Lesions:**
 - Involvement of Nucleus Ambiguus in Medulla.
 - Cause bilateral VFP and other CN involvement.
 - **High Vagal Lesions:**
 - At skull base and Parapharyngeal space.
 - Cause unilateral VFP and other branches of Vagus nerve.
 - **RLN Lesions:**
 - Cause isolated unilateral VFP.

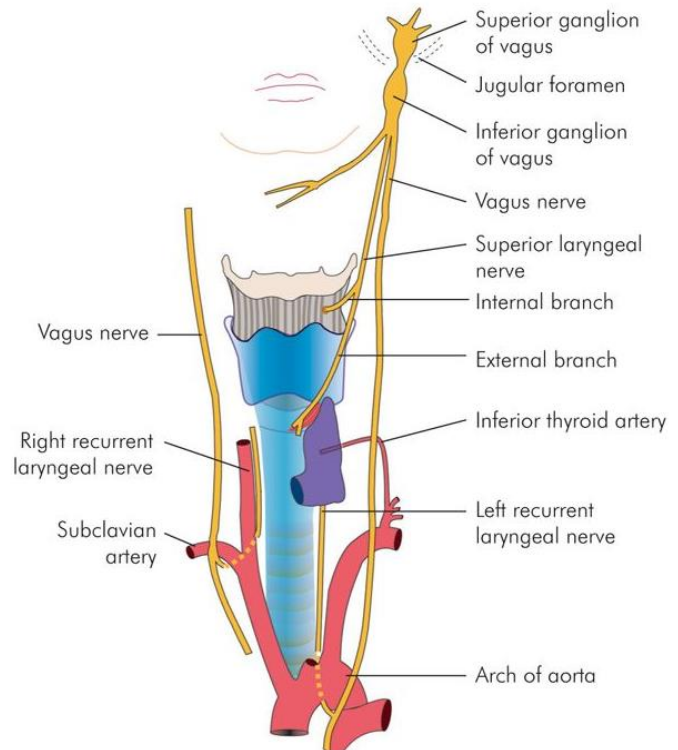


Table 59-1. Causes of combined paralysis (high vagal lesions)

<i>Intracranial</i>	Tumours of posterior fossa
	Basal meningitis (tubercular)
<i>Skull base</i>	Fractures
	Nasopharyngeal cancer
	Glomus tumour
<i>Neck</i>	Penetrating injury
	Parapharyngeal tumours
	Metastatic nodes
	Lymphoma

- **Positions of Vocal Folds:**

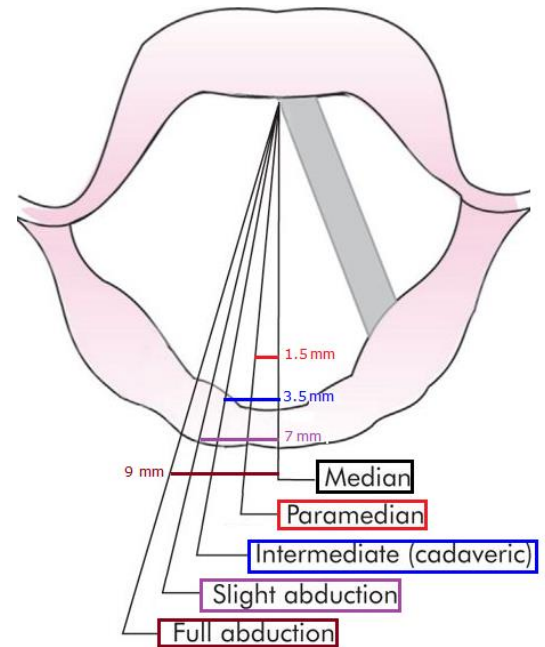
- Does not necessarily predict site of the lesion.
- Caused mainly by degree of Reinnervation and Synkinesis.

1. Median Position:

- In the Midline.
- Normally occurs during:
 - Phonation.

2. ParaMedian Position:

- 1.5 mm from Midline.
- Normally occurs during:
 - Strong whispering.
- Abnormally occurs during:
 - Isolated RLN Paralysis.
 - Partial ADduction is preserved from:
 1. CricoThyroid (Intact SLN).
 2. InterArytenoid (Receives Bilateral innervation)



3. Cadaveric (Intermediate) Position:

- 3.5 mm from Midline.
- Neutral position of Cricoarytenoid joint.
- ABduction and ADduction take place from this position
- Abnormally occurs during:
 - Complete Paralysis of both RLN and SLN.

4. Gentle ABduction Position:

- 7 mm from Midline.
- Normally occurs during:
 - Quiet Respiration.

5. Full ABduction Position:

- 9.5 mm from Midline.
- Normally occurs during:
 - Deep Respiration.

Unilateral Vocal Fold Paralysis (UVFP):

- **Unilateral RLN Paralysis:**
 - Ipsilateral paralysis of **ALL** intrinsic laryngeal muscles **EXCEPT** Cricothyroid and InterArytenoid.
 - Vocal cord situation:
 - ParaMedian Position.
 - Loss of ABduction on Deep inspiration.
 - Preservation of Partial ADduction from action of:
 1. CricoThyroid (Intact SLN).
 2. InterArytenoid (Receives Bilateral RLN innervation)
 - In Adult:
 - **Left** RLN paralysis is more common because it has a much longer course (Greater risk of injury).
 - In Pediatric:
 - **Right** RLN is affect more by Congenital anomalies because of its shorter length (more susceptible to traction forces).
- **Unilateral SLN Paralysis:**
 - Cricothyroid muscle Paralysis and Ipsilateral anaesthesia of Larynx Above VF.
 - Isolated lesions of SLN are rare.
 - Usually part of combined paralysis.
- **Unilateral Complete (RLN and SNL) Paralysis:**
 - Paralysis of **ALL** laryngeal muscles on one side **EXCEPT** Interarytenoid.
 - Receives Bilateral innervation from both RLN.

TABLE 69.1 UNILATERAL VOCAL FOLD IMMOBILITY: CAUSES	
Cause	Percentage
Iatrogenic (surgical trauma)	
Nonthyroid	30.6
Thyroid	15.7
Malignancy	
Lung	6.6
Nonlung	6.9
Idiopathic	17.6
Neurologic	7.9
Intubation	4.4
Nonsurgical trauma	2.2
Aortic/cardiac	0.6
Other	12.6

Adapted from Rosenthal LHS, Benninger MS, Deeb RH. Vocal fold immobility: a longitudinal analysis of etiology over 20 years. *Laryngoscope* 2007;117:1864–1870, with permission.

Causes of Unilateral Vocal Fold Paralysis (UVFP):

1. Surgical Trauma/Iatrogenic (45%):

- Most common cause of UVFP.
 - **Non-Thyroid:**
 - Most common iatrogenic cause.
 - **Anterior C-spine Surgery:**
 - Most common non-thyroid surgery cause.
 - Higher rate of persistent RLN palsy (11%).
 - Cardiothoracic surgery
 - Carotid Endarterectomy
 - Esophagectomy
 - Thymectomy
 - Neck dissection
 - Mediastinoscopy
 - **Thyroid:**
 - Overall rate of temporary RLN paralysis is (5%)
 - Overall rate of permanent RLN paralysis is (1%)

**TABLE
69.2**

SURGICAL OR IATROGENIC CAUSES OF VFP

Surgery or Procedure	Mechanism of Nerve Injury or Relevant Anatomy
Anterior cervical spine	Retraction; stretch injury of RLN (right more common) (6)
Esophagectomy	RLN injury in tracheoesophageal groove
Carotid endarterectomy	Vagal injury during dissection
Mediastinoscopy	RLN injury, usually left
Coronary artery bypass grafting	1. Retraction or direct injury to vagus or RLN during internal mammary artery harvest for grafting (8) 2. Hypothermic nerve injury from ice cardioplegia (7)
Pulmonary resection	Usually left upper lobe or RLN injury
Endotracheal intubation	Possible pressure neuropraxia from compression of anterior rami of RLN caused by a high-riding endotracheal cuff in the subglottis (9)

RLN, recurrent laryngeal nerve.

2. Neoplastic (25%):

- 2nd most common cause of UVFP.
 - **Bronchogenic Cancers:**
 - Most common neoplastic cause.
 - Cause Left RLN Paralysis.
 - **Non- Bronchogenic Cancers:**
 - Skull base.
 - Thyroid
 - Laryngeal
 - Esophageal
 - Mediastinal

3. Idiopathic (20%):

- Diagnosis of exclusion, once detailed history and appropriate imaging studies fail to demonstrate a cause.
- May be caused by (HSV1) of vagus nerve.
 - Inflammatory neuropathy similar to Bell palsy.
 - Spontaneous recovery in 50%.
 - May take up to 12 months to resolve.

4. Non-Surgical Trauma:

- **Blunt and penetrating injuries of Skull base and Neck.**
- **Intubation:**
 - Post-intubation RLN Neurapraxia.
 - Secondary to excessive cuff inflation which placed in proximal subglottis resulting in a pressure induced neurapraxia of RLN.
 - Anterior Rami of RLN which pierce the larynx in subglottis just at level below TVFs.
 - Should be differentiated from Arytenoid cartilage dislocation or subluxation by:
 - Laryngeal Electromyography (LEMG).
 - Direct palpation under GA.

5. Neurologic:

- Usually cause VFP in addition to other neurological symptoms and other CNs involvement.
 - Brainstem stroke
 - Arnold Chiari malformations
 - ALS
 - Guillain-Barre syndrome
 - Parkinson
 - Myasthenia gravis
 - Multiple sclerosis
 - Postpolio syndrome

6. Systemic Diseases:

- Rare.
- Cause immobility by either paralysis or joint fixation.
 - Sarcoidosis
 - Tuberculosis
 - Rheumatoid arthritis
 - hypothyroid myxedema

7. Medications:

- Rare.
- Vinca Alkyls (vincristine and vinblastine) and Cisplatin through Neurotoxicity.
- Resolve 4-6 weeks after meds stopped.

Table 59-3. Causes of recurrent laryngeal nerve paralysis (low vagal trunk or recurrent laryngeal nerve)

Right	Left	Both
• Neck trauma	I. <i>Neck</i>	
• Benign or malignant thyroid disease	• Accidental trauma	
• Thyroid surgery	• Thyroid disease (benign or malignant)	Thyroid surgery
• Carcinoma cervical oesophagus	• Thyroid surgery	Carcinoma thyroid
• Cervical lymphadenopathy	• Carcinoma cervical oesophagus	Cancer cervical oesophagus
	• Cervical lymphadenopathy	Cervical lymphadenopathy
	II. <i>Mediastinum</i>	
• Aneurysm of subclavian artery	• Bronchogenic cancer	
• Carcinoma apex right lung	• Carcinoma thoracic oesophagus	
• Tuberculosis of cervical pleura	• Aortic aneurysm	
• Idiopathic	• Mediastinal lymphadenopathy	
	• Enlarged left auricle	
	• Intrathoracic surgery	
	• Idiopathic	

- **Differential diagnosis of UVFP:**
 - CA subluxation or dislocation:
 - Diagnosed by:
 - No history of surgical iatrogenic trauma.
 - History of trauma (including intubation).
 - LEMG
 - DLB and EUA:
 - Anterior displacement of Arytenoid.
 - CA joint palpation for passive mobility.
- **Clinical Picture of Unilateral VFP:**
- Asymptomatic (1/3 of cases).
- **Voice:**
 - Hoarse-breathy dysphonia due to insufficient glottic closure.
 - Weak fatigued voice (Decrease MPT).
 - Diplophonic voice (Each cord at unique tension and frequency).
 - Gradually improves due to compensatory mechanisms:
 - Hyper-Adduction of healthy vocal fold which crosses Midline to meet the paralyzed one.
 - Supraglottic hyperfunction leads to a characteristic rough, pitch-locked, low-frequency voice.
 - In cases of SLN involvement:
 - Low pitch which cannot be raised due to loss of VF tension.
- **Airway:**
 - Ineffective cough due to insufficient glottic closure.
 - Can't do Valsalva maneuver.
 - No Airway Obstruction or Stridor.
- **Aspiration:**
 - Aspiration of liquid due to ineffective cough.
 - In cases of SLN involvement:
 - Loss of ipsilateral glottic and supraglottic sensation.

- **Evaluation of Unilateral VFP:**
- **General ENT H&N Examination:**
 - Neck and Thyroid masses.
 - Complete CN examination especially CN-XI and CN-XII.
 - Palatal movement:
 - Palatal paralysis with ipsilateral VFP indicate high vagal lesion.
 - In case of palatal paralysis, uvula is deviated away from the side of the lesion.
 - In cases of VFP, MPT is reduced to 10 seconds or less.
 - Shorter values indicate more severe glottic incompetence, worse voice and increased vocal fatigue.
- **Endoscopy:**
 - Flexible laryngoscopy is the only method to view vocal fold mobility in its natural state.
 - **"ee-sniff" maneuver:**
 - Patient alternates between phonating "e" and sniffing vigorously to adduct and abduct vocal folds maximally which allows an excellent way to judge the degree of paresis or paralysis.
 - **Signs:**
 - Unilateral immobility of vocal fold during phonation (Adduction) and inspiration (ABduction).
 - Small degree of ADduction during phonation is expected in complete unilateral VFP due to bilateral innervation of InterArytenoid muscle.
 - Any ABduction during inspiration implies incomplete paralysis "paresis".
 - Paralyzed vocal fold can be in a variety of positions including median, paramedian and cadaveric position.
 - Evaluate presence of compensatory mechanisms:
 - Hyper-Adduction of healthy vocal fold which crosses midline to meet the paralyzed one.
 - Supraglottic hyperfunction which may obscure vocal fold movement.
 - Eliminated by humming or with prolonged phonation of "who".
 - Anterior displacement (overhanging) of ipsilateral Arytenoid cartilage.
 - Presence of this sign in traumatic cases should rise suspicion of Arytenoid dislocation.
 - Evaluate the glottic closure and presence of glottic gap.
 - **In cases of isolated SLN paralysis:**
 - Askew position of glottis with posterior commissure is pointing to Paralyzed side.
 - Bowing deformity of vocal fold.



Left vocal fold paralysis close to the open (breathing) position. Note the thin bowed appearance of the paralysed fold.



Left vocal fold paralysis in voicing with poor closure. Note the recruitment of surrounding muscles.



Paralysed right vocal fold: paralysed fold lies between the open and closed position in breathing.



Well compensated right vocal fold paralysis in voicing with good closure. Note the minimal posterior gap and the compensatory movement of the left fold across the midline.

- **Stroboscopy:**

- Better Evaluation.
- Findings:
 - Incomplete closure or a large glottal gap in uncompensated VFP.
 - Increase amplitude of vibration because of the atrophic "floppy" nature of denervated vocalis muscle.

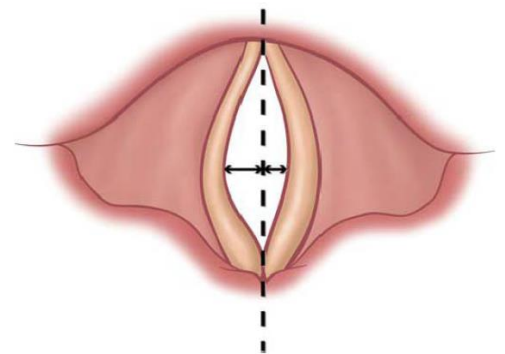
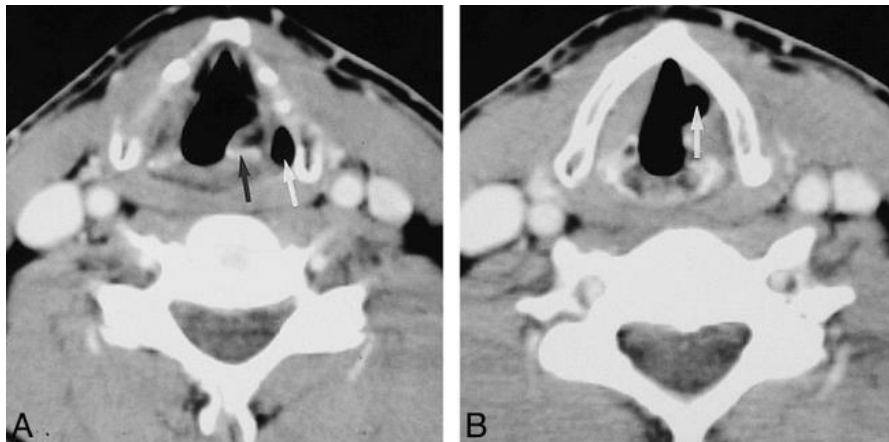


Figure 69.6 Illustration of laryngovideostroboscopy during phonation. The vocal folds are in the open phase at maximal amplitude. Note the increased amplitude (excursion of the vocal fold laterally from the midline) of the left vocal fold compared with the right. This is consistent with an atrophic, paretic left vocal fold. From Rosen CA, Simpson CB. *Operative techniques in laryngology*. New York, NY: Springer, 2008, with permission.

- **Imaging:**
 - CT with contrast or MRI:
 - From Skull base to Chest
 - Gold standard to evaluate lesions along course of Vagus Nerve in cases of idiopathic UVFP where no cause has been found.
 - **Signs of UVFP on CT:**
 - Ipsilateral short vocal fold.
 - Ipsilateral anterior dislocation of arytenoid.
 - Ipsilateral distention of piriform fossa with air.
 - Ipsilateral laryngeal ventricle dilatation.



- CXR:
 - Most lesions in chest that is big enough to cause VC paralysis can be seen on routine CXR.
- MBS and FEES:
 - Evaluate extent of Aspiration.
- **Laryngeal Electromyography (LEMG):**
 - Evaluates integrity of laryngeal neuromuscular system in by recording action potentials generated in laryngeal muscles during voluntary and involuntary contraction.
 - Most effective between 1-6 months post onset of paralysis.
 - Tests 3 laryngeal muscles:
 - 1. ThyroArytenoid (TA):**
 - Evaluation of RLN.
 - Needle placement is confirmed by vocalisation.
 - 2. Posterior CricoArytenoid (PCA):**
 - Evaluation of RLN.
 - Needle placement is confirmed by sniffing.
 - 3. CricoThyroid (CT):**
 - Evaluation of SLN.
 - Needle placement is confirmed by glide from one pitch to another.

- Indications of LEMG in VFP:

- 1. Neuropathy vs. Mechanical Fixation:**

- Vocal fold Paralysis:
 - EMG findings of Neuropathy.
 - CricoArytenoid Fixation:
 - Normal EMG findings.

- 2. Localization of lesion (SLN vs RLN):**

- CricoThyroid for SLN Paralysis
 - ThyroArtyenoid for RLN Paralysis

- 3. Prognosis of Vocal fold Paralysis:**

- Good Prognosis:
 - Presence of Motor-Unit Potentials.
 - Poor Prognosis:
 - Absence of Motor-Unit Potentials.
 - Fibrillation Potentials

- 4. Detects Synkinesis:**

- When ThyroArtyenoid activated by Sniffing or Posterior CricoArytenoid activated by vocalisation.

- Interpretation of LEMG:

- 1. Normal LEMG:**

- Bi or Triphasic Motor-Unit Potentials.
 - Seen in patients with CricoArytenoid Fixation.

- 2. Neuropathy:**

- Fibrillation Potentials:
 - Abnormal spontaneous activity at rest.
 - Due to ongoing axonal degeneration.
 - Starts after 3 weeks of injury.
 - Persist until Re-innervation occurs or until the motor endplates of the muscle degenerate (Myopathic injury).
 - Sharp waves:
 - Lower Frequency
 - Normal Amplitude

- 3. Muscle Re-innervation after Neuropathic injury:**

- Polyphasic Potentials
 - > 4 phases.
 - Due to Not well synchronized muscle fibers.

- 4. Myopathy:**

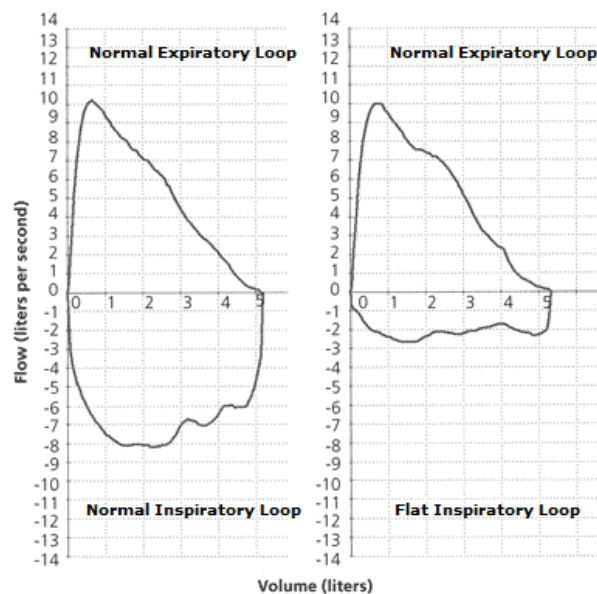
- Decreased functioning muscle fibers.
 - Polyphasic.
 - Low Amplitude.
 - Low Duration.





TABLE 61.3 CLASSIFICATION OF LARYNGEAL ELECTROMYOGRAPHY FINDINGS IN LARYNGEAL PARALYSIS				
Class	Spontaneous Activity	Recruitment	Motor Unit Morphology	Interpretation
I	Absent	Normal	Normal	Normal
II	Absent	Reduced	Low-amplitude polyphasic	Reinnervation
III	Absent	Reduced	Giant polyphasic units	Old injury
IV	Present	Reduced	Polyphasic units	Equivocal
V	Present	None	Fibrillations, positive sharp waves	Denervation

- **Pulmonary Function Test (PFT):**
 - PFT with a flow-volume loop is used to confirm VFP and to compare pre and post op results.
 - Flow-volume loop include forced inspiratory and expiratory maneuvers.
 - Extra-thoracic upper airway obstruction:
 - Normal expiratory loop.
 - Flat inspiratory loop.



- **Management of Unilateral VFP:**

1. **Conservative with Speech Therapy:**

- Watchful waiting with speech therapy for 6-12 months before surgical intervention is considered.
 - Healthy vocal fold may compensate the loss of function of paralyzed vocal fold by moving across midline to provide adequate voice quality and no aspiration.
- Early surgical intervention without waiting for conservative management is indicated for the following cases:
 1. Clinical Aspiration
 2. Vocal professional
 3. Tran-section nerve injury
 4. LEMG findings of poor prognosis

TABLE 69.3 PATIENT FACTORS AFFECTING TREATMENT STRATEGY FOR VOCAL FOLD PARALYSIS	
Patient Factor	Influence on Treatment (Early Surgical Intervention vs. Observation)
Presence of clinical aspiration	Favors early surgical intervention
Nature of nerve injury (transection, stretch, or unknown)	Nerve cut: favors early treatment ^a Nerve intact or stretch injury: favors conservative treatment (observation vs. temporizing injection laryngoplasty)
Vocal demands of patient	Nonvocal professional or limited voice use: favors observation Vocal professional—favors early surgical treatment
Medical comorbidity	Minimal comorbidity: all options open Significant comorbidity: favors local anesthetic (office-based injections vs. ML in OR)
LEMG findings	Good prognosis: favors observation or temporary injection Poor prognosis: favors early ML ^a or permanent injection

^aIn general, it is recommended that 2 to 3 months pass after nerve transection or injury before permanent surgery is performed to allow for muscular atrophy of the vocal fold to occur, to lessen the need for revision surgery months later.
LEMG, laryngeal electromyography; ML, medialization laryngoplasty; OR, operating room.

2. **Surgical Intervention (Medialization Producers):**

- Indications:
 - Failure of conservative management with speech therapy.
 - Early surgical intervention:
 1. Clinical Aspiration
 2. Vocal professional
 3. Tran-section nerve injury
 4. LEMG findings of poor prognosis

- Aim of the surgical intervention for UVFP is to Medialize the paralyzed vocal fold without compromising the airway.

- **Methods of Medialization Producers:**

1. Injection Thyroplasty
2. Type I Thyroplasty
3. Arytenoid Adduction
4. Re-innervation Procedures

- **Injection Thyroplasty:**

- General indications of VF injection in laryngology:

- o Unilateral VFP.
- o Sulcus vocalis.
- o Vocal fold atrophy.
- o Vocal fold scar.

- Injection augmentation of paralyzed vocal fold to medialize it.

- Site of injection:

- o **Temporary injection materials:**

- 2 points:

1. Lateral aspect of TA muscle at the mid-membranous part of VF.
2. Lateral to vocal process at posterior VF.

- o **Permanent injection materials:**

- Far lateral in Paraglottic space to avoid an irregular epithelial lining of vocal fold surface.

- o Avoid injections into:

- Superficial layer of lamina propria.
- Vocal fold ligament

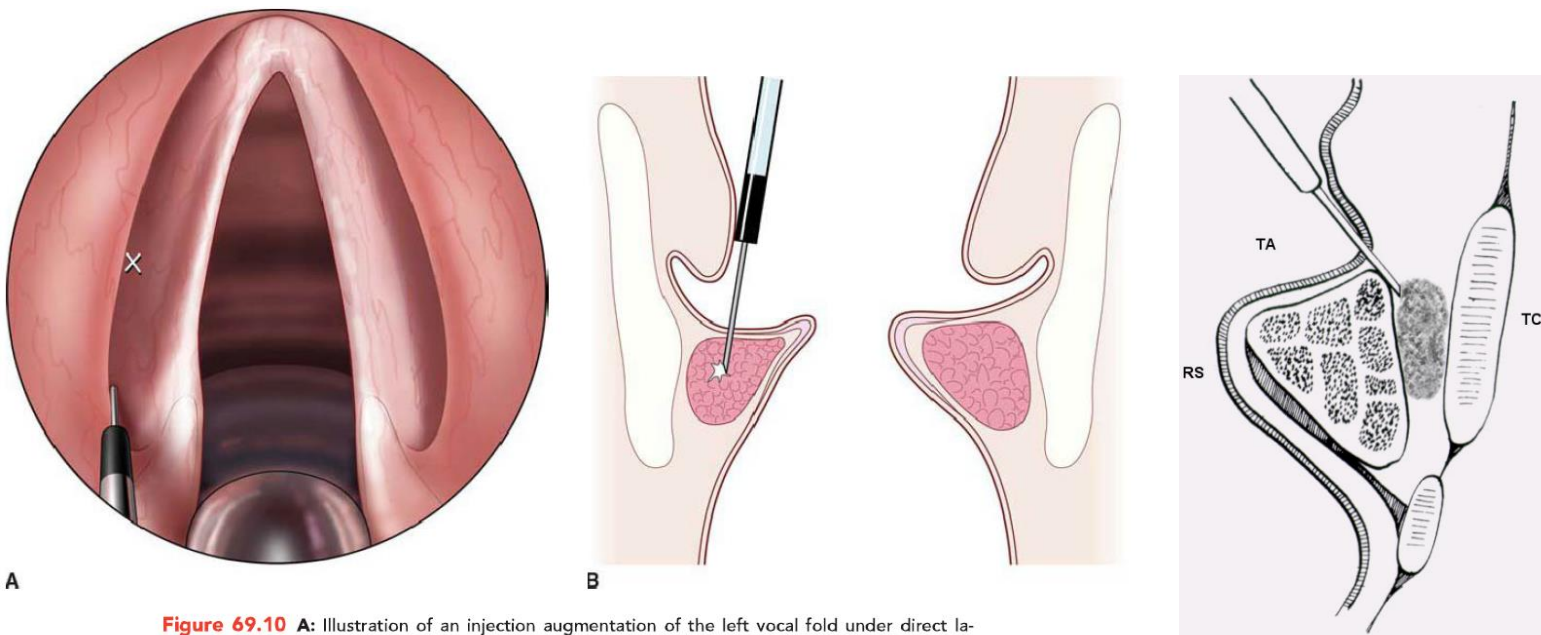


Figure 69.10 A: Illustration of an injection augmentation of the left vocal fold under direct laryngoscopy. "X" marks the injection site seen on direct visualization, at the posterior aspect of the vocal fold, lateral to the vocal process. B: The depth of injection into the TA muscle is depicted on coronal section. The needle tip should be deep into the vocal fold, and not superficially in Reinke space. From Rosen CA, Simpson CB. *Operative techniques in laryngology*. New York, NY: Springer, 2008, with permission.

- **Injection materials are classified into:**
 1. Temporary Materials
 2. Permanent Materials

TABLE 69.4 LARYNGEAL INJECTABLES			
Material	Length of Effect	Advantages	Disadvantages
Gelfoam	4–6 wk	Long track record	1. Short duration 2. Must use 18 g
Cymetra (Micronized AlloDerm)	2–4 mo	No allergy testing	1. More prep time 2. Expensive 3. Unpredictable
Fat	2 yr+ (Possibly permanent)	Autologous	1. Time/morbidity from harvest
Teflon	Forever	Forgiving Long lasting	2. Unpredictable 1. Granuloma 2. VF stiffness Risk of VF stiffness with superficial injection
Radiesse (calcium hydroxylapatite)	18 mo	1. 24 g needle 2. Long lasting	
Radiesse Voice Gel (carboxymethylcellulose)	1–3 mo	1. 24 g needle 2. Temporary/trial VFI	
Restylane (hyaluronic acid)	2–4 mo	1. 25 g needle (office) 2. Temporary/trial VFI	Few data

- **Temporary (Re-absorbable) Materials:**
 - Indication:
 - Patients with temporary UVFP to bridge the time until recovery.
 - UVFP with good prognosis on LEMG.
 - It can be done under LA as office based procedure or under GA for better control during injection.
 - Temporary injection materials:
 - 1. Fat:**
 - Advantages:
 - Autologous.
 - Excellent biocompatibility.
 - Simple and minimal costs.
 - Disadvantages:
 - Unpredicted resorption rate.
 - Harvesting time.
 - 2. Calcium Hydroxylapatite (Radiesse):**
 - Synthetic material identical to bone tissue.
 - Advantages:
 - Excellent biocompatibility.
 - Lasts for 18 months (Longest duration)
 - 3. Hyaluronic Acid (Restylane):**
 - Advantages:
 - Lasts 2–4 months.
 - No need for Allergy Test pre-injection

4. Cymetra (Collagen formed of micronized homologous alloderm):

- Advantages:
 - Excellent biocompatibility.
 - Lasts 2-4 months.
 - No need for Allergy test pre-injection
- Disadvantages:
 - Needs time for preparation before injection.
 - High costs.

- Permanent Materials:

- Indication:
 - Patients with expected permanent UVFP.
 - Tran-section nerve injury.
 - UVFP with poor prognosis on LEMG.
- It should be done under GA for better control during injection (Irreversible effect).
- Permanent injection materials:
 - 1. Teflon:**
 - Advantages:
 - Permanent.
 - Disadvantages (Not anymore used):
 - Risk of Granulation tissue formation (50%).
 - Risk of Migration.
 - Risk of Airway obstruction.

2. VOX (Silicone Paste):

- Considered a reasonable alternative to framework surgery for patients with permanent UVFP.
- Advantages:
 - Permanent.
 - Excellent biocompatibility
 - No risk of Airway obstruction.
 - No risk of Granulation tissue formation.
 - No risk of Migration to distant organs.
 - No need for Allergy Test pre-injection
- Disadvantages:
 - Needs High-pressure administration device for injection.

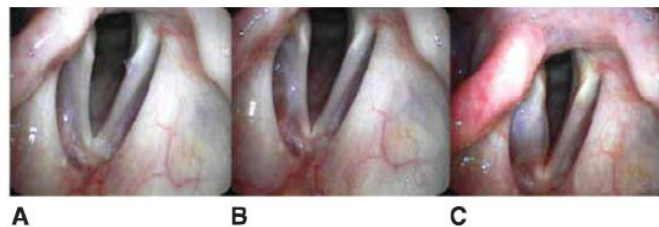
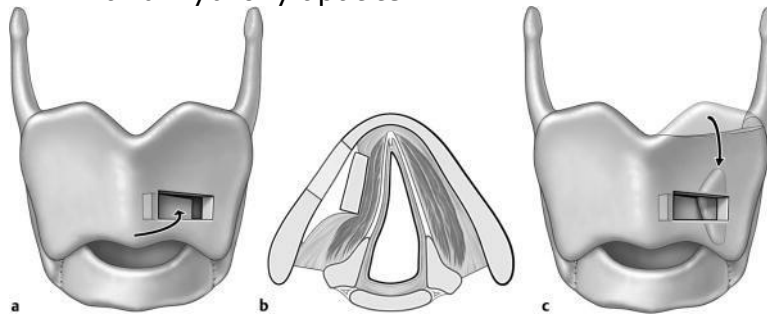


Figure 69.11 Fiberoptic laryngoscopy view from a right vocal fold injection augmentation. **A:** Preinjection. **B:** Initial injection. **C:** Postinjection. From Rosen CA et al. Advances in office-based diagnosis and treatment in laryngology. *Laryngoscope* 2009;119:S185–214, with permission.

- **Type I Thyroplasty (Medialization Laryngoplasty):**

- Laryngeal Framework Surgery.
- Gold standard for long-term treatment of unilateral VFP.
- Medialize paralyzed vocal fold from external approach and work through Thyroid cartilage.
 - o Small window is incised and removed from Thyroid cartilage.
 - o Implant is placed through the window to medialize the paralyzed VC.
 - o Most common implant used is a Silastic block, Gore-Tex and Hydroxylapatite.



- **Advantages:**

- o Reversible procedure.
- o Good voice outcome
- o Can be performed under LA.
- o 3D increase in vocal fold size
 - Anterior-posterior
 - Medial-lateral
 - Superior-inferior.
- o Does not hinder mobility of vocal fold.

- **Disadvantages:**

- o Avoid implant too anterior or too superior
- o Post- op Paraglottic edema and submucosal hemorrhage.
 - Lasts 2-6 weeks before resolving.

- **Post-op Care:**

- o Post-implant intubation must be done with care.
 - 6 months waiting period for elective surgery.
 - 6.0 ETT to avoid inducing laryngeal edema.

- **Complications:**

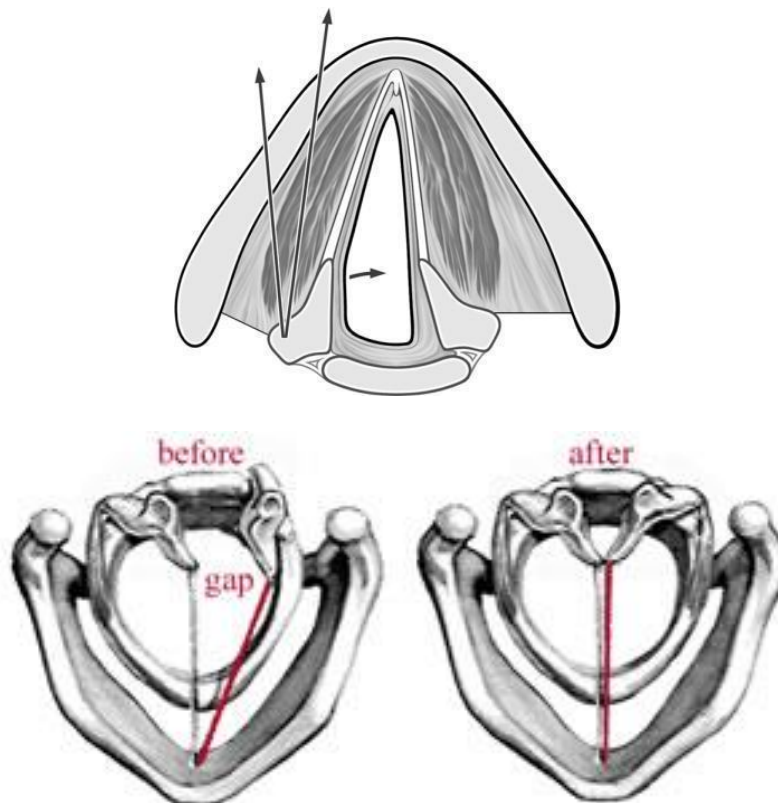
- o Persistent dysphonia.
- o Airway obstruction.
- o Implant migration or extrusion.
- o Hematoma.
- o Infection

- **Contraindications:**

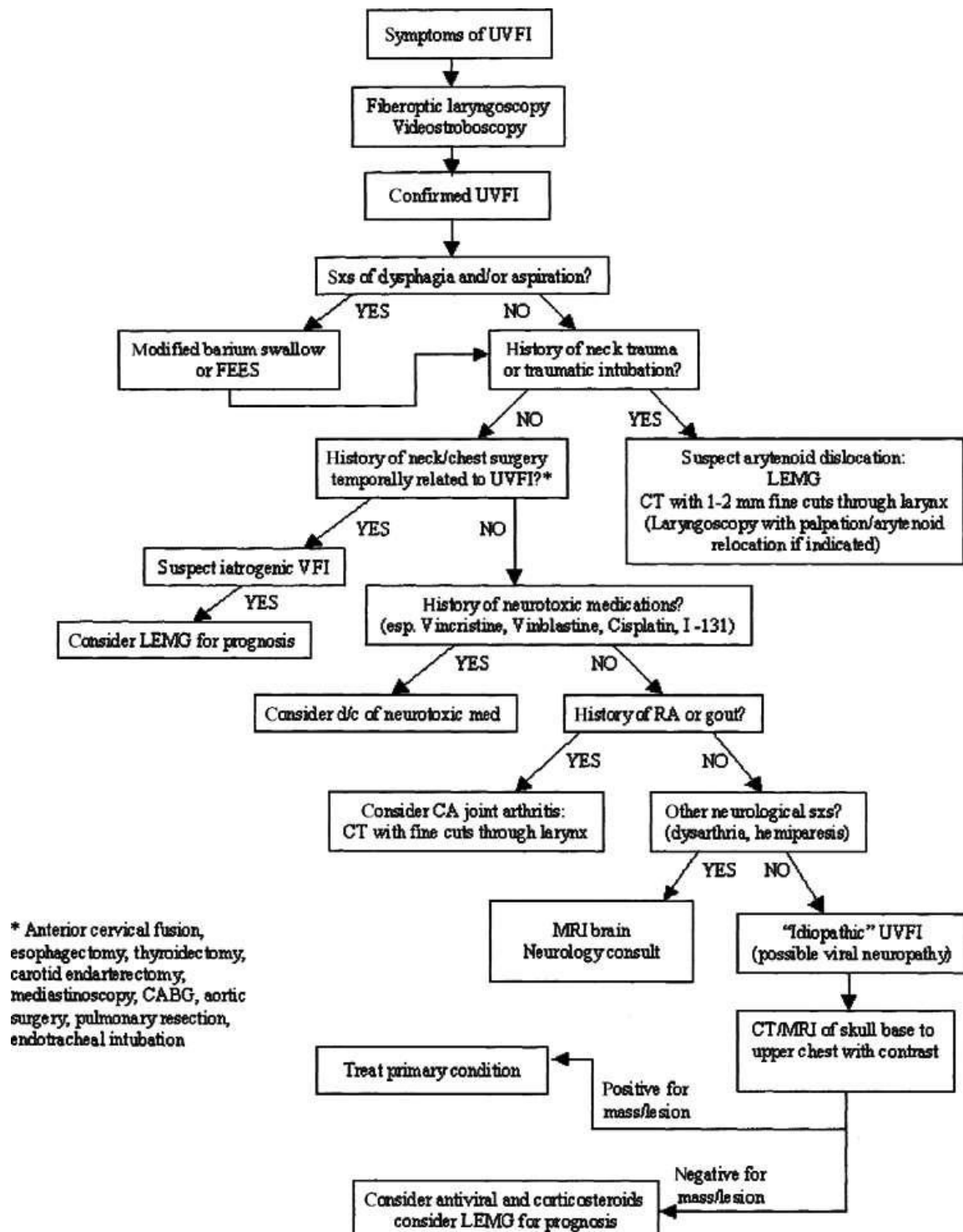
- o Patients with hemilaryngectomy.
- o Previous laryngeal irradiation due to extensive scarring.

- **Arytenoid Adduction:**

- Laryngeal Framework Surgery.
- Placing Arytenoid cartilage in the most favorable position for rehabilitation of paralyzed vocal fold.
 - o Placing a suture through Muscular process of Arytenoid.
 - o Into Thyroid lamina Anteriorly.
 - o Simulates Lateral CricoArytenoid contraction.
- Indicated as an adjunct to Type I Thyroplasty in the following:
 1. Large posterior glottic gap.
 - Vocal processes will not touch in midline during phonation.
 2. Asymmetric vocal fold levels.
 3. Patients with MPT < 5 sec.
- Results:
 - o Lower position of vocal process.
 - o Medialize and stabilize vocal process.
 - o Rotate Arytenoid.
 - o Lengthens paralyzed vocal fold.



- **Key differences between Injection Thyroplasty and Laryngeal Framework Surgery:**
 1. Injection Thyroplasty is less effective at closing large (3 mm or greater) glottic gaps, especially posterior membranous region of the vocal folds.
 2. Injection Thyroplasty may be less precise than framework surgery
 3. Injection Thyroplasty is more minimally invasive approach and can be carried out in a clinic-based setting.
- **Reinnervation Procedures:**
 - Indicated for unilateral permanent VFP.
 - Contraindications:
 - Fixation of Arytenoid cartilages
 - Disruption of Ansa Cervicalis.
 - Advantages:
 - Maintains muscle tone.
 - No foreign body reaction.
 - Best preservation of mucosal wave.
 - Most physiologic.
 - Disadvantages:
 - Does not result in active ADduction and ABduction.
 - Long operative time.
 - Delayed results (up to 6 months).
 - High technical skill.
- Ansa cervicalis nerve has similar fiber composition (myelinated vs. unmyelinated) to RLN making it compatible for RLN grafting.
- Types of Reinnervation Procedures:
 1. End-to-end anastomosis of RLN.
 2. Ansa cervicalis to main trunk of RLN.
 3. Anastomosis of Ansa cervicalis to Adductor branch of RLN.
 4. Ansa cervicalis Neuromusclar pedicle (omohyoid) to TA muscle.
 5. Vagus, Ansa, or Phrenic nerve fibers or motor end plate insertion into Thyroarytenoid.
- In a recent multicenter randomized trial, Paniello et al. directly compared medialization laryngoplasty and laryngeal reinnervation and found no difference between these procedures.



Bilateral Vocal Fold Paralysis (BVFP):

- Bilateral vocal fold immobility (BVFI) can be caused by:
 - Bilateral vocal fold paralysis (BVFP)
 - Posterior glottic stenosis (PGS) with or without CA joint fixation.
- It is important to distinguish clinically between these two entities to successfully manage BVFI.
- **Bilateral RLN Paralysis:**
 - Bilateral paralysis of **ALL** intrinsic laryngeal muscles **EXCEPT** Cricothyroid.
 - Vocal cords situation:
 - ParaMedian Position.
 - Loss of ABduction on Deep inspiration.
 - Preservation of Partial ADduction from action of:
 - CricoThyroid (Intact SLN).
- **Bilateral SLN Paralysis:**
 - Bilateral paralysis of Cricothyroid muscles and bilateral anaesthesia of Larynx Above VC.
- **Bilateral Complete (RLN and SNL) Paralysis:**
 - Paralysis of **ALL** laryngeal muscles on both.
 - Causes are similar to RLN paralysis.

Table 59-3. Causes of recurrent laryngeal nerve paralysis (low vagal trunk or recurrent laryngeal nerve)

Right	Left	Both
• Neck trauma	I. <i>Neck</i>	
• Benign or malignant thyroid disease	• Accidental trauma	
• Thyroid surgery	• Thyroid disease (benign or malignant)	Thyroid surgery
• Carcinoma cervical oesophagus	• Thyroid surgery	Carcinoma thyroid
• Cervical lymphadenopathy	• Carcinoma cervical oesophagus	Cancer cervical oesophagus
	• Cervical lymphadenopathy	Cervical lymphadenopathy
	II. <i>Mediastinum</i>	
• Aneurysm of subclavian artery	• Bronchogenic cancer	
• Carcinoma apex right lung	• Carcinoma thoracic oesophagus	
• Tuberculosis of cervical pleura	• Aortic aneurysm	
• Idiopathic	• Mediastinal lymphadenopathy	
	• Enlarged left auricle	
	• Intrathoracic surgery	
	• Idiopathic	

Causes of Bilateral Vocal Fold Paralysis (BVFP):

1. Surgical Trauma/Iatrogenic (55%):

- Most common cause of BVFP.
- Most commonly occur post

Thyroid surgery.

- Patient develops stridor immediately on extubation or several hours after the procedure.

2. Neoplastic:

- Bronchogenic
- Thyroid
- Laryngeal
- Esophageal

3. Neurologic:

- Brainstem stroke
- CNS neoplasms
- Arnold Chiari malformations
- Hydrocephalus
- Guillain-Barre syndrome
- Parkinson
- Myasthenia gravis
- Multiple sclerosis
- Postpolio syndrome

4. Traumatic

5. Idiopathic

- In cases where no clear-cut and immediate surgically induced (neck or chest) RLN trauma exists, three primary questions help to sort out the direction of subsequent workup:
 - History of rheumatoid arthritis ?
 - History of blunt or penetrating neck trauma ?
 - Endotracheal intubation in non-neck or -chest cases ? especially if a history of prolonged ventilatory dependence exists.
- If any of the above questions are positive, a high likelihood exists that CA fixation or Posterior glottic stenosis the cause of the bilateral immobility, not paralysis.
- If the answer to all three questions above is negative,. then BVFP is almost certainly present, and represents a manifestation of progressive neurologic, congenital, endocrine, metabolic, toxic, neoplastic or inflammatory condition.

**TABLE
69.5**

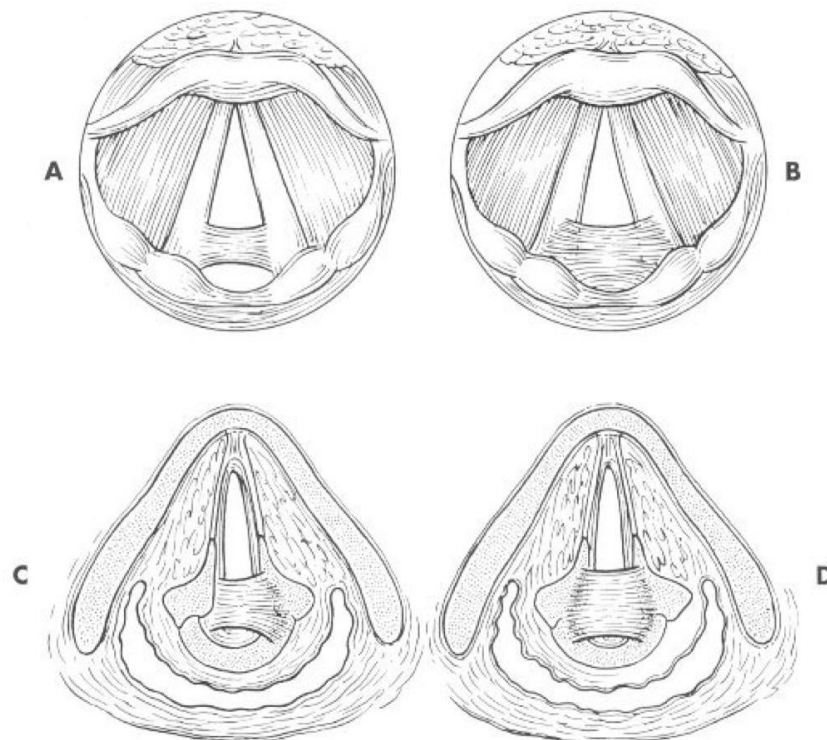
ETIOLOGY OF BVFP AND BILATERAL VOCAL FOLD IMMOBILITY (BVFI)

Etiology	Percentage
BVFP^a	
Iatrogenic (surgical trauma)	55.5
Thyroid surgery	48.6
Nonthyroid surgery	6.9
Malignancy (lung/nonlung)	9.7
Intubation	9.7
Idiopathic	8.3
Neurologic	6.9
Trauma	1.4
Other	8.4
BVFI^b	
Iatrogenic	25.7
Malignancy	17
Neurologic	12.8
Intubation	15.4
Idiopathic	12.8
External trauma	11.1
Rheumatoid arthritis or inflammatory disease	3.4
Radiation therapy	1.7
TOTAL	100

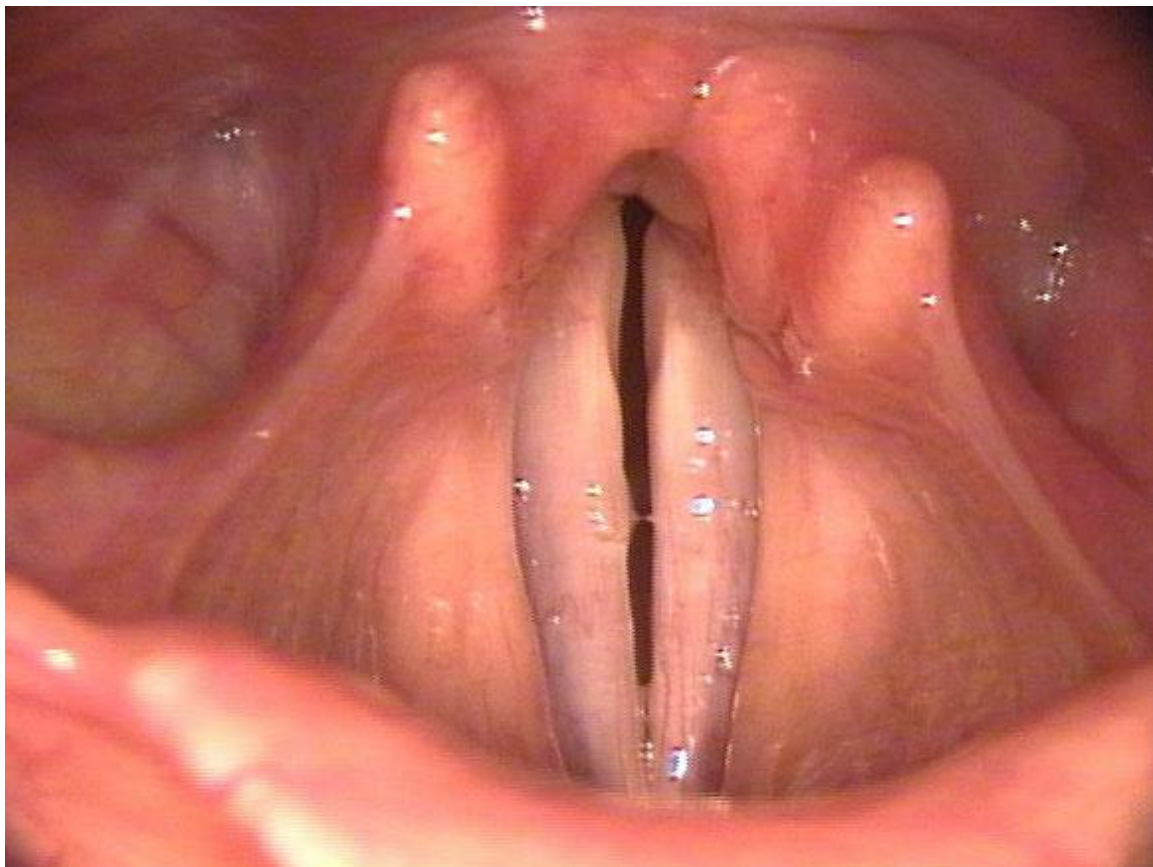
^aAdapted from (3) Rosenthal LHS, Benninger MS, Deeb RH. Vocal fold immobility: a longitudinal analysis of etiology over 20 years. *Laryngoscope* 2007;117:1864–1870, with permission.

^bAdapted from Benninger MS, Gillen JB, Altman JS. Changing etiology of vocal fold immobility. *Laryngoscope* 1998;108:1346–1349, with permission.

- **Differential diagnosis of BVFP:**
 - Posterior glottic stenosis (PSG):
 - Diagnosed by:
 - No history of surgical iatrogenic trauma.
 - LEMG
 - DLB and EUA:
 - Inspection for InterArytenoid scarring.
 - Palpation of vocal folds for passive mobility.
 - Posterior commissure and contralateral arytenoid cartilage will be moved to midline during vocal process palpation because of entire posterior glottic complex being fused with scar tissue.
 - CA Fixation:
 - Diagnosed by:
 - No history of surgical iatrogenic trauma.
 - History of RA.
 - LEMG
 - DLB and EUA:
 - CA joint palpation for passive mobility.



- **Clinical Picture of Bilateral VFP:**
- **Voice:**
 - Normal voice due to narrow glottis.
 - Maybe dysphonic.
- **Airway:**
 - Airway obstruction
 - Inspiratory or Biphasic Stridor.
 - Vocal folds typically drift to midline soon after neural injury (can be delayed for days or weeks in some cases).
- **Aspiration:**
 - Aspiration of liquid due to ineffective cough.
- **Evaluation of Bilateral VFP:**
- **Endoscopy:**
 - Bilateral immobility of vocal folds during phonation and inspiration.
 - Paralyzed VC in median or paramedian position.
 - 1-3 mm slit-like glottic chink.



- **Management of Bilateral VFP:**
- The goal is to lateralize vocal fold for airway improvement without compromising voice and causing aspiration as possible.
- Surgical interventions for BVFP cannot simultaneously result in improvement in airway without dysphonia (except Tracheostomy).
- **Initial Management of the Airway:**
 - Main determinant for intervention is status of patient's airway.
 - In patient with respiratory distress, Endotracheal intubation is the preferred initial management.
 - Gives time before decision can be made regarding short- or long-term airway management strategies.
 - In cases of BVFP directly after Thyroidectomy:
 - Intubation with sedation in ICU.
 - IV corticosteroids.
 - Trial of controlled extubation in OR after 5-10 days later (Bilateral neuropraxic injury).
 - If the patient is still symptomatic:
 - Secure airway with Tracheostomy or other temporizing measures.
- **Temporizing (Reversible) Treatment of BVFP:**
 1. Tracheostomy
 2. Endo-Extralaryngeal Suture Lateralization
 3. Laryngeal Botox Injection

1. Tracheostomy:

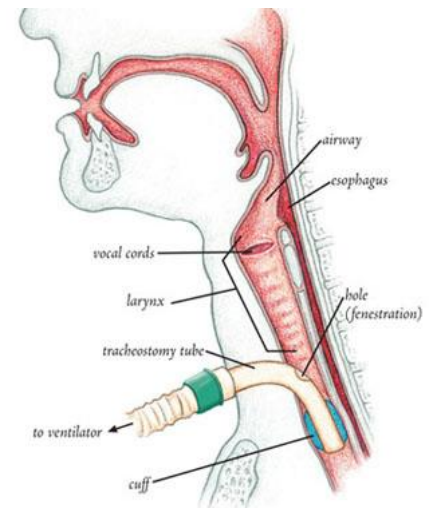
- Excellent treatment to secure the airway until adequate time passes for spontaneous recovery to occurs (8-12 months).
- In long-standing cases of BVFP, the choice is between a permanent Tracheostomy with a speaking valve or a surgical procedure to Lateralize vocal folds.
- Patients must undergo lateralizing procedure (at least 4 months) before decannulation.

- **Advantages:**

- Immediate relief of airway restriction.
- Can be performed under LA
- Larger airway caliber and better voice quality compared to other lateralization producers.

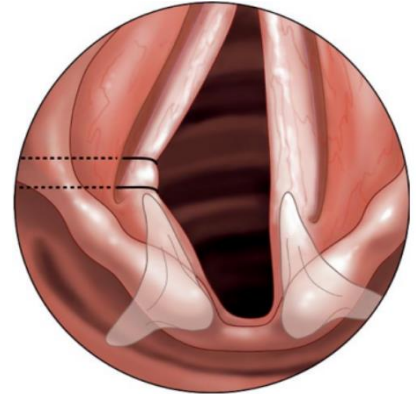
- **Disadvantages:**

- Creation of a stoma that has both cosmetic and long-term care problems.
- The need to occlude the tube or wear a speaking valve to phonate.



2. Suture Lateralization:

- Viable option for temporizing patient's airway in select cases of BVFP.
- Technique:
 - Non-absorbable suture placed endoscopically above and below posterior vocal fold at region of the vocal process.
 - Suture can be brought out over thyroid ala, or through the skin of the neck and secured over a button externally.
 - Suture tension can be adjusted intra-op and post-op to fine tune the degree of vocal fold lateralization or airway aperture.



3. Laryngeal Botox Injection:

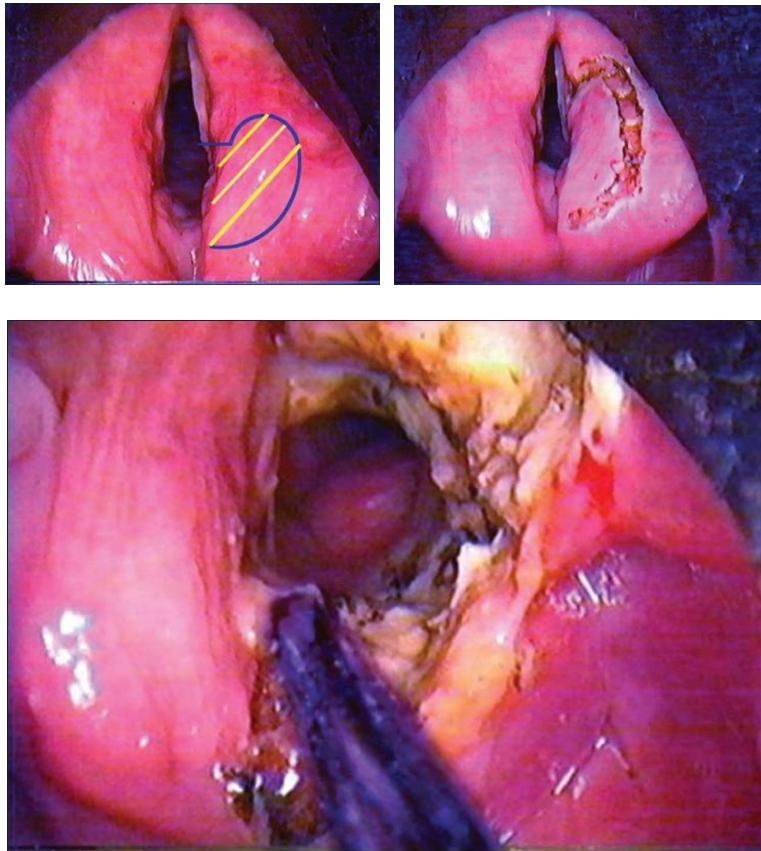
- Indicated in selected cases of BVFP where airway obstruction is not imminent and airway symptoms are mainly exertional.
- Injection of 2.5 mouse units in each vocal fold into TA-LCA muscular complex.
- May result in minor airway improvement.
- Lasts 2-4 months.
- Side effects:
 - Breathy dysphonia
 - Aspiration of liquids

- **Long-Term Surgical Treatment of BVFP:**

1. Laser Arytenoidectomy
2. Transverse Cordotomy
3. Arytenoid Abduction
4. Laryngeal Pacing

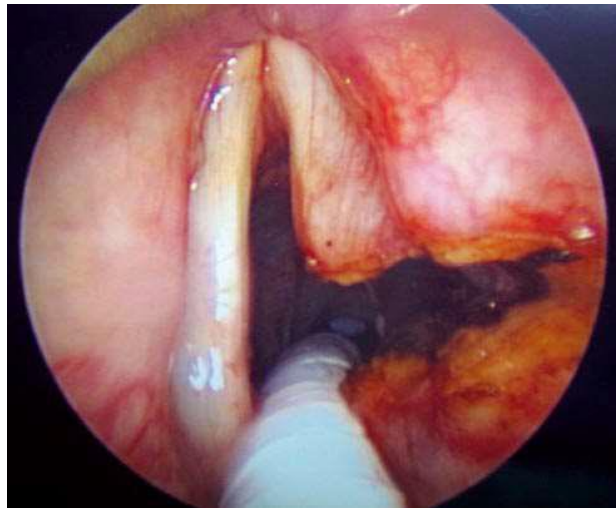
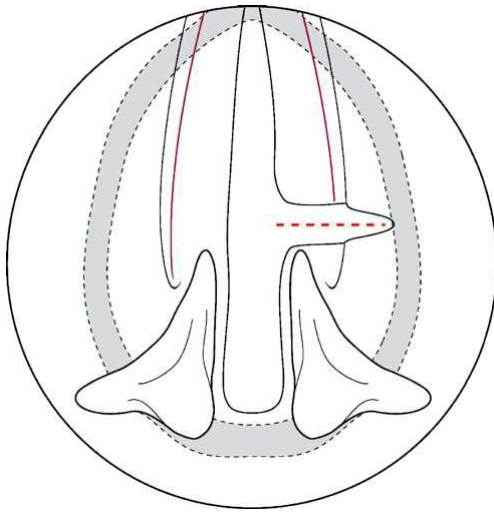
1. Laser Arytenoidectomy:

- Done using endoscopic CO2 laser
 - Increased precision.
 - Better hemostasis.
- **Total Arytenoidectomy:**
 - Entire Arytenoid cartilage is ablated.
 - Small wedge of posterior vocal fold is removed.
 - Not used much now given other options.
 - Good voice results.
 - 85% success rate.
- **Partial Arytenoidectomy:**
 - Preserves the lateral aspect of Arytenoid.
 - **Advantages:**
 - Reduce risk of aspiration.
 - Voice is better than Total Arytenoidectomy because there is no violation of membranous vocal fold.



2. Transverse Cordotomy:

- Done using endoscopic CO2 laser.
- Transverse cut anterior to vocal process quite far laterally 3–4 mm (cut through false cord as well).
- Result is cut ThyroArytenoid muscle which scars anteriorly over 4-6 weeks.
- Advantages:
 - Easy.
 - Comparable airway outcome to laser arytenoidectomy.
 - Reasonable conversational voice (breathy and rough).



3. Arytenoid Abduction:

- External approach procedure that simulates action of Posterior cricoarytenoid muscle (PCA).
- Suture is placed in muscular process and anchored to inferior cornu of thyroid cartilage.
- Externally rotates the arytenoid to move vocal process laterally and rostrally.
- Aim to enlarge glottic airway while allowing continued vocal fold adduction during phonation.
- Technically more difficult than other endoscopic procedures.

4. Laryngeal Pacing:

- Most promising treatment for BVFP.
- Implantation of programmable pulse generator beneath the skin and delivers electrical stimulation to PCA muscle during inspiration once triggered by an electrode on diaphragm.
- Improves ventilation without impairing the voice.

Pediatric Vocal Fold Paralysis (VFP):

- **Unilateral Vocal Fold Paralysis (UVFP):**
 - Less common in newborns than bilateral VFP.
 - **Causes:**
 - **In Newborns:**
 - Direct injury during a difficult delivery.
 - **In Infants:**
 - Surgical iatrogenic injury to vagus or RLN.
 - Cardiovascular surgery (Most common).
 - Esophageal surgery for atresia with tracheo-esophageal fistula.
 - **Clinical picture:**
 - Mild, position-dependent inspiratory stridor.
 - Hoarse, breathy cry.
 - Potential feeding difficulties (aspiration).
 - **Treatment:**
 - **Conservative:**
 - Watchful follow-up with dietary recommendations.
 - Does not require any therapy in majority of cases as natural improvement in voice occurs over time.
 - **Instructions:**
 - Infant should be fed on affected side.
 - Fluids should be thickened with powder to allow for adequate nutritional intake and weight gain.
 - During sleep, the same position contributes to decrease inspiratory stridor.
 - **Surgical Intervention (Medialization Producers):**
 - Indicated for older children and adolescents when speech therapy did not improve dysphonia.
 - **Injection Thyroplasty:**
 - Only medialization producer indicated in old children.
 - Temporary materials should be injected only.
 - Permanent materials should be avoided because of their irreversible and unpredictable long-term effects.
 - **Tracheotomy:**
 - Indicated only in:
 - Respiratory distress.
 - Persistent aspiration.

- **Bilateral Vocal Fold Paralysis (BVFP):**
 - 2nd most common congenital laryngeal anomaly.
 - Accounts for more than 50% of all pediatric VFPs.
 - Less common in newborns than bilateral VFP.
 - Causes:
 - **Neurological:**
 - Arnold–Chiari II malformation (1/3 of cases)
 - Hydrocephalus
 - Myelomeningocele
 - Intracerebral haemorrhage
 - **Traumatic:**
 - Birth trauma is the most common cause of traumatic BVFP.
 - Iatrogenic surgical trauma.
 - **Idiopathic:**
 - Recover spontaneously in 50% of the cases within 1-2 years.
 - Clinical picture:
 - High-pitched stridor.
 - Signs of respiratory distress:
 - Suprasternal and chest retractions.
 - Exacerbations is noted during increased airway demands.
 - Normal or near normal cry.
 - 45% association with upper airway diseases:
 - Laryngomalacia.
 - Tracheo(broncho)malacia
 - Subglottic stenosis
 - Diagnosis:
 - MRI is necessary to detect any CNS pathology.
 - Awake transnasal flexible laryngoscopy (TNFL) to confirm the clinical diagnosis.
 - MLB under GA:
 - Essential to confirm the diagnosis and assessment of entire upper airway:
 1. Assessing vocal fold mobility when awake office TNFL has failed.
 2. Differentiating BVFP from PGS especially in case of prior endotracheal intubation.
 3. Searching for associated lesions of the upper airway

- **Treatment of BVFP:**
 - If urgent stabilization of the airway is required:
 - Infant should be intubated first.
 - MRI to rule out Type II Arnold–Chiari malformation with hydrocephalus.
 - Shunt procedure can be performed by neurosurgeon to decrease high ICP and stretching of the vagus nerve.
 - Subsequent recovery of vocal fold movements alleviates need for tracheostomy.
 - **Tracheostomy:**
 - Required only in 50% of patients with BVFP.
 - Indications of airway stabilization:
 - Failure to thrive
 - Spells of apnoea
 - Cyanotic attacks.
 - **PICU monitoring with nasal CPAP and enteral feeding:**
 - Alternative method of tracheostomy.
 - Aims to buy time until spontaneous recovery of vocal fold function.
 - Continue for several weeks, representing a prolonged period of discomfort for the child.
 - **Long-term Intubation:**
 - Alternative method of tracheostomy.
 - With periodic reassessment of Vocal fold mobility.
 - 50% of patients recovered within 1st 3 months of life.
 - Advantage of avoiding Tracheostomy.
 - Risk of developing Posterior glottic stenosis (PGS).

- **Lateralization procedures for BFVP:**
 - Done only after the child reaches 2 years of age.
 - Spontaneous recovery occurs within 1-2 years in 50% of idiopathic and iatrogenic BVFP cases.
 - Decannulation rate following surgery remains the most common outcome measure.
 - **Open surgery:**
 - Excellent decannulation rate.
 - Either Postero-lateral Approach or Laryngofissure Approach:
 - **Arytenoidectomy:**
 - Removal of Arytenoid.
 - **Arytenoidopexy:**
 - Laterally sutured Arytenoid laterally to Thyroid ala.
 - **Arytenoidectomy with suture lateralization:**
 - Combined procedure.
 - Superior to endoscopic-CO2 laser arytenoidectomy.

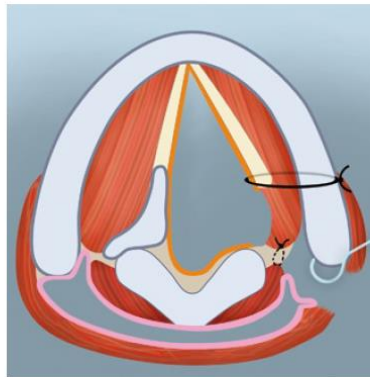


Fig. 7.2 External lateral approach for arytenoidectomy with suture lateralisation of the vocal process

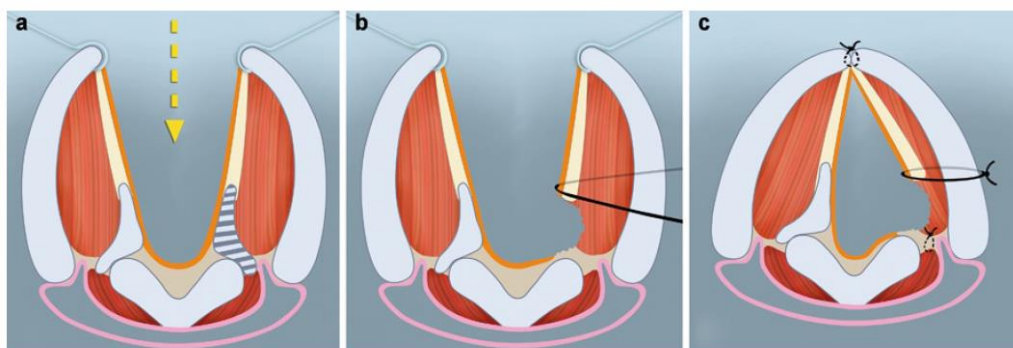


Fig. 7.3 Laryngofissure approach for arytenoidectomy, with suture lateralisation of the vocal process: (a) Laryngofissure. (b) Arytenoidectomy and vocal cord lateralisation. (c) End result

- **Endoscopic surgery:**
 - Laser Arytenoidectomy
 - Transverse Cordotomy
 - Arytenoid Abduction
 - Laryngeal Pacing

1. Laser Arytenoidectomy:

- Done using endoscopic CO₂ laser on Tracheostomised children.
- Combined with transverse cordotomy to increase posterior glottic chink.

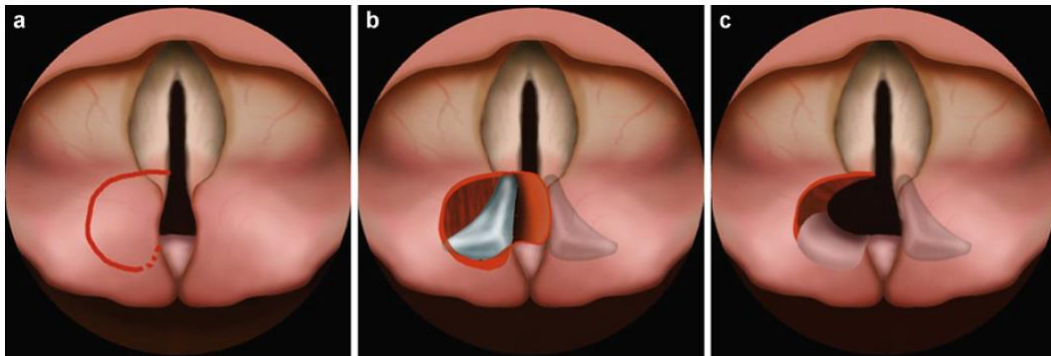


Fig. 7.4 Submucosal CO₂ laser arytenoidectomy: (a) Mucosal flap design. (b) Medial reflexion of the mucosal flap with exposure of the arytenoid. (c) Posterior reflexion of the mucosal flap into the arytenoidectomy bed

2. Transverse Cordotomy:

- Done using endoscopic CO₂ laser on Tracheostomised children.
- Combined with total arytenoidectomy to increase posterior glottic chink.

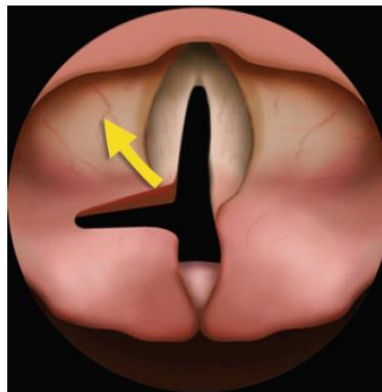


Fig. 7.6 CO₂ laser posterior cordotomy: a simple transverse section from the vocal process of the arytenoid to the lateral thyroid cartilage creates a wedge-opening of the glottis with anterior retraction of the thyro-arytenoid muscle (*arrow*)

3. Vocal Fold Lateralization:

- Reversible procedure to avoid tracheotomy.
- Endo-extralaryngeal suture lateralization of one arytenoid is performed.

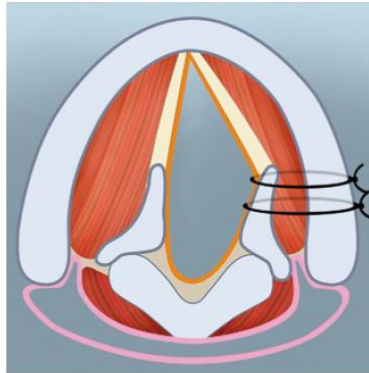
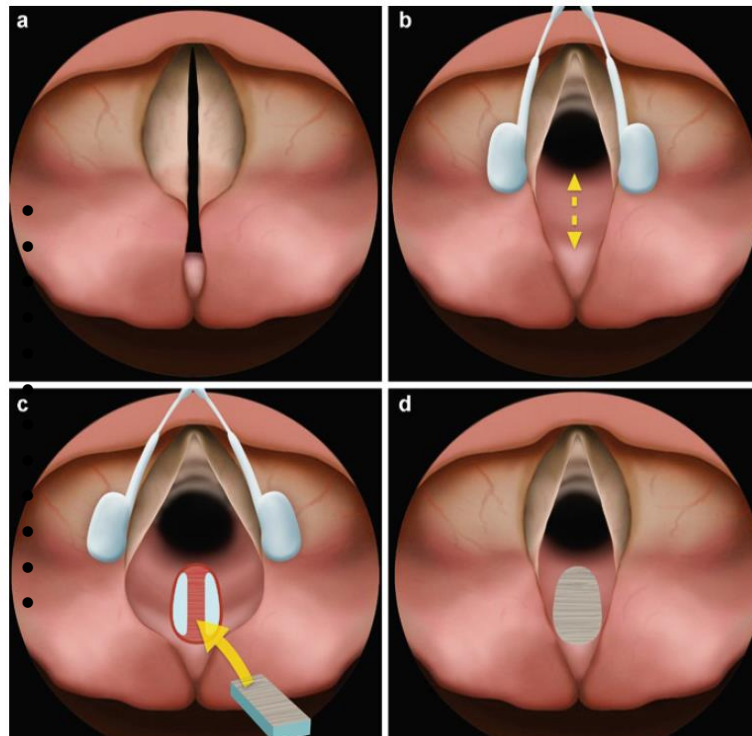


Fig. 7.7 Endoscopic vocal cord lateralisation with the Lichtenberger endo-extra-laryngeal suture technique: a non-resorbable 4.0 prolene suture is placed around the vocal process of the arytenoid and tied on the external aspect of the thyroid cartilage. A second stitch is often used to improve lateralisation

4. Endoscopic Posterior Cricoid Split and Rib Grafting:

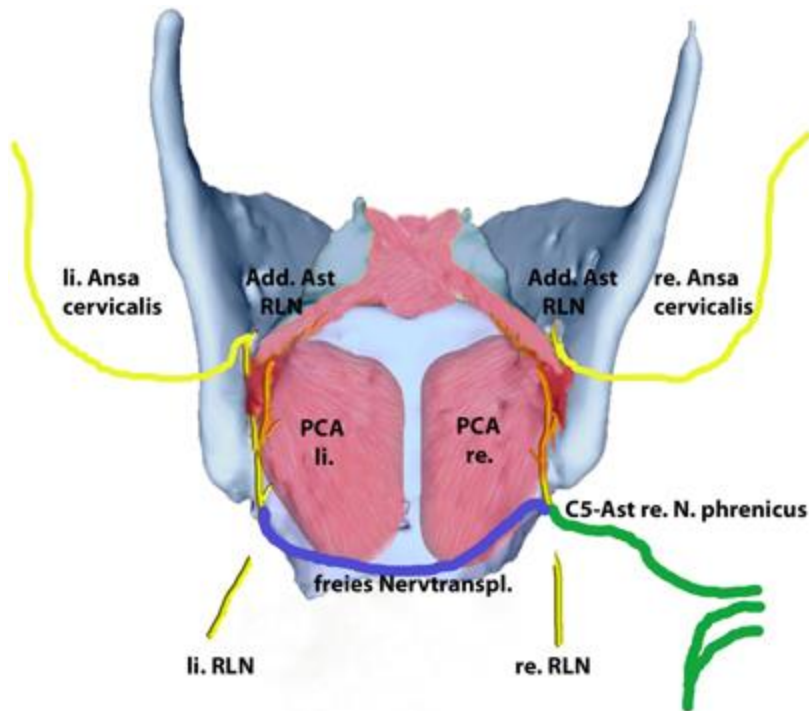
- Posterior enlargement of interarytenoid space by CO₂ laser division of cricoid plate and endoscopic interposition of a costal cartilage graft.
- Advantages:
 - Integrity of vocal folds and arytenoid is preserved
 - Anterior laryngeal commissure is not damaged
 - Does not compromise further surgery
 - No irreversible damage to laryngeal tissues

Fig. 7.8 Endoscopic posterior cricoid split with costal cartilage grafting (Ingkis technique [30]): (a) Bilateral vocal cord paralysis. (b) Exposure of the posterior glottis with Lindholm false cord retractor. The arrow shows the extent of the cricoid split. (c) Complete posterior midline cricoid split with the CO₂ laser and costal cartilage ready to be placed between the two cricoid laminae. (d) End result: the interarytenoid distance is improved as compared to the initial condition



5. Reinnervation of Posterior Cricoarytenoid (PCA):

- Reinnervation using Ansa cervicalis nerve-muscle pedicle transfer to the posterior cricoarytenoid muscle.
- Not routinely used in clinical practice due to technical difficulties and inconsistent results.



BENIGN VOCAL FOLD LESIONS

- Includes:
 - Keratotic lesions
 - Infectious (Fungal) lesions
- **Keratosis:**
- Benign (pre-malignant) epithelial hyperkeratotic lesions.
- Potential risk for malignancy transformation:
 - Mild/moderate dysplasia **10%**
 - Severe dysplasia /CIS **30%**
- Includes:
 1. Leukoplakia
 2. Erythroplakia
- Risk factors (Laryngeal irritation):
 1. Smoking
 2. LPR
 3. Vocal abuse or misuse
 4. Glottal insufficiency
- Diagnosis:
 - Microlaryngoscopy with excisional biopsy to rule out a malignant process.
- Treatment:
 - Avoidance of risk factors.
 - **Sub-epithelial cordectomy (Type 1)**
 - Monitor regularly (3-6 months) and repeat biopsy with any changes in nature, size or location.

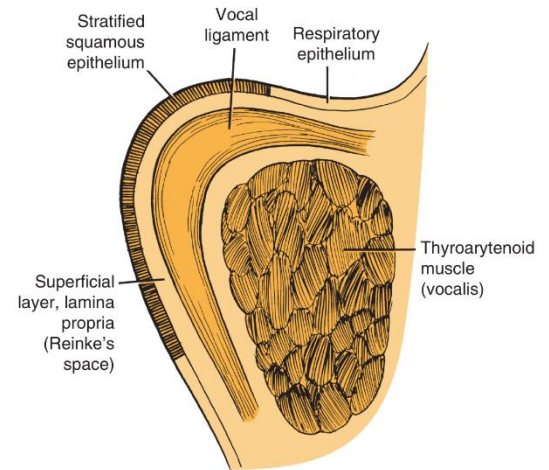
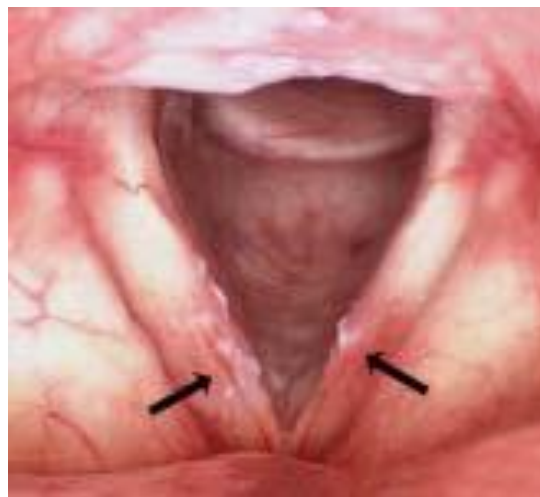


FIGURE 61-1. Cross section of the vocal fold.



- **Fungal Laryngitis:**
- Risk factors:
 - Immunocompromised patient:
 - Diabetes
 - HIV
 - Chronic use of corticosteroids (Systemic or **inhalers**).
 - Radiation
 - Poor nutrition
 - Debilitating illnesses
 - Long-term antibiotics
- Clinical picture:
 - Dysphonia
 - Cough
 - Aspiration
 - Odynophagia
 - Mucositis
- Diagnosis:
 - Endoscopy and biopsy and culture of the white patches (may be confused with malignancy).
- Pathogens:
 - Candidiasis (Moniliasis):
 - Most common.
 - Adherent, friable, cheesy, white plaques, spread from oral cavity.
 - Aspergillosis:
 - Allergic, noninvasive, or invasive forms.
 - Blastomycosis:
 - Red laryngeal ulcers or miliary nodules on vocal fold.
- Treatment:
 - Effectively treated with oral antifungal medications.



BENIGN VOCAL FOLD TUMORS

- Divided into:
 1. Non-neoplastic (Reactive).
 2. Neoplastic.

Non-neoplastic	Neoplastic
• Solid ○ Vocal nodules ○ Vocal polyp ○ Reinke's oedema ○ Contact ulcer ○ Intubation granuloma ○ Leukoplakia ○ Amyloid tumours • Cystic ○ Ductal cysts ○ Saccular cysts ○ Laryngocele	• Squamous papilloma ○ Juvenile type ○ Adult-onset type • Chondroma • Haemangioma • Granular cell tumour • Glandular tumours • Rhabdomyoma • Lipoma • Fibroma

- **Non-neoplastic (Reactive) Vocal Fold Tumors:**
- Lamina propria of vocal fold is a key component to vocal fold vibration.
- Voice quality depends on the elastic properties of the lamina propria.
- Non-neoplastic tumors results as a reaction from laryngeal irritation such as vocal trauma, surgery, smoking or LPR.
- Maximal vocal fold collision occurs in the striking zone at the mid-membranous vocal fold (junction of anterior 1/3 and posterior 2/3 of the vocal fold).
- **Types:**
 - Vocal fold nodules
 - Vocal fold polyps
 - Vocal fold cysts
 - Reinke's edema
 - Sulcus vocalis
 - Vocal fold scar
 - Vocal fold granuloma
 - Capillary ectasias

- **Vocal fold nodules (Singer's nodules):**
 - More common in young women, children and patients with voice abuse.
 - Professional singers, teachers, other occupations with high voice demands.
 - Bilateral chronic symmetric inflammatory tissue at junction of anterior and middle third of vocal fold (striking zone).
 - Always bilateral and chronic by definition.
 - Pathophysiology:
 - Forceful or prolonged vibration causes localized vascular congestion with edema at midportion membranous (vibratory) portion of vocal folds.
 - Long-term vocal abuse leads to hyalinization of superficial lamina propria and thickening of basement membrane zone (unlike polyp).
 - Stroboscope:
 - Normal or minimal reduction of mucosal wave.
 - Hour glass appearance.
 - Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - **Voice therapy:**
 - Main treatment.
 - Reduce in size and disappearing of the nodules in response to behavioral modification of voice use (75%).
 - **Surgical therapy:**
 - Indicated for large or persistent nodules after minimum of 3 months of voice therapy.
 - Followed by post-op voice therapy.

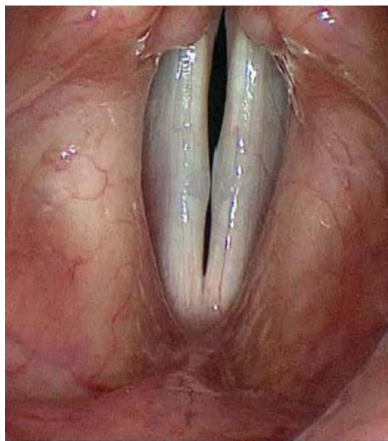


Figure 68.1 Vocal fold nodules.

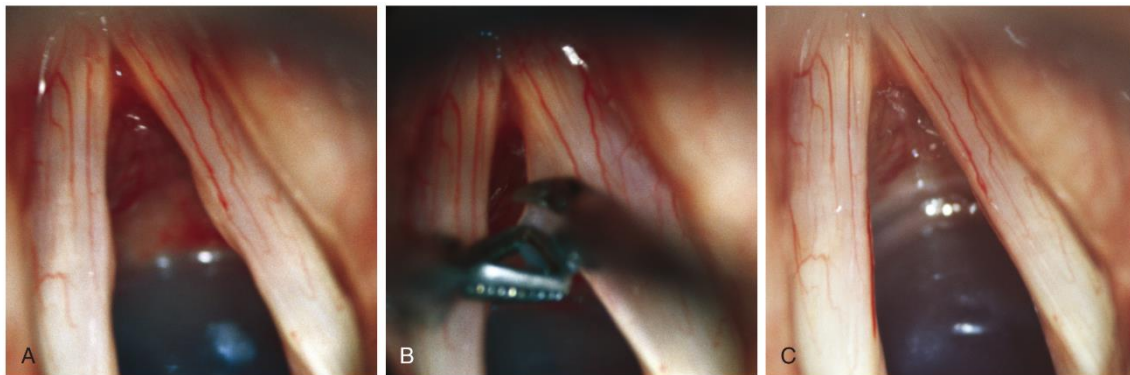
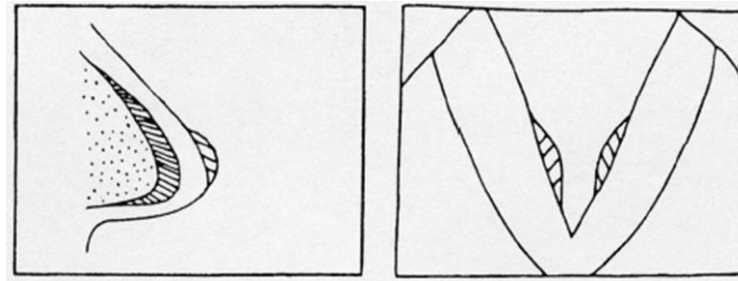


FIGURE 61-8. The operative sequence in a professional musical theater actress who had been experiencing vocal symptoms and limitations compatible with fusiform vocal nodules for more than 2 years. **A**, The operative view after many months of conservative management. Not all fusiform swellings are reversible with conservative measures alone. **B**, A polypoid nodule is grasped superficially and tented medially with Bouchayer forceps. Scissors that curve away from the vocal fold are used for removal. The nodule is thus removed in a very superficial plane, which minimizes the risk of scar between the remaining and regenerated mucosa and the underlying vocal ligament. **C**, Vocal fold appearance after excision. The patient experienced dramatic normalization of vocal capabilities, and no evidence of scarring was found on postoperative stroboscopic examination. The dilated capillaries may predispose to recurrent nodule formation and can be spot coagulated with a microspot laser.

- **Vocal fold polyp:**

- Typically unilateral, but bilateral polyps can occur (10%).
- Located at junction of anterior and middle third of vocal fold (striking zone) in patients with voice abuse.
- Classification:
 - 1. Pedunculated**
 - 2. Sessile (Broad-base)**
- Types:
 - 1. Hemorrhagic (submucosal bleeding):**
 - Might be associated with anticoagulant use.
 - Results from capillary rupture and focal accumulation of blood after extreme vocal effort
 - No thickening of basement membrane.
 - 2. Hyalinized**
 - 3. Gelatinous**
 - 4. Fibrous**
- Stroboscope:
 - Normal or minimal reduction of mucosal wave.

○ Treatment:

▪ **Medical therapy:**

- Anti-reflux medications.
- Stop smoking.

▪ **Voice therapy:**

- Typically no improvement with voice therapy.
- Small early hemorrhagic polyp may resorb after many months of conservative measures.

▪ **Surgical therapy:**

- **Main treatment.**
- Surgical excision is required to return the vocal fold to its normal appearance vibratory function.
- Followed by post-op voice therapy.

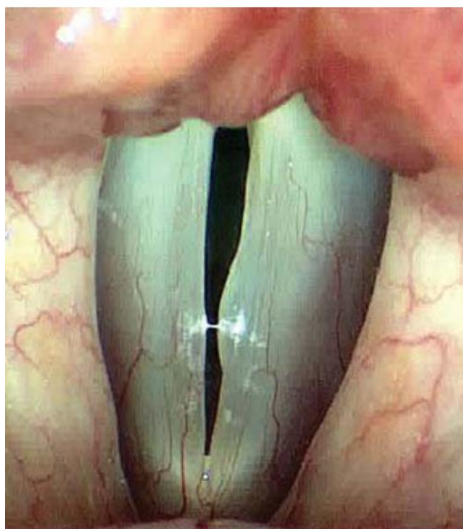
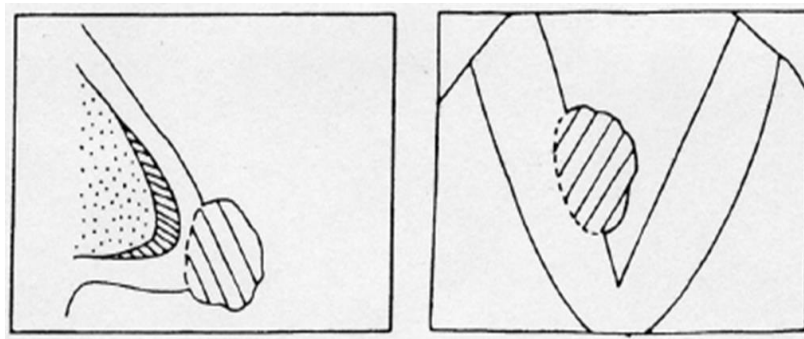


Figure 68.2 Vocal fold polyp.

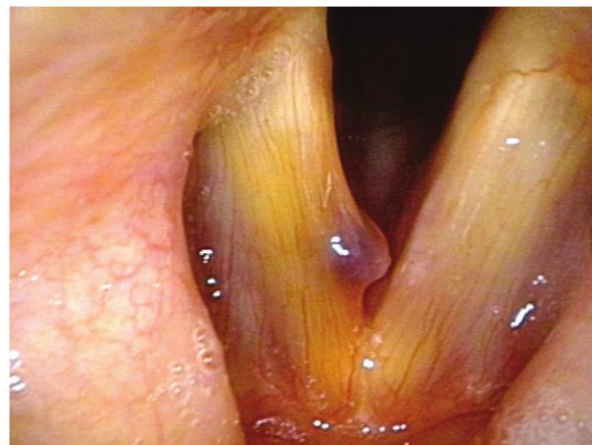


FIGURE 61-11. Hemorrhagic polyp, right fold. Note the blood-blister appearance. Recent further bleeding is evident from the yellowish discoloration of the upper surface of the fold because of breakdown products of a bruise, estimated to have occurred 2 weeks earlier. Hemorrhagic polyps sometimes rebruise intermittently.

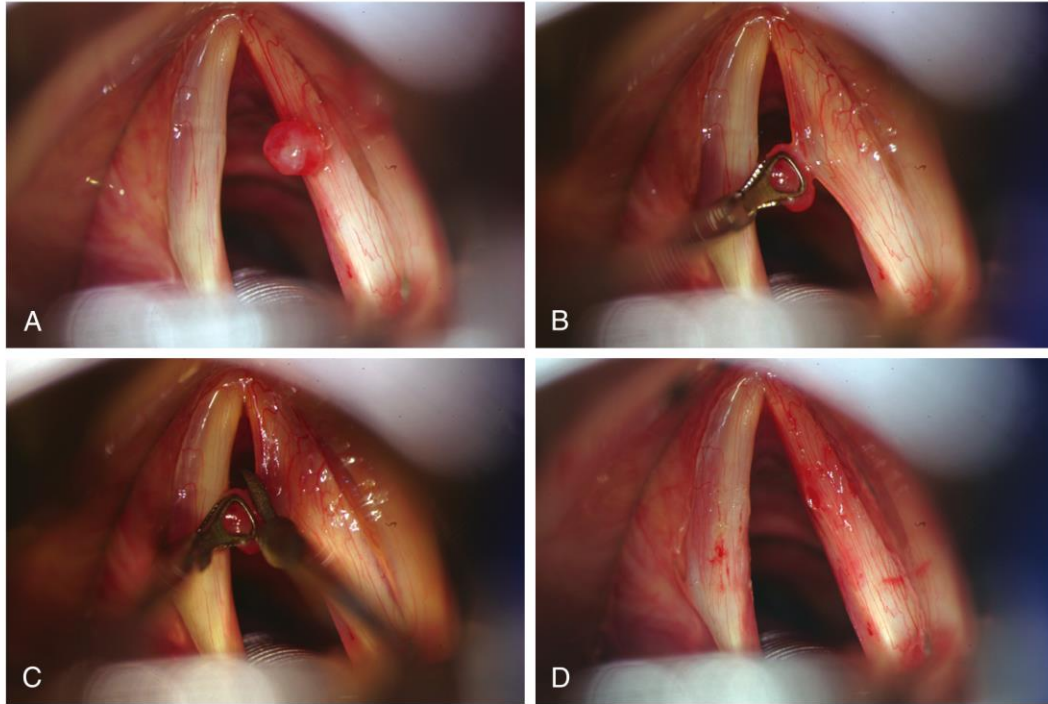


FIGURE 61-12. **A**, Hemorrhagic polyp, right vocal fold. **B**, Polyp is grasped with right-turning heart-shaped forceps to reveal pedunculation and flexibility of the mucosa. **C**, At the moment of excision with a left-turning scissors. **D**, Tiny residual wound. This patient's voice was entirely normalized, including the upper voice.

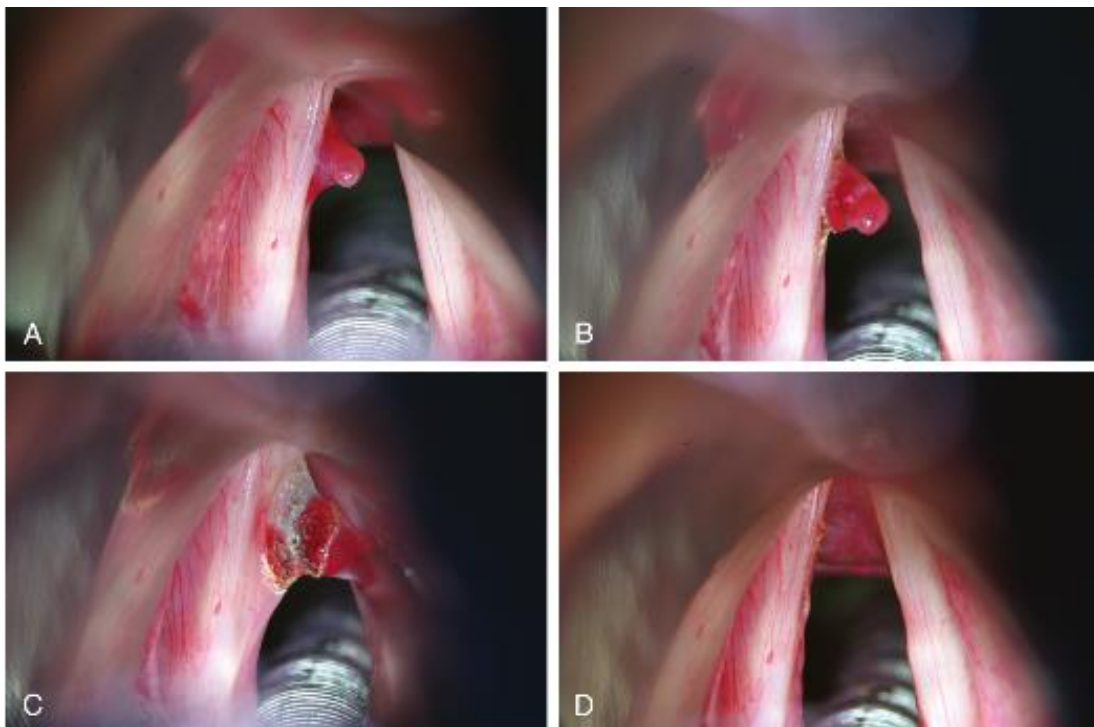


FIGURE 61-13. **A**, Hemorrhagic polyp, left fold, with broad attachment and "shoulders" rather than a stalklike attachment. **B**, Beginning of excision, starting with broad-based anterior and posterior shoulder elements. **C**, Laser dissection directed to the thrombosed contents of the polyp and sparing much of the stretched overlying mucosa. **D**, Resultant linear wound after removal. Because of the remaining layers of Reinke's space (superficial lamina propria), adherence to vocal ligament does not occur, and vibratory ability is normalized, including at high pitch.

- **Vocal fold cyst:**

- Typically unilateral subepithelial cyst.
- Located at junction of anterior and middle third of vocal fold (striking zone).
- Types:
 - 1. Mucus retention cyst:**
 - Arise spontaneously when mucus gland duct becomes obstructed and retains glandular secretions.
 - 2. Epidermal cyst:**
 - Contains accumulated keratin.
 - Results from a nest of epithelial cells buried congenitally in subepithelial layer **or** from healing of mucosa injured by voice abuse over buried epithelial.
 - Cyst may rupture spontaneously.
 - If the resulting opening is small in relation to overall size the cyst, epidermal debris may be retained and create an open cyst.
 - If the opening is as large as the cyst, the resulting empty pocket becomes a glottis sulcus.
- Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - **Voice therapy:**
 - Typically no improvement with voice therapy.
 - Epidermal cysts may improve with voice therapy.
 - **Surgical therapy:**
 - **Main treatment.**
 - Micro-flap excision of the cyst with its capsule.
 - Followed by post-op voice therapy.



Figure 68.4 Vocal fold cyst-subepithelial.

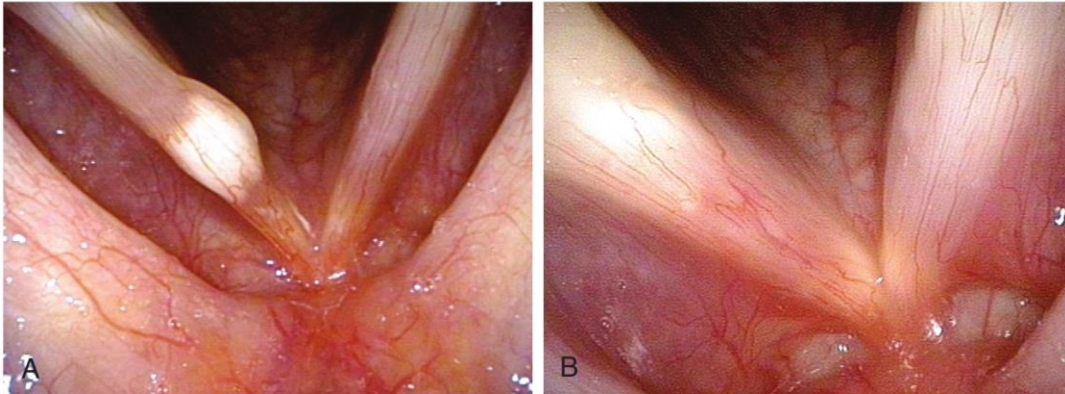
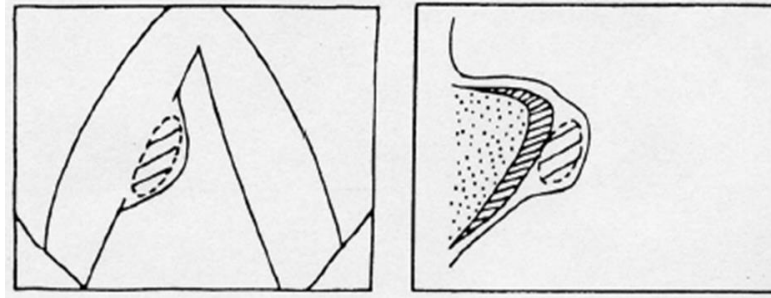


FIGURE 61-14. A, Epidermal cyst, right vocal fold. Note the white submucosal mass predominantly on the upper surface of the fold but with bilateral free margin elevation as well. **B,** After submucosal dissection and removal of cyst. In some similar cases, free margin swelling remains because the margin cannot be straightened (i.e., redundant mucosa that had been stretched over the cyst cannot be removed) at the same time as cyst removal through an upper vocal fold surface incision. In this case, the margin was straight, and vocal fold oscillatory ability markedly improved but was not normal at very high pitches. The voice was highly improved overall.

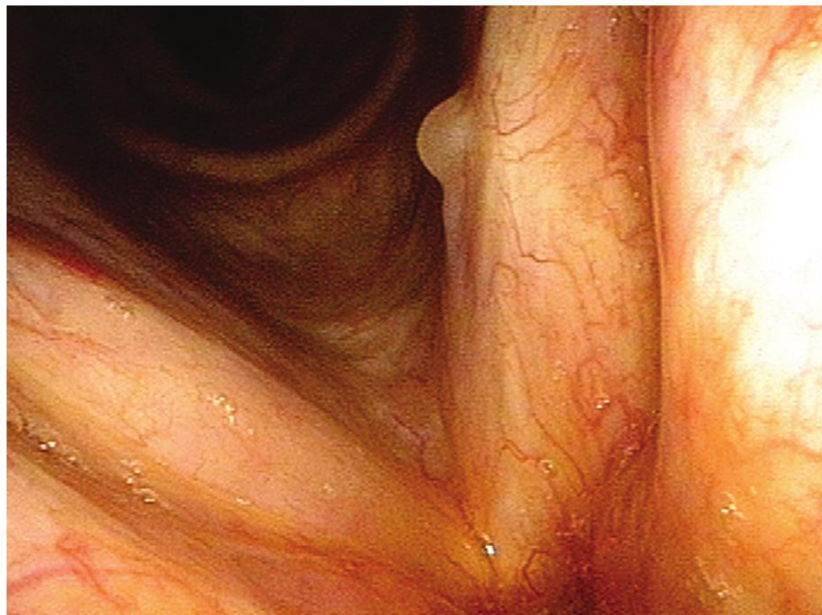


FIGURE 61-15. Mucus retention cyst after laser excision of early vocal fold cancer, left vocal fold. Note capillary reorientation, which is typical after full-thickness mucosal excision. The small projecting lesion could be mistaken as a polyp. Instead, it is the result of plugging of a tiny mucus gland just below the free margin of the vocal folds during mucosal regeneration. A polyp is not consistent with this man's very quiet nature and minimal vocal commitments. Note that the lesion is below the point of maximum contact and vibratory injury that would produce a polyp. This man's voice is excellent.

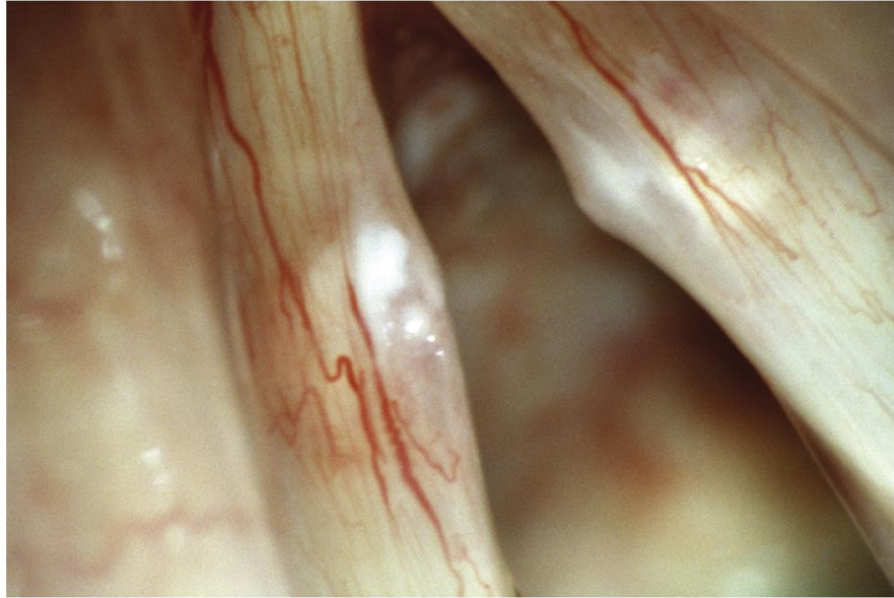


FIGURE 61-16. Bilateral open cysts. Because the openings are small in relation to the size of the cysts, partial emptying of the keratin contents causes a mottled appearance.



FIGURE 61-17. **A**, Mucus retention cyst of right vocal fold. Yellowish spherical mass shines through overlying mucosa and was causing the patient severe hoarseness. Incision to enter the fold is made on the dotted line. **B**, Near completion of dissection of the cyst from its final attachments using curved scissors. **C**, After cyst removal. The patient's voice sounded virtually normal in the recovery room, although upper voice was still abnormal.

- **Reinke's edema (Bilateral Diffuse Polyposis/Smoker's Polyps):**
 - Collection of gelatinous inflammatory exudate within subepithelial space of Reinke (SLP).
 - Results in diffuse polypoid changes of the vocal folds that become permanent.
 - Typically occurs bilaterally.
 - Develops in only a small percentage of high risk patients:
 - Smokers
 - Voice abusers
 - LPR
 - Not all patients must have all three etiologic factors present, but typically patients will have two of these three when Reinke edema is present.
 - Clinical Presentations:
 - Lowered voice pitch and harshness.
 - Woman with Rinke's edema may complain of being called "sir" on the phone.
 - Stroboscope:
 - Diffuse symmetrical pale swellings of the superior surface and the margins of the vocal folds.
 - Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - Elimination of the causative factors for Reinke edema has a significant impact upon long term prognosis after surgery.
 - **Voice therapy:**
 - Short-term voice therapy is appropriate to introduce optimal vocal behavior and reduce the polyps' turgidity with modest improvement in vocal functioning.
 - **Surgical therapy:**
 - **Main treatment.**
 - Vocal fold stripping and removal of gelatinous material in Reinke's space with preserving enough mucosa for epithelialization.
 - Only one vocal fold is operated at a time
 - Followed by post-op voice therapy.
 - Success rate of surgical treatment of Reinke edema is usually favorable when redundant mucosa is carefully excised.
 - Recurrence or lack of voice improvement can be problematic in some patients.

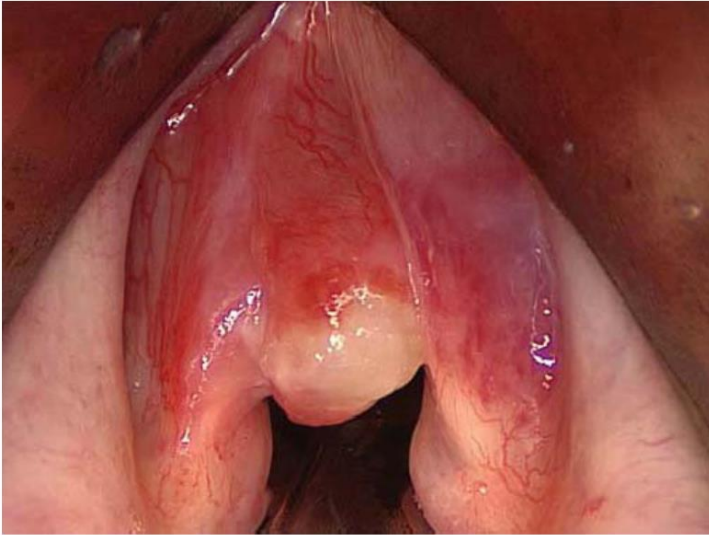


Figure 68.18 Reinke edema of the bilateral true vocal folds, left greater than right.

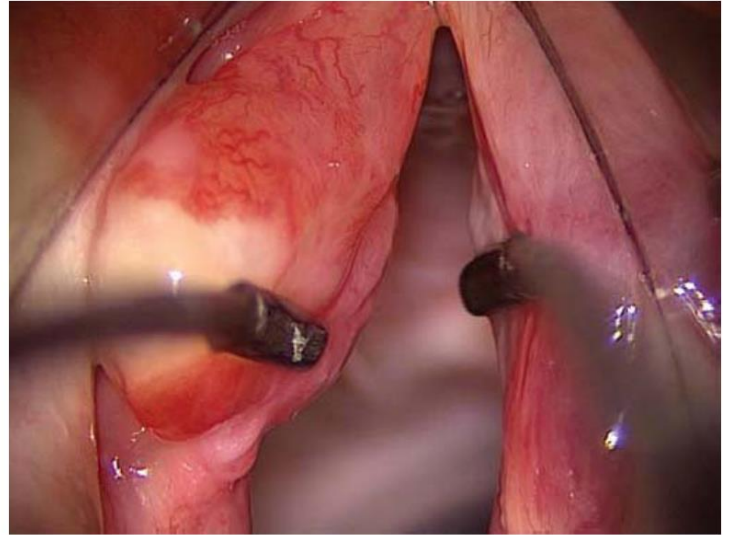
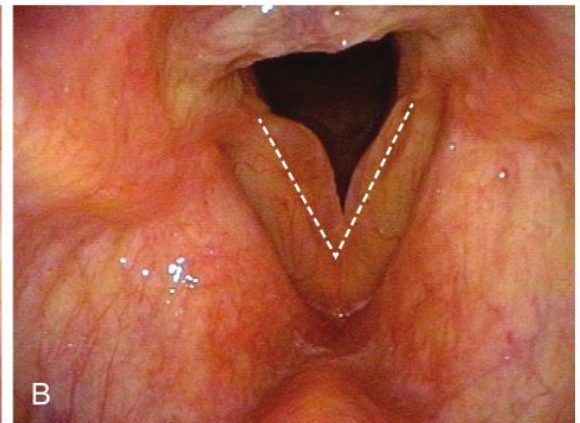


Figure 68.19 Displacement of left true vocal fold Reinke edema from Figure 68.18.

FIGURE 61-22. Bilateral diffuse polypoidosis. **A**, Quiet breathing under standard light. **B**, Elicited inspiratory phonation (same patient) draws in and thereby reveals the edematous mucosa, which is greater on the right than on the left. *Dashed lines* indicate location and contour of free margin, had these vocal folds been normal.



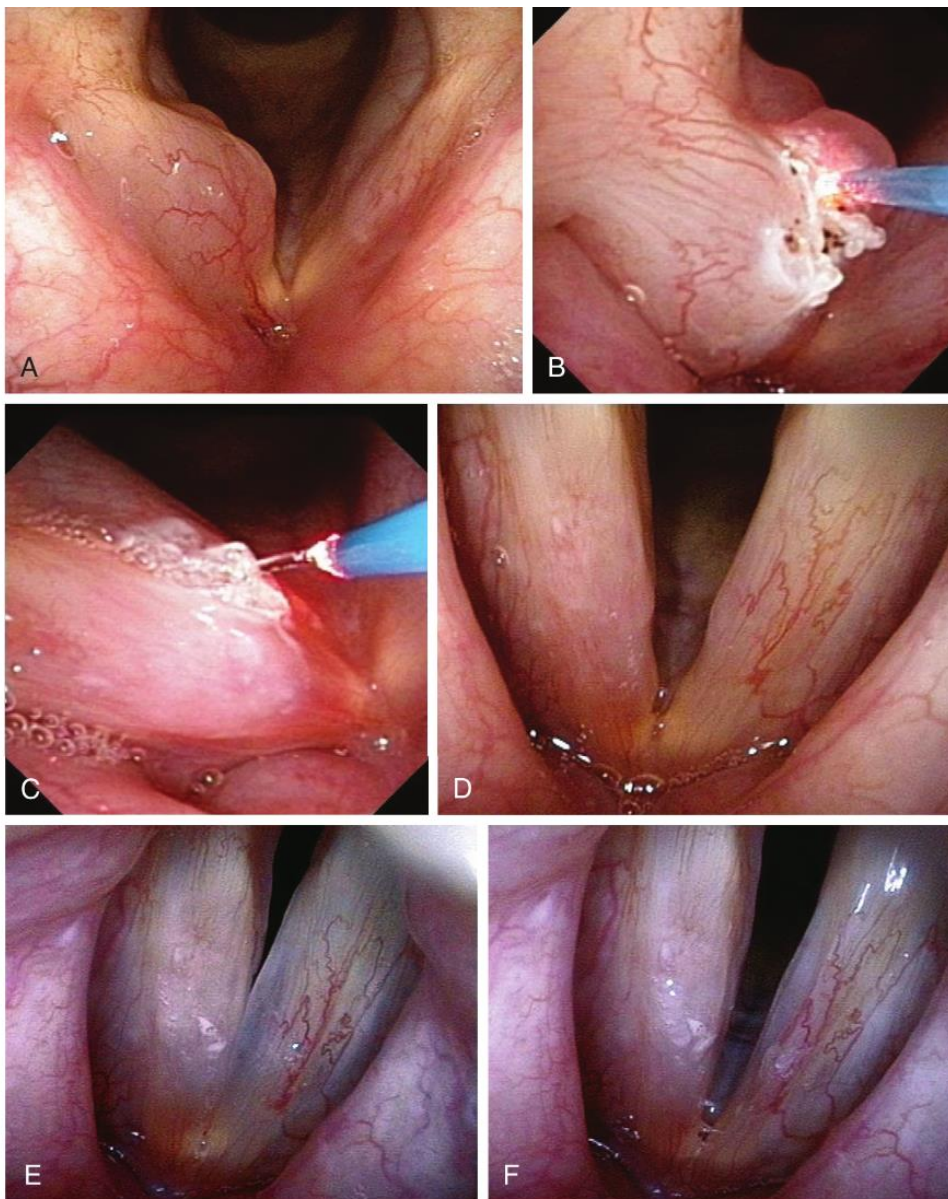


FIGURE 61-23. Smoker's polyp operative sequence. **A**, Voice laboratory view of smoker's polyps; right fold is much more pronounced than the left. **B**, During first thulium laser coagulation. Note attempt to pull the redundant tissue medially from the underlying vocal ligament. **C**, Six weeks postsurgery, during the second thulium laser treatment. **D**, Twelve weeks postsurgery. Early postlaser inflammatory reaction is still evident, but the voice is dramatically improved. **E**, During strobe light and chest phonation, closed phase. **F**, Open phase as the patient finishes phonation and begins to separate the folds. Note slight edema in Reinke's space (translucence) of the left (unoperated) fold. Right fold oscillated well at low frequency and less well at high frequency in this early postoperative examination.

- **Sulcus Vocalis (Glottis Sulcus):**

- Epithelium-lined depression parallel to free edge of the membranous vocal folds (not extending beyond vocal process).
- Leads to stiffening of the mucosa and inhibiting the oscillation which leads to dysphonia.
- More common in vocal overusers.
- Suspected when the voice is worse than the laryngoscopic exam.
- Types:

1. Congenital:

- Abnormal cry and hoarseness since birth.
- Typically have a very severe form of sulcus vocalis.

2. Acquired:

- Resulted from epidermal cyst that has spontaneously emptied, leaving the collapsed depression behind to form a sulcus.
- Mucosal bridge is the result of two parallel sulci that arise from a single cyst and diagnosed by intra-op palpation and pulling medially.

○ Stroboscope:

- Groove along the free edge of membranous vocal fold.
- Incomplete glottic closure.
- Adynamic segment.
- Should be differentiated from pseudosulcus, in which the sulcus is extending past the vocal process.
- Microlaryngoscopy is often required for definitive diagnosis, because the lips of the sulcus are not always visible with inspiratory phonation.

○ Ford Classification:

▪ **Type I:**

- Depression extends into superficial lamina propria only, not reaching into vocal ligament.
- Physiologic type.

▪ **Type II:**

- Depression extends into vocal ligament (involves superficial and intermediate lamina propria).
- Moderate dysphonia

▪ **Type III:**

- Depression extends into vocalis muscle.
- Deep pit like appearance (True sulcus).
- Marked voice impairment.



Figure 68.13 Sulcus vocalis, bilateral.



Figure 68.14 Sulcus vocalis, bilateral. (After right deep true vocal fold injection augmentation with Radiesse Voice Gel.)

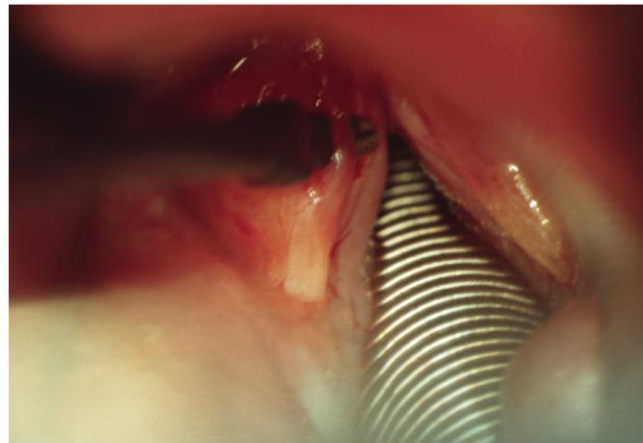
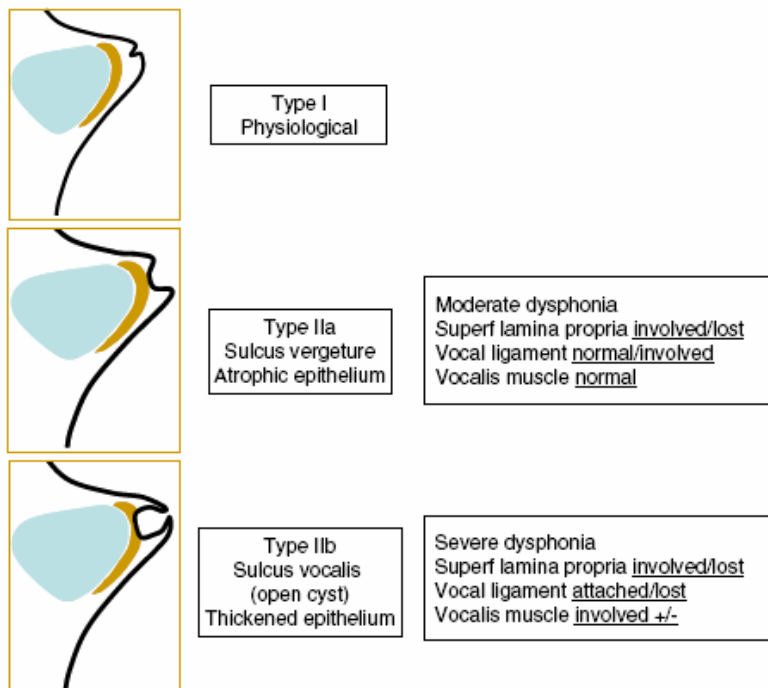


FIGURE 61-20. Left vocal fold mucosal bridge. If an epidermal cyst opens in two places and parallels the margin of the vocal fold, the mucosa between the openings becomes a bridge. In this case, the forceps enters the upper (lateral) opening and exits the lower (medial) opening.



- Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - **Voice therapy:**
 - Short-term voice therapy is appropriate preoperative to introduce optimal vocal behavior.
 - **Surgical therapy:**
 - **Main treatment.**
 - Approaches:
 - 1. Vocal fold Augmentation:**
 - Indicated in patients with sulcus vocalis and glottal insufficiency.
 - Vocal fold injection or medialization laryngoplasty.
 - 2. Collagen injection into superficial lamina propria:**
 - Indicated in patients with sulcus vocalis without glottal insufficiency.
 - Address the vibratory characteristics of the lamina propria.
 - Superficial vocal fold injection of collagen-based materials is limited by technical difficulty in depositing the material in the area of sulcus vocalis
 - 3. Direct implantation of fat:**
 - Indicated in patients with sulcus vocalis without glottal insufficiency.
 - Address the vibratory characteristics of the lamina propria.
 - Direct implantation of fat into lamina propria via a microflap approach or Gray mini-thyrotomy.
 - Provides excellent vibratory characteristic to the involved vocal fold.
 - Technically demanding and difficult surgery.

4. Mucosal slicing technique of Pontes:

- Indicated in patients with sulcus vocalis without glottal insufficiency.
- Address the vibratory characteristics of the lamina propria.
- Aim to re-organize lines of stress throughout the damaged lamina propria by making multiple parallel mucosal slices of varying lengths.
- Post-op recovery is prolonged with 9-12 months is required for voice improvement.

5. Sulcus excision:

- Indicated in patients with sulcus vocalis without glottal insufficiency.
- Address the vibratory characteristics of the lamina propria.
- Technically demanding and involves considerable disturbance of mucosa.
- Includes cordal injection to make sulcus shallower and to accomplish hydrodissection followed by dissection of the invaginated mucosal pocket from underlying fold ligament followed by excision of the sulcus.
- Post-op results depend on surgical skills and thickness of the mucosa (the thickest the better).

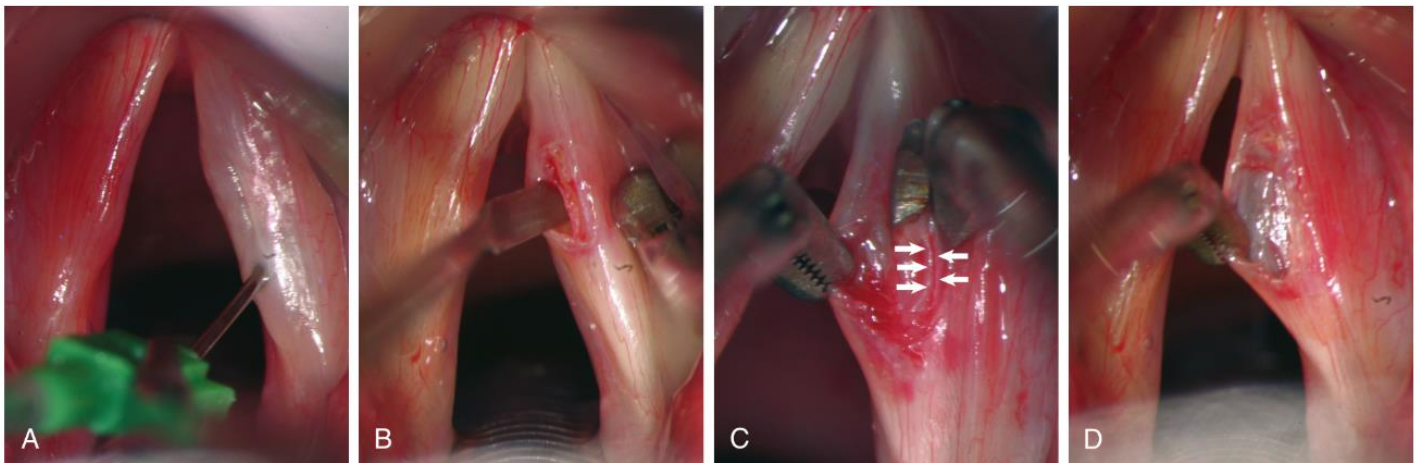


FIGURE 61-19. Glottic sulcus. **A,** At beginning of surgery, the fold is infiltrated with lidocaine/epinephrine to provide hydrodissection and to expand the mucosa. Line of sulcus is seen proceeding anteriorly from the point of needle entry. **B,** An elliptic incision has been made around the lips of the sulcus. **C,** Right-curved alligator clip tents the medial mucosal flap. Arrows indicate the fine line that represents the opening into the sulcus. Curved scissors dissect the anterior aspect of the sulcus pocket from underlying vocal ligament. **D,** After the sulcus pocket is removed, gossamer mucosa is tented medially to show remaining flexibility. The voice is expected to be improved, but normal upper voice capabilities are only sometimes achieved.

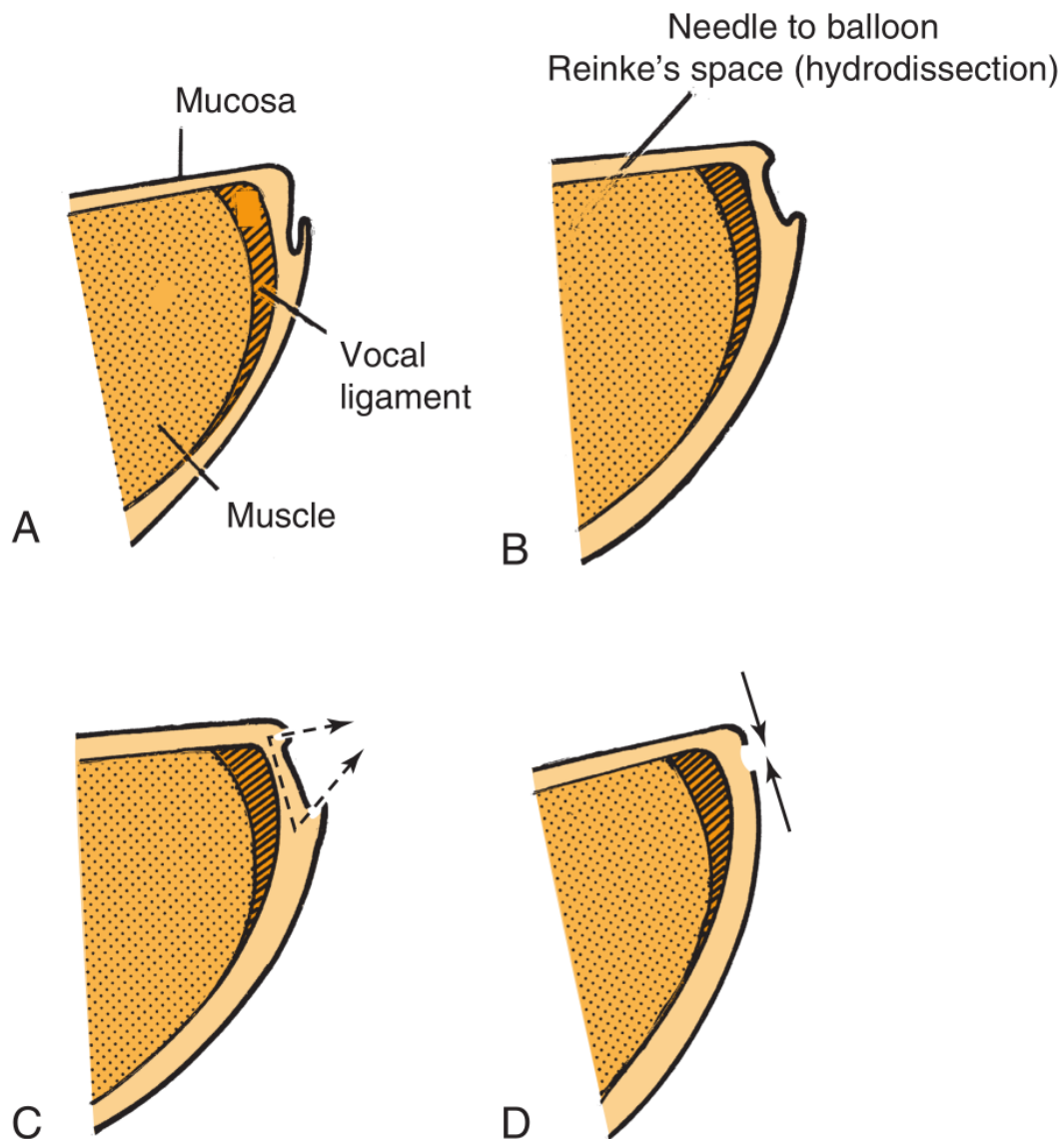


FIGURE 61-21. Schematic of the removal of a glottic sulcus. **A**, Vocal fold coronal section shows the sulcus. **B**, Injection of 1% lidocaine with epinephrine into Reinke's space spreads the lips of the sulcus. **C**, Incisions at the sulcus lips and dissection off the vocal ligament. **D**, After removal of the sulcus.

- **Vocal fold scar:**

- Fibrous scarring of lamina propria that result in poor vocal fold vibratory function.
- Causes:
 - **Voice Abuse:**
 - Repeated hemorrhage into the vocal fold results in serial deposition of fibrous scar tissue, which eventually replaces the lamina propria.
 - **Iatrogenic:**
 - Imprecise vocal fold surgery.
 - Can results in permanent post-op dysphonia and maybe worse than pre-op condition.
- Scarring leads to either expansion of the lamina propria from the fibrous tissue or loss the lamina propria due to adhering of the mucosa to underlying vocal ligament (resembles sulcus vocalis).
- Post-op scarring is avoided by:
 - Precise surgical technique
 - Early post-op graduated resumption of voice use
- Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - **Voice therapy:**
 - Trial of voice therapy is needed before any surgical consideration.
 - **Surgical therapy:**
 - Post-op scarring may diminish slowly over many months and one should wait for 9-12 months before doing second surgery.
 - Treated surgically like sulcus vocalis.

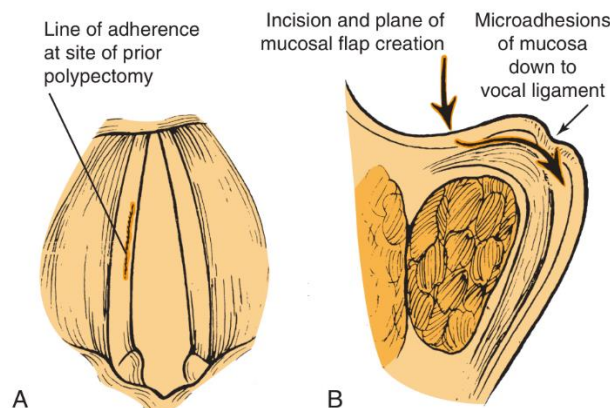


FIGURE 61-25. Schematic of mucosal adhesion to underlying vocal ligament and one surgical option. **A**, The operative view shows a longitudinal scar. **B**, Cross-sectional view shows the surgical approach to release microadhesion of mucosa from the vocal ligament. Such a patient may have only modest effect on vocal capability but a more noticeable relief from aberration such as diplophonia.

- **Vocal fold granuloma:**

- Results from inflammation or injury of the thin mucosa and perichondrium overlying posterior cartilaginous glottis.
- Speaking voice is not always affected because the membranous vocal folds may be unaffected by the granuloma.
- Causes:

- **Trauma:**

- Voice abusers:
 - Chronic forceful apposition of both arytenoids at onset of voicing.
- Chronic cough:
 - Chronic forceful apposition of both arytenoids.
- Intubation:
 - Direct abrasion of arytenoid perichondrium.
 - Injury to the mucosa of the arytenoids from coughing on an endotracheal tube.
 - Long-term pressure necrosis of vocal process area.
- Surgery:
 - Injury to the mucosa of vocal fold.

- **LPR:**

- Increase the inflammation at the vocal process.

- Stroboscope:

- Erythema, ulceration or granuloma at the vocal process.
- Large pedunculated granuloma may flip above and below the vocal folds with respiration.

- Treatment:

- **Medical therapy:**

- Anti-reflux medications.
- Stop smoking.
- Oral antibiotics for recent intubation granuloma
- Steroids:
 - Systemic oral steroids.
 - Inhaled oral steroids (Triamcinolone 150 µg TID for 6-8 weeks).
 - Office-based intra-lesion injection of steroid.
- Resolution of granuloma occurs spontaneously over 3-6 months without any intervention.

- **Voice therapy:**

- Aim to abolish throat clearing, raise average pitch for speech to minimize chronic forceful apposition of both arytenoids.

- **Surgical therapy:**

- Last resort.
- Indicated for persistent symptomatic granuloma.
- Steroid injection is done at granuloma base before its removal.

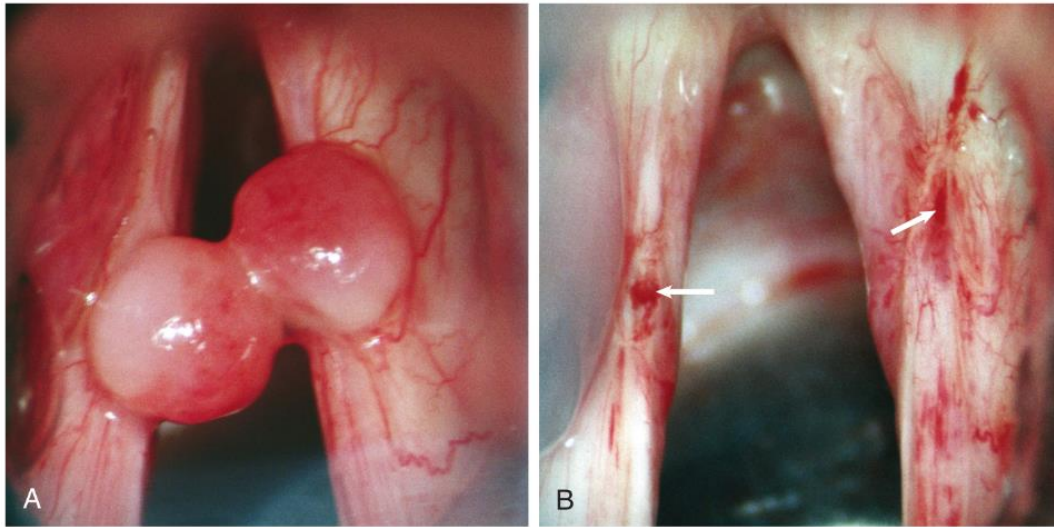


FIGURE 61-24. Iatrogenic mucosal scarring. This patient underwent bilateral vocal fold stripping elsewhere for persistent dysphonia, which was subsequently diagnosed as spasmodic dysphonia. The patient was reportedly aphonic for many weeks postoperatively. **A**, This operative photograph was taken 4.5 months after the original surgery. Granulomas are highly pedunculated and may have eventually detached or regressed spontaneously. Note the medial-to-lateral reorientation of vocal fold capillaries, a common finding after vocal fold stripping. **B**, The same patient after granuloma removal. Attachment points of the granulomas are marked at arrows. In this view, the vocal folds are rolled superiorly and considerable scarring is evident, particularly on the right vocal fold.

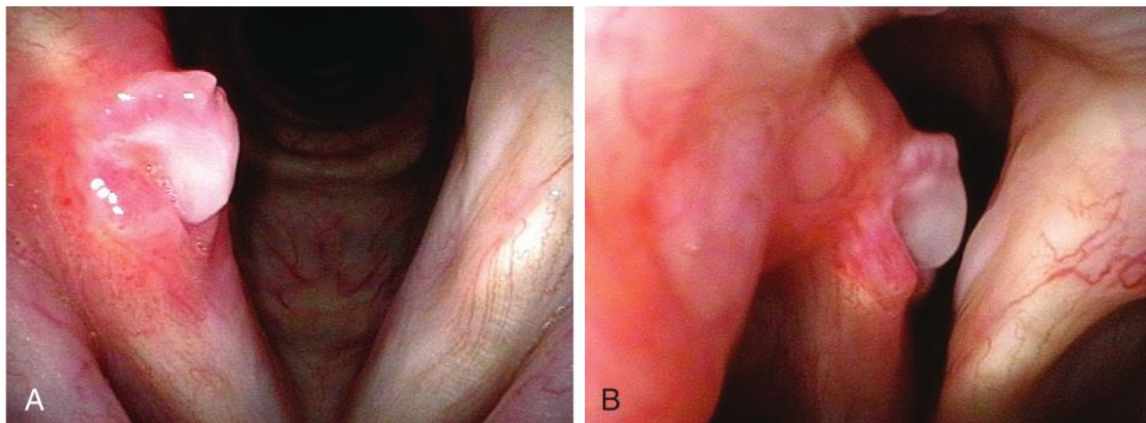


FIGURE 61-26. **A**, Contact granuloma, right posterior vocal fold. Note bilobularity and surrounding inflammation (erythema). **B**, Same patient, as folds are arriving at phonatory contact. The medial surface of the left arytenoid cartilage will fit into the cleft between the two lobes of the contact granuloma.

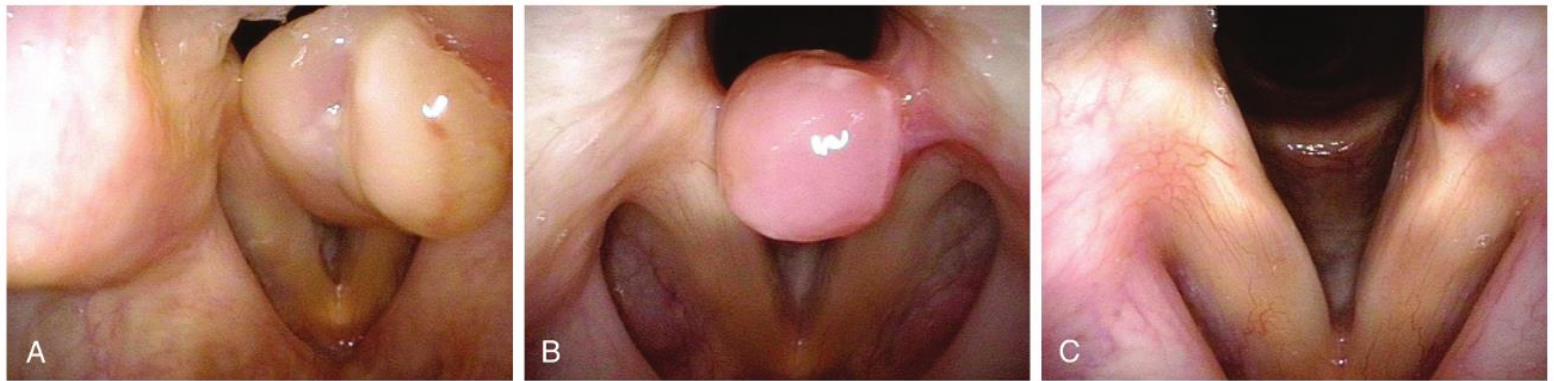


FIGURE 61-27. **A**, Large, bilobed contact granuloma, left fold, with significant pedunculation. Voice is surprisingly unaffected by the pedunculation and the deep cleft between the lobes. **B**, This granuloma was allowed to mature and detach spontaneously. Here, a few months later, the inferior lobule has detached, and a single spherical and highly pedunculated granuloma remains. **C**, Several months later in the same patient, the remaining granuloma has detached, leaving a characteristic bruise at its base. This mark may remain visible for many months.

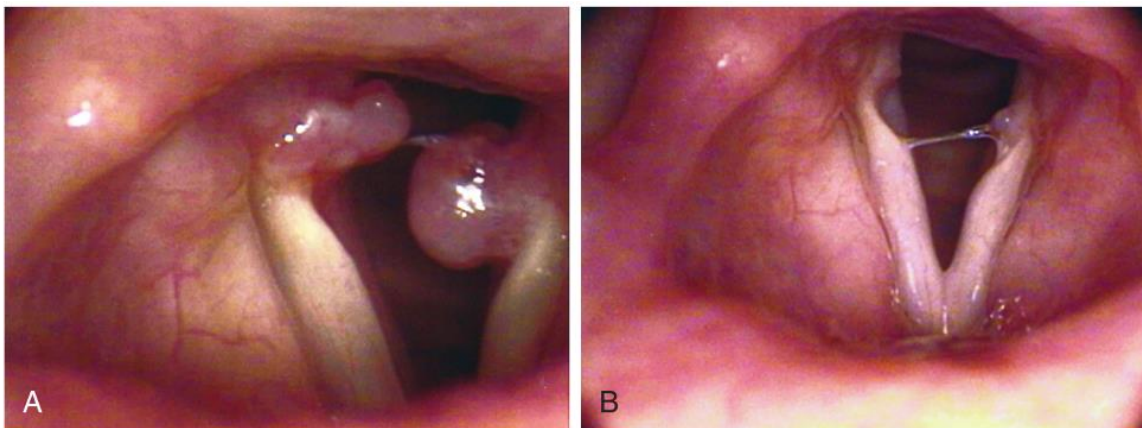


FIGURE 61-28. Granulomas after long-term intubation. **A**, Evidence of posterior commissure divots from pressure necrosis of the endotracheal tube, along with reparative granulomas. **B**, Same patient a few months later. Without any intervention, granulomas have matured, pedunculated, and spontaneously detached, leaving the divots more visible.

- **Capillary (vascular) ectasias:**

- Abnormal dilated subepithelial blood vessels within vocal fold.
- Parallel to longitudinal axis of vocal fold.
- Aberrant clusters of dilated capillaries are called capillary lake.
- Associated with:
 - Vocal fold lesions.
 - Phonotrauma.
- Repeated vibratory micro-trauma lead to capillary angiogenesis and formation of abnormally dilated capillaries which increase mucosa's vulnerability to further vibratory trauma.
- Increase risk of:
 - Mucosal swelling
 - Hemorrhagic polyp
 - Vocal fold hemorrhage.
- Without mucosal swelling, voice is normal.
- Treatment:
 - **Medical therapy:**
 - Anti-reflux medications.
 - Stop smoking.
 - Stop anticoagulant drugs if medically appropriate.
 - Increase the severity of the bruising but not the incidence of hemorrhage.
 - **Voice therapy:**
 - Patients are warned about sudden explosive use of the voice.
 - **Surgical therapy:**
 - Does not require surgical treatment unless:
 - History of documented vocal fold hemorrhage
 - Located along striking edge of the vocal fold
 - Surgical ablation is be done pulsed angiolytic laser ablation (KTP).
 - Chromophore for these lasers is hemoglobin rather than water; as with Co2 laser, resulting in more precise delivery of laser energy to the abnormal blood vessel with less thermal injury risk.



FIGURE 61-9. **A**, The abducted breathing position with standard light. This is called a "capillary lake." **B**, Prephonatory instant with standard light in the same patient shows slight projection from free margin. **C**, The condition resolved after surgical ablation, the voice normalized, and mucosal oscillation was preserved to the highest vocal range.

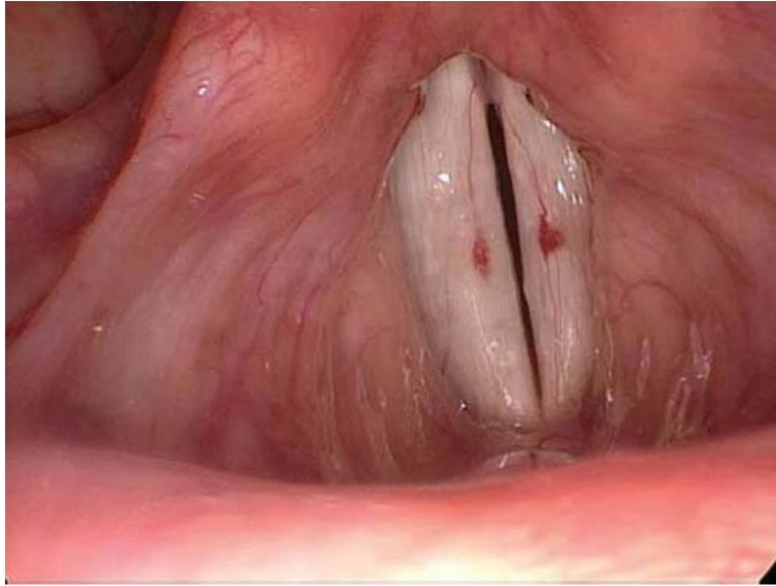


Figure 68.17 Vascular lesions of the bilateral true vocal folds.

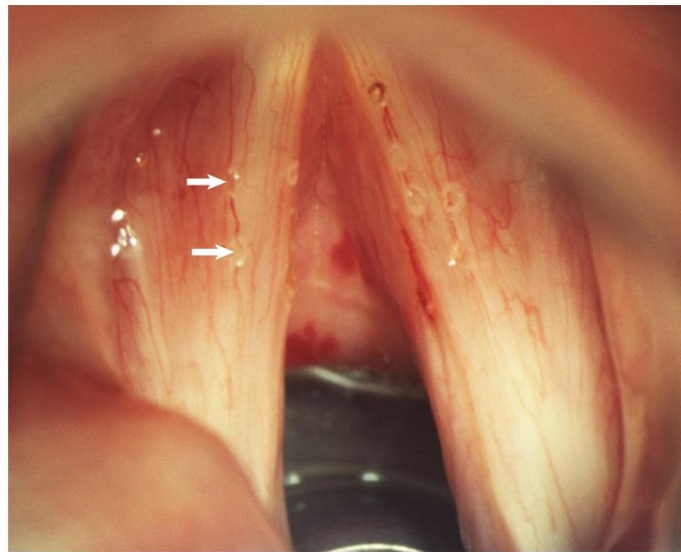


FIGURE 61-10. Ectatic capillaries need not be ablated in their entirety. Instead, flow is stopped with spot coagulations (*arrows*) along the course of the capillary. Within 3 weeks, capillary “segments” disappear.

- **Neoplastic Vocal Fold Tumors:**
- Benign vocal fold neoplasms are rare.
- It should be differentiated from non-neoplastic mucosal reactive disorders in response to chronic vocal injury.
- Includes:
 - Recurrent Respiratory Papillomatosis
 - Laryngeal Hemangiomas
 - Rhabdomyoma
 - Lipoma
 - Pleomorphic adenomas
 - Oncocytic Neoplasms of the Larynx
 - Chondroma
 - Granular Cell Neoplasms
 - Neurofibroma
 - Neurilemmoma

Non-neoplastic	Neoplastic
● Solid ○ Vocal nodules ○ Vocal polyp ○ Reinke's oedema ○ Contact ulcer ○ Intubation granuloma ○ Leukoplakia ○ Amyloid tumours ● Cystic ○ Ductal cysts ○ Saccular cysts ○ Laryngocele	● Squamous papilloma ○ Juvenile type ○ Adult-onset type ● Chondroma ● Haemangioma ● Granular cell tumour ● Glandular tumours ● Rhabdomyoma ● Lipoma ● Fibroma

- **Recurrent Respiratory Papillomatosis (RRP):**
- Most common benign laryngeal neoplasm (80%).
- Second most common cause of childhood hoarseness.
 - o After vocal cord nodules.
- Wart-like, irregular and exophytic lesions of the airway.
- Difficult to treat because of its tendency to recur and spread throughout the aerodigestive tract.
- Location:
 - o **At squamo-ciliary junctions of the upper airway:**
 - Limen vestibule
 - Nasopharyngeal surface of soft palate
 - Laryngeal surface of epiglottis
 - Upper and lower margins of ventricle
 - Undersurface of vocal folds
 - Tracheostomy stoma (iatrogenic squamo-ciliary junction/squamous metaplasia)
 - Carina and bronchial spurs

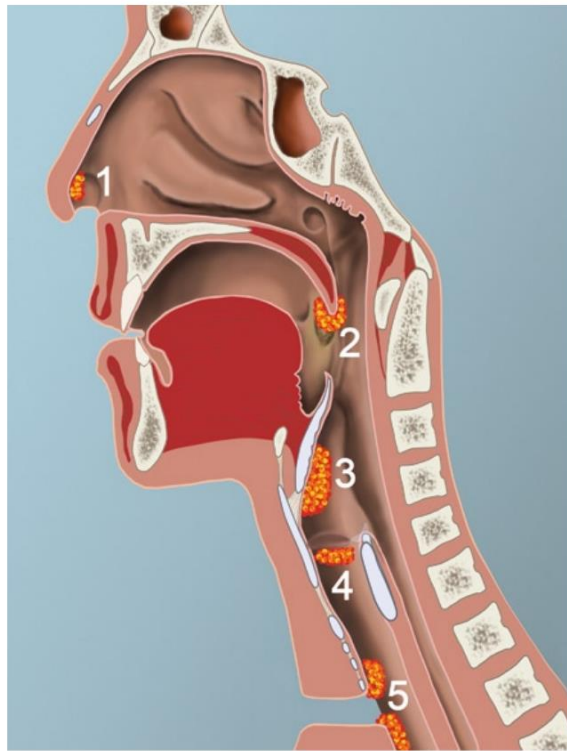


Fig. 16.2 Prevalence of human papillomavirus exophytic growth at the squamo-ciliary junctions of the upper airways: 1. Limen vestibuli nasi. 2. Naso-pharyngeal surface of soft palate. 3. Supraglottis 4. Vocal cords. 5. Areas of squamous metaplasia due to chronic trauma (e.g., tracheostoma, cannula tract in the trachea, carina and bronchial spurs)

- Types:

1. Juvenile onset RRP (< 12 years):

- Most common and severe form.
- Incidence of 4.3 cases of RRP per 100,000 children.
- Most often diagnosed between 2-4 years of age.
- Youngest age diagnosed with JoRRP was 1 day-old.
- Multiple and diffuse involvement.
- Rapidly recurrent.
- Multiple surgical intervention required.
 - Mean number of procedures are 20 per child.
 - Average of 4 procedures per year.
 - Children diagnosed before 3 years of age:
 - Require > 4 surgical procedures per year.
 - Involvement of ≥ 2 anatomical sites.
- Most common HPV subtype (11).

2. Adult onset RRP (>12 years):

- Less common and severe form.
- Incidence of 1.8 cases of RRP per 100,000 adults.
- Most often diagnosed between 20-40 years of age.
- Oldest patient diagnosed with AoRRP was 84 year-old.
- Single and localized involvement.
- Slowly recurrent.
- Less surgical intervention required.
 - 50% of patients with AoRRP will require < 5 surgical procedures in lifetime.
- Most common HPV subtype (6).

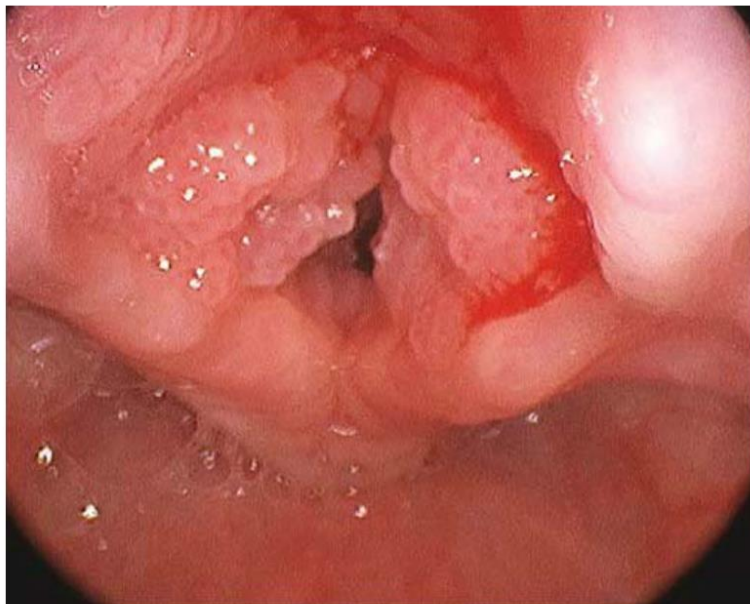


Figure 94.2 Papilloma lesions involving the laryngeal inlet.

- Etiology:

○ **Infection with Human papilloma virus (HPV):**

- Double stranded DNA virus.
- HPV infects stem cells within basal layer of the mucosa then it can either be actively expressed, or exist as a latent infection in the epithelium.
- HPV DNA can be detected in normal-appearing mucosa in RRP patients who have been in remission for years.
- Reactivation of viral expression can occur any time following establishment of a latent infection.
- Subtypes (by PCR):
 - 100 different subtypes have been identified.
 - Same subtypes responsible for genital warts.
 - Most common subtypes (6 & 11).
 - Most aggressive subtype (11).
 - Highest risk of malignant transformation (16 & 18).
 - Intermediate risk of malignant transformation (31 & 33).

- Histopathology:

- **Fingerlike projections** of non-keratinized stratified squamous epithelium supported by highly vascularized connective tissue stroma.

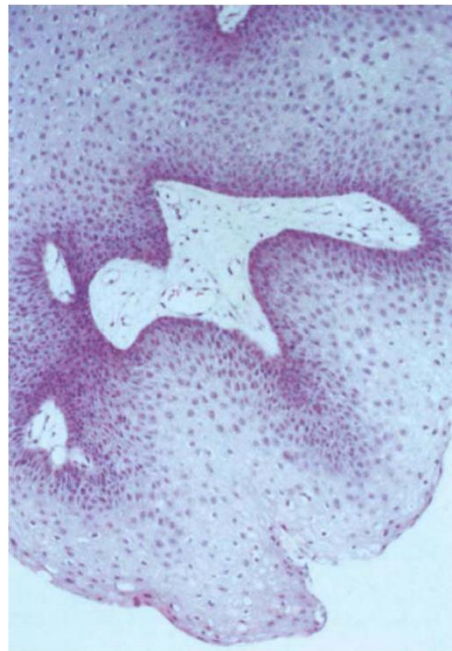


Figure 94.1 Histologic section of papilloma, demonstrating fingerlike projection of nonkeratinized stratified squamous epithelium and vascularized connective tissue stroma.

- Mode of transmission (Unclear):
 - **Vertical transmission:**
 - Major mode of transmitting the infection in children.
 - **Risk factors of acquiring JoRRP:**
 1. Mothers with active condylomata (50%)
 2. Mother age < 20 years
 3. Newly acquired genital HPV lesions.
 4. Vaginal delivery.
 5. First born (prolonged delivery and viral exposure).
 6. Low socioeconomic status.
 - However, few children exposed to genital warts at birth actually develop RRP.
 - Estimated risk of transmission = 1 in 400
 - Incidence with c-section is less but still can happen, suggesting blood transmission through umbilical cord.
 - Caesarean section was not found to be protective against JoRRP.
 - **Secondary factors playing role in vertical transmission:**
 1. Patient immunity (HLA , reduction CD4/CD8 ratio)
 2. Timing, length, and volume of virus exposure
 3. Local traumas (intubation, extra-esophageal reflux)
 - **Sexual transmission:**
 - Some data suggested that patients with AoRRP are exposed to HPV later in life than JoRRP.
 - **Risk factors of acquiring AoRRP:**
 1. Multiple sexual partners
 2. Higher frequency of oral sex
 - However, HPV has the capability to form latent infections in the basal cell layer of otherwise healthy mucosa and it has been suggested that AoRRP may represent a reactivation of HPV infection acquired during birth instead of a de novo exposure during adulthood.

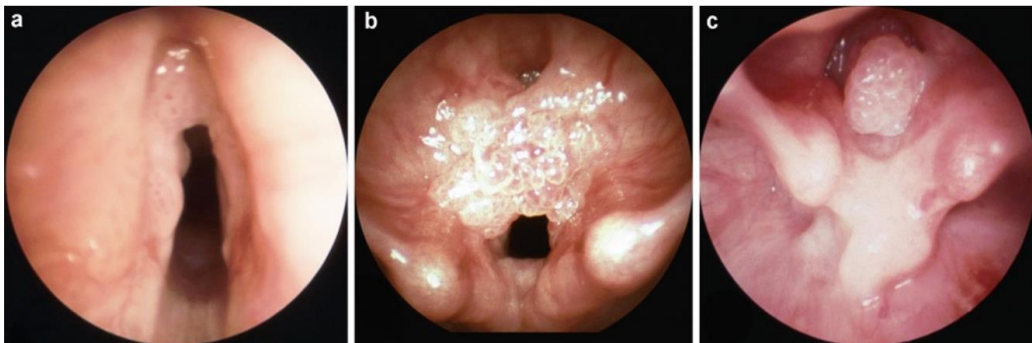


Fig. 16.3 Macroscopic features of juvenile-onset recurrent respiratory papillomatosis: (a) Sessile implantation of papillomas on the left vocal cord. Prior to suction of the secretions, the papillary nature of the lesion is not conspicuous. (b) Irregular exophytic clusters of papillomas with multiple implantation sites. (c) Pedunculated papillomas of the right supraglottis and retrocricoid region

- Clinical picture:
 - Typical delay of one year between symptoms onset and the diagnosis of RRP.
 - 75% of patients diagnosed before 5 years of age.
 - Misdiagnosed initially as having asthma, croup, or chronic bronchitis.
 - Triad:
 1. Progressive hoarseness (initially)
 2. Inspiratory progressing to biphasic stridor
 3. Respiratory distress
 - Associated symptoms:
 - Chronic cough
 - Recurrent pneumonia
 - Feeding difficulties
 - Allergic symptoms
 - Vocal abuse
 - Presence of hereditary congenital anomalies
 - Malignant transformation into SCC **(2.3%)**
 - Death secondary to:
 - Complication of frequent surgical procedures
 - Respiratory failure from distal disease progression.
- Diagnosis:
 - **Flexible scope:**
 - Appearance:
 - Glistening white irregular growths
 - Pedunculated or sessile
 - Friable and bleeding easily
 - **Carpet variant:**
 - Seen in AoRRP.
 - Does not show the typical exophytic growth.
 - Showing velvety appearance of red dots projection from the surface which represents the fibrovascular core of each papilloma.
 - Location:
 - Anterior part of true vocal folds, false folds, or epiglottis.
 - Trauma (iatrogenic or by acid reflux) to area adjacent to areas of frank papilloma can lead to implantation of papilloma.
 - Pattern:
 - Diffuse involvement in JoRRP
 - Limited and localized in AoRRP
 - **CXR for Pulmonary involvement.**
 - **MLS and Biopsy for Histopathology.**

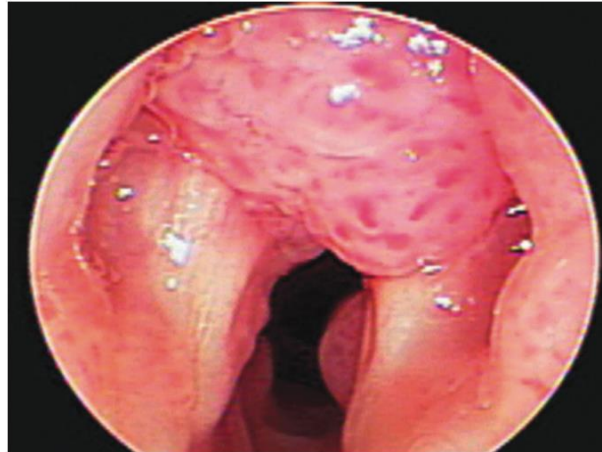


FIGURE 204-2. Gross appearance of respiratory papillomas during laryngoscopy. Exophytic fronds are visible above and below the levels of true vocal cords, with a darker fibrovascular core in each papilloma.

- Clinical course of JoRRP:
 - **High aggressive JoRRP (20%):**
 - Onset before the age 3 years.
 - HPV-11 subtype.
 - Risk of developing distal airway disease
 - 8% in the trachea
 - 3% in the bronchi
 - 3% in the lungs
 - Requires > 40 lifetime procedures.
 - Requires tracheostomy (15% of all JoRRP cases)
 - Likelihood for mortality.
 - **Intermediate aggressive JoRRP (50%):**
 - Most common course.
 - Required > 5 lifetime procedures.
 - Disease aggressiveness is easily controlled.
 - **Low aggressive JoRRP (25%):**
 - Required < 5 lifetime procedures.
 - Disease aggressiveness is easily controlled.
 - Some patients experience spontaneous remission.

- Scoring system for disease severity in RRP:
 - o **Coltrera-Derkay staging system**

COLTRERA-DERKAY STAGING AND SEVERITY SCHEME³⁵

A. Clinical Score

1. Describe the patient's voice today:
normal___(0), abnormal___(1), aphonic___(2)
2. Describe the patient's stridor today:
absent___(0), present with activity___(1), present at rest___(2)
3. Describe the urgency of today's intervention:
scheduled___(0), elective___(1), urgent___(2), emergent___(3)
4. Describe today's level of respiratory distress:
none___(0), mild___(1), moderate___(2), severe___(3), extreme___(4)

Total Clinical Score (Questions 1 through 4) = ___

B. Anatomical Score

For each site, score as: 0 = none, 1 = surface lesion, 2 = raised lesion, 3 = bulky lesion

LARYNX:

Epiglottis:	Lingual surface___	Laryngeal surface___
Aryepiglottic folds:	Right___	Left___
False vocal cords:	Right___	Left___
True vocal cords:	Right___	Left___
Arytenoids:	Right___	Left___
Anterior commissure_____		
Posterior commissure_____		
Subglottis_____		

TRACHEA:

Upper one-third_____	
Middle one-third_____	
Lower one-third_____	
Bronchi:	Right___ Left___
Tracheotomy stoma_____	

OTHER:

Nose_____	
Palate_____	
Pharynx_____	
Esophagus_____	
Lungs_____	
Other_____	Total Anatomical Score _____

C. Total Score = Total Anatomical Score plus Total Clinical Score

- Extra-laryngeal spread of RRP:
 - Occurs in 30% of patients with JoRRP.
 - Occurs in 15% of patients with AoRRP.
 - Most frequent sites of spread:
 1. Oral cavity
 2. Trachea
 3. Bronchi.

- Risk factors for aggressive RRP disease:
 1. Age < 3 years
 2. HPV 11 subtype
 3. Extensive airway with extra-laryngeal spread
 4. Frequent recurrence after treatment

- Risk factors for distal dissemination of RRP:
 1. Age < 3 years
 2. HPV 11 subtype
 3. Prolonged tracheotomy
 4. Presence of subglottic papillomas at time of tracheotomy
 5. Prolonged ET intubation (Bronchopulmonary dysplasia)
 6. Jet ventilation
 7. ?? Reflux disease
 8. ?? Immunity status

- Risk factors for malignant transformation (Carcinogenic factors):
 1. HPV 16 and 18 subtypes
 2. Smoking
 3. Radiation
 4. Cidofovir injection (Not proven yet)

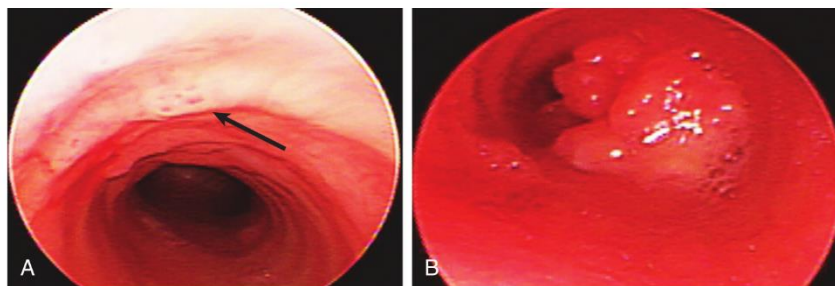


FIGURE 204-5. Tracheal spread of recurrent respiratory papillomatosis. Note nodular, exophytic lesions (*arrow*) in the distal trachea (**A**) and massive, exophytic, obstructing papillomas in another patient (**B**).

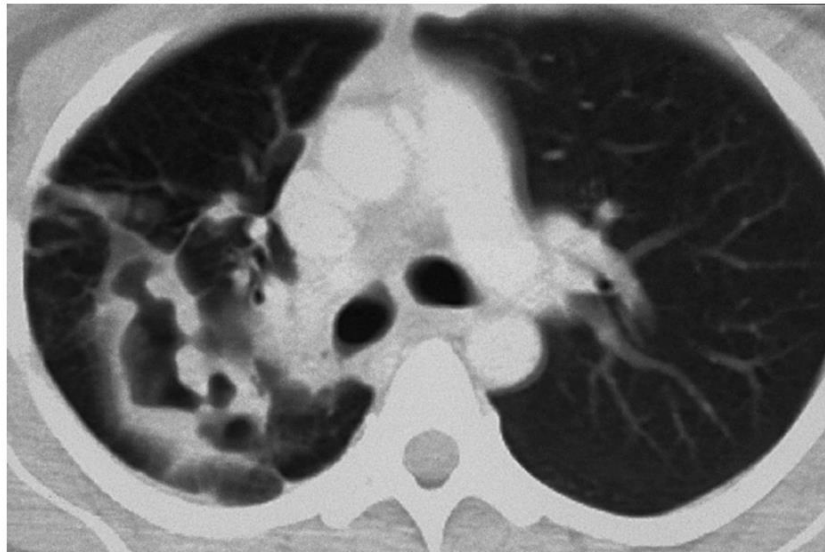


FIGURE 204-6. Pulmonary spread of recurrent respiratory papillomatosis. Note lytic, cavitary lesions on computed tomography.

- Prevention of RRP:

1. Quadrivalent HPV vaccine (Gardasil):

- Vaccine against HPV subtypes (6,11,16,18).
- Most effective if administered to individuals who have not yet become sexually active.
- CDC's Advisory Committee on Immunization Practices recommended to administer it to all boys and girls 11-12 years of age.
- Used mainly for prevention of:
 - Cervical cancer
 - Adenocarcinoma in situ
 - Vulvar and vaginal intraepithelial neoplasias
 - Genital warts
- It also holds promise to eliminate maternal and paternal reservoir of HPV and lead to a near eradication of RRP caused by HPVs 6 and 11.
- Long-term results of widespread vaccination have not been established yet.

2. Bivalent HPV vaccine:

- Vaccine against HPV subtypes (16,18).
- Effective in preventing incident and persistent cervical HPVs 16 and 18 infections.
- It may reduce incidence of HPV-associated head and neck cancers.
- However, it does affect the vertical transmission of HPVs 6 or 11 from mother to child.

- Treatment:
- During viral latency, HPV DNA can be detected in normal-appearing mucosa in RRP patients who have been in remission for years.
- Unknown stimuli can result in reactivation and clinical recurrence anytime following years of remission.
- To “cure” RRP, it is necessary to modulate host response to the virus and to eliminate latent infection which have not been achieved.
- **To date, there is no current cure for RRP.**
 - **Medical therapy:**
 - Anti-reflux medications.
 - Rarely, papillomata may regress spontaneously, especially at puberty.
 - **Surgical therapy:**
 - Goals:
 - Provide an adequate airway
 - Improve voice quality
 - Facilitate disease remission
 - Total eradication of all papillomas by aggressive surgical treatment should not be attempt as it may lead to cicatricial sequelae such as vocal cord synechia or laryngotracheal stenosis which are more problematic than the primary disease itself.
 - Precise endoscopic assessment using 0°, 30°, 70° telescopes to confirm the papilloma implantation sites prior to the surgical removal of papillomas.

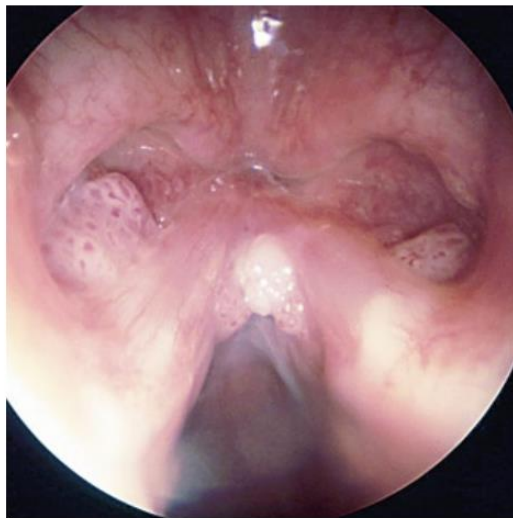


Fig. 16.4 Yield of angulated telescopes to assess the full extent of juvenile-onset recurrent respiratory papillomatosis: With the 70°-angle rod-lens telescope, papillomas are detected in both laryngeal ventricles and the anterior subglottis. None of these lesions were easily visible with the 0° telescope

- Anesthesia techniques:
 - **Limited disease:**
 - ET intubation with smallest possible tube.
 - **Diffuse disease:**
 - Aim to provide a free operative field.
 - Total intravenous anesthesia (TIVA) with:
 - Insufflation technique.
 - Intermittent apneas.
 - Jet ventilation (ventilating catheter is inserted into subglottis or percutaneous trans-tracheal catheter).
 - ET intubation with ETT can be done followed by removal of gross disease then extubation and removal of the residual posterior disease with apneic technique.
- Surgical techniques:
 - **Cold Steel Excision (Phonomicrosurgery):**
 - Lowest risk of scar formation in vocal folds.
 - **Microdebrider (Skimmer):**
 - Powered-instrument with a special toothless oscillating blade has a side-hole coupled to suction.
 - Avoid damaging the delicate vocal folds.
 - Advantages:
 - Fast
 - Cost-effective
 - No risk of thermal injury to surrounding tissues.
 - Less pain
 - Better for diffused disease.
 - Anterior commissure mucosa should always be preserved to prevent synechia formation.



- **Laser ablation:**
 - Better for limited disease.
 - Types:
 - **Carbon Dioxide (CO2):**
 - Absorbed by water in the tissues.
 - Resulting in destruction and cauterization of tissue surfaces.
 - Risk of thermal injury to adjacent tissues.
 - Water-targeted lasers give immediate visible debulking of lesions.
 - Previously, it was used only with rigid laryngoscopy and micromanipulator.
 - Now, flexible CO2 laser is available.
 - **Potassium Titanyl Phosphate (KTP):**
 - Angioleptic laser.
 - Absorbed by oxyhemoglobin.
 - More precise delivery of laser energy to blood vessel feeding the lesions with less thermal injury risk.
 - Photoangiolytic lasers (PDL/KTP) limited in depth of penetration.
 - Delivered with rigid and flexible methods.
 - Advantages:
 - Precise.
 - Excellent hemostatic ability
 - Disadvantages:
 - Slow
 - Expensive
 - Increase operative time
 - Risk of thermal injury to surrounding tissues.
 - Risk of infection transmission to the surgeon (laser smoke was found to contain active viral DNA)
 - Risks of lasers (fire, eye injury)

- Special conditions:
 - **Limited laryngeal involvement:**
 - Typically dysphonia is the main complaint.
 - Usually the patient can be intubated with small ETT or the ventilation can be done with insufflation technique or intermittent apneas technique.
 - When the disease is limited, laser is utilized to remove the papillomatosis.
 - When the disease is diffuse or multifocal, microdebrider is utilized to remove the papillomatosis.
 - Optimal voice quality, rather than total eradication of all papillomas, should be the desired outcome.
 - **Diffuse disease involving the anterior and posterior commissures::**
 - Typically shortness of breath and respiratory distress is the main complaint.
 - Ventilation is usually done with insufflation technique or intermittent apneas technique or jet ventilation.
 - Microdebrider is utilized to quickly remove the papillomatosis and open the airway.
 - Anterior and posterior laryngeal commissures should left untouched to avoid formation of synechia and cicatricial sequelae.
 - Cidofovir is injected into residual papillomas.
 - Some surgeons may do staged papilloma removal for disease in anterior commissure can be done to prevent apposition of two raw mucosal surfaces and minimize risk of web formation.
 - Goals of therapy in extensive disease:
 - Create safe and patent airway
 - Reduce tumor burden
 - Decrease spread of the disease
 - Optimize voice quality
 - Increase time interval between surgical procedures.

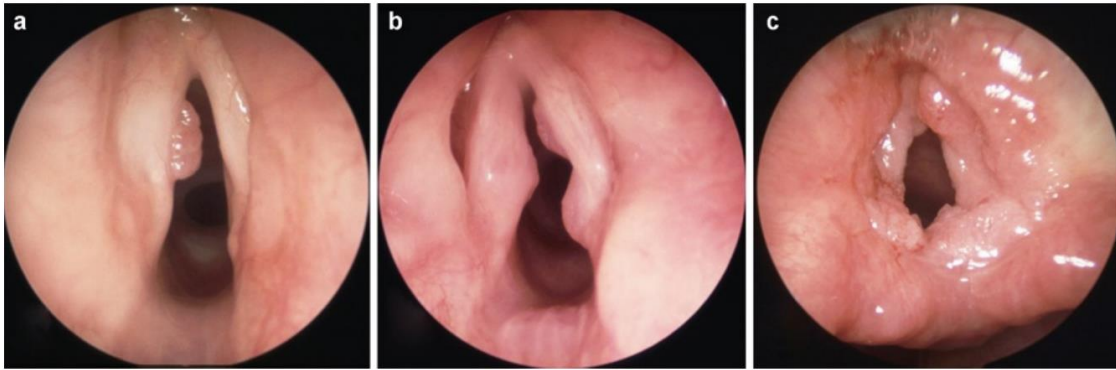


Fig. 16.7 Juvenile-onset recurrent respiratory papillomatosis – limited laryngeal disease: (a) Single papilloma at the free border of the left vocal cord. (b) Multiple sites of papillomatosis on both vocal cords. (c) More spreading disease involving the anterior and posterior laryngeal commissures

- **Aggressive disease:**

- Concurrent cidofovir injections should be done at the first session of debulking in order to avoid tracheostomy.
- If there is early recurrence of dyspnoea, securing the airway with a tracheostomy is necessary.
- The aim is to maintain a safe lower airway while controlling disease progression using adjuvant therapies.
- Tracheotomy should be avoided unless absolutely necessary as it may activate or contribute to distal spread of the disease.
- During surgical debridement on tracheostomised patient, tracheostomy cannula is removed intermittently and microdebrider probe is passed through stoma to remove tracheal papillomas at tracheostomy site.
- Lower airways can be easily reached to the carina and main stem bronchi.
- Decannulation should be considered once the disease is managed effectively.

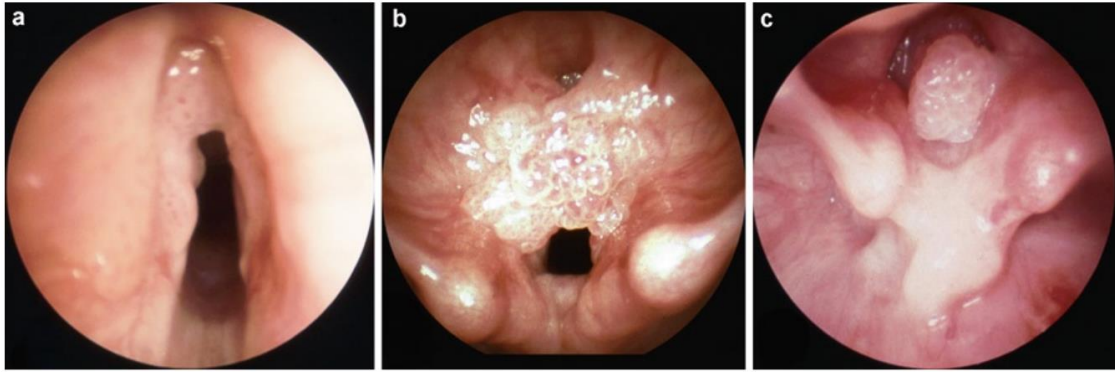


Fig. 16.3 Macroscopic features of juvenile-onset recurrent respiratory papillomatosis: (a) Sessile implantation of papillomas on the left vocal cord. Prior to suction of the secretions, the papillary nature of the lesion is not conspicuous. (b) Irregular exophytic clusters of papillomas with multiple implantation sites. (c) Pedunculated papillomas of the right supraglottis and retrocricoid region

Fig. 16.5 Aggressive airway papillomatosis in a 2-year-old child (same patient as in Fig. 16.9): (a) Tracheal involvement at the distal tip of the tracheostomy cannula: Squamous cell metaplasia is probably responsible for implantation of papillomas at this level. (b) Subtotal obstruction of the right intermediate bronchus by a pedunculated papilloma

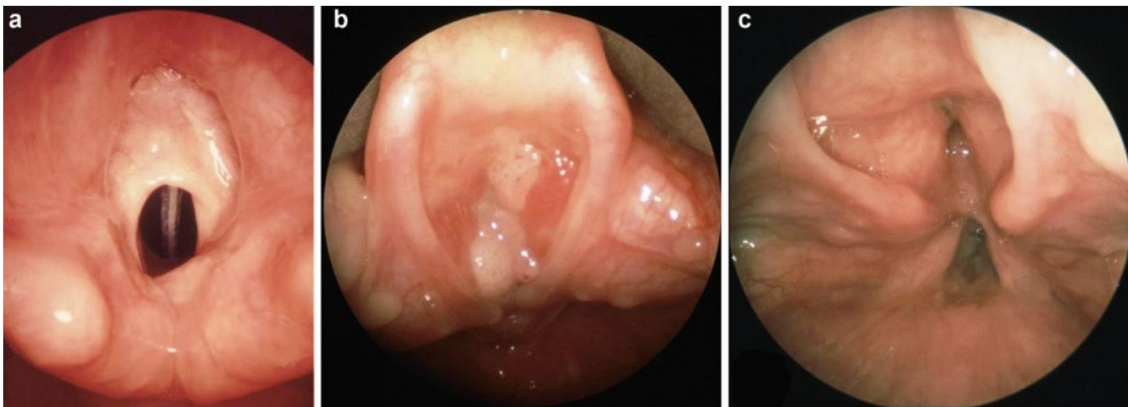
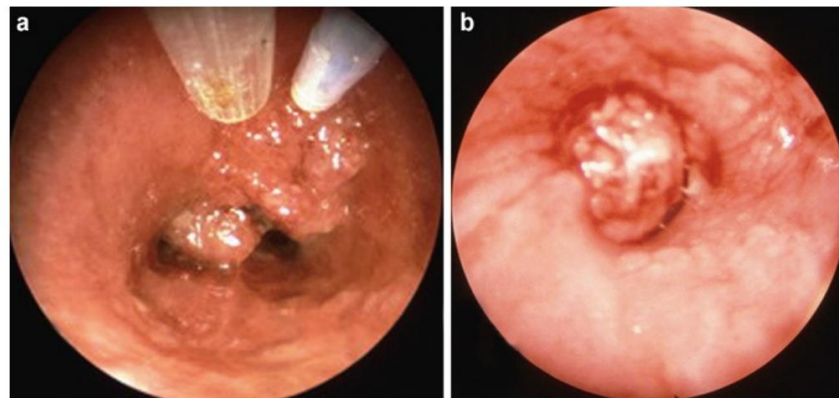
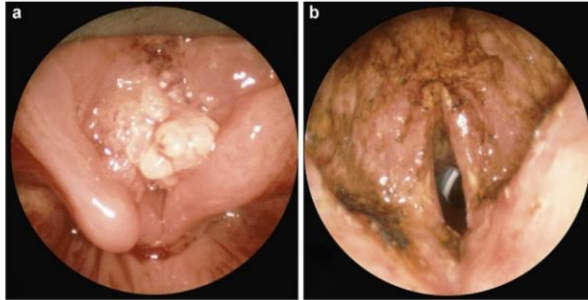


Fig. 16.6 Unacceptable cicatricial sequelae of treatment for juvenile-onset recurrent respiratory papillomatosis: (a) Severe glottic synechia. (b) Complete transglottic stenosis with residual clusters of papillomas. (c) Severe pharyngeal and laryngotracheal stenoses with pharyngotracheal fistula

Fig. 16.8 Obstructive clusters of papillomas obscuring the glottis: (a) Preoperative view: no visible residual lumen upon endoscopy. No tracheotomy. (b) Immediate postoperative view: complete superficial removal of supraglottic papillomas, and preserved vocal cords and subglottis. The transtracheal catheter for jet ventilation is seen in the cervical trachea



- **Adjuvant therapy:**
 - 20% of JoRRP patients will require adjuvant therapy.
 - **Indications:**
 - 1. Location:**
 - Anterior commissures.
 - Posterior commissures.
 - 2. Aggressive disease**
 - JoRRP < 3 years of age.
 - HPV 11 subtype
 - Airway compromise requiring tracheostomy
 - Distal disease spread
 - 3. Recurrence disease:**
 - > 4 surgical procedures/year.
 - **Types:**
 - 1. Cidofovir:**
 - Antiviral agent.
 - Most frequently used adjuvant drug.
 - Intra-lesional injection (off-label use) followed by surgical depulking.
 - Normal mucosa surrounding sites initial papilloma implantations is injected to reduce the viral load.
 - More effective in 'virgin papillomas' than in recurrent papillomas embedded in cicatricial tissue due to the leakage of injected solution out of dense scar tissue.
 - Complete resolution in 60% of patients.
 - Partial response in 35% of patients.
 - **Dose:**
 - Maximum recommended dose is 2mg/kg at concentration of **7.5mg/ml**.
 - Maximum dose of 25mg per session.
 - **Period:**
 - 5 sessions with interval of 2–3 weeks.
 - **Side effects:**
 - Dysplasia (2.7%)
 - Nephrotoxic

2. Alpha-2A Interferon:

- Prevents HPV viral replication and penetration into host cells.
- Response rates ranging from 30–60%.
- Subcutaneous injections.
- Dose:
 - 5 million IU/m² body surface/day.
- Period:
 - BID for 1 month and then 3 times a week for 6 months.
- Side effects:
 - Not tolerated by small children because of the need for repeated shots.
 - Fever and flu-like symptoms.
 - Hepatotoxic.
 - Leukopenia spastic diplegia
 - Febrile seizures
 - Thrombocytopenia

3. Bevacizumab (Avastin):

- Antineoplastic monoclonal antibody.
- Blocks angiogenesis by inhibiting vascular endothelial growth factor A (**VEGF-A**).
- Synergistic effects when combined with angiolytic lasers (KTP/PDL).
- 90% reduction in recurrence after five treatments was demonstrated.
- Intralesional injection
- Dose:
 - 2.5-5mg in children.
 - Up to 12.5mg in adults.

4. Indole-3-carbinol (I3C):

- Over-the-counter oral nutritional supplement.
- Found in cruciferous vegetables such as broccoli and cabbage.
- Potent inducer of cytochrome P450-regulated metabolism of estrogen.
- Estrogen increases HPV gene expression and allows for epithelial proliferation.
- Slow disease progression in 70% of patients.
- Dose:
 - 100–200 mg/day.

5. Photodynamic Therapy (PDT):

- Based on transfer of energy to a photosensitive drug.
- Intravenous injection of Dihematoporphyrin Ether (DHE) which has tendency to concentrate within papillomas.
- Followed by photoactivation with argon pump dye laser.
- Results in small but significant in decreasing RRP growth.
- Dose:
 - 4.25mg/kg of DHE.
- Side effects:
 - Marked photosensitivity for 2-8 weeks

6. Anti-Reflux medications:

- Recurrent or chronic exposure to acid cause inflammatory changes to epithelial lining of aerodigestive tract.
- Complete or partial control of RRP was achieved when reflux was improved.
- Optimal control of extraesophageal reflux disease has been advocated as a means of improving outcomes, along with surgical therapy.

7. Mumps/MMR vaccine:

- Intralesional injection of mumps vaccine and measles/mumps/rubella vaccine has been investigated in an open-label single-center trial with moderate success.
- These positive results, have not been reproduced by other investigators.

8. HSP-E7 (Heat Shock Protein):

- Recombinant fusion protein.
- Composed of heat shock protein (HSP65) from Mycobacterium bovis and E7 gene product of HPV-16.
- Used successfully as a novel vaccine in patients with anogenital warts.
- Ongoing clinical trials for RRP.

9. Celecoxib (Celebrex):

- Selective COX-2 inhibitors.
- HPV-infected cells was found to behave differently regarding regulation of cyclooxygenase-2 (COX-2).
- Ongoing clinical trials for RRP.

10. Propranolol:

- Initial studies showed that oral administration of the drug as an adjuvant improve voice and increase intervals between OR.
- Need further studies to confirm its efficacy for RRP.

11. Acyclovir:

- Inhibits thymidine kinase which is an enzyme not encoded by HPV.
- Not recommended in the treatment of juvenile respiratory papillomatosis.

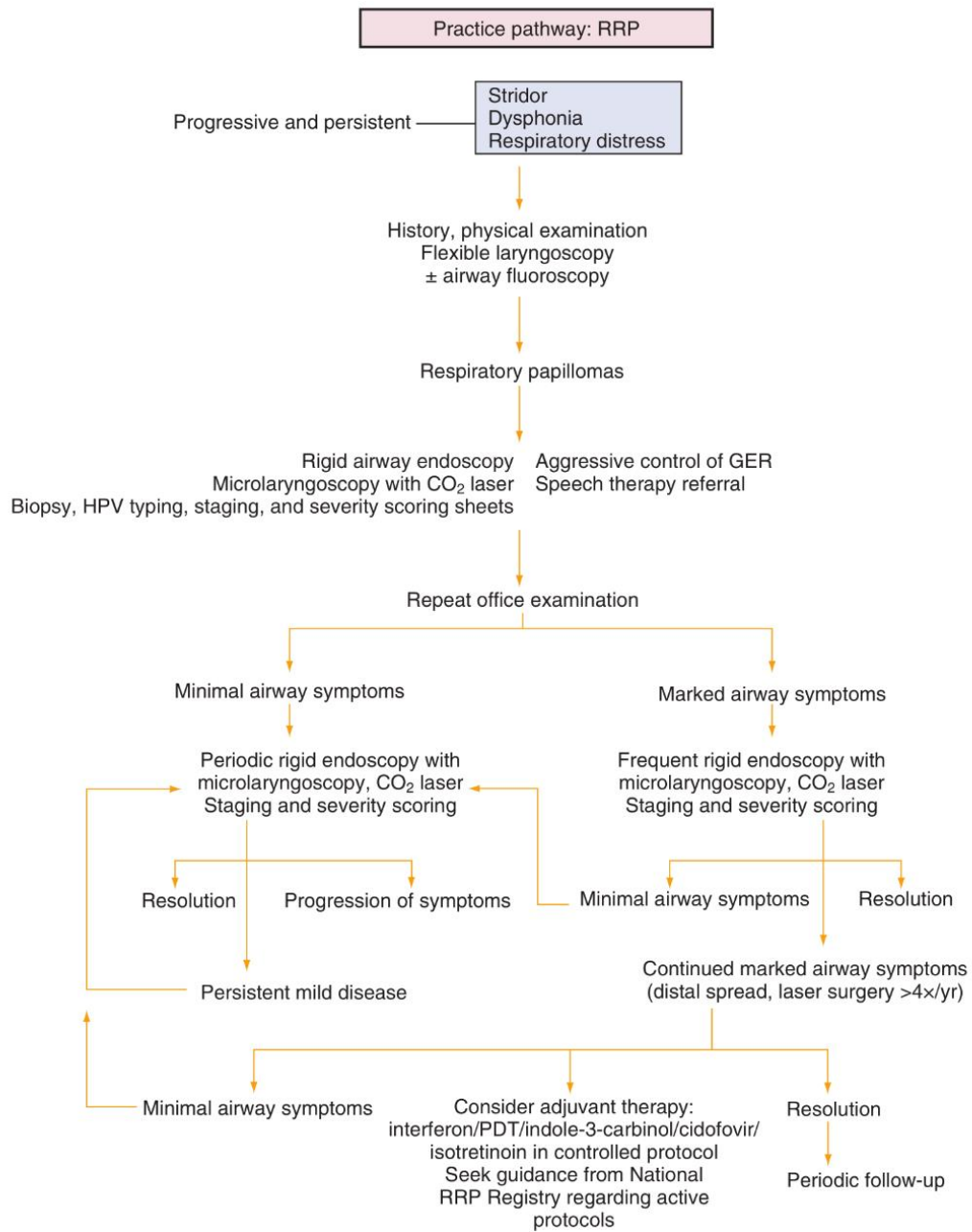


FIGURE 204-7. Practice pathway for recurrent respiratory papillomatosis. GER, gastroesophageal reflux; HPV, human papillomavirus; PDT, photodynamic therapy; RRP, recurrent respiratory papillomatosis.

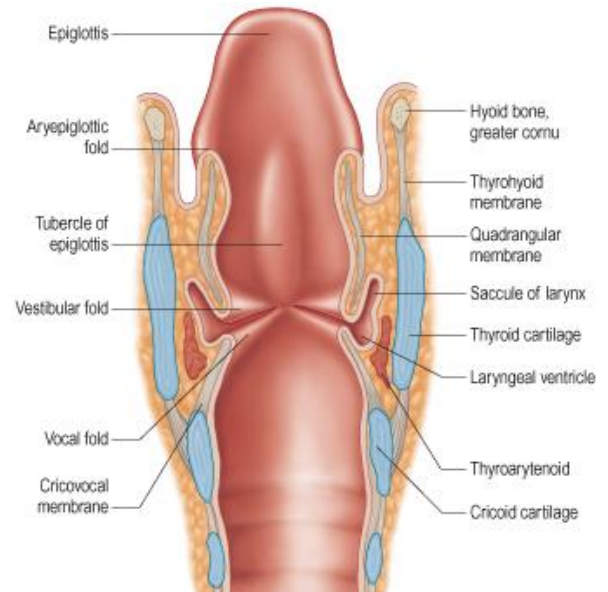
- **Laryngeal Hemangioma:**
 - See other lecture for more details.
 - Most common airway location is subglottic.
 - Adult hemangiomas involve vocal cord or supraglottic larynx.
 - They are cavernous type and cannot be treated with laser.
 - They are left alone if asymptomatic.
 - For larger ones causing symptoms, steroid or radiation therapy may be employed.

- **Chondroma:**
 - More common in men (40-60 age group)
 - Most commonly arises from internal posterior cricoid cartilage (hyaline cartilage).
 - May also arise from thyroid, arytenoid, epiglottic cartilage (fibroelastic cartilage)
 - Symptoms:
 - o Insidious hoarseness from vocal fold restriction, dyspnea from subglottic lesions, dysphagia from posterior cricoid lesions, globus sensation
 - Examination:
 - o Lesion is smooth, firm, fixed tumor, normal mucosa
 - Diagnosis:
 - o Endoscopic wedge biopsy
 - o CT neck (calcification)
 - Treatment:
 - o Complete excision via endoscopic or external approach (depending on the size of lesion)

- **Granular Cell Tumor (Granular Cell Myoblastoma)**
 - 3% risk of malignant degeneration
 - Arise from Schwann cells in the posterior aspect of true vocal fold or arytenoids (originally believed to arise from myoblasts).
 - May also be found on tongue, skin, breast, and subcutaneous tissue.
 - Symptoms:
 - o Insidious hoarseness
 - Examination:
 - o Lesion is small, sessile, gray mass.
 - Diagnosis:
 - o Endoscopic biopsy
 - Treatment:
 - o Complete excision via endoscopic or external approach (depending on the size of lesion)

Saccular disorders:

- Ventricle of larynx opens into **Saccul**.
 - Blind-ended pouch ascends forwards from ventricle between vestibular fold and thyroid cartilage.
 - 60-70 mucous glands open onto its luminal surface help to lubricate vocal cords which lack mucous glands.
- **Laryngocele:**
- Laryngocele is **Expanding air-filled cyst** formed in Laryngeal Saccul with communication to laryngeal lumen (patent orifice).
- **Laryngopyocele** is **infected pus-filled** laryngocele.
- Types of laryngocele:
 - **External Laryngocele:**
 - Laryngocele sac protrudes through Thyrohyoid membrane at point of entry superior laryngeal vessels and Internal branch of superior laryngeal nerve.
 - Presents as Lateral Neck mass.
 - More common.
 - **Internal Laryngocele:**
 - Laryngocele sac remains within thyroid cartilage.
 - Expands into Paraglottic space and extend superiorly to expand Aryepiglottic fold and reach Vallecula.
 - Less common.
 - **Combined.**
- Pathophysiology:
 - Congenital (Extremely rare)
 - Acquired due to expansion from prolonged increased intra-glottic pressure of laryngeal saccul (glass blowers, trumpet players).



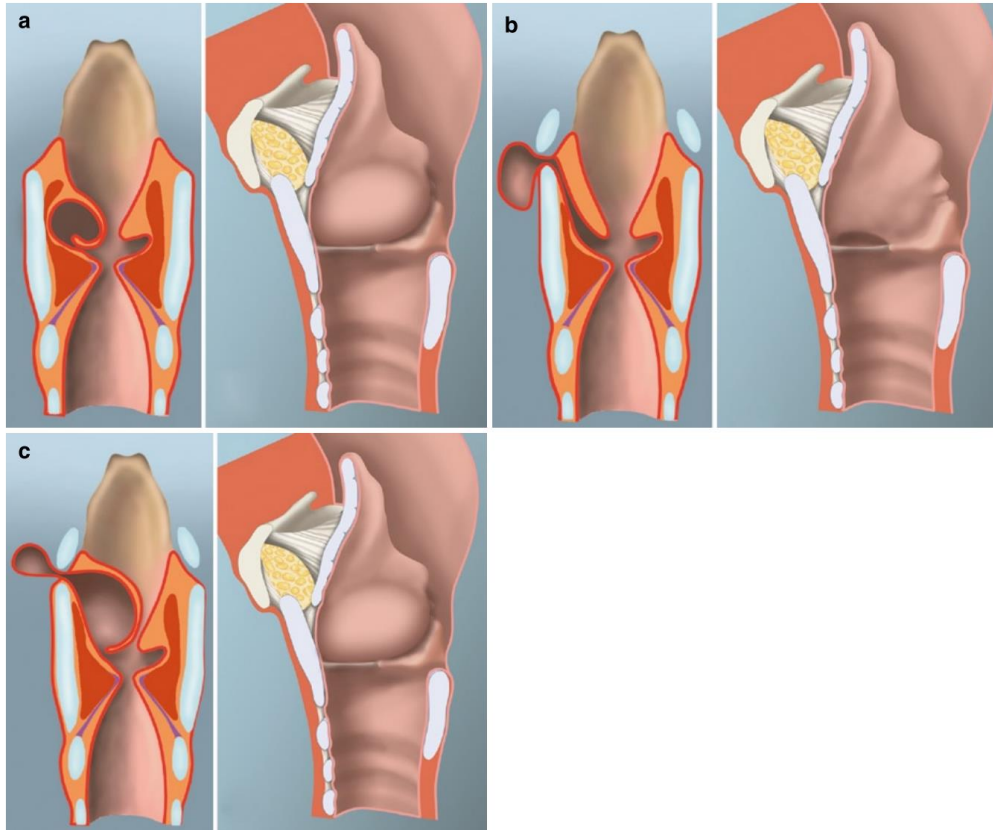


Fig. 11.3 Diagram of laryngoceles: (a) Internal laryngocele (b) External laryngocele (c) Combined laryngocele (Adapted from Holinger [7]. With permission)

- **Saccular Cyst:**
- Saccular Cyst is **Mucus-filled dilation** of Saccule without communication to laryngeal lumen (blocked orifice).
- Mucopyoceles is **infected pus-filled** saccular cyst.
- Pathophysiology:
 - Congenital (rare but more common than congenital laryngocele)
 - Acquired due to obstruction of saccular orifice by inflammation, scarring or by **laryngeal carcinoma**.
- Types of Saccular cyst:
 - **Lateral saccular cyst:**
 - Most common,
 - Large cyst extends postero-superiorly into false vocal folds and aryepiglottic fold.
 - Causing respiratory symptoms in neonates.
 - **Anterior saccular cyst:**
 - Less common.
 - Small cyst extends medially into laryngeal lumen.
 - Obstructing anterior portion of the vocal fold.
 - Causing hoarseness mainly.

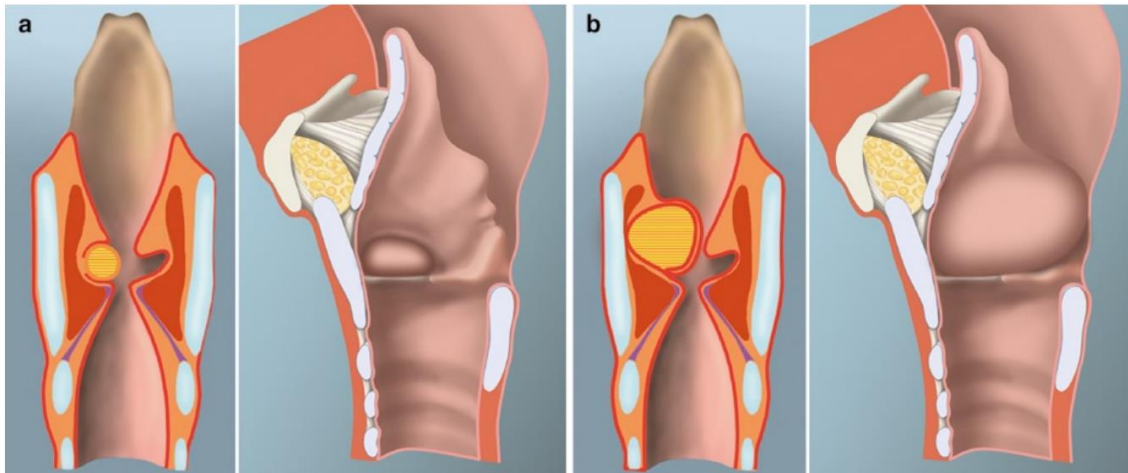
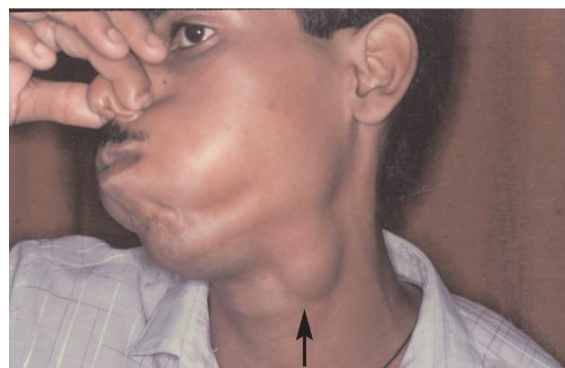
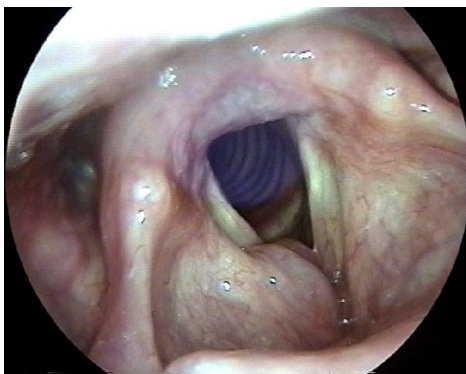


Fig. 11.2 Diagram of saccular cysts of the larynx: (a) Anterior saccular cyst (b) Lateral saccular cyst (Adapted from Holinger [7]. With permission)

- Clinical Picture of saccular cyst and laryngocele:
 - Hoarseness:
 - Most common complaints in adults.
 - Inspiratory stridor
 - Weak cry
 - Dysphagia
 - Lateral Neck mass (Laryngocele).
 - Compressible.
 - Increases in size with increasing intra-laryngeal pressure (Valsava).



- Clinical Picture of saccular cyst and laryngocele:
 - 1. Flexible Nasopharyngoscopy:**
 - Submucosal laryngeal swelling.
 - 2. U/S Neck.**
 - 3. CT Neck:**
 - Presence of air within the lesion differentiates Laryngocele from Saccular cysts.

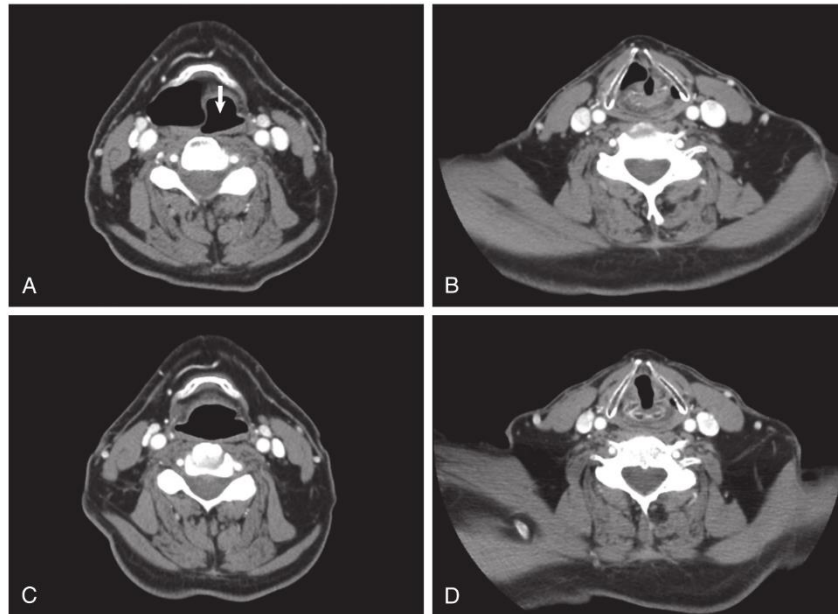


FIGURE 61-33. Laryngocele (air-filled variant of saccular cyst) in preoperative and postoperative computed tomography scans. **A**, Preoperative view of a large air-filled sac that pushes the epiglottis (arrow) to the patient's left. **B**, At the level of the ventricle, the opening to the dilated saccule is shown. **C**, Postoperative view. **D**, Postoperative view.

- Clinical Picture of saccular cyst and laryngocele:
 - **Endoscopic Aspiration:**
 - High recurrence rate.
 - **Endoscopic Marsupialization.**
 - **Endoscopic Excision:**
 - Mainstay of treatment.
 - Low recurrence rate.
 - Co2 laser can be used.
 - **Trans-cervical open approach:**
 - For external or combined laryngocele.
 - Can be used for recurrent internal disease (laryngofissure).

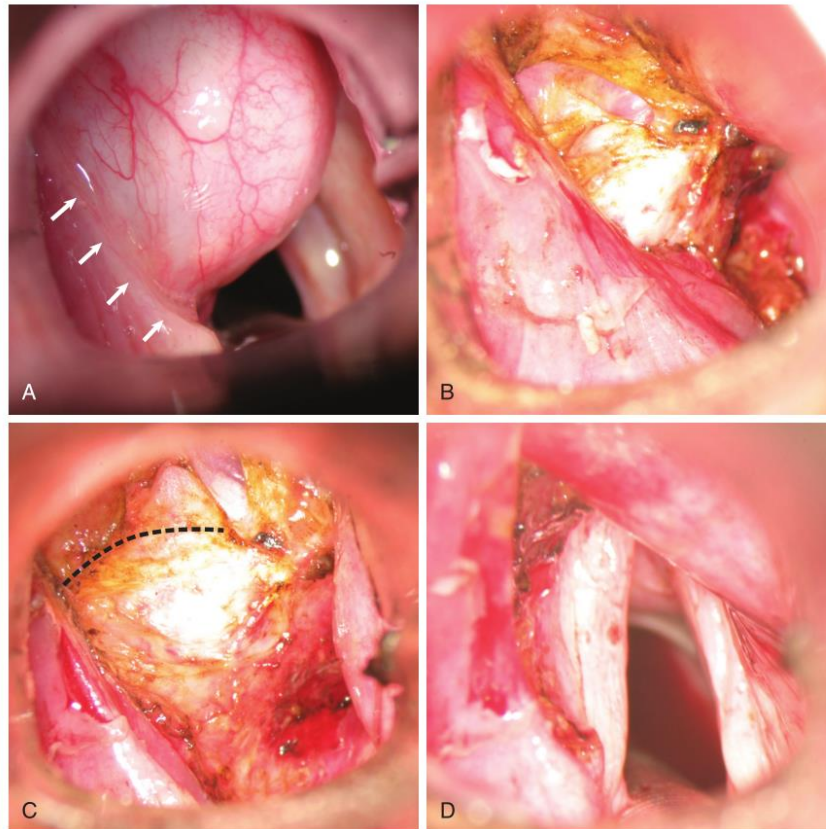


FIGURE 61-30. Lateral saccular cysts may not present as obviously into the laryngeal vestibule as this one does, but its lateral extent has dissected over the top of the thyroid cartilage. **A**, Note margin of false fold (*line of arrows*). **B**, Removal begins with excision of the false fold margin in order to dissect downward to the lining of the saccule. **C**, After removal. Note upper border of inner surface of thyroid cartilage (*dotted line*); the distal end of the laryngoscope is aimed laterally toward the neck contents. **D**, In-line view of vocal folds at conclusion of surgery.

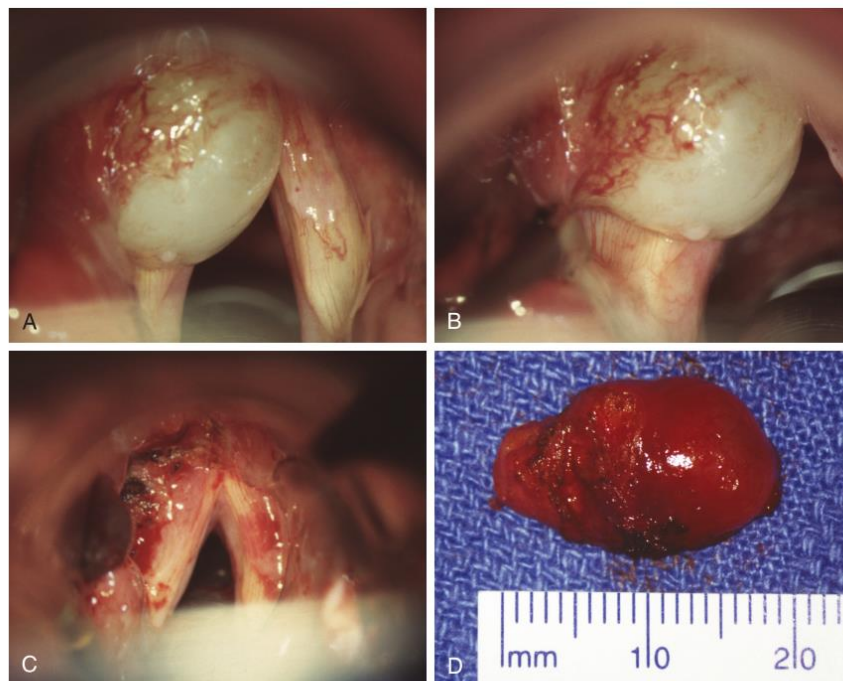
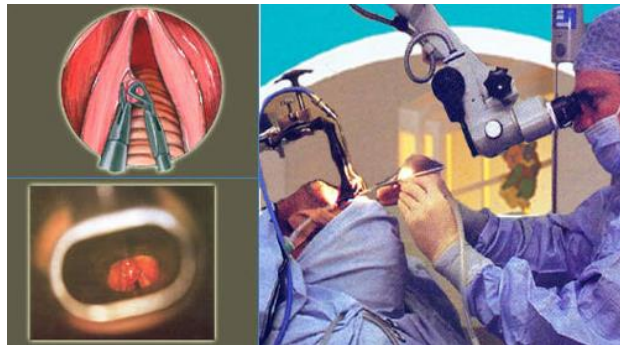


FIGURE 61-32. Anterior saccular cyst. **A**, This cyst presents only into the laryngeal vestibule and has not dissected outside the larynx. **B**, Forceps retracted to show true vocal fold below. **C**, Laryngoscopic view just after excision. **D**, Intact anterior saccular cyst.

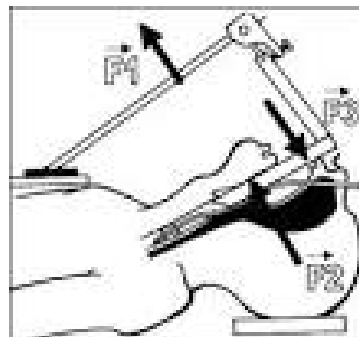
Phono-Microsurgery:

- Elective operation involve precision microsurgical removal of benign vocal fold pathology, commonly from subepithelial space of the vocal fold.
- Based on vocal fold physiology (Hirano cover-body theory).
- Primary goal is to improve voice quality.
 - o By limiting dissection to as superficial plane as possible and to maximally preserve epithelial and lamina propria.
- Indicated after failure of all nonsurgical treatment modalities.
- **Pre-operative considerations:**
 - o Avoid:
 - NSAID or anticoagulation medications as possible.
 - Vocal abuse immediately before surgery.
 - o Voice therapy:
 - Pre-op voice therapy for at least 1-2 sessions.
 - Extremely important in pre-op preparation due to:
 - Psychological preparation for surgery and postoperative voice rest.
 - Modify and improve improper speaking technique
 - o Stroboscopy:
 - All patients should have preoperative stroboscopy.
 - Surgeon should review the most recent stroboscopy in OR to correlate stroboscopic findings with surgical findings.
 - o Consent:
 - Risks of general anesthesia
 - Injury to dentition
 - Injury to lingual nerve
 - Numbness of tongue and change in taste sensation
 - Temporary lasting form 2 weeks to maximum of 1 month.
 - Risk of no improvement or reduction of voice quality (1-2%).
 - Failure of microflap to appropriately redrape.
 - Postoperative granuloma formation



- **Equipment:**

- Laryngoscopes:
 - The larger the laryngoscope the better.
 - Allows improved exposure and access to surgical field.
 - Suspension of laryngoscope is a basic aspect of phonosurgery.
- Telescopes:
 - 30- and 70-degree telescopes
 - 4mm in diameter and 30 cm long
 - Should be used immediately before phonosurgical incision
 - Provide multiple angles of visualization for the vocal folds and its related pathology.
- Microscope:
 - Should be of highest quality and compatible with a CO2 laser micromanipulator attachment.
 - Typical focal length of lens used on surgical microscope for phonosurgery is **400 mm** which allows adequate space between proximal end of laryngoscope and the microscope for placement and use of hand instruments for phonosurgery.
- Instruments:
 - Microelevators
 - Cup forceps
 - Scissors
 - Curved alligators
 - Small suctions (3, 5, and 7 French)
 - Microflap retraction (triangular/ Bouchayer forceps)
 - Microsurgical knife (sickle or diamond shaped)
- Laser:
 - **CO2 laser with the micromanipulator:**
 - Used to incise vocal fold tissue
 - Allows an "instrument free" approach
 - Risks of thermal injury
 - Expensive
 - **Angiolytic lasers:**
 - KTP and PDL
 - Mainly used in papilloma disease



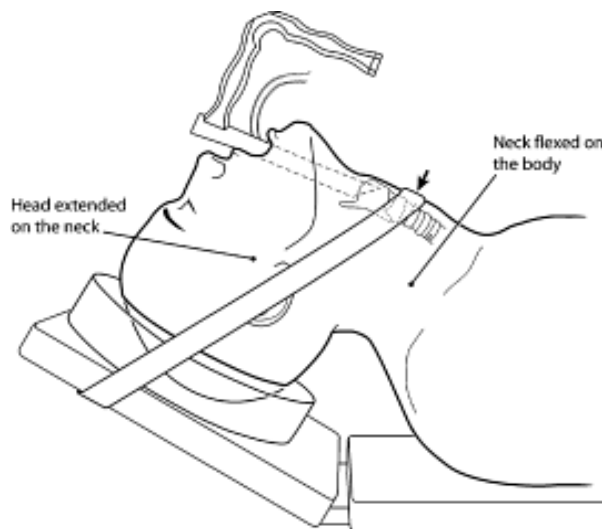
- **Surgical approach:**

○ Anesthesia:

- General anesthesia with muscle paralysis.
- Pre-op IV steroids and glycopyrrolate
- Ventilation options:
 - Endotracheal intubation:
 - Best performed with small (5 or 6) specialized endotracheal tube.
 - Provides both a still operating field and complete control of airway.
 - If ETT obstruct the surgical site, it can be repositioned or completely removed.
 - Insufflation method.
 - Apneic method.
 - Jet ventilation:
 - Should be done only as needed.
 - Best done with a midtracheal jet ventilation catheter.
 - Tracheal jet ventilation is preferred to supraglottic jet ventilation because it provides the surgeon with less vibration and desiccation of the vocal fold tissues.

○ Patient position:

- Neck flexed on the body and head is extended.
- Optimal position for exposure of endolarynx with laryngoscope.
- Neck flexion is achieved by using articulated head on OR table with the head extension on the neck being done by the surgeon, and then secured with the suspension device
- Shoulder roll places the patient in a suboptimal position for optimal laryngoscope placement should not be used.



○ Laryngoscope position:

- Dental and alveolar ridge protection.
- Laryngoscope should be positioned immediately above the vocal fold resulting in retraction of false vocal fold tissues.
- Avoid contacting superior surface of vocal fold.
- Suspension device should be positioned to provide upward and forward (caudal) suspension of laryngoscope in the larynx.
- A Velcro strap or silk tape is applied to external neck (cricoid or trachea) to improve endolaryngeal exposure if needed.
- Trendelenburg or reverse trendelenburg position of the OR table is utilized to improve the surgical view on microscope.

○ Surgical techniques:

- Examination with telescope should be done initially.
- Microscope is used for surgery.
- Microflap approach to submucosal pathology:
 - Making incision through epithelium immediately lateral to vocal fold pathology to minimize disruption of normal mucosa.
 - Minimal disrupting of surrounding tissue to vocal fold pathology.
 - After incision, submucosal dissection is done with small curved elevator in a superficial plane between vocal fold pathology and underlying epithelium and between vocal fold pathology and vocal ligament.
 - Submucosal pathology is removed and sent for pathologic examination.
 - Microflap is then re-draped and palpation of free edge of vocal fold is done to determine if any residual submucosal pathology remains.
 - Topical application of 1/10,000 epinephrine on cottonoid is achieved to improve hemostasis.

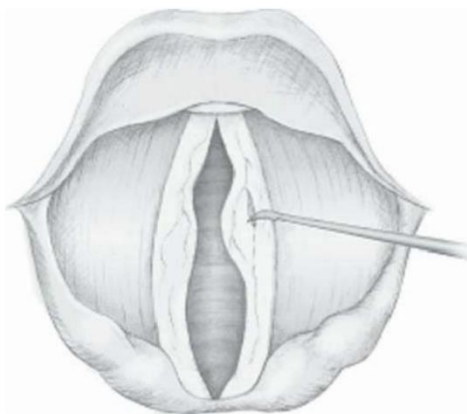


Figure 68.20 Incision immediately lateral to submucosal pathology for microflap approach for removal of the vocal fold lesion.

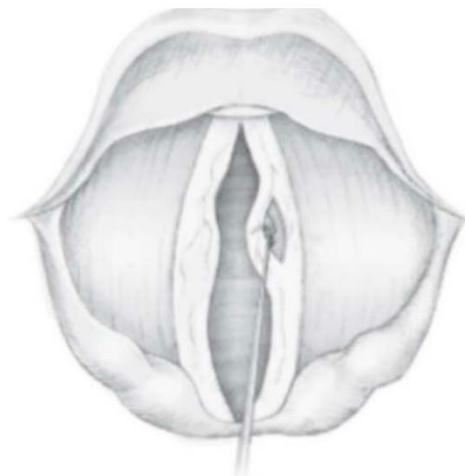


Figure 68.21 Microflap elevation.

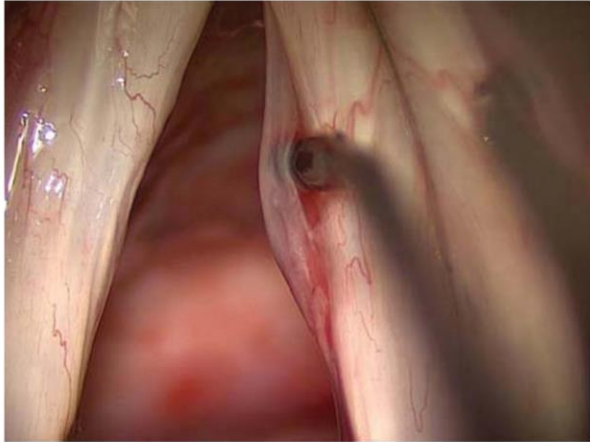


Figure 68.22 Careful elevation of the microflap overlying the vocal fold cyst-ligament. Same cyst also seen in Figures 68.5 and 68.6.

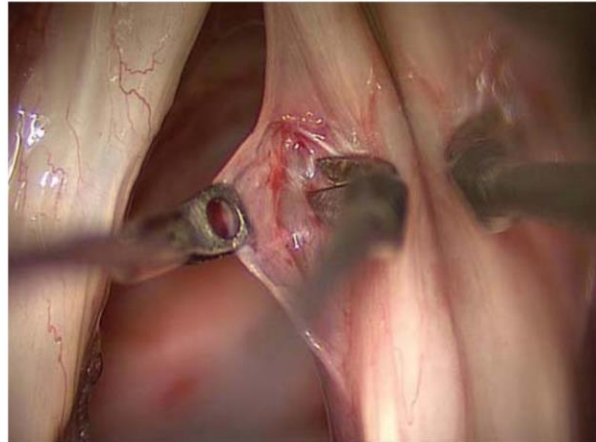


Figure 68.24 Release of fibrous bands over the vocal fold cyst-ligament with microcurved scissors.

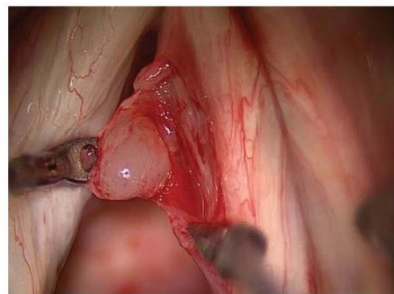


Figure 68.26 Vocal fold cyst-ligament undergoing final release with microcurved scissors.



Figure 68.25 Difficult dissection of vocal fold cyst with adherent bands to the vocal ligament.

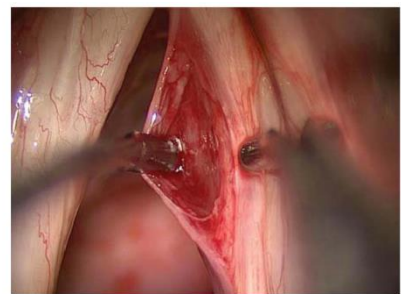


Figure 68.27 Inspection of the microflap and vocal ligament after phonomicrosurgical excision of a vocal fold cyst-ligamentous.

- **Postoperative care:**

- Voice rest:
 - Range from 2-14 days post-op depending on nature of the surgery, compliance of the patient and surgeon's philosophy and previous experience.
- Anti-reflux medications:
 - Continue empiric treatment with PPI and GERD behavior modification to minimize possibility of acid reflux.
- Postoperative stroboscopy:
 - Done at the end of strict voice rest period.
 - Evaluate recovery and healing process of vocal fold.
 - Recovery is demonstrated by return of mucosal pliability.
 - Stroboscopy helps to identify postoperative scar formation.
- Voice therapy:
 - Important to assist the patient in transitioning from complete voice rest to light voice use to ensure using the optimal postoperative voice technique to facilitate healing and prevent injury.

- **Complications of suspension laryngoscopy:**

1. Mucosal injury:

- Most common complication **(75%)**.
- Mostly, minor injuries that heals spontaneously within few days.
- More severe injury can lead to hematoma, mainly at the tongue.
- Right side is the most common side.
- Types:
 - Erosions
 - Fissures
 - Bleeding
 - Hematoma
- Locations:
 - Oral cavity **(50%)**
 - Oropharynx **(30%)**
 - Lips **(20%)**
 - Hypopharynx

2. Dental injury:

- Occurs in **10%** of all patients.
- Most common injury is loosening of one or more teeth.
- Most common location is upper front teeth.
- Right side is the most common side.

3. Nerve injury:

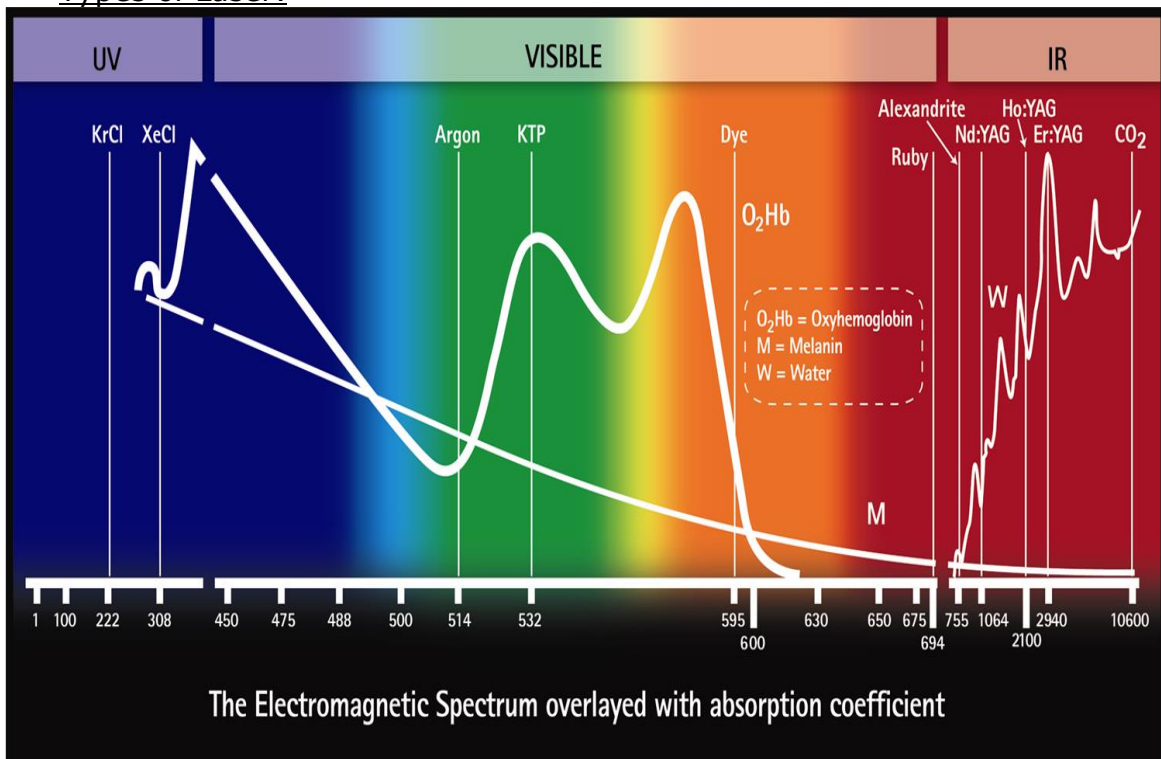
- Occurs in **5%** of all patients.
- Most are temporarily due to neuropraxic injury.
- Related mainly to the length of suspension time
- Location:
 - **Lingual nerve:**
 - Most common nerve injured (2%).
 - Caused by stretching of lingual nerve by pressure of suspended laryngoscope on the tongue or retrolingual region.
 - Tongue numbness and change in taste sensation.
 - Mainly at the right side.
 - Lasting for 4 weeks.
 - **Hypoglossal nerve:**
 - Occurs in 1% of all patients.
 - Caused by compression of hypoglossal nerve at the tongue base between the hyoid and the laryngoscope blade.
 - Tongue weakness.
 - Mainly at the right side.
 - Lasting for 8 weeks.



Typical complications after suspension laryngoscopy. **A)** Massive hematoma of tongue. **B)** Erosion of lateral part of tongue. **C)** Swelling of lower lip. **D)** Severe tooth loosening at front part of maxilla.

Laser Surgery:

- **Light Amplification by Stimulated Emission of Radiation.**
- Principle:
 - Normally, an atom is in a stable form, i.e. the electrons equal to the number of protons, are revolving around the nucleus in a fixed orbit.
 - When given energy, electrons change their orbits away from the nucleus and the atoms are then called 'excited' but this excited state of atoms does not last long.
 - The atoms soon release their absorbed energy automatically (spontaneous emission) and return to their original state.
 - If photons are made to strike these excited atoms, the decay of the atoms is accelerated and both the incident and the absorbed photons are released (stimulated emission).
 - This stimulated radiation is amplified with the help of mirrors.
 - Thus, lasers are electromagnetic radiations with specific wave length depending upon the type of lasing medium such as argon, carbon dioxide or Nd:YAG.
- Clinical Application:
 - Functions of the laser beams:
 - Vaporize the tissue.
 - Cut the tissue.
 - Coagulate the tissue.
 - The clinical applications of different laser types depend on their wave length and special absorptive powers of the tissues on which they are used.
- Types of Laser:



- **Argon Laser:**

- Wave length 488-514 nm.
- Blue in color.
- Passes through clear fluid.
- Absorbed by Hgb, thus good for pigmented tissues.
- It is mainly used for photocoagulation such as of port wine stain, hemangioma, and telangiectasia.
- It is also used for retinal lesions and microsurgery of the ear.

- **KTP-532 Laser:**

- Wave function 532 nm.
- Falls in visible spectrum (blue-green).
- Absorbed by Hgb, thus good for pigmented tissues.
- Angiolytic Laser used mainly for Hemangioma and RRP (synergic effect if used with Avastin).
- Can be transmitted by flexible endoscopes.

- **Nd:YAG Laser:**

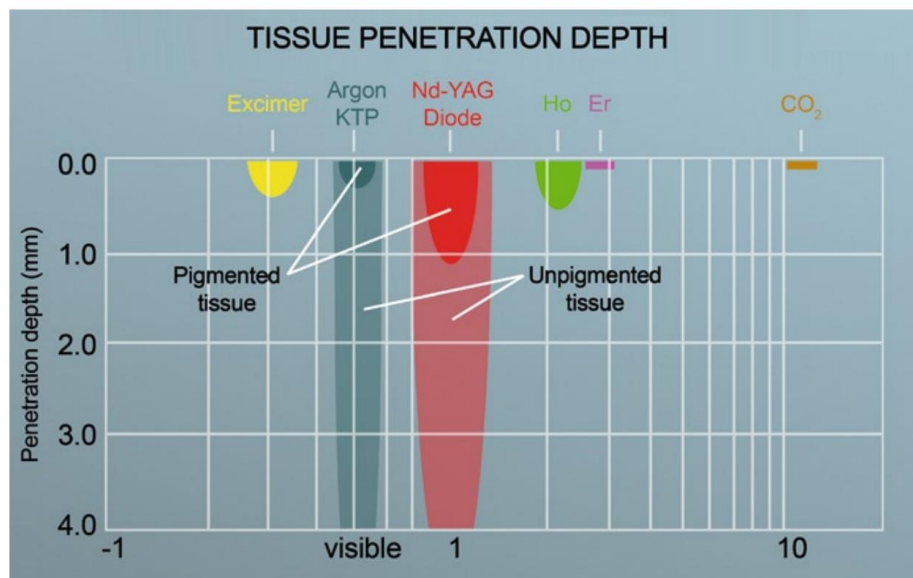
- Wave length 1060 nm.
- Invisible.
- Transmits through clear fluids.
- Has effective coagulative properties.
- Deep zone of coagulation (4 mm).
- Can be transmitted by flexible endoscopes.
- It can be used with CO2 laser in the treatment of tracheal and endobronchial tumors.

- **CO2 Laser:**

- Wave length 10600 nm.
- Invisible.
- Requires aiming beam of helium-neon laser.
- Strongly absorbed by water independent of tissue color.
- Absorbed by Cornea in the eyes, and regular opticals can reflect it.
- Minimal thermal effect on surrounding tissues.
- Very effective to vaporize tissues and give a bloodless field.
- Depth of penetration is 0.1 mm.
- Can coagulate vessels up to 0.5 mm.
- Recently, can be transmitted by flexible endoscopes (Omniguide).
- It is the most commonly used laser in ENT surgery and ideal Laser to be used in laryngeal surgery:
 - Precise cut.
 - Minimal thermal effect.

Table 5–1. Currently used lasers with their electromagnetic spectrum and penetration depth.

Electromagnetic Spectrum	Laser Type	Wavelength	Penetration Depth
Visible lasers	Argon	514 nm	0.8 mm
	KTP-532	532 nm	0.9 mm
	Flashlamp-Excited Dye	577 nm	0.9 mm
Near infrared lasers	Nd: YAG	1060 nm	4.0 mm
Infrared lasers	Ho: YAG	2100 nm	0.4 mm
	Er: YAG	2940 nm	3.0 μ m
	CO ₂	10,600 nm	30.0 μ m

Fig. 4.25 Penetration depth of different lasers into mucous membranes: The wavelength of the laser and the type of biological tissue determine the penetration depth (see text)**Table 4.3** Properties of different lasers

Type of laser		Vaporise	Cut	Coagulate
CO ₂ :	10,600 nm	++	++	±
Argon :	514 nm	±	±	±
KTP :	532 nm			
Nd-Yag :	1,064 nm	±	–	++
Diode :	980 nm			

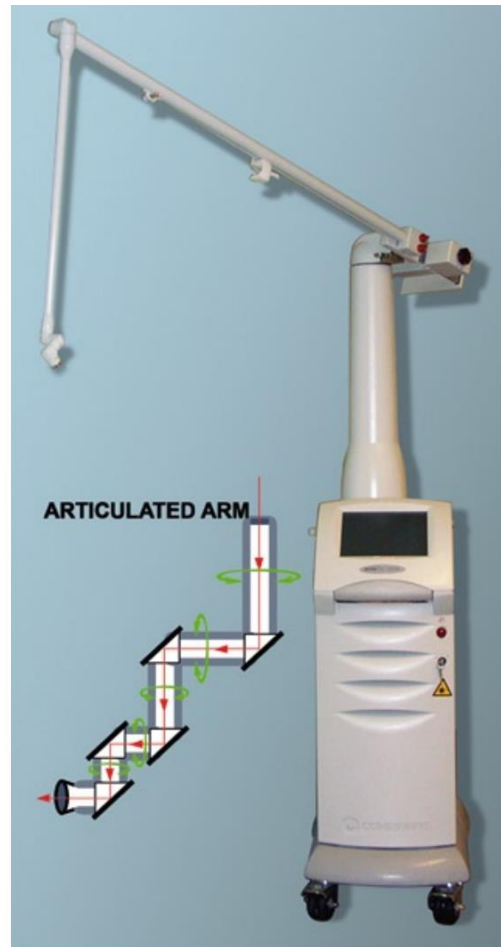


Fig. 4.30 CO₂ laser and articulated arm: Ultrapulse CO₂ laser console and diagram of the articulated arm with mirrors at each knuckle joint

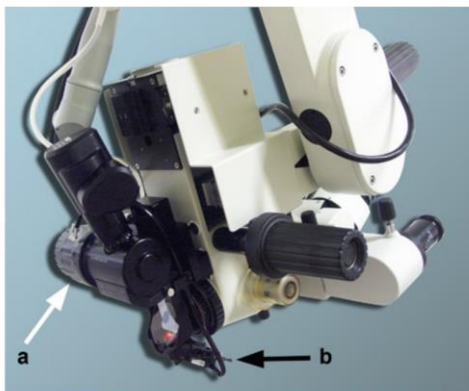


Fig. 4.31 Computer-controlled digital AcuBlade micromanipulator for the CO₂ laser, coupled to the Wild-Leica® microscope: (a) The side knob is used to focus the laser beam on the target (white arrow). (b) The central joystick is used to move the laser beam in the operative field (black arrow)

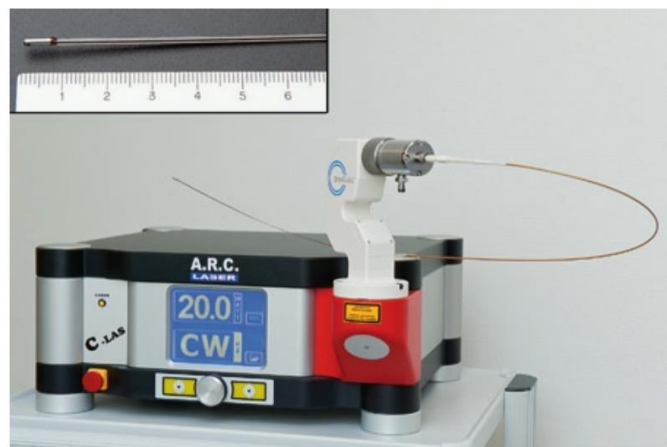


Fig. 4.32 CO₂ laser console with the omniguide CO₂ laser fibre

Safety precautions (Surgical)
• Place laser warning signs outside the operating room • Cover windows and securely close doors while the laser is “on” or in “standby” mode • Personnel and patients must use eye protection specific for CO₂ laser; it should also be available at entrances to the room; standard prescription eye-glasses are sufficient for CO₂ laser; side-guards are recommended • Tape the patient’s eyes shut and apply moist pads • Do not use alcohol or flammable cleaning prep solutions or in the operating room • Use flame retardant materials and drapes • Keep an open container filled with water or saline immediately available to douse a laser fire • Placed moistened swabs/sponges adjacent to the path of the laser beam to protect surrounding tissues and structures • Minimize the possibility of a “blow-torch effect” by carefully protecting endotracheal tubes with wet cloth or neuro patties • The locking key to laser machine should be accessible only to persons trained in the use of laser; it should not be stored in or on the laser machine, but kept in secure location; some lasers have electronic keypads • Keep the laser turned off or in “standby” mode unless in use • Instruments should have brushed, beaded or sand-blasted surfaces to prevent reflection of the laser beam • The surgeon should give the anaesthetist prior warning to reduce the FiO₂ before the laser is activated
Safety precautions (Anaesthetic)
• “Nonflammable” endotracheal tubes may be used (all tubes are flammable) • Fill the cuff of the endotracheal tube with saline coloured with methylene blue (Senior author (WS) prefers air, not saline) • Maintain inspired oxygen (FiO₂) as low as clinically feasible (<30%FiO₂) • Wait a few minutes for the oxygen concentration in the airways to drop before approving activation of the laser • Avoid nitrous oxide if possible
Laser airway fire
• Turn off laser at emergency switch • Turn off all anaesthetic gases • Remove swabs and flammable materials from the airway • Immediately remove the endotracheal tube if intubated • Pour saline down the airway
Fire extinguished
• Re-establish ventilation • Use air/oxygen at lowest FiO₂ that maintains oxygenation • Avoid 100% O₂ if possible • Examine endotracheal tube to see if fragments left in airway • Consider bronchoscopy

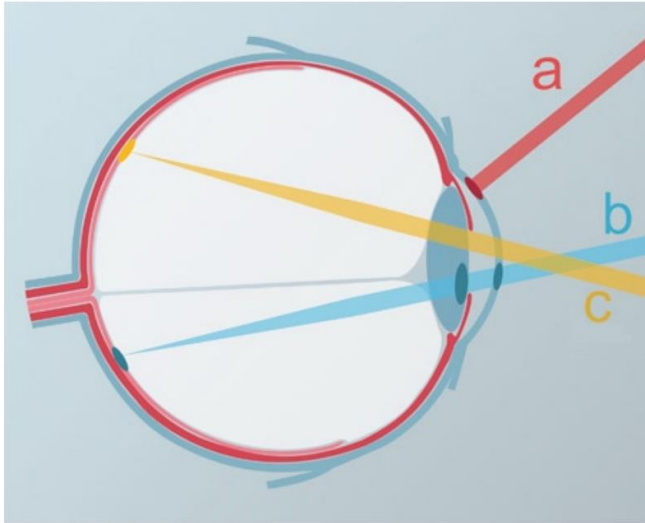


Fig. 4.36 Eye damage resulting from various laser wavelengths: (a) Far-infrared laser beams (CO_2 , erbium and holmium) (*red*) are fully absorbed by water-rich tissues. They are damaging to the cornea. (b) Near infrared lasers (Nd-YAG, diode) (*blue*) are partially absorbed by clear aqueous tissues and pigmented tissues. They may damage the cornea, lens and retina. (c) Lasers working in the visible range (argon, KTP) (*yellow*) are fully transmitted through the transparent aqueous tissues of the eye. They are strongly absorbed by pigmented tissues, thus damaging the retina

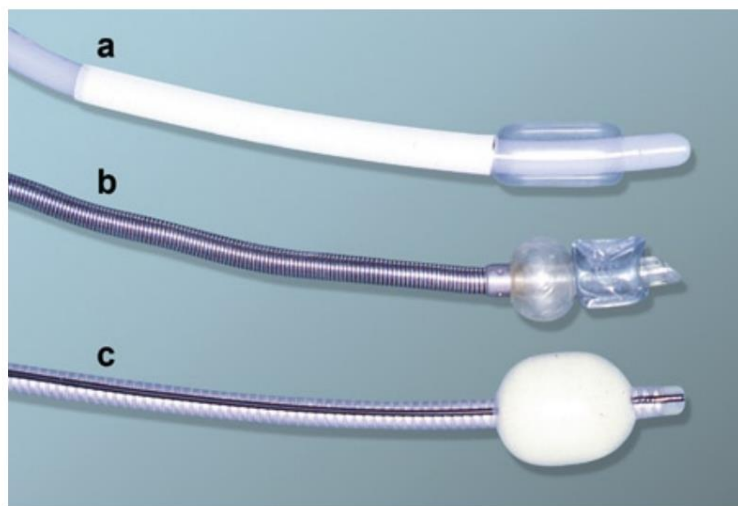


Fig. 4.39 Laser-safe ET tubes: (a) Laser shield tube: This tube is less safe than the Oswald-Hunton, Mallinkrodt or Bivona tubes. (b) Mallinkrodt metallic tube with double cuffs. (c) Bivona metallic tube with foam-cuff

- **In case of Fire during Laryngeal Laser surgery:**
 - Shout **FIRE!**
 - Stop ALL gases including oxygen.
 - Remove damaged ET tube.
 - Douse the fire with saline.
 - Mask ventilate with 100% O₂.
 - Re-intubate with patient paralyzed to facilitate reintubation.
 - Intraoperative bronchoscopy to remove any charred debris and assess extent of damage.
 - Keep the patient intubated and admit him to ICU.
 - Intravenous steroids and antibiotics.
 - Intra-operative re-examination of the airway 3-5 days to assess extent of further airway compromise.
- **Source of ignition:**
 - CO₂ laser.
- **Combustible material:**
 - Endotracheal tube.
 - Anesthetic gas.
 - Dry swab, pledges, tapes, drapes, gum shield, NGT, non-metallic tracheostomy tube, fluid containing alcohol, petroleum jelly, anesthetic tubing and connections.
- **Combustion –supporting atmosphere :**
 - Oxygen, Nitrous oxide.
 - Inert gases (Nitrogen, Helium).

Endoscopic Laser Cordectomy:

- European Laryngological Society Classification:
 - **Type I / Sub-epithelial Cordectomy:**
 - Endoscopic resection of:
 - 1. Vocal cover** (Epithelium and Reinke's space).
 - From vocal process to anterior commissure.
 - Sparing vocal ligament and muscle.
 - Indications:
 - **Diagnostic:**
 - Pre-malignant lesions.
 - **Therapeutic:**
 - Pre-malignant lesions.
 - Carcinoma in situ without microinvasion.

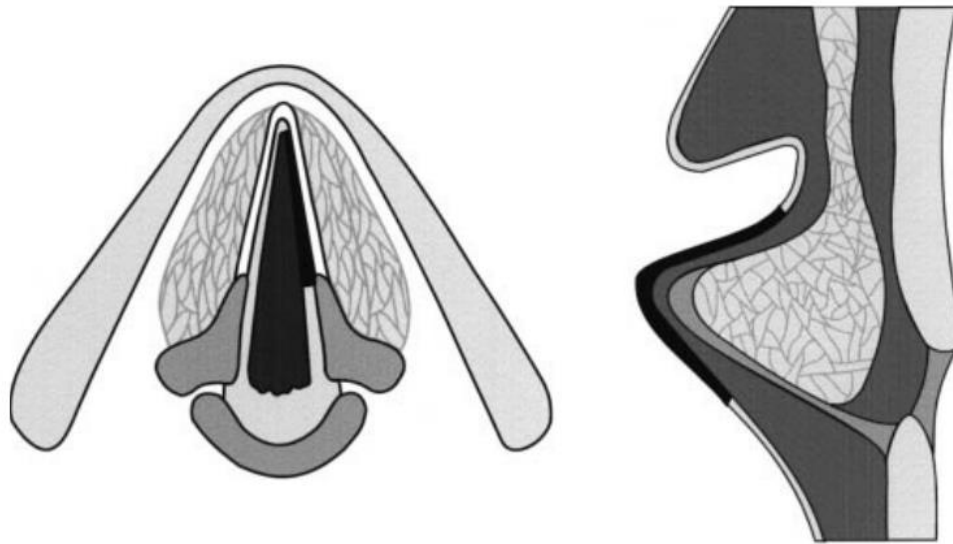


Fig. 1 a, b Subepithelial cordectomy (type I)

- **Type II / Sub-ligamental Cordectomy:**
 - Endoscopic resection of:
 1. Vocal fold cover
 2. Vocal ligament
 - From vocal process to anterior commissure.
 - Sparing vocal muscle.
 - Indications:
 - **Diagnostic:**
 - Severe leukoplakia with:
 - Clinical signs of neoplastic transformation or deep infiltration.
 - Stroboscopic "vibratory silence".
 - **Therapeutic:**
 - Carcinoma-in-situ with microinvasion.
 - Microinvasive carcinoma

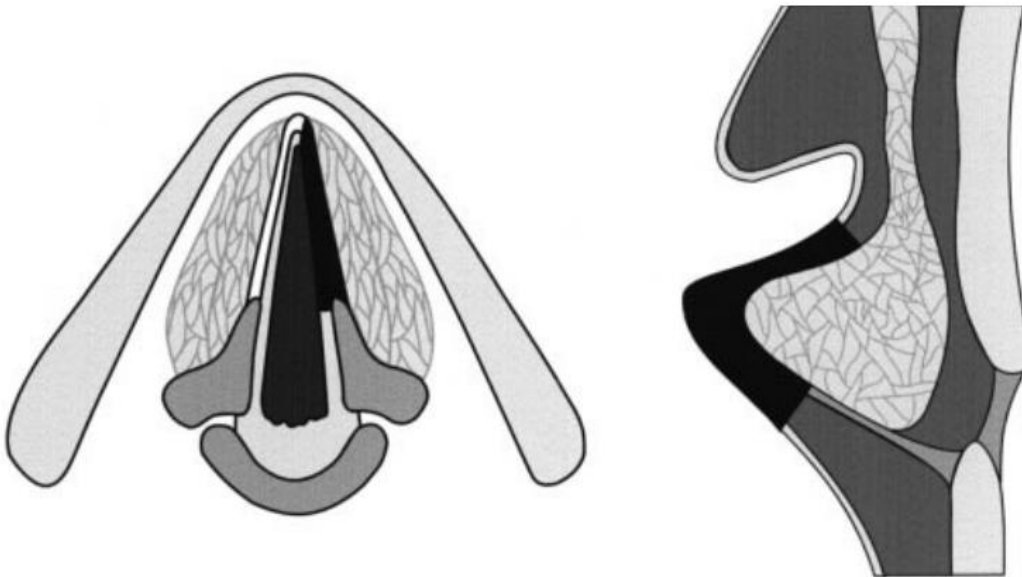


Fig. 2 a, b Subligamental cordectomy (type II)

- **Type III / Trans-muscular Cordectomy:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Part of vocalis muscle
 4. -/+ vestibulectomy if needed for exposure
 - From vocal process to anterior commissure.
 - Indications:
 - **Therapeutic:**
 - Small superficial cancer of mobile vocal fold

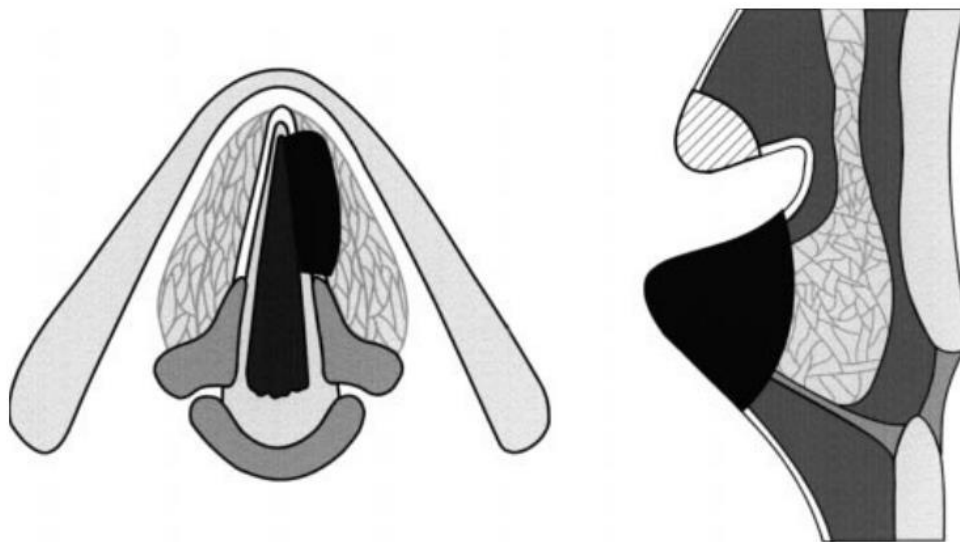


Fig. 3a, b Transmuscular cordectomy (type III). In order to expose the entire vocal fold, partial resection of the ventricular fold may be necessary (hatched area)

- **Type IV / Total Cordectomy:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 - From vocal process to anterior commissure.
 - Indications:
 - **Therapeutic:**
 - Unilateral mobile vocal fold cancer (**T1a**)

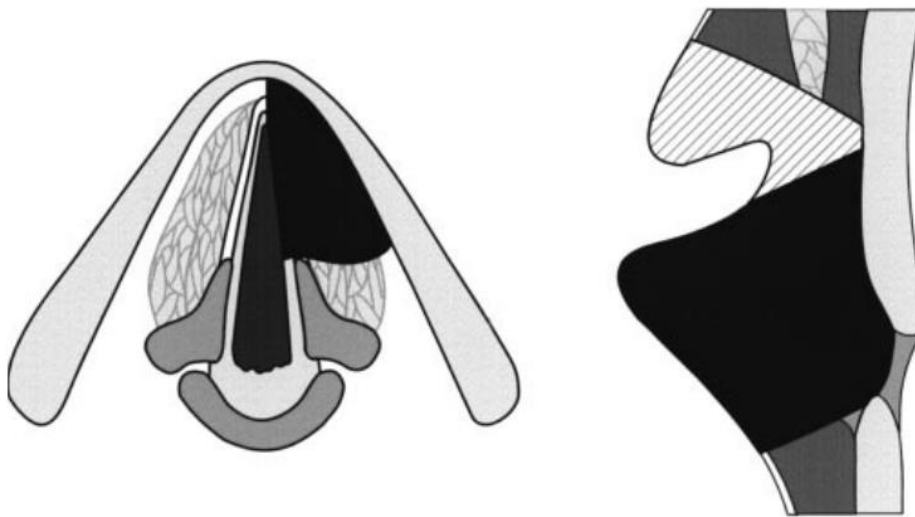


Fig.4 a, b Total or complete cordectomy (type IV). The ipsilateral ventricular fold can be removed partially or totally to ensure complete resection of the vocal fold (hatched area)

- **Type Va / Extended Cordectomy to Contralateral Vocal Fold:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 5. Contralateral vocal fold (partial or complete)
 - From vocal process to anterior commissure.
 - Indications:
 - **Therapeutic:**
 - Bilateral mobile vocal fold cancer (**T1B**) superficially reaching anterior commissure without deep infiltration.

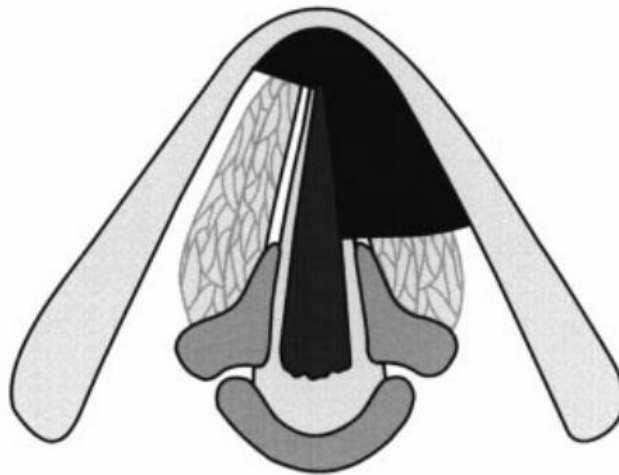


Fig.5 Extended cordectomy encompassing the contralateral vocal fold (type Va). The extent of the resected contralateral vocal fold depends on the extent of the tumor

- **Type Vb / Extended Cordectomy to arytenoid cartilage:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 5. Arytenoid cartilage
 - Indications:
 - **Therapeutic:**
 - Unilateral vocal fold cancer (**T1**) involving the vocal process with mobile arytenoid.



Fig.6 Extended cordectomy encompassing the arytenoid (type Vb)

- **Type Vc / Extended Cordectomy to Ventricular Fold:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 5. Ventricular fold
 - From vocal process to anterior commissure.
 - Indications:
 - **Therapeutic:**
 - Ventricular cancer
 - Transglottic cancers that spread from the vocal fold to the ventricle.

Fig.7 Extended cordectomy encompassing the ventricular fold (type Vc). The inferior resection of the vocal fold is maximum

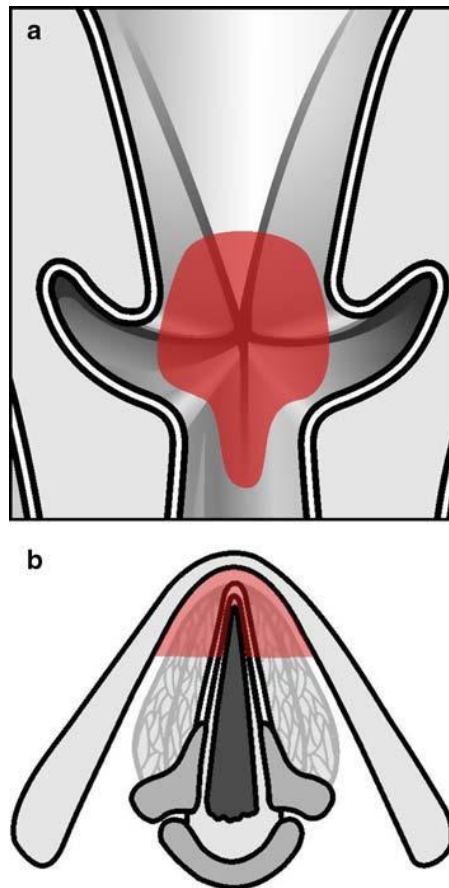


- **Type Vd / Extended Cordectomy to Subglottis:**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 5. Subglottis
 - From vocal process to anterior commissure.
 - Indications:
 - **Therapeutic:**
 - Tumor extended to subglottis (**T2**)

Fig.8 Extended cordectomy encompassing the subglottis to a distance of 1 cm (type Vd)



- **Type VI / Anterior Bilateral Cordectomy (Commissurectomy):**
 - Endoscopic resection of:
 1. Vocal cover
 2. Vocal ligament
 3. Full depth of vocalis muscle
 4. Vestibulectomy
 - At the anterior commissure area.
 - Indications:
 - **Therapeutic:**
 - Tumor originate from anterior commissure without extension to thyroid cartilage.



Tracheostomy

- Tracheostomy is making an opening in the anterior wall of trachea and converting it into a stoma on the skin surface.
- Sometimes, the term **Tracheotomy** has been interchangeably used but the latter actually means opening the trachea (=cutting), which is a step in the tracheostomy operation.

❖ **Functions of Tracheostomy**

- **1. Alternative pathway for breathing:**
 - This circumvents any obstruction in the upper airway from lips to the tracheostome.
- **2. Improves alveolar ventilation:**
 - In cases of respiratory insufficiency, alveolar ventilation is improved by:
 - Decreasing the dead space by 30-50% (normal dead space is 150 ml).
 - Reducing the resistance to airflow.
- **3. Protects the airways**
 - By using **cuffed** tube, tracheobronchial tree is protected against aspiration of:
 - Pharyngeal secretions, as in case of bulbar paralysis or coma.
 - Blood, as in hemorrhage from pharynx, larynx or maxillofacial injuries. With tracheostomy, pharynx and larynx can also be packed to control bleeding.
- **4. Permits removal of tracheobronchial secretions:**
 - When patient is unable to cough as in coma, head injuries, respiratory paralysis; or when cough is painful, as in chest injuries or upper abdominal operations, the tracheobronchial airway can be kept clean of secretions by repeated suction through the tracheostomy.
- **5. Intermittent positive pressure respiration (IPPR):**
 - If IPPR is required beyond 72 hours, tracheostomy is superior to intubation.
- **6. To administer anesthesia:**
 - In cases where endotracheal intubation is **difficult or impossible** as in laryngopharyngeal growths or trismus.

Indications of Tracheostomy

Respiratory obstruction	Retained secretions (Pulmonary toilet)	Respiratory insufficiency (Prolonged ventilatory support)
• <u>Infections:</u> • Acute laryngo-tracheo-bronchitis (Croup). • Acute epiglottitis. • Diphtheria. • Ludwig's angina. • Peritonsillar, retropharyngeal or parapharyngeal abscess. • Tongue abscess. • <u>Trauma:</u> • External injury of larynx and trachea. • Trauma due to endoscopies, especially in infants and children. • Fractures of mandible or maxillofacial injuries. • <u>Neoplasms:</u> • Benign and malignant neoplasms of larynx, pharynx, upper trachea, tongue and thyroid. • <u>Foreign body in the larynx.</u> • <u>Edema of the larynx.</u> • Due to steam, irritant fumes or gases, allergy (angioedema or drug sensitivity), radiation. • <u>Bilateral abductor paralysis.</u> • <u>OSA.</u> • <u>Congenital anomalies:</u> • Laryngeal web, cysts, tracheo-esophageal fistula.	• <u>Inability to cough:</u> • Coma. • Paralysis of respiratory muscle (spinal injuries, polio, Guillain-Barre syndrome, myasthenia gravis) • Spasm of respiratory muscles, tetanus. • <u>Painful cough:</u> • Chest injuries, multiple rib fractures, pneumonia. • <u>Aspiration of pharyngeal secretions:</u> • Bulbar polio, polyneuritis, bilateral laryngeal paralysis.	• <u>Respiratory insufficiency.</u> • Chronic lung conditions emphysema, chronic bronchitis, bronchiectasis, atelectasis

- ❖ Prior to the early 1960s, short-term tracheostomies were mainly used to treat airway obstructions due to acute infections (epiglottitis, pharyngeal abscesses or laryngotracheo-bronchitis) or trauma (foreign bodies).
- ❖ The duration of ET intubation before a tracheostomy is recommended and varies widely and must be decided on a case-by-case basis, depending on the nature and prognosis of the primary disease, as well as the presence of comorbidities.
- ❖ **Types of Tracheostomy**
 - Emergency tracheostomy
 - Elective tracheostomy
 - Percutaneous dilatational tracheostomy
 - Mini tracheostomy (cricothyroidotomy)

Emergency Tracheostomy	Elective Tracheostomy
• Indicated in patients with <u>complete airway obstruction</u> and there is an urgent need to establish the airway. • Done under Local anesthesia when endotracheal intubation is not possible or feasible in such cases.	• Done under General anesthesia and the patient is already intubated with endotracheal tube. • All operative surgical facilities are available. • Can be temporary or permanent (depends on the reason). <u>It is of two types:</u> • <u>Therapeutic:</u> ○ Respiratory obstruction. ○ Remove tracheobronchial secretions. ○ Assisted ventilation. • <u>Prophylactic:</u> ○ Guard against anticipated <u>respiratory obstruction</u> or <u>aspiration</u> of blood or pharyngeal secretions such as "extensive surgery of tongue, floor of mouth, mandibular resection or laryngofissure".

- Tracheostomy has also been divided into high, mid or low:

1. High Tracheostomy:

- Done at the 1st tracheal ring, above the level of thyroid isthmus (isthmus lies against II, III and IV tracheal rings).
- Tracheostomy at this site can cause perichondritis of the cricoid cartilage and subglottic stenosis and is always avoided.
- Indications of High Tracheostomy:
 - Patients with advanced laryngeal cancer presenting with airway obstruction because total laryngectomy will be done including the stoma site a fresh stoma will be made in a clean area lower down.
 - Pediatric patients with Subglottic stenosis and expected to need CTR because the stoma site will be removed with stenotic segment ultimately.

2. Mid Tracheostomy (Standard):

- Preferred site.
- Done through the 2nd or 3rd tracheal rings.
- Entail division of the thyroid isthmus or its retraction upwards or downwards to expose this part of trachea.
- Not preferred site for:
 - Patients with advanced laryngeal cancer presenting with airway obstruction because the increased risk of stoma recurrence.
 - Pediatric patients with Subglottic stenosis and expected to need CTR because it will require resecting more tracheal segments to include the stoma site.

3. Low Tracheostomy:

- Done through 6th or 7th tracheal ring.
- Trachea is deep at this level and close to several large vessels
- Also there are difficulties with tracheostomy tube which impinges on suprasternal notch.
- Indications of Low Tracheostomy:
 - Intrathoracic tracheal stenosis.
 - Patients with advanced laryngeal cancer presenting with airway obstruction.
 - Pediatric patients with Subglottic stenosis and expected to need CTR.

Table 21.2 Location of tracheotomy

• Tracheotomy for ventilatory support or aspiration (normal airway) – Third or fourth tracheal ring
• Tracheotomy for incipient laryngotracheal stenosis – First tracheal ring - or – Sixth or seventh tracheal ring
• Tracheotomy for incipient tracheal stenosis – Through tracheal stenosis
• Tracheotomy for intrathoracic tracheal stenosis – Sixth or seventh tracheal ring with long cannula to stent the distal stenosis
• Tracheotomy for recurrent stenosis at former tracheostomy site – Through former tracheostomy

Fig. 14.18 Correct placement of tracheostomy with impending laryngotracheal stenosis: (a) Tracheostoma situated immediately below the cricoid ring to maximally preserve normal trachea. (b) Tracheostoma situated at the sixth–seventh or eighth tracheal ring to spare sufficient length of normal trachea between the stenosis and the tracheostomy

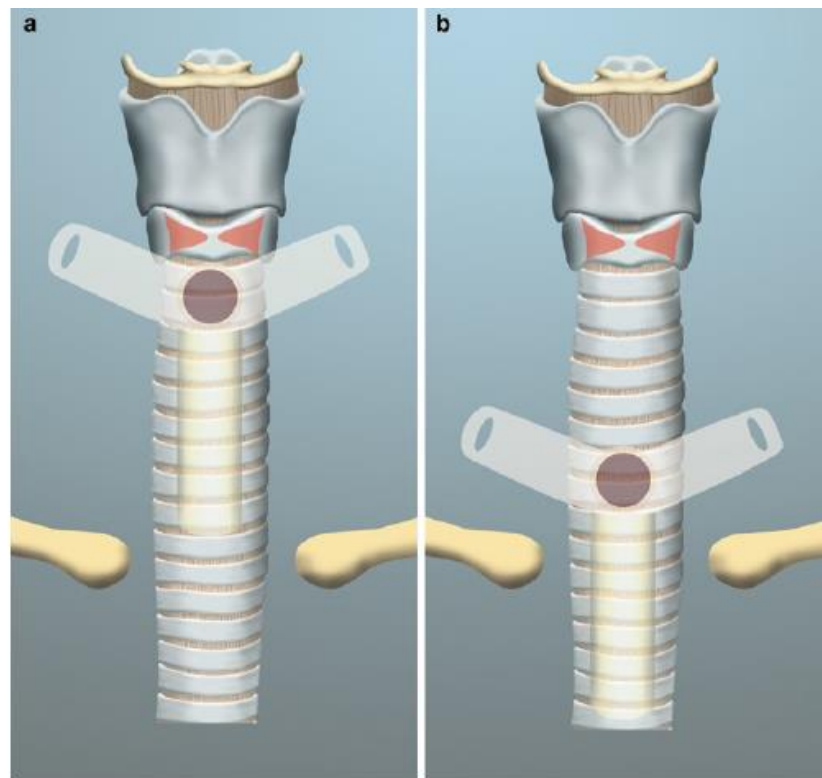


Fig. 14.19 Correct placement of tracheostomy when subglottic stenosis is associated with tracheal stenosis: (a) Cervical cuff or post-tracheostomy stenosis: the new tracheostomy must be placed in the stenotic segment. (b) Substernal cuff or cannula tip stenosis: the new tracheostomy must be placed just above the stenosis, which is stented by the tracheostomy cannula

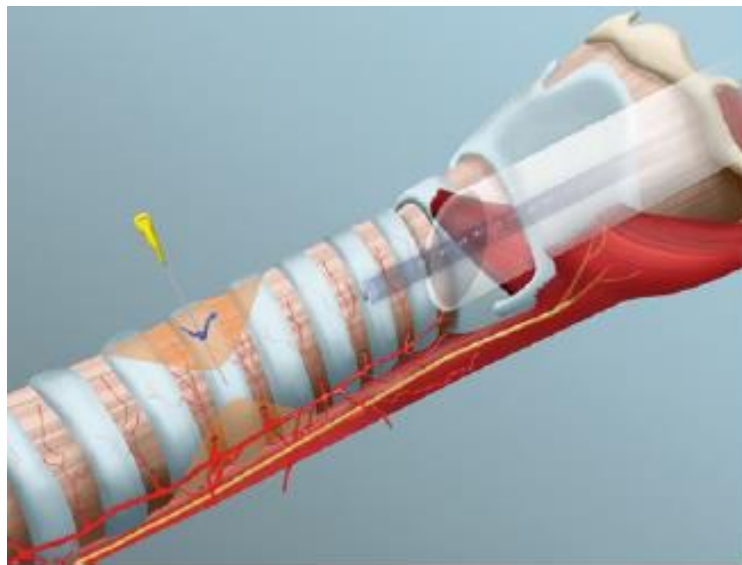
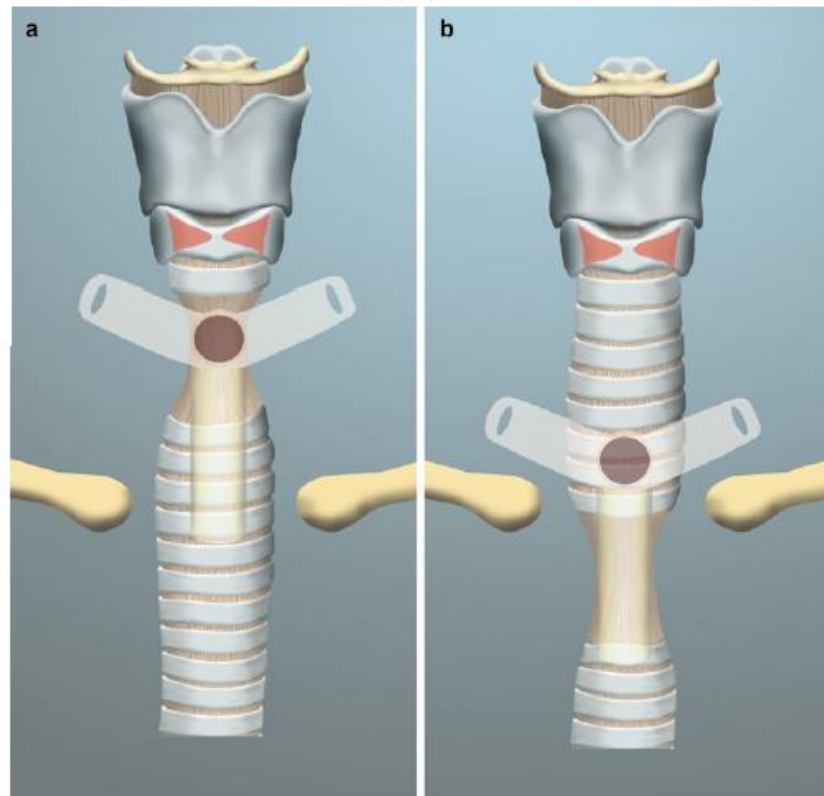


Fig. 22.2 Location of a tracheal stenosis not clearly identifiable externally: A fine needle is inserted through the middle of the stenosis under visual bronchoscopic guidance, and a stitch is temporarily placed as a landmark

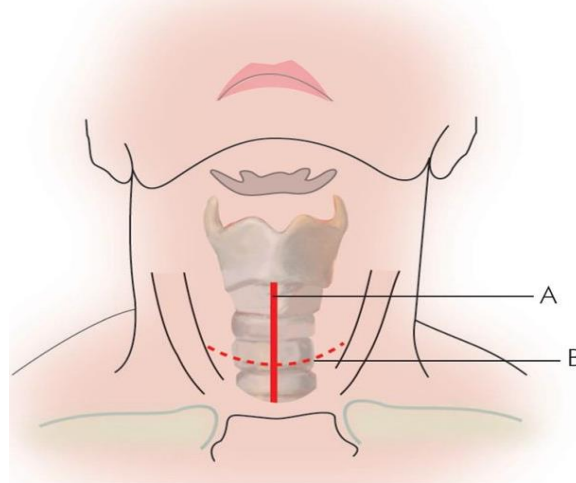
❖ Technique of Tracheotomy

- ETT or rigid ventilating bronchoscope should be done before tracheostomy.
- Position: supine with neck is extended "brings the trachea forward".

➤ Steps of Operation

1. Incision:

- Transverse incision, 5 cm long, made 2 fingers' breadth above the sternal notch can be used in **elective procedures** "cosmetically better scar".
- Vertical incision is made in the midline of neck, extending from cricoid cartilage to just above the sternal notch. This is the most favored incision and can be used in **emergency and elective procedures**. It gives rapid access with minimum of bleeding and tissue dissection.



Dhingra: Diseases of Ear Nose and Throat, 5th Edition
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Figure 63.1 Skin incisions in tracheostomy. (A) Vertical midline incision. (B) Transverse incision.

2. After incision, tissues are dissected in the midline. Dilated veins are either displaced or ligated "most of the dissection is performed bluntly with S-shaped retractors, great care is taken to stay in the midline".

3. Strap muscles are separated in the midline and retracted laterally.

4. Thyroid isthmus is displaced upwards or divided between the clamps, and suture-ligated by running sutures bilaterally to obtain perfect haemostasis, and the lower thyroid veins must be tied, if they cannot be displaced laterally. When an innominate artery is located aberrantly high in the neck, it can be protected by an inferiorly pedicled sternohyoid muscular flap, and by placing the stoma higher in the neck.

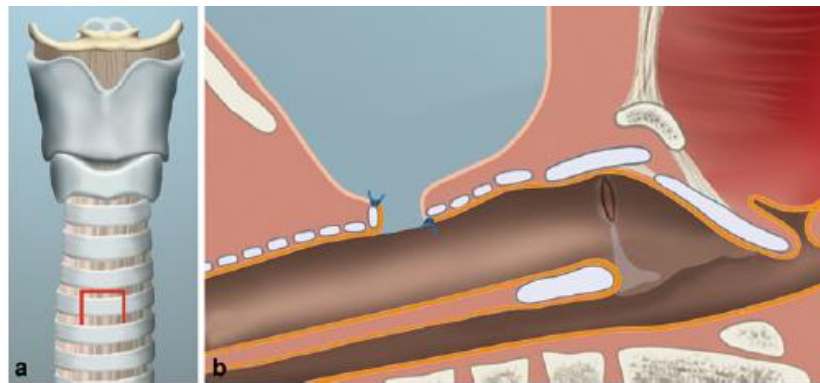
5. A few drops of 4% lignocaine are injected into the trachea to suppress the cough when trachea is incised (if tracheostomy done under LA).

6. Trachea is fixed with a hook and opened with incision in the region of 3rd and 4th or 3rd and 2nd rings. This is then converted into a circular opening. The first tracheal ring is never divided as perichondritis of cricoid cartilage with stenosis can result.

➤ Bjork flap:

- Inferiorly based tracheal incision.
- Facilitate reinsertion of the tracheostomy tube while it is being changed or during accidental extubation.
- Prevents anterior accidental subcutaneous dislodgment of the cannula, prevents air and mucus leaks around the stoma, possibly leading to subsequent subcutaneous emphysema or pneumomediastinum.
- Risk of Peristomal cellulitis.
- High chance of tracheo-cutaneous fistula post decannulation.
- Not preferred in pediatric tracheostomy.

Fig. 21.1 Björk flap for paediatric tracheostomy: (a) Only one single ring is incised. The width of the flap should not be larger than its length to obtain a square opening. (b) The neck-skin is sutured to the Björk flap inferiorly and all around the tracheal window. The cannula calibrates the opening to its own size



"The debate is still ongoing as to whether a vertical or horizontal tracheal incision, with or without flap, should be made. In a study including 93 tracheostomised children with various tracheal incisions, Mac Rae et al. reported no difference in results or complications when comparing the different types of tracheal incisions. The basic principle consists of incising as few tracheal rings as possible"

7. The ETT is withdrawn to the level of the tracheostomy, Tracheostomy tube of appropriate size is inserted and connected to the anesthesia tube in order to ensure adequate bilateral lung ventilation, The flanges of the cannula are fixed around the patient's neck.

8. Skin incision should not be sutured or packed tightly as it may lead to development of subcutaneous emphysema "secure stoma in the event of accidental extubation following surgery".

9. Gauze dressing is placed between the skin and flange of the tube around the stoma.

Box 21.1 Surgical Highlights for Tracheotomy

- Select a proper site for tracheotomy tube placement depending on the indication.
- Regardless of the technique selected, perform a limited incision of the trachea and suture the skin around the edges of the tracheal opening.
- A 'one ring' Björk flap prevents anterior accidental subcutaneous dislodgment of the cannula.
- The final tracheostoma must be calibrated to the size of the cannula rather than that of the tracheal incision.
- Always check adequate positioning of the tracheostomy tube with respect to the distal trachea and carina.
- The first tube change is made 1 week after the operation in a fully equipped endoscopy suite.
- Tracheo-innominate artery fistula and tracheo-oesophageal fistula are extremely rare in children, but must be recognised and treated quickly.
- Tracheostoma closure should be performed surgically after a difficult laryngotracheal reconstruction in order to optimise the tracheal calibre.
- When closing a tracheostoma surgically, always place stitches in the cranio-caudal axis to restore the Roman vault of the trachea.
- Diagnose any localised malacia at the stoma site prior to tracheostomy closure.
- A malacic tracheostoma requires either a resection and anastomosis or a tracheoplasty with ACCG.

- ✓ The position of the distal tip of the tracheostomy tube, which should rest at least **two to three** rings above the **carina**, is checked using a flexible fibre-optic bronchoscope.
- ✓ A chest X-ray is taken during the immediate postoperative period in order to rule out a pneumomediastinum or pneumothorax.
- ✓ Antibiotics are administered for 48 h following surgery, and a gentamycin-corticosteroid ointment (Diprogenta®) is applied around the stoma twice daily to protect the skin from soaking in mucus secretions.

❖ Tracheostomy in Infants and Children

- Prolonged intubation generally preferred over tracheotomy in **neonates up to 6 months** (high risk of subglottic stenosis).
- May consider performing tracheotomy with a rigid bronchoscope to provide rigidity and facilitate dissection
- Consider endoscopy every 3 months for pediatric tracheotomy to evaluate for stenosis.
- Important conditions requiring tracheostomy in this age group.

Table 63-2. Common indications of tracheostomy in infants and children

Infants below 1 year (mostly congenital lesions) • Subglottic haemangioma • Subglottic stenosis • Laryngeal cyst • Glottic web • Bilateral vocal cord paralysis
Children (mostly inflammatory or traumatic lesions) • Acute laryngo-tracheo-bronchitis • Epiglottitis • Diphtheria • Laryngeal oedema (chemical/thermal injury) • External laryngeal trauma • Prolonged intubation • Juvenile laryngeal papillomatosis

- Great care and caution is required when doing tracheostomy in infants and children:
 1. Trachea of infants and children is **soft and compressible** and its identification may become difficult and the surgeon may easily displace it and go deep or lateral to it injuring **recurrent laryngeal nerve** or even the **carotid**.
 2. During positioning, **do not extend** the neck too much as this **pulls structures from chest into the neck** and thus injury may occur to **pleura, innominate vessels and thymus or the tracheostomy opening may be made too low near suprasternal notch**.
 3. Before incising trachea, silk sutures (**stay sutures**) are placed in the trachea, on either side of midline.
 4. Tracheal lumen is small, **do not insert knife too deep**; it will injure posterior tracheal wall or even oesophagus causing tracheo-oesophageal fistula.
 5. Trachea is simply incised, without excising a circular piece of tracheal wall.
 6. Avoid infolding of anterior tracheal wall when inserting the tracheostomy tube.
 7. Selection of tube is important. It should be of **proper diameter, length and curvature**. A long tube impinges on the carina or right bronchus. With high curvature, lower end of tube impinges on anterior tracheal wall while upper part compresses the tracheal rings or cricoid.
 8. Use soft silastic or portex tube. Metallic tubes cause more trauma.
 9. Take a post-operative X-ray of the neck and chest to ascertain the position of the tracheostomy tube.

Procedures for Immediate Airway Management

- When airway obstruction is so marked as to allow no time to do an orderly tracheostomy, following measures are taken:

1. Jaw thrust:

- Lifting the jaw forward and extending the neck improves the airway by displacing the soft tissues.

2. Oropharyngeal airway:

- Ventilation can be carried out by face mask.
- Ambo bag can be used for inflation of air or oxygen.

3. Nasopharyngeal airway (trumpet):

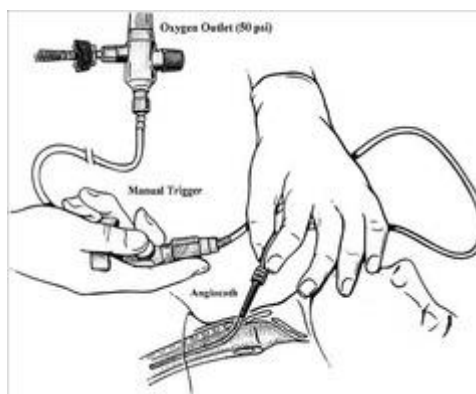
- It is inserted **transnasally** into the **posterior hypopharynx** and relieves soft tissue obstruction caused by the tongue and pharynx.
- It is better tolerated than oropharyngeal airway in awake patients.

4. Laryngeal mask airway:

- Fits over the laryngeal inlet.
- Oxygen can be delivered directly into the trachea.
- Though most commonly used for non-emergent airway control, it can be used as an alternative if standard mask ventilation is inadequate and intubation unsuccessful

5. Trans-tracheal jet ventilation:

- It is an invasive procedure.
- An intravenous catheter of 12 or 14 gauge with a syringe attached is inserted into the cricothyroid membrane and directed caudally.
- Once intraluminal placement is confirmed by aspiration, needle is withdrawn leaving the catheter in position and jet ventilation started.
- Expiration of air should be insured otherwise pulmonary barotrauma with pneumothorax, pneumomediastinum and surgical emphysema can result.

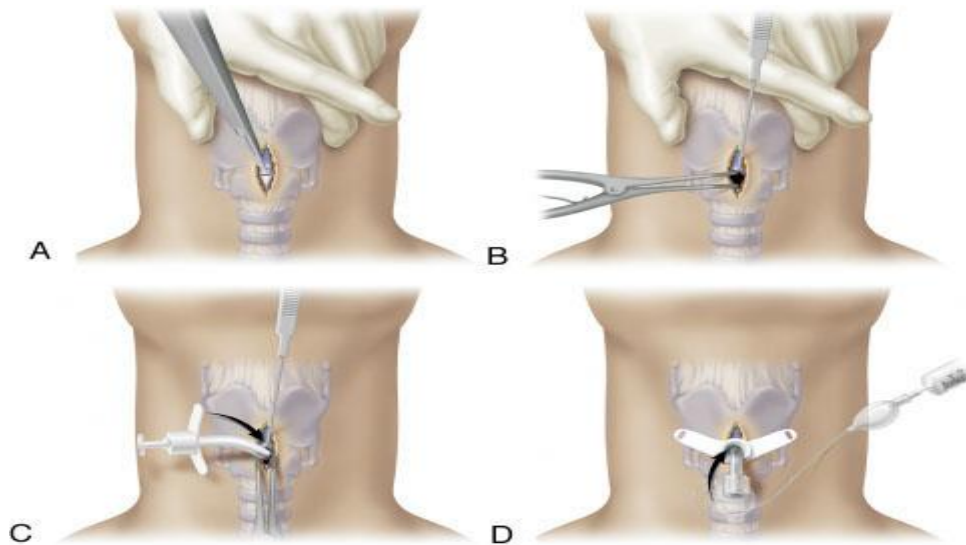


6. Endotracheal intubation:

- This is the most rapid method.
- This helps to avoid a hurried tracheostomy in which complication rate is higher.
- After intubation, an orderly tracheostomy can be performed.

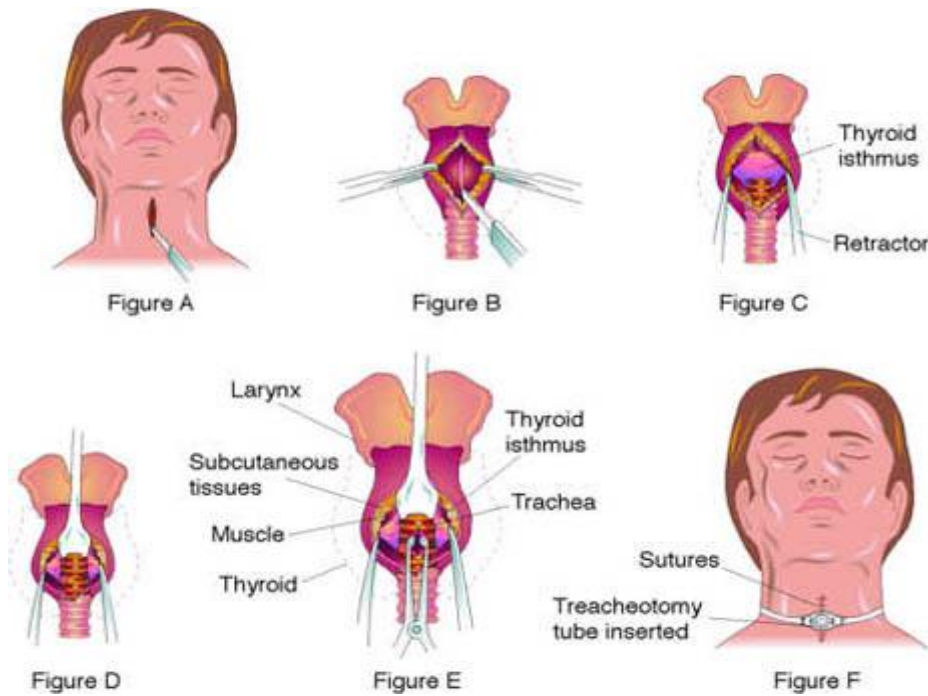
7. Cricothyrotomy (Mini tracheostomy):

- This is a procedure for opening the airway through the cricothyroid membrane.
- Patient's head and neck is extended.
- Lower border of thyroid cartilage and cricoid ring are identified.
- Skin in this area is incised **vertically** and then cricothyroid membrane cut with a **transverse incision**.
- This space can be kept open with a small tracheostomy tube or by inserting the handle of knife and turning it at right angles if tube is not available.
- It is essential to perform an orderly tracheostomy as soon as possible because **perichondritis**, **subglottic oedema** and **laryngeal stenosis** can follow prolonged laryngotomy.



8. Emergency Tracheostomy (Slash):

- Patient's neck is extended.
- Trachea identified and fixed between surgeon's left thumb and index finger.
- Vertical skin incision is made from **lower border of thyroid to suprasternal notch cutting through skin and subcutaneous tissues**.
- Lower border of cricoid cartilage is identified and a transverse incision made in pretracheal fascia.
- Thyroid isthmus dissected down to expose upper three tracheal rings.
- Vertical tracheal incision is made in 2nd and 3rd rings, opened with a haemostat and the tube inserted.
- Bleeding can be controlled by packing with gauze.
- Emergency tracheostomy on a struggling patient with inadequate lighting, suction and instruments is fraught with many complications.
- If possible, an endotracheal tube should be put for a more orderly procedure to be carried out.

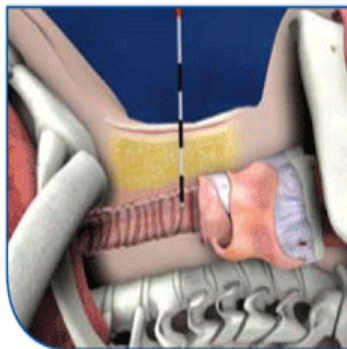


Percutaneous Dilatational Tracheostomy:

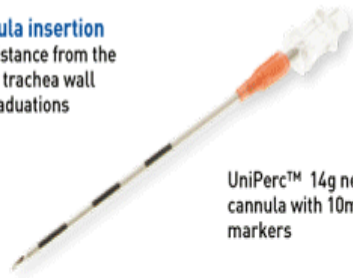
- This type of tracheostomy is done in ICU where patient is already intubated and being monitored.
- It is done under sedation.
- Neck is extended with a pad under the shoulders.
- Neck is prepared and draped and 1.5-2 cm incision is made 2 cm below the lower border of cricoid.
- Trachea is exposed by dissection and palpation.
- Thyroid isthmus is pushed down.
- Now a small caliber flexible bronchoscope, to which a camera has been attached, is passed through the endotracheal tube to monitor the passage of the needle, guide wire and dilator/s.
- It is important to enter the trachea in the midline and avoid any lateral entry.
- Entry into the trachea is made between second and third rings.
- After dilatation tracheostomy tube is inserted.

<https://www.youtube.com/watch?v=6TVOSb6sqL8>

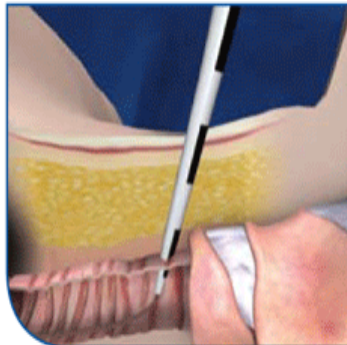
- Advantages of the procedure include:
 - No need to transport the patient to operation theatre
 - Avoiding OR expenses.
 - Avoiding ICU nosocomial infections to be carried to OR.
 - Earlier discharge of patient from ICU.



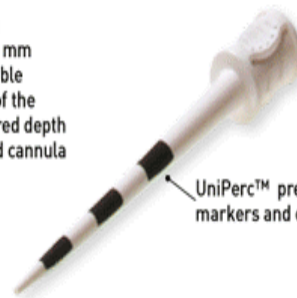
Needle and cannula insertion
 – establishes the distance from the skin to the anterior trachea wall using the 10mm graduations



UniPerc™ 14g needle and cannula with 10mm graduated markers



Pre-dilator insertion
 – the corresponding 10 mm graduated markers enable a confident placement of the pre-dilator to the required depth measured at needle and cannula insertion stage



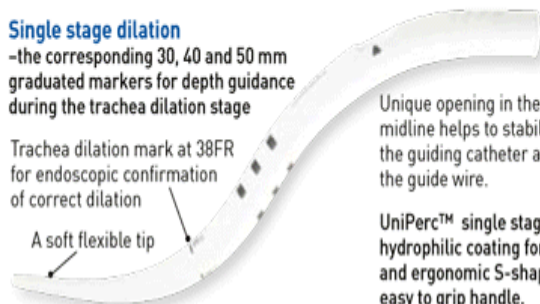
UniPerc™ pre-dilator with 10mm markers and enhanced grip



Single stage dilation
 –the corresponding 30, 40 and 50 mm graduated markers for depth guidance during the trachea dilation stage

Trachea dilation mark at 38FR for endoscopic confirmation of correct dilation

A soft flexible tip



Unique opening in the midline helps to stabilise the guiding catheter and the guide wire.



UniPerc™ single stage dilator – with hydrophilic coating for smooth insertion and ergonomic S-shape design with easy to grip handle.

❖ **Post-operative Care:**

1. Constant supervision

- After tracheostomy, constant supervision of the patient for bleeding, displacement or blocking of tube and removal of secretions, tight tracheotomy ties, is essential.
- Accidental decannulation occur despite these preventive measures, parents and nurses must be instructed to stretch the skin around the stoma to prevent inward collapse during inspiration, thus alleviating respiratory distress.

2. Suction:

- Tracheotomy tubes disrupt ciliary function, decrease subglottal pressure required for an adequate cough, and increase risk of microaspiration.
- Requires regular suctioning of the tracheal airway, especially for the first few days.
- Depending on the amount of secretion, suction may be required every half an hour or so; use sterile catheters.
- Suction injuries to tracheal mucosa should be avoided" **trauma to the carina and bronchi can lead to granulation Tissue formation & bleeding**".
- Suction should be limited to the tracheostomy tube length in order to avoid severe coughing spells and trauma to the distal airway, this is done by applying suction to the catheter only when withdrawing it.

3. Prevention of crusting, mucous plug and tracheitis:

- This is achieved by proper humidification.
- If crusting occurs, a few drops of normal or hypotonic saline or Ringer's lactate are instilled into the trachea every 2-3 hours to loosen crusts.
- A mucolytic agent such as acetylcysteine solution, can be instilled to liquify tenacious secretions or to loosen the crusts.

4. Care of tracheostomy tube:

- Inner cannula should be removed and cleaned as and when indicated for the first 3 days.
- Outer tube, unless blocked or displaced, should not be removed for 1 week to allow a track to be formed when tube placement will become easy.
- If cuffed tube is used, it should be periodically deflated to prevent pressure necrosis or dilatation of trachea (subglottic stenosis, tracheal-innominate artery erosion, tracheomalacia)
- Cuff pressure should be less than capillary pressure (**<25** cm H₂O) to prevent mucosal necrosis and development of subglottic stenosis.
- The frequency of tracheostomy tube changes varies on an individual basis. Initially, once per week is sufficient, provided that the airway, stoma and peritracheostomal skin are not infected or inflamed.

5. Skin Care:

- Cuff dressings may be considered to prevent skin breakdown.

6. Feeding:

- No solid food ingestion when cuff is inflated; capping tracheotomy tube facilitates swallowing.

❖ **Complications:**

Intraoperative	Early Postoperative	Late Postoperative
<u>1. Bleeding:</u> ○ From injury to major vessels or thyroid tissue. <u>2. Apnea:</u> ○ This follows opening of trachea in a patient who had prolonged respiratory obstruction. ○ This is due to sudden washing out of CO₂ which was acting as a respiratory stimulus. ○ Treatment is to administer 5% CO₂ in oxygen or assisted ventilation. <u>3. Pneumothorax:</u> ○ Due to injury to apical pleura. <u>4. Injury to recurrent laryngeal nerves.</u> <u>5. Aspiration of blood.</u> <u>6. Injury to esophagus:</u> ○ Occurs with tip of knife while incising the trachea and may result in tracheoesophageal fistula.	<u>1. Bleeding:</u> ○ Reactionary or secondary. <u>2. Tube displacement.</u> <u>3. Tube Blockage.</u> <u>4. Postoperative pulmonary edema:</u> ○ From release of prolonged obstruction. ○ Treated with positive pressure ventilation. <u>5. Subcutaneous emphysema.</u> <u>6. Tracheitis and tracheobronchitis with crusting in trachea.</u> <u>7. Atelectasis and lung abscess.</u> <u>8. Local wound infection and granulations.</u>	<u>1. Bleeding:</u> ○ From erosion of major vessel (Tracheoinnominate artery Fistula). <u>2. Subglottic stenosis:</u> ○ From perichondritis of cricoid cartilage. <u>3. Tracheal stenosis:</u> ○ From tracheal ulceration and infection. <u>4. Tracheo-esophageal fistula:</u> ○ Due to prolonged use of cuffed tube or erosion of trachea by the tip of tracheostomy tube. <u>5. Failed decannulation:</u> ○ Seen commonly in infants and children. ○ Caused by Tracheomalacia, suprastomal collapse or granulation tissue. <u>6. Persistent tracheocutaneous fistula.</u> <u>7. Tracheostomy scar and Keloid formation.</u>

Table 21.3 Complications of paediatric tracheotomy

• Early complications of tracheotomy – Haemorrhage – Subcutaneous emphysema, pneumomediastinum and pneumothorax – Local infection – Accidental decannulation, tubal obstruction
• Late local complications of tracheostomy – Suprastomal collapse and granuloma – Tip of cannula granuloma or stenosis – Granulation tissue along stoma tract – Tracheal innominate artery fistula (rare) – Tracheo-oesophageal fistula (rare) – Lower airway infection, pneumonia – Accidental decannulation, tubal obstruction
• Post-decannulation complications – Tracheomalacia at tracheostomy site – A-frame tracheal deformity – Tracheal stenosis

❖ **Early Complications:**

- Most often caused by technical errors during the surgery, or poor tracheostomy tube care during the early postoperative period.
- Acute hemorrhage may be prevented by attention to hemostasis throughout the procedure.
- Most common early complications of pediatric tracheostomy is tube issues (blockage and extrusion).

❖ Late Complications:

○ Granulation tissue:

- Severe granulation tissue formation along the stoma tract is generally due to insufficient tracheostomy care and poor hygiene at home.
- The exuberant granulation tissue is often infected, which necessitates a short hospital stay to remove it under general anesthesia using bipolar coagulation and biopsy forceps.
- Aggressive local treatment with a gentamycin-corticosteroid ointment (Diprogenta®) along with frequent tracheostomy tube changes is required until the stoma tract is fully epithelialized.
- Any lower airway infection must be treated with systemic antibiotics based on bacteriological culture and sensitivity test results.

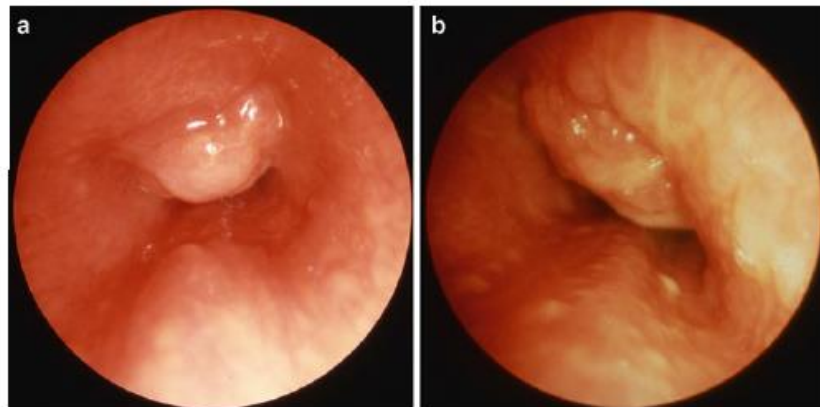
▪ Suprastomal granulation tissue:

- Most frequently seen in very young children with long-term tracheostomy tubes.
- **Remain asymptomatic until decannulation**

▪ Cannula tip granulation tissue:

- Lead to symptoms such as hemoptysis and airway obstruction.
- Potentially life-threatening complication.
- Treated with rigid bronchoscopy in order to reopen the distal airway then non-cuffed endotracheal tube must be used to bypass the stenotic segment during the healing phase.

Fig. 21.2 Complications of tracheostomy: (a) Suprastomal collapse and granuloma. (b) Tip of cannula granuloma creating an asymmetrical distal tracheal stenosis



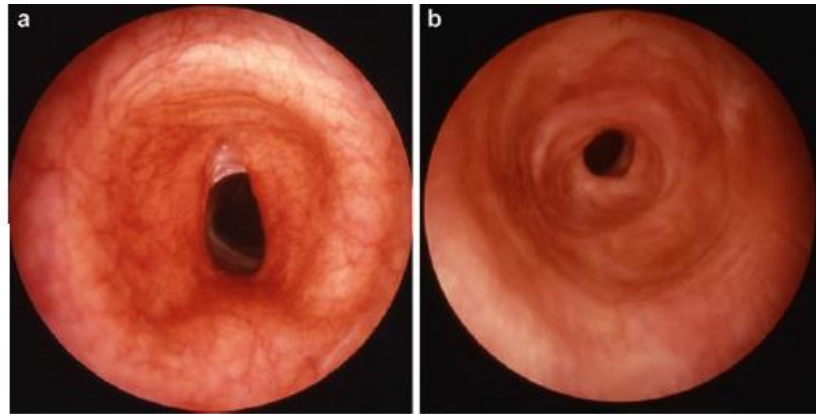


Fig. 21.3 Sequelae of tracheostomy: (a) A-frame deformity and malacia at the former tracheostoma site. (b) Distal annular cicatricial stenosis resulting from circumferential tip of cannula lesions

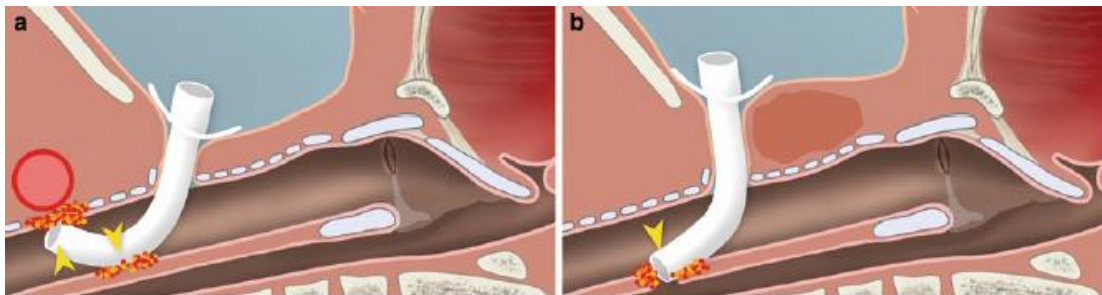
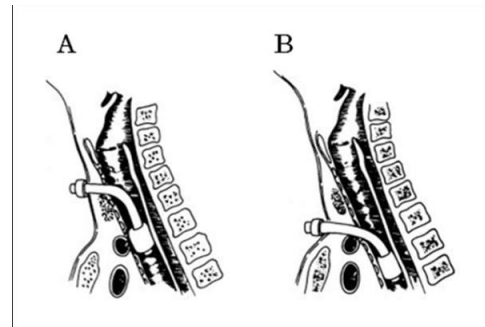


Fig. 21.4 Extremely rare complications of tracheostomy: (a) Ill-placed cannula impinging on the anterior tracheal wall, with possible tracheo-innominate artery fistula. (b) Ill-placed cannula impinging on the posterior tracheal wall, due to a cervical mass, with possible tracheo-oesophageal fistula

○ Tracheo-innominate artery fistula:

- Life-threatening complication.
- Occurs in < 1% of patients.
- Most commonly within 3-4 weeks postoperatively.
- Risk factors:

1. Low Placement
2. High pressure Cuffed tube
3. Excessive tube movement
4. Excessive Hyperextension
5. Malnutrition
6. Radiation
7. DM
8. Steroid
9. Infection



○ Clinical signs of possible TIF:

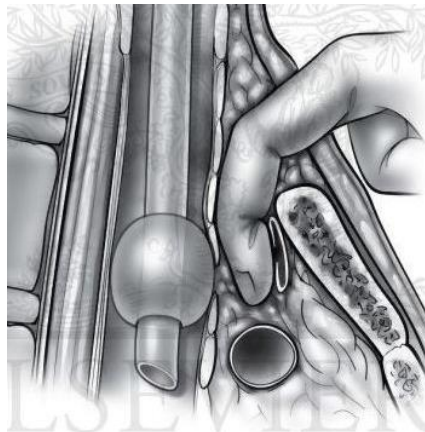
1. Sentinel bleeding
2. Pulsating tracheostomy tube

○ Diagnosis of suspected TIF:

- Bedside fiberoptic exam after slowly withdraw the tube:
 - Bleeding granulation tissue at this level is indicative of an imminent TIF.
- CT Angiography/Angiogram.
- DLB.



- Emergency Treatment of TIF:
 - Over-inflate the cuff of tracheostomy tube.
 - Remove tracheostomy tube and do ET intubation.
 - Finger pressure against sternum while patient is intubated till reaching the OR for definitive management.



- Definitive treatment of TIF:
 - **Median sternotomy + Ligation of innominate artery:**
 - Performed by cardiothoracic and vascular surgery teams.
 - The anterior tracheal defect is closed and covered with an inferiorly pedicled sternohyoid or intercostal muscle.
 - Other options:
 - Direct repair
 - Bypass
 - **Endovascular Stenting**

- Tracheo-esophageal fistula:
 - At times, a cuffed cannula in a patient requiring ventilatory support may squeeze the posterior membranous trachea against an inappropriately hard or large nasogastric tube, thereby inducing pressure necrosis, leading to a tracheo-esophageal fistula.
 - The use of low pressure, high-volume cuffed tracheostomy tubes and soft nasogastric tubes has reduced the frequency of such complications.
 - Inexplicable increase in tracheal secretions or the occurrence of lung infection should suggest the development of TOF.
- Tracheo-cutaneous fistula:
 - Persistent stoma after 6 months of decanulation.
 - Occurs in up-to 30% of patients post decanulation.
 - Risk Factors:
 1. Long duration of Trach > 1 year
 2. Bjork flap
 3. Obesity
 4. Proximal AW obstruction
 5. Malnutrition
 6. Radiation
 7. Infection
 - Treatment:
 - **DLB:**
 - To rule out suprastomal obstruction (collapse or granulation tissue).
 - **Primary Closure:**
 - Elliptical incision.
 - Resection of fistula track.
 - Close trachea defect.
 - Insert Drain.
 - Strap muscle flap.

❖ **Tracheostomy Tubes:**

- Main components of a tracheostomy tube are universal across the range of designs.
- Arc shaped tube shaft.
- Designed as either a single cannula or dual cannula (inner and outer) tracheostomy tube.
- It comes either with or without a cuff.
- For ease of insertion it is supplied with an obturator.
- The neck flange helps secure the tracheostomy tube to the skin of the neck and stabilize its position.



✓ **Tracheostomy tube dimensions:**

- Length and diameter of trachea are roughly proportional to the size of the individual.
- A tracheostomy tube should be selected according to the outer diameter, the inner diameter and the length of the tube, rather than the manufacturer's "size", which is not standardized between models nor manufacturers. i.e. a "size 8" from one manufacturer is likely to have different dimensions to a "size 8" from another.
- Outer diameter:
 - Should be about $\frac{2}{3}$ to $\frac{3}{4}$ of the tracheal diameter.
 - A tube should be no wider than necessary in order to minimize trauma to the tracheal wall and long term complications.
- Inner diameter:
 - Influence the work of breathing in a spontaneously breathing patient and in turn the course of weaning from the ventilator.
- Tube length:
 - The ideal length is such that the tube tip lies a few centimeters above the carina.
 - A tube which is too short carries a higher risk of accidental decannulation or partial airway obstruction due to poor positioning.
 - A tube which is too long may impinge on the carina leading to discomfort and coughing.

- Single cannula Tracheostomy tube:
 - It has only one un-fenestrated cannula.
 - Pediatric tracheostomy tubes usually comes with single cannula.
 - Higher risk of obstruction than double cannula tubes.
 - Requires regular care with suction and tube change to avoid the risk of obstruction from crusts.
- Double cannula Tracheostomy tube:
 - Safer than single cannula tubes as the inner cannula may be removed quickly in the event of obstruction.
 - Preferred for **stable patients with long-standing tracheostomy tube.**
 - Fenestrated tubes may be used to facilitate speech.
- Fenestrated tracheostomy tube:
 - Double cannula tracheostomy tube.
 - Outer cannula is having fenestrae to allow exhaled airflow to reach vocal cords and facilitate speech.
 - It is supplied with 2 types of inner cannulae:
 - One with a fenestration to promote air flow and speech.
 - One without a fenestration for suctioning.
 - Indications:
 - **Stable patients with long-standing tracheostomy tube to facilitate speech.**
 - **Patients undergoing weaning from tracheostomy,** as they facilitate speech and reduce the work of breathing in comparison to non-fenestrated tubes.
 - Contraindications:
 - Newly-formed stomas due to the risk of surgical emphysema during positive pressure ventilation.
- ✓ Un-cuffed tracheostomy tubes:
 - Reduces the risk of tracheal damage caused by inflation of the cuff.
 - Aid swallowing and communication with the concomitant use of a speaking valve.
 - Indications:
 - **Stable patients with long-standing tracheostomy who can protect their own airway, have an adequate cough reflex and can manage their own secretions.**
 - Frequently used for patients being cared for in the community or a hospital ward (who doesn't require positive ventilator support).
 - A double cannula un-cuffed tube is preferred for safety and comfort as removal of the inner cannula for cleaning is not traumatic to the patient.

- Cuffed tracheostomy tubes:
 - Provide an airtight seal to prevent escape of air around the tube.
 - Advantages:
 - Facilitates positive pressure ventilation and prevent air leak around the tube.
 - Reduce the risk of aspiration.
 - Indications:
 - Patients requiring positive pressure ventilation (ICU setting).
 - Patients with high risk of aspiration.
 - Cuff pressure should NOT exceed 25 cmH₂O to minimize the risks of both tracheal wall injury and aspiration (From obstruction of the esophagus located posteriorly).
 - Common causes of excessive cuff pressure include:
 - Overinflated cuff
 - Undersized tracheostomy tube
 - Poor tube positioning
 - Reduced lung compliance.
- Standard length tracheostomy tubes:
 - Designed to accommodate patients with normal airway anatomy.
 - However, the length and angulation of standard design tracheostomy tubes may be too short and unsuitable for some critical care patients, risking complications.
- Long tracheostomy tubes:
 - Available with a fixed or adjustable flange (fixed or adjustable length).
 - Fixed longer length tubes may be elongated in either the proximal portion (between the stoma and the trachea) or the distal portion of the tube (within the trachea).
 - Extra proximal length is needed for patients with deep set tracheas (large neck due to obesity, goiter, neck mass)
 - Extra distal length is needed for patients with tracheal problems but normal neck anatomy (tracheomalacia, tracheal stenosis).
 - A flexible (reinforced) tracheostomy tube with an adjustable flange can be used in any of the above patients.



❖ **Tracheostomy Care:**

○ **Infection Control:**

- Presence of the tracheostomy tube, secretions and fresh stoma site in an already debilitated and possibly immuno-compromised patient all increase the risk of infection.
- For changing the tracheostomy tube or a dressing, gloves should be sterile.
- For suctioning, gloves can be clean rather than sterile.
- Aprons should be worn at all times and changed between procedures.
- Eye protection should be worn for suctioning, dressing changes and tube changes or where there is any risk a patient may cough secretions towards the carer.

○ **Dressing:**

- Secretions that collect above the cuff may ooze out of the stoma site and cause wetness around the tracheostomy site, this can cause irritation leading to skin maceration and excoriation.
- The site should be assessed at least once in every 24 hours.
- Broad, soft adjustable tracheostomy tube holder should be used (allow for two fingers to fit under the holder).
- The dressing and tracheostomy holder may need to be changed more frequently if they become soiled.
- Red, excoriated or exuding stomas should be swabbed.

○ **Humidification:**

- A tracheostomy bypasses the normal upper airway mechanisms for humidification, filtration and warming of inspired gases.
- This results in increased viscosity of mucus secretions, which depresses ciliary function and therefore mucociliary transport.
- This leads to an increased risk of infection, impaired secretion removal and microatelectasis.
- Failure to provide adequate humidification to address these issues can lead to obstruction of the major airways and tracheostomy tube blockage.
- Methods:
 - Adequate systemic hydration.
 - Heat Moisture Exchangers (HME).
 - Saline nebulizers (q4-6 hourly) for patients with thick/dry secretions.
 - Acetylcysteine nebulizers (mucolytic) for very difficult to clear secretions.



○ **Suctioning:**

- Gently introduce the suction catheter to the length of the tracheostomy ONLY.
- Stop advancing when the patient coughs or when resistance is felt.
- Slowly withdraw the catheter by and apply negative pressure by occluding the side port of the catheter.
- Continue to withdraw the catheter applying continuous suction until it is fully removed.
- Do not apply suction on insertion.
- Do not suction for more than 15 seconds (from insertion to end of withdrawal).
- If the patient has a fenestrated tracheostomy tube, ensure a non-fenestrated inner tube is in situ.
- Ideal catheter size = (Size of Tracheostomy tube - 2) x 2
- Ensure the oxygen/ventilation source is reapplied immediately and attention is paid to oxygen saturations post procedure, considering hyperoxygenation if needed.
- Instillation of normal saline, to facilitate sputum clearance, is not recommended practice, instead, humidification of inspired gases and saline nebulizers (0.9 or 5%) should be considered.

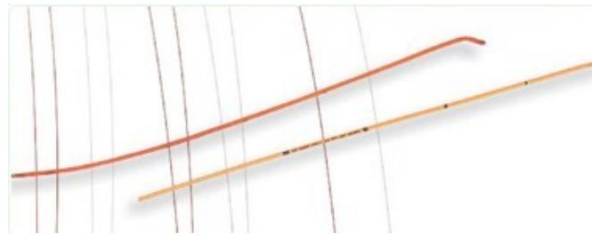
○ **Oral Feeding:**

- Not all patients with tracheostomies will have swallowing problems.
- Oral intake should be considered and offered only when the tracheostomy cuff is deflated.
- Swallowing assessment is needed in prior to starting oral feeding in the following conditions:
 - Neurological involvement (bulbar involvement).
 - Head & Neck surgery.
 - Evidence of aspiration of food/fluid/oral secretions on tracheal suctioning.
 - Persistent wet or weak voice when cuff is deflated and speaking valve or decannulation cap in place.
 - Coughing in relation to oral intake.
 - Oxygen desaturation in conjunction with oral intake.
 - Patient anxiety or distress during oral intake.
- Management of dysphagia:
 - Swallowing exercises.
 - Downsizing tracheostomy tube.
 - Diet modification.
 - Postural maneuvers.
 - Non-oral feeding.

❖ **Tracheostomy Tube changing:**

○ **Routine Changes:**

- Routine tube changes should be performed in the morning of working days (not evening or weekends).
- 1st Tracheostomy tube change:
 - Performed after stoma formation (7-10 days post-op).
- Subsequent Tracheostomy tube changes:
 - Recommendations for the frequency of changing tracheostomy tubes are unsupported by the literature.
 - In practice the frequency of tracheostomy tube change should be assessed on an individual patient basis.
- It is a two person technique, with one person supporting the tube and the patient and the other performing the change.
- In patients who are at risk of aspiration it is recommended that any enteral feed be stopped 3-4 hours prior to the procedure and the enteral tube aspirated immediately prior to the procedure.
- The procedure used for changing any tracheostomy tube will depend on the circumstances of that change.
- There are two commonly used methods:
 - **Guided exchange:**
 - Using a tube exchange device.
 - Usually required for early changes and for patients with a high risk of airway loss.
 - Steps:
 - Check emergency equipment.
 - Semi-recumbent position.
 - Pre-oxygenate.
 - Ensure assistant is clear regarding what is expected of them.
 - Check and lubricate tube.
 - Ask assistant to suction if required, remove old dressing, inner cannula and tapes and support the tube.
 - Ask assistant to deflate the cuff with suction applied.
 - Insert exchange device to length of tube.
 - Remove old tube over exchange device.
 - Insert new tube over exchange device
 - Check for airflow through tube.
 - Inflate cuff.
 - Remove exchange device.
 - Replace inner cannula
 - Check patient is stable (and cuff pressure).



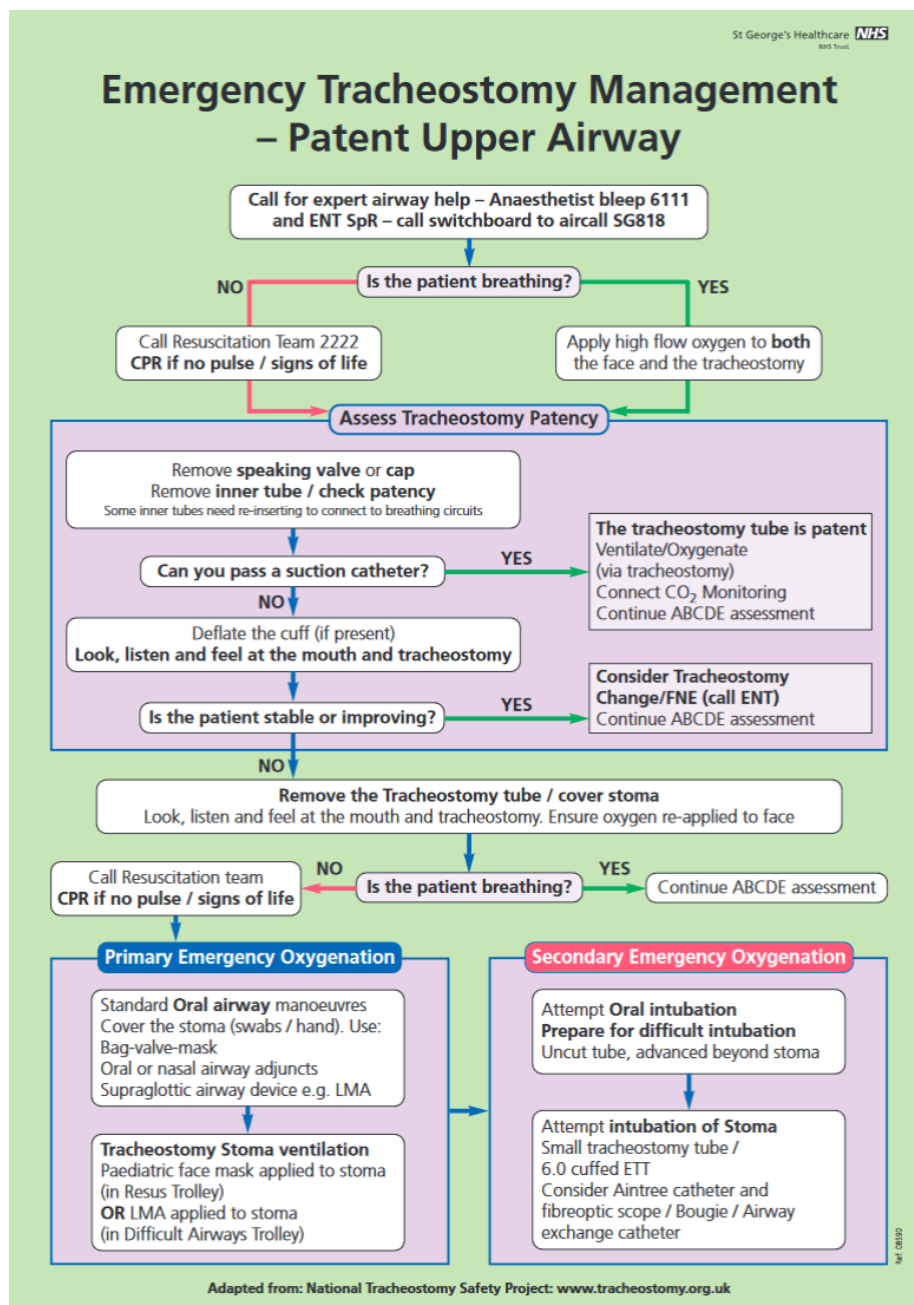
Portex® Tracheal Tube Inducer and Guide



Airway Exchange Guidewire

- **Blind exchange:**
 - Using an obturator.
 - Required for patients with formed stomas and a low risk of airway loss.
 - Steps:
 - Check emergency equipment.
 - Semi-recumbent position.
 - Pre-oxygenate.
 - Ensure assistant is clear regarding what is expected of them.
 - Check and lubricate tube.
 - Ask assistant to suction if required, remove old dressing, inner cannula and tapes and support the tube.
 - Ask assistant to deflate the cuff.
 - Remove tube on expiration.
 - Insert new tube over obturator.
 - Remove obturator.
 - Check for airflow through tube.
 - Inflate cuff.
 - Replace inner cannula
 - Check patient is stable (and cuff pressure).

- **If unable to re-insert tube successfully or the patient become compromised:**
 - Call for help immediately (Senior ENT specialist/ Anesthetist/Resus team) as appropriate to the situation to assist and/or orally intubate where appropriate.
 - Maintain oxygenation via stoma and nose and mouth with a facemask.
 - Use tracheal dilator and attempt to re-insert tube.
 - Reposition patient neck and attempt to re-insert tube.
 - Consider using a smaller size tube.



❖ **Tracheostomy Weaning and Decannulation:**

- Tracheostomy tube should NOT be kept longer than necessary.
- Prolonged use of tube leads to tracheobronchial infections, tracheal ulceration, granulations, stenosis and unsightly scars.
- Criteria required prior to decannulation:
 1. **Original indication for tracheotomy must be resolved.**
 2. Conscious and alert patient.
 3. Hemodynamically stable.
 4. Not requiring positive pressure ventilation.
 5. Supplemental oxygen requirement is < 40%.
 6. Minimal secretions requiring occasional suctioning.
 7. Good cough to clear chest secretion.
 8. Normal bedside swallowing evaluation.
 9. Patent upper airway:
 - Can be examined by either:
 - Noninvasively by fully deflating the cuff (if any) and plugging the tube opening by gloved finger:
 - If upper airway is patent, the patient will phonate without any stridor or distress.
 - Unable to phonate or presence of stridor or distress, indicate upper airway obstruction requiring further endoscopic examination.
 - Flexible endoscope examination of upper airway and vocal cords.
- Steps of decannulation:
 1. Cuff-deflation and observe for 24 hours (If the patient is on cuffed-tracheostomy tube).
 2. Downsizing the tracheostomy tube to uncuffed +/- fenestrated tube (allows adequate airflow through and around the tracheostomy tube into mouth/nose) and observe for 24 hours.
 3. Tolerating speaking valve placement for at least 4 hours.
 4. Capping of the tracheostomy tube and observe initially for 12 hours (daytime) then 24 hours if tolerated.
 5. If tolerated, decannulation if tracheostomy tube can be done safely and patient is observed for 24 hours.
 6. Wound is taped with occlusive dressing.

- Important points:
 - Decannulation process should start in morning time.
 - The patient should be closely monitored throughout the weaning process for signs of clinical deterioration.
 - STOP CRITERIA:
 - HR > 120
 - RR > 35
 - SpO₂ < 90%
 - FiO₂ > 40%
 - Tracheostomy set, small size tracheostomy tube and equipment for re-intubation should be always available bedside.
 - Healing of the stoma wound after decannulation takes place within a few days or a week.
- Possible reasons for failure of decannulation:
 - Persistence of condition for which tracheostomy was done.
 - Obstructing granulations around the stoma or below it where tip of the tracheostomy tube had been impinging.
 - Tracheal edema or subglottic stenosis.
 - Incurving of tracheal wall at the site of tracheostome.
 - Tracheomalacia (as the tracheostomy tube may work as a stent, preventing collapse of the tracheal wall in tracheomalacia).
 - Psychological dependence on tracheostomy and inability to tolerate the resistance of the upper airways.
- A case of difficult decannulation may require DLB to evaluate for airway obstruction and dynamic collapse.
- Post-decannulation instructions:
 - The dressing covering the stoma should be changed only if it becomes wet, or begins to leak or lift away from the skin.
 - The patient should provides support for the wound by placing a finger on top of the dressing when talking or coughing.



TRACHEOSTOMY SIZING CHART

PORTEX® BLUE LINE ULTRA and PORTEX SUCTIONAID TRACHEOSTOMY TUBES

Size	ID (mm)	ID with Inner Cannula (mm)	OD (mm)	Length (mm)	Cuff OD(mm)
6	6.0	5.0	9.2	64.5	20.0
7	7.0	5.5	10.5	70.0	24.0
7.5	7.5	6.0	11.3	73.0	30.0
8	8.0	6.5	11.9	75.5	30.0
8.5	8.5	7.0	12.6	78.0	30.0
9	9.0	7.5	13.3	81.0	30.0
10	10.0	8.5	14.0	87.5	30.0



AIR in cuff
Suctionaid model has above cuff line

PORTEX® BLUE LINE ADJUSTABLE FLANGE TRACHEOSTOMY TUBES

Size	ID (mm)	OD (mm)	Length (mm)	Cuff OD(mm)
6	6.0	8.3	59-81	20.0
7	7.0	9.7	64-84	24.0
8	8.0	11.0	73-97	30.0
9	9.0	12.4	78-111	30.0
10	10.0	13.7	84-123	30.0



AIR in cuff

PORTEX®UNIPERC™ADJUSTABLE FLANGE TRACHEOSTOMY TUBES

Size	ID (mm) without inner cannula	ID with Inner Cannula (mm)	OD (mm)	Length (mm)
7	9.3	7.0	11.6	62
8	10.3	8.0	12.6	68
9	11.3	9.0	13.6	74



AIR in cuff – Has inner cannula

BIVONA® (TTS* and CUFFLESS) TRACHEOSTOMY TUBES

Size	ID (mm)	OD (mm)	Length (mm)	Cuff (mm)
6	6.0	8.8	70.0	11.0
7	7.0	10.0	80.0	12.0
7.5	7.5	10.5	80.0	12.0
8	8.0	11.0	88.0	13.0
8.5	8.5	11.8	88.0	13.0
9	9.0	12.3	98.0	N/A



WATER in cuff

BIVONA® AIRE CUFF TRACHEOSTOMY TUBES

Size	ID (mm)	OD (mm)	Length (mm)	Cuff (mm)
6	6.0	8.8	70.0	
7	7.0	10.0	80.0	21.0
8	8.0	11.0	88.0	25.0
9	9.0	12.3	98.0	28.0
	9.5	13.3	98.0	30.0



AIR in cuff

BIVONA® HYPERFLEX ADJUSTABLE FLANGE TRACHEOSTOMY TUBES

Size	ID (mm)	OD (mm)	Maximum Length (mm)	Cuff(mm)
6	6.0	9.2	110.0	11
7	7.0	10.6	120.0	12
8	8.0	11.7	130.0	13
9	9.0	12.9	140.0	14.5



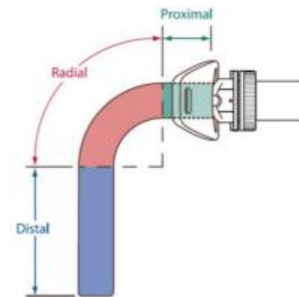
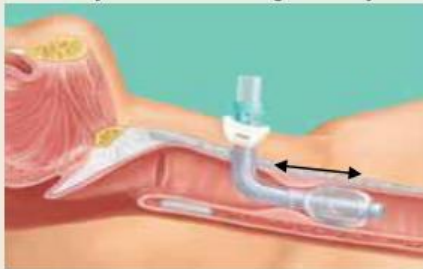
WATER in cuff
Only use in ICU/HDU

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SHILEY®XLT™ TRACHEOSTOMY TUBES

Description							Catalog Number		
Extension	I.D.	O.D.	Total Length	Proximal Length	Radial Length	Distal Length	Cuffed	Cuffless	Disposable Inner Cannula
Distal	5.0 mm	9.6 mm	90 mm	5.0 mm	37.0 mm	48.0 mm	50XLCD	50XLUD	50XLIN
Proximal	5.0 mm	9.6 mm	90 mm	20.0 mm	37.0 mm	33.0 mm	50XLCP	50XLUP	50XLIN
Distal	6.0 mm	11.0 mm	95 mm	8.0 mm	38.0 mm	49.0 mm	60XLCD	60XLUD	60XLIN
Proximal	6.0 mm	11.0 mm	95 mm	23.0 mm	38.0 mm	34.0 mm	60XLCP	60XLUP	60XLIN
Distal	7.0 mm	12.3 mm	100 mm	12.0 mm	39.0 mm	49.0 mm	70XLCD	70XLUD	70XLIN
Proximal	7.0 mm	12.3 mm	100 mm	27.0 mm	39.0 mm	34.0 mm	70XLCP	70XLUP	70XLIN
Distal	8.0 mm	13.3 mm	105 mm	15.0 mm	40.0 mm	50.0 mm	80XLCD	80XLUD	80XLIN
Proximal	8.0 mm	13.3 mm	105 mm	30.0 mm	40.0 mm	35.0 mm	80XLCP	80XLUP	80XLIN

**Shiley XLTD – 'extra length distally'**

For long tracheal anatomy, tracheal stenosis or malacia

Shiley XLTP – 'extra length proximally'

For swollen or thick neck anatomy

SHILEY™ FLEXIBLE TRACHEOSTOMY TUBES (CN/UN)

Size (mm)	ID (mm) without inner cannula	ID with Inner Cannula (mm)	OD (mm)	Length (mm)
4	6.5	5.5	9.4	62
5	7.0	6.0	10.1	68
6	7.5	6.5	10.8	74
7	8.0	7.0	11.4	77
8	8.5	7.5	12.2	79
9	9.0	8.0	12.7	79
10	10.0	9.0	13.8	79

**SHILEY™ STANDARD TRACHEOSTOMY TUBES (DCT/DFEN/DCFS/DCFN)**

Size	ID (mm)	OD (mm)	Length (mm)
4	5.0	9.4	62
6	6.4	10.8	74
8	7.6	12.2	79
10	8.9	13.8	79



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COOK® VERSA™ TRACHEOSTOMY TUBE

Size(mm)	ID (mm)	ID with Inner Cannula (mm)	OD (mm)	Length (mm)	Cuff OD(mm)	Angle (°)	Colour code
7	7.0	6.0	10.0	78.0	25.0	96	Green
8	8.0	7.0	11.0	86.0	28.0	96	White
9	9.0	8.0	12.0	98.0	30.0	98	Blue

Current as at 23/02/2015

**TRACOE® COMFORT**

Reference Numbers and Measurements: REF 101-REF 105, REF 114, REF 115

Size	① ID-OC (mm)	② OD-OC bc (mm)	③ OD-OC bf (mm)	④ ID-IC (mm)	Lengths (mm)	
					⑤ TL	⑥ AB
03	3.5	4.8	6.4	2.6	42	46
04	4.0	5.6	7.3	3.2	50	55
05	5.0	6.7	8.7	4.0	52	57
06	6.0	7.6	10.0	4.8	54	60
07	7.0	8.8	10.5	5.8	57	65
08	8.0	10.0	11.3	6.8	61	70
09	9.0	11.4	12.6	7.6	65	75
10	10.0	12.4	13.7	8.7	74	85
11	11.0	13.1	15.0	9.7	76	87
12	12.0	14.2	16.2	10.8	78	90
13	13.0	15.4	17.2	11.8	87	100
14	14.0	16.6	18.2	12.6	91	105

① ID-OC: inside diameter (clear width) at bottom of outer cannula; ② OD-OC bc: outside diameter at bottom of outer cannula; ③ OD-OC bf: outside diameter of outer cannula behind the flange; ④ ID-IC: inside diameter at bottom of inner cannula; ⑤ Length TL: length along centre line from start of neck flange to bottom of tube; ⑥ Length AB: length along outer bend (greatest length)

• The bending angle is 90° in all comfort tubes.

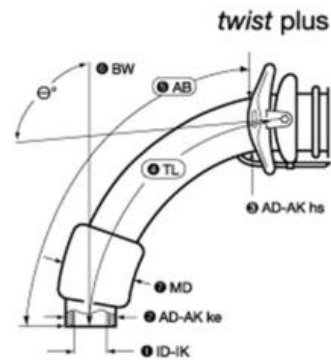
► To order, specify: REF + size (e.g. REF 106-05).

**TRACOE® TWIST PLUS**

technical data TRACOE twist plus Tracheostomy Tube, Double Fenestrated, with Low Pressure Cuff**

Size	ID-IK	AD-AK ke	AD-AK hs	Length TL	Length AB	BW	MD	REF-No.
	mm	mm	mm	mm	mm	°	mm	
07	7.0	9.8	10.1	85	91	100	26	REF 312-07
08	8.0	10.8	11.1	88	95	100	28	REF 312-08
09	9.0	11.8	12.1	90	99	100	30	REF 312-09
10	10.0	12.8	13.1	92	102	100	32	REF 312-10

ID-IK: inside diameter (clear width) at bottom of inner cannula
AD-AK ke: outside diameter at bottom of outer cannula
AD-AK hs: outside diameter of outer cannula behind the flange
Length TL: along centre line from start of neck flange to bottom of tube
Length AB: length along outer bend from start of neck flange to bottom of tube
BW: bending angle
MD: cuff diameter



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Table 2 A guide to the size of paediatric tracheostomy tubes, bronchoscopes and endotracheal tubes

			Preterm-1 month	1-6 months	6-18 months	18 mths - 3 yrs	3-6 years	6-9 years	9-12 years	12-14 years		
Trachea (Transverse Diameter mm)			5	5-6	6-7	7-8	8-9	9-10	10-13	13		
PLASTIC	Great Ormond Street	ID (mm)		3.0	3.5	4.0	4.5	5.0	5.5	6.0	7.0	
		OD (mm)		4.5	5.0	6.0	6.7	7.5	8.0	8.7	10.7	
	Shiley	Size		3.0	3.5	4.0	4.5	5.0	5.5	6.0	6.5	
		ID (mm)		3.0	3.5	4.0	4.5	5.0	5.5	6.0	6.5	
		OD (mm)		4.5	5.2	5.9	6.5	7.1	7.7	8.3	9.0	
	*Cuffed Tube Available	Length (mm) Neonatal		30	32	34	36					
		Paediatric		39	40	41*	42*	44*	46*			
		Long Paediatric							50*	52*	54*	56*
	Portex (Blue Line)	ID (mm)		3.0	3.5	4.0	4.5	5.0	5.0	6.0	7.0	
		OD (mm)		4.2	4.9	5.5	6.2	6.9	6.9	8.3	9.7	
	Portex (555)	Size		2.5	3.0	3.5	4.0	4.5	5.0	5.5		
		ID (mm)		2.5	3.0	3.5	4.0	4.5	5.0	5.5		
		OD (mm)		4.5	5.2	5.8	6.5	7.1	7.7	8.3		
		Length Neonatal		30	32	34	36					
		Paediatric		30	36	40	44	48	50	52		
	Bivona	Size	2.5	3.0	3.5	4.0	4.5	5.0	5.5			
		ID (mm)	2.5	3.0	3.5	4.0	4.5	5.0	5.5			
		OD (mm)	4.0	4.7	5.3	6.0	6.7	7.3	8.0			
	All sizes available with Fome Cuff, Aire Cuff & TTS Cuff	Length Neonatal	30	32	34	36						
		Paediatric	38	39	40	41	42	44	45			
		ID (mm)	2.5	3.0	3.5	4.0	4.5	5.0	5.5			
	Bivona Hyperflex	Usable Length (mm)	55	60	65	70	75	80	85			
		ID (mm)	2.5	3.0	3.5	4.0	4.5	5.0	5.5			
	Bivona Flexlend	ID (mm)	2.5	3.0	3.5	4.0	4.5	5.0	5.5			
		Shaft Length (mm)	38	39	40	41	42	44	45			
Flexlend Length (mm)		10	10	15	15	17.5	20	20				
SILVER	Alder Hey	FG		12-14	16	18	20	22	24			
	Negus	FG			16	18	20	22	24	26	28	
	Chevalier Jackson	FG		14	16	18	20	22	24	26	28	
	Sheffield	FG		12-14	16	18	20	22	24	26		
		ID (mm)		2.9-3.6	4.2	4.9	6.0	6.3	7.0	7.6		
Cricoid (AP Diameter)	ID (mm)		3.6-4.8	4.8-5.8	5.8-6.5	6.5-7.4	7.4-8.2	8.2-9.0	9.0-10.7	10.7		
	Bronchoscope (Storz)	Size		2.5	3.0	3.5	4.0	4.5	5.0	6.0	6.0	
		ID (mm)		3.5	4.3	5.0	6.0	6.6	7.1	7.5	7.5	
		OD (mm)		4.2	5.0	5.7	6.7	7.3	7.8	8.2	8.2	
	Endotracheal Tube (Portex)	ID (mm)	2.5	3.0	3.5	4.0	4.5	5.0	6.0	7.0	8.0	
		OD (mm)	3.4	4.2	4.8	5.4	6.2	6.8	8.2	9.6	10.8	

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Laryngospasm

- Laryngospasm is one of the complication seen in the perioperative period especially during induction of
- Anesthesia or during extubation.
- It consists of prolonged glottis closure reflex mediated by the superior laryngeal nerve
- The common inciting factors are hyperactive airway like in case of upper respiratory tract infection.
- **Other common triggering factors are:**
 - Painful stimulation
 - Primary vagal hypertonicity
 - Insufficient depth of anaesthesia on endotracheal intubation
 - Light anaesthesia on tracheal extubation
 - Combination of either preceding with or without some irritant such as blood, mucus, laryngoscope blade, suction catheter, surgical debris or other foreign body

It can be serious causing fatal cardiac or cerebral complications.

ANATOMY AND MECHANISM OF LARYNGOSPASM

Laryngospasm involves three structures:

- The aryepiglottic folds
- False vocal cords
- The true vocal cords

The muscles most involved in are the:

- Lateral cricoarytenoid and the thyroarytenoids (adductors of the glottis)
- The cricothyroid (a tensor of the vocal cords).

During a laryngospasm either the true vocal cords alone or the true and false vocal cords both become apposed in the midline and close the glottis.

The recurrent nerve supplying all intrinsic muscles other than the cricothyroid, which is supplied by the external branch of the superior laryngeal nerve.

- Stimulation of the **nasal mucosa, soft palate and the pharynx.**
- mediated a minor portion of the pharyngeal inhibitory reflex, but that the main component was mediated through the pharyngeal branch of the vagus nerve.
- Stimulation of the **epiglottis and larynx.**
- The internal branch of the superior laryngeal nerve innervates the larynx from its superior boundaries to the level of the true vocal cords.
- Whereas below the vocal cords level the recurrent laryngeal nerve carries the sensory elements.
- The entrance to the larynx has receptors which form a protective mechanism and have greatest degree of sensitivity.
- **The posterior aspect of the true vocal cords, which is more exposed to foreign material than the anterior aspect, was a region of greater distribution of nerve endings than the anterior aspect.**

- Simulation of the **tracheobronchial tree**
- Mechanical stimulation of the larger passages elicits a forced expiratory response and showed that the afferent nerve fibers are in the vagus
- Apnea and bronchospasm occurred even though an isolated segment of the trachea with its nerve and blood supply was intact, and was exposed to ether.
- This suggested stimulation of the chemoreceptors in the lung.

- **Abdominal viscera and diaphragm** stimulation
- Pressure, tension and friction applied to the deep surfaces of the parietal peritoneum caused periods of apnea and the efferent pathway was said to be in the intercostals nerves.

The reflex bronchiolar constriction was best observed by stimulation of the nasal mucous membrane

LARYNGEAL CLOSURE AND ITS FUNCTION

- There is double valve mechanism with in the larynx which is capable of controlling both the entrance and the exit of air
- When the **true vocal** cords were in apposition they would prevent the **entrance** of air, but not its exit,
- Whereas apposition of **false vocal** cords was capable of preventing even a powerful current of air passing through from below. (**exit**)

- The posterior cricoarytenoid muscles are normally in a state of partial contraction, which is tonic in nature and that the afferent impulses involved in this reflex are conducted along the vagus.
- The adductor muscles have no role in respiration but protect the lower airway against foreign body and in modified forms of expiration as in coughing and laughing

CLINICAL SIGNIFICANCE OF LARYNGOSPASM

Incidence of 0.78%-5% depending on:

- Surgical type
- Patient age
- Preexisting condition
- Anesthetic technique

Laryngospasm is the occlusion of the glottis due to contraction of the intrinsic muscles of the larynx, which is essentially considered a protective reflex to prevent any foreign body reaching the tracheobronchial tree and lungs.

Bronchospasm is the contraction of the bronchial musculature which causes constriction of the smaller air passages and may be associated with laryngospasm in some cases.

Causes:

- Direct irritation of the vocal cords (ex. irritating vapor)
- Mechanical stimulation of the epiglottis during attempts at intubation, especially in light planes of anaesthesia (barbiturates)
- Respiratory infection
- Elongate uvula
- Different triggers, such as asthma, allergies, exercise, irritants (smoke, dust, fumes), stress, anxiety or commonly or GERD
- Respiratory infection dramatically increases the incidence of laryngospasm especially in children.
- Application of topical lidocaine 4% to the larynx at the time of intubation has shown to decrease the incidence of laryngospasm in tonsillectomy patients
- Laryngospasm has been described with the elongated uvula as sleep-related and evoked
- by distal esophageal afferent and even post-operatively
- causing pulmonary edema
- The development of marked negative intrathoracic pressures due to airway obstruction is
- believed to be the primary pathological event in the development of pulmonary edema

LARYNGOSPASM IN CHILDREN

The incidence in children being higher especially in infants 1-3 months.

It can persist to cause:

- Hypoxia, hypercapnea, cyanosis, desaturations, arrhythmias, pulmonary edema, bronchospasm, cardiac arrest or gastric aspiration
- It is often self limiting as hypoxia and carbon-di-oxide retention abolishes the reflex

Precipitating factors were:

- Majorly in children with respiratory tract
- Extubation
- Presence of a nasogastric tube
- Oral endoscopy
- Esophagoscopy and infections
- Some authors have proposed laryngeal spasm to be a complication of **barbiturate** induced parasymphetic
- Activity
- Amongst the inhalational agents **isoflurane** showed greater incidence of laryngospasm than halothane, enflurane and sevoflurane

DDX of laryngospasm:

- Bronchospasm
- Supraglottic obstruction
- A psychogenic cause
- In anxious adolescents and young adults (in response to exercise and emotional stress)
- A paradoxical vocal cord movement (post-extubation vocal cord dysfunction)
- Episodic laryngeal spasm subsequent to superior laryngeal nerve injury after thyroid surgery
- Other causes to be excluded are foreign body, epiglottic impaction, laryngeal edema and tracheal spasm or collapse.

Incomplete airway obstruction or partial obstruction is generally associated with an audible inspiratory or expiratory sound => **stridor**.

If the obstruction worsens:

Tracheal tug and paradoxical respiratory movements of the thorax and the abdomen develop.

PRECIPITATING RISK FACTORS

❖ Patient-related

- URTI
- Asthma
- Irritable airway
- Chronic smoking (need abstinence at least 48hrs)
- Passive smoking
- GERD
- Long uvula
- Upper airway anomalies
- Hyperventilation (activation of laryngeal thermo/chemoreceptors)

❖ Anesthesia related

- Inadequate depth
 - Less experience anesthesia
 - Use of laryngeal mask
 - Volatile anesthetic like isoflurane
 - Intravenous induction with barbiturates
 - Mucus, secretions, blood, laryngoscope, suction catheter or any other foreign body with light anesthesia
- Ketamine although not usually associated with laryngospasm, produces secretions which can play a trigger by irritating the vocal cords.
 - Laryngospasm is seen more with sevoflurane than propofol induction.

❖ Surgery related

- Upper airway surgery (21-26% more than in adenotonsillectomy)
- Thyroid surgery with damage to superior laryngeal nerve
- Iatrogenic removal of parathyroid, hypocalcemia then spasm
- Reflux spasm after esophageal stimulation

❖ Rare association

- febrile non-hemolytic transfusion reaction
- Inapparent regurgitation and aspiration of gastric contents
- Parkinson
- Acute withdrawal of medication

Management

- If not promptly managed effectively may lead to increased morbidity and mortality.
- These patients can deteriorate easily.

❖ Incomplete airway obstruction:

- Remove irritant stimulus (eliminating surgical stimulation of visceral nerve endings),
- Deepen anaesthetic plane, apply jaw thrust maneuver,
- Insert an oral or a nasal airway
- Provide gentle continuous positive airway pressure with 100% oxygen.
- Periosteal pain caused by pressing on the styloid process) helps relaxing the vocal cords by the autonomic nervous systems

If not relieved by above maneuvers suspect complete laryngospasm

❖ Complete obstruction

- Deepen anaesthesia with intravenous or non-irritant inhalational agents. (**Propofol** 0.8mg/kg show rapid action with success of 77%)
- **If no success**, use of suxamethonium (**succinylcholine**) in the dose 0.1-3mg/kg intravenous or 4mg/kg intramuscular or even intralingual, or itraosseous (if no intravenous access available)
- Dislocation of the temporomandibular joint anteriorly by application of pressure to the ascending rami of the mandible lengthens the thyrohyoid muscle and unfolds the soft supraglottic tissue.
- Followed with mask ventilation or if needed tracheal intubation relieves laryngospasm

Forced inflation of pharynx distends the pyriform fossa which subsequently presses the aryepiglottic folds more tightly against each other.
This further causes stomach inflation rather than of the lungs.
Avoided with good jaw thrust

A small dose of **succinylcholine** intravenously causes relaxation immediately facilitating intubation of the larynx.

Pulmonary edema has been reported after intramuscular injection.

If this fails **atropine** and succinylcholine is given intravenous

- On becoming hypoxic and having bradycardia, the child may need to be **intubated** without muscle relaxation, then to wait for the effects of succinylcholine.
- The vocal cords can be **sprayed** with **lidocaine**, in order to bring relaxation and facilitate intubation.

If these methods fail; emergency cricothyrotomy or emergency tracheostomy may be required.

- Various other studies done, showed that extubation or removal of LMA in anaesthetized state is associated with far fewer complications than in awake state.
- Despite the fact that deep extubation might afford some protection against coughing and straining, the risk of aspiration and inadequate airway protection in this vulnerable period is a cause of concern.
- Removing the tube while the lungs are inflated by positive pressure, thus decreasing the adductor response of the laryngeal muscles and subsequently reducing the incidence of laryngospasm.

Summery of medications might help during spasm:

- **Propofol** (depresses airway reflexes)
- Anticholinergics which reduce secretions like atropine
- Nebulised **lidocaine/xylocain** (reducing sensitivity of upper airway reflexes)
- Cocaine
- Benzodiazepines like diazepam (decrease upper airway reflexes)
- Doxapram in dose 1.5mg/kg over 20 secs (increased respiratory drive from medulla)
- Nitroglycerine (vasodilator) stimulates formation of cGMP which is a mediator for non-adrenergic and non-cholinergic nerves which cause relaxation of the airway smooth muscles
- Succinylcholine (intramuscular blocker)
- **In case of recurrent** postoperative laryngospasm, a differential of psychogenic/hysterical stridor must be considered and often reassurance and benzodiazepines help
- Instructions to breathe in a slow pattern minimizes inspiratory obstruction.
- On laryngoscopy, these may present with fully adducted vocal cords, hypopharyngeal spasm and paradoxical vocal cord movement.

Finally, laryngospasm was successfully treated with [superior laryngeal nerve blocks](#) (1999).

Failed intubation, increasing hypoxaemia and difficult ventilation in the paralysed anaesthetised patient: Rescue techniques for the "can't intubate, can't ventilate" situation

failed intubation and difficult ventilation (other than laryngospasm)

Face mask
Oxygenate and Ventilate patient
Maximum head extension
Maximum jaw thrust
Assistance with mask seal
Oral $\pm$ 6mm nasal airway
Reduce cricoid force - if necessary

failed oxygenation with face mask (e.g. SpO₂ < 90% with FiO₂ 1.0)

call for help

LMA™ Oxygenate and ventilate patient
Maximum 2 attempts at insertion
Reduce any cricoid force during insertion

succeed

Oxygenation satisfactory and stable: Maintain oxygenation and awaken patient

"can't intubate, can't ventilate" situation with increasing hypoxaemia

Plan D: Rescue techniques for "can't intubate, can't ventilate" situation

or

Cannula cricothyroidotomy

Equipment: Kink-resistant cannula, e.g. Patil (Cook) or Ravussin (VBM)
High-pressure ventilation system, e.g. Manujet III (VBM)

Technique:

1. Insert cannula through cricothyroid membrane
2. Maintain position of cannula - assistant's hand
3. Confirm tracheal position by air aspiration - 20ml syringe
4. Attach ventilation system to cannula
5. Commence cautious ventilation
6. Confirm ventilation of lungs, and exhalation through upper airway
7. If ventilation fails, or surgical emphysema or any other complication develops - convert immediately to surgical cricothyroidotomy

fail

Surgical cricothyroidotomy

Equipment: Scalpel - short and rounded (no. 20 or Minitracheal scalpel)
Small (e.g. 6 or 7 mm) cuffed tracheal or tracheostomy tube

4-step Technique:

1. Identify cricothyroid membrane
 2. Stab incision through skin and membrane
Enlarge incision with blunt dissection (e.g. scalpel handle, forceps or dilator)
 3. Caudal traction on cricoid cartilage with tracheal hook
 4. Insert tube and inflate cuff
- Ventilate with low-pressure source
Verify tube position and pulmonary ventilation

Notes:

1. These techniques can have serious complications - use only in life-threatening situations
2. Convert to definitive airway as soon as possible
3. Postoperative management - see other difficult airway guidelines and flow-charts
4. 4mm cannula with low-pressure ventilation may be successful in patient breathing spontaneously

Difficult Airway Society guidelines Flow-chart 2004 (use with DAS guidelines paper)



Laryngitis:

Acute Laryngitis:

- Acute laryngitis may be infectious or non-infectious.
- **Etiology**
 - The infectious type is more common and usually follows upper respiratory infection.
 - To begin with, it is viral in origin.
 - o Pathogens: rhinovirus (most common), parainfluenza, respiratory syncytial virus, adenovirus, influenza virus, pertussis.
 - Soon bacterial invasion takes place with Strept. pneumoniae, H. influenzae and haemolytic streptococci or Staph. aureus.
 - The non-infectious type is due to vocal abuse, allergy, thermal or chemical burns to larynx due to inhalation or ingestion of various substances, or laryngeal trauma such as endotracheal intubation.
- **Clinical Features**
 - Symptoms are usually abrupt in onset and consist of:
 1. Hoarseness which may lead to complete loss of voice.
 2. Discomfort or pain in throat, particularly after talking.
 3. Dry, irritating cough which is usually worse at night.
 4. General symptoms of head, cold, rawness or dryness of throat, malaise and fever if laryngitis has followed viral infection of upper respiratory tract.
 - Laryngeal appearances vary with severity of disease.
 - In early stages, there is erythema and edema of epiglottis, aryepiglottic folds, arytenoids and ventricular bands, but the vocal cords appear white and near normal and stand out in contrast to surrounding mucosa, betraying the degree of hoarseness patient has.
 - Later, hyperaemia and swelling increase.
 - Vocal cords also become red and swollen.
 - Subglottic region also gets involved.
 - Sticky secretions are seen between the cords and interarytenoid region.
 - In case of vocal abuse, submucosal haemorrhages may be seen in the vocal cords.
- **Treatment**
 - Vocal rest is the most important single factor.
 - o Use of voice during acute laryngitis may lead to incomplete or delayed recovery.
 - Avoidance of smoking and alcohol.
 - Steam inhalations to soothe and loosen viscid secretions.
 - Cough sedative to suppress troublesome irritating cough.
 - Antibiotics when there is secondary infection with fever and toxæmia or purulent expectoration.
 - Analgesics to relieve local pain and discomfort.
 - Steroids are useful in laryngitis following thermal or chemical burns.

Acute Epiglottitis (Supraglottic Laryngitis)

- Acute inflammatory condition confined to supraglottic structures (Epiglottis, aryepiglottic folds and arytenoids).
- There is marked edema of these structures which may obstruct the airway.

- **Etiology:**
- It is a serious condition and affects children of 2-7 years of age but can also affect adults.
- Common Pathogens:
 - o H. influenzae (classic pediatric disease) rare since Hib vaccine.
 - o S. pneumoniae, S. aureus, β -hemolytic Streptococcus

- **Clinical Features**
- Onset of symptoms is abrupt with rapid progression.
- Sore throat and dysphagia are the common presenting symptoms in adults.
- Dyspnoea and stridor are the common presenting symptoms in children.
- They are rapidly progressive and may prove fatal unless relieved.
- Fever may go up to 40°C. It is due to septicaemia. Patient's condition may rapidly deteriorate.

- **Examination**
- Depressing the tongue with a tongue depressor may show red and swollen epiglottis. Indirect laryngoscopy may show edema and congestion of supraglottic structure.
- This examination is avoided for fear of precipitating complete obstruction.
- It is better done in operation theatre where facilities for intubation are available.
- Lateral soft tissue X-ray of neck may show swollen epiglottis (thumb sign).

- **Treatment**
- Avoid aggravating patient and precipitating airway collapse (if high clinical suspicion and patient symptomatic, establish airway in operating room).
- For mild symptoms and stable patient with diagnosis in question, consider plain x-ray, flexible laryngoscopy to rule out epiglottic/supraglottic edema.
- Establish Emergency Airway: intubation performed in the operating room with preparation for a tracheotomy.
- Endoscopy: examine and culture epiglottis
- Postoperative Care: monitored bed, parenteral antibiotics and corticosteroids for 7–10 days, consider extubation when clinical improvement and decreased epiglottic edema.

Acute Laryngo-Tracheo-Bronchitis (Croup):

- Inflammatory condition of the larynx, trachea and bronchi.
- More common than acute epiglottitis.
- Most common infectious cause of stridor in children
- **Etiology**
 - Mostly, it is viral infection (parainfluenza type I and II) affecting children between 6 months to 3 years of age during fall and winter seasons.
 - Male children are more often affected.
 - Secondary bacterial infection by Gram positive cocci soon supervenes.
- **Pathology**
 - The loose areolar tissue in the subglottic region swells up and causes respiratory obstruction and stridor.
 - This, coupled with thick tenacious secretions and crusts, may completely occlude the airway.
 - Primarily involves the subglottic region
- **Symptomatology**
 - Disease starts with gradual onset of upper respiratory infection with hoarseness and croupy cough.
 - There is fever of 39-40°C.
 - This may be followed by difficulty in breathing and inspiratory type of stridor.
 - Respiratory difficulty may gradually increase with signs of upper airway obstruction, i.e. suprasternal and intercostal recession.
 - Plain neck x-rays ("steeple" sign = narrowed subglottis on PA neck x-ray),
- **Treatment**
 - Hospitalization is often essential in severe cases because of the increasing difficulty in breathing.
 - Humidification helps to soften crusts and tenacious secretions which block tracheobronchial tree.
 - Parenteral fluids are essential to combat dehydration.
 - Steroids, e.g. hydrocortisone 100 mg i.v. may be useful to relieve edema.
 - Racemic epinephrine administered via a respirator is a bronchodilator and relieve dyspnea.
 - Antibiotics not required unless suspect bacterial superinfection.
 - For severe cases not improving with medical management, intubation/tracheostomy is done.
 - Recurrent attack of croup required DLB later on to rule out subglottic stenosis.

Table 57-1. Differences between acute epiglottitis and acute laryngo-tracheo-bronchitis in children

	Acute epiglottitis	Acute laryngo-tracheo-bronchitis (or group)
• Causative organism	<i>Haemophilus influenzae</i> type B	Parainfluenza virus type I and II
• Age	2-7 years	3 months to 3 years
• Pathology	Supraglottic larynx	Subglottic area
• Prodromal symptoms	Absent	Present
• Onset	Sudden	Slow
• Fever	High	Low grade or no fever
• Patient's look	Toxic	Non-toxic
• Cough	Usually absent	Present, (Barking seal-like)
• Stridor	Present and may be marked	Present
• Odynophagia	Present, with drooling of secretions	Usually absent
• Radiology	*Thumb sign on lateral view	Steeple sign on anteroposterior view of neck
• Treatment	Humidified oxygen, third generation cephalosporin (ceftriaxone) or amoxicillin	Humidified O ₂ tent, steroids

*Examination of larynx and radiographs are avoided lest complete obstruction is precipitated. Examination is done in the operation theatre where immediate intubation can be done.

TABLE 10–2. Contrasting Acute Laryngotracheobronchitis (Croup) and Epiglottitis

	Acute Laryngotracheobronchitis	Acute Epiglottitis
Pathogen	Parainfluenza virus 1	<i>Haemophilis influenzae b</i>
Age	<5 years old	2–6 years old
Location	subglottic	supraglottic
Onset	gradual (days)	sudden onset (hours)
Cough	barky	normal
Posture	supine	upright
Drooling	no	yes
Fever	low-grade	high fevers
Radiographs	steeple sign	thumbprinting
Treatment	supportive	airway management and antibiotics

Bacterial Tracheitis (Exudative Tracheitis, Membranous Laryngotracheobronchitis):

- **Pathophysiology:**
 - Bacterial superinfection following viral URI prodrome.
- **Clinical picture:**
 - Thick secretions in airway, fibrinous sloughing (exudative) membrane in trachea, high fever, tender larynx and trachea.
- **Diagnosis:**
 - Clinical suspicion.
 - Lateral neck x-ray may show shaggy appearance of the trachea (membranes).
 - Flexible laryngoscopy may reveal exudative membranes in the subglottis and upper trachea.
 - Microlaryngoscopy and bronchoscopy
- **Treatment:**
 - Aggressive pulmonary hygiene.
 - Parenteral antibiotics
 - Microlaryngoscopy and bronchoscopy with membrane clearing and culture.
 - May require intubation

Laryngeal Edema:

- It involves the supraglottic and subglottic region where laryngeal mucosa is loose.
- Edema of the vocal cords occurs rarely because of the sparse subepithelial connective tissue.
- **Etiology**
 1. Infections:
 - Acute epiglottitis, laryngo-tracheo-bronchitis, tuberculosis or syphilis of larynx.
 - Infection in neighbourhood, e.g. peritonsillar abscess, retropharyngeal abscess and Ludwig's angina.
 2. Trauma:
 - Surgery of tongue, floor of mouth, laryngeal trauma, foreign body, endoscopy especially in children, intubation, thermal or caustic burns or inhalation or irritant gases or fumes.
 3. Neoplasms:
 - Cancer of larynx or laryngopharynx often associated with deep ulceration.
 4. Allergy:
 - Angioneurotic oedema, anaphylaxis.
 5. Radiation:
 - For cancer of larynx or pharynx.
 6. Systemic diseases:
 - Nephritis, heart failure, or myxoedema.

- **Symptoms and Signs**

- Airway obstruction Degree of respiratory distress varies. Tracheostomy may become essential.
- Inspiratory stridor.
- Indirect laryngoscopy shows oedema of supraglottic or subglottic region. Children may require direct laryngoscopy.

- **Treatment**

- If there is airway obstruction, intubation of larynx or tracheostomy will be immediately required.
- Less severe cases are treated conservatively and treatment will depend on the cause.
- An injection of adrenaline (1:1000) 0.3-0.5 ml i.m., repeated in 15 minutes if necessary, is useful in allergic or angioneurotic edema.
- Steroids are useful in epiglottitis, laryngo-tracheo-bronchitis or edema due to traumatic allergic or post-radiation causes.

Chronic Laryngitis:

- Diffuse inflammatory condition symmetrically involving the whole larynx (true cords, ventricular bands, interarytenoid region and root of the epiglottis).

- **Etiology**

1. It may follow incompletely resolved acute simple laryngitis or its recurrent attacks.
2. Presence of chronic infection in paranasal sinuses, teeth and tonsils and the chest are important contributory causes.
3. Occupational factors, e.g. exposure to dust and fumes such as in miners, stokers, gold or iron smiths and workers in chemical industries.
4. Smoking and alcohol.
5. Persistent trauma of cough as in chronic lung diseases.
6. Vocal abuse.

- **Clinical Features**

- Hoarseness. This is the commonest complaint. Voice becomes easily tired and patient becomes aphonic by the end of the day.
- Constant hawking. There is dryness and intermittent tickling in the throat and patient is compelled to clear the throat repeatedly.
- Discomfort in the throat.
- Cough. It is dry and irritating.

- **Laryngeal examination**

- There is hyperaemia of laryngeal structures.
- Vocal cords appear dull red and rounded.
- Flecks of viscid mucus are seen on the vocal cords and interarytenoid region.

- **Treatment**

- Eliminate infection of upper or lower respiratory tract Infection in the sinuses, tonsils, teeth or chronic chest infection (bronchitis, bronchiectasis, tuberculosis, etc.) should be treated.
- Avoidance of irritating factors, e.g. smoking, alcohol or polluted environment, dust and fumes.
- Voice rest and speech therapy Voice rest has to be prolonged for weeks or months. Patient should receive training in proper use of voice.
- Steam inhalations They help to loosen secretions and give relief.
- Expectorants They help to loosen viscid secretions and give relief from hawking.

Riyadh et al. Notes	Disease	Specific site	Finding
	Tuberculosis	Mostly on post 1/3 of larynx " Interarytenoid" turban epiglottis " laryngeal surface"	diffusely edematous and hyperemic mucosa; can have granular exophytic lesions airway obstruction (late)
	Wegener	25% involve larynx subglottis	Late → granulomatous ulcers Subglottic stenosis
	Rheumatoid arthritis	25% involve larynx arytenoid fixation 86 % and VC bowing	+/- submucosal nodules " Bamboo nodule" red swollen mucosa over arytenoids
	Relapsing polychondritis	50% have laryngeal/tracheal involvement Subglottic stenosis	
	Amyloidosis	Supraglottic " venricul + FVC" TVC	pink/gray, smooth, submucosal nodule; cobblestone-like; diffuse mucosal thickening
	SLE	glottis and CA joints " paralysis" subglottic stenosis	33 % get laryngeal involvement
	Sarcoidosis	supraglottic "epiglottis"	1-5% laryngeal involvement
	Syphilis		2 stage → erythematous papules and ulcers 3 stage → gumma formation → fibrosis, chondritis & stenosis

Leprosy	supraglottic	nodular and edematous mucosa with ulceration; stenosis
RhinoScleroma	subglottis	A/W obstruction
Histoplasmosis		superficial granulomas ; complications stenosis
Blastomycosis		small, red, granular lesions

Laryngopharyngeal Reflux

GERD:

- Most common chronic disease of adults in the USA (30%)
- up to 50 reflux episodes a day (below pH 4) are considered normal
- Disease considered when reflux is prolonged and/or there is a breakdown in the defense mechanisms.

LPR:

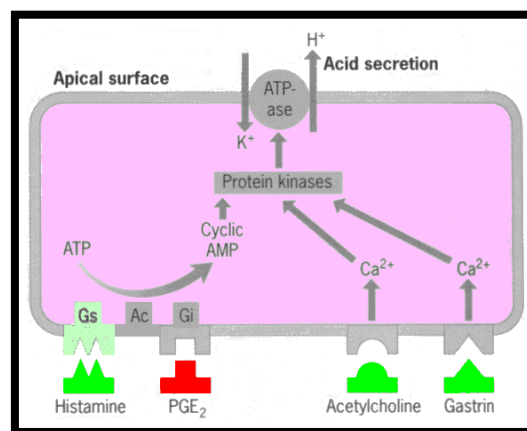
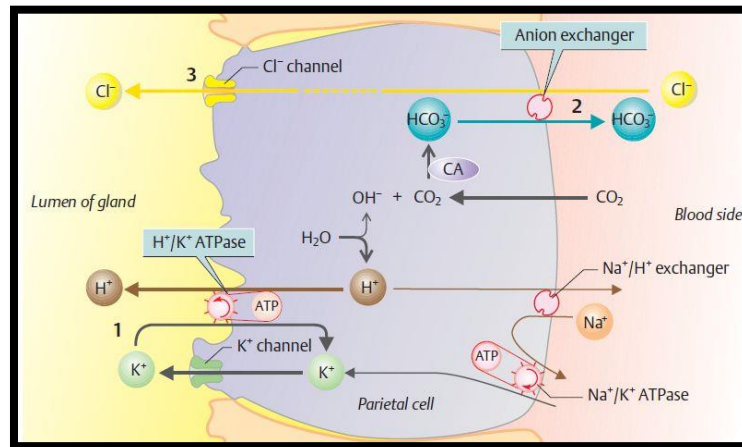
- Other aliases: extraesophageal reflux, reflux laryngitis, posterior laryngitis
- Contributes up to 50% of laryngeal complaints
- Backflow of gastric contents into larynx, pharynx, and upper aerodigestive tract
- Upper esophageal sphincter (UES) dysfunction
- Common in adult children and infants.
- Present in 4-10% of those with GERD
- About 20-70% with LPR have symptoms of GERD
- Laryngopharynx appears to be more sensitive to injury by gastric refluxate.
- 3 reflux episodes per week or less into the laryngopharynx results in severe damage of the laryngeal epithelium.
- One or more reflux events per 24-hour period are significant for diagnosis of LPR.

Upper Esophageal Sphincter

- C-shaped sling attached to cricoid cartilage
 - Cricopharyngeus
 - Thyropharyngeus
 - Proximal cervical esophagus
- Innervated by pharyngeal plexus
 - Vagus nerve
 - Superior laryngeal nerve
 - Recurrent laryngeal nerve
 - Glossopharyngeal nerve
 - Sympathetics from superior cervical ganglion
 - Nucleus:
 - Motor: nucleus ambiguus
 - Afferent: nucleus tractus solitaries
 - Relaxation of UES

GASTRIC REFLUX CONTENTS AND PHYSIOLOGY

- **Gastric Acid**
 - produced by parietal cells
 - approximately pH 2
 - proton pump H^+/K^+ ATPase
 - denature of food protein
 - chief cells secrete pepsinogen; activated by acid to pepsin; breakdown of amino acids bonds (proteolysis)
 - Gastric acidic inhibits growth of many microorganisms
- **Secretion of Gastric acid**
 - 3 Phases: Cephalic Phase 30%; Gastric Phase 60%; Intestinal Phase 10%
 - The release histamine (ECL) is the most important positive regulation mechanism of the secretion of gastric acid
 - stimulated by gastrin and acetylcholine
 - inhibited by somatostatin
 - in duodenum, gastric acid is neutralized by $NaHCO_3$



- **Pepsin**
 - Maximally active at PH 2; inactive at PH 6.5; denatured at PH 8
 - biomarker for reflux

- Bile Salts:
 - acts as a surfactant, helping to emulsify the fats in food to form micelles
 - alkaline bile neutralize stomach acid
 - known to cause esophagitis
 - likely to play a role in reflux-attributed laryngeal injury
- Trypsin
 - secreted into the duodenum
 - hydrolyze peptides into amino acids
 - trypsin can stimulate the production of inflammatory mediators

PATHOPHYSIOLOGY

- Gastric refluxate, containing acid, pepsin, bile, and trypsin
- LPR patients have intact esophageal defense mechanisms
- UES closes prevent refluxate from entering the laryngopharynx
- Reflux cause injury by several mechanisms:
 - micro aspiration theory: direct contact of acid and pepsin with the epithelium
 - trauma theory: an additional factor, such as vocal abuse or concomitant viral infection, is necessary to induce mucosal lesions
 - esophageal-bronchial reflex theory: acid in the distal esophagus stimulates vagally mediated reflexes that cause chronic cough, which in turn causes laryngeal symptoms and lesions

Symptoms of LPR

- **Key Symptoms:** Dysphonia, Throat clearing, Chronic cough, Dysphagia, Globus pharyngeus, Excessive phlegm, Upright (daytime) reflux, Heartburn (uncommon), Dyspnea.
- Other: Asthma exacerbation (wheezing), Ear pain, Halitosis, Laryngospasm, Neck pain, Odynophagia, PND, Voice complaints (Breaks, Fatigue, Longer warmup time, Loss of upper singing range)

Signs of LPR

- Inflammation such as erythema and edema of the larynx, especially in the postcricoid and hypopharyngeal areas.
- Vocal fold edema.
- diffuse laryngeal erythema and edema
- Pseudosulcus vocalis
- Obliteration of the laryngeal ventricles
- Thickened laryngopharyngeal mucus
- Granulomata
- can be due to other cause of Laryngitis (not specific)

OTHER CLINICAL IMPLICATIONS

- Mechanism: either by direct contact of refluxate and resulting inflammation of the mucosa or via a vagally mediated neurogenic process
- **Otitis Media:** studies showed
 - high rate of acid reflux in patients with OM
 - presence of pepsin within the middle ear
 - presence of bile acids in effusions
 - clearing of effusions with antireflux therapy alone
- **Rhinosinusitis:** studies showed
 - 78% of patients had evidence of reflux
 - Adult with GER more likely to have a history of sinusitis
 - high prevalence of GER with sinusitis in the pediatric population
 - children with medically refractive CRS experienced relief of symptoms and avoided operation after acid suppressive therapy
- **Laryngotracheal stenosis:**
 - Reflux has long been suggested to have a significant role in development of and worsening of LTS
 - Reflux suggested to have a role in the cause of posterior glottic stenosis
 - Over 3/4 of those with LTS had abnormal pH studies
 - 80-100% of patients with **Laryngomalacia** has reflux
 - Reflux acts as a synergistic factor that stimulates granulomatous disease to react and subsequently result in the development of stenosis
 - In most studies, patients taking H2 blockers or PPIs show enhancement of surgical outcomes.
 - Nissen fundoplication is commonly recommended for those with LTS and confirmed EER
- **sleep disordered breathing**
- **Laryngitis, Laryngospasm**
- **Pharyngitis**
- **chronic cough**
- **Lower airway problems** such as asthma, chronic obstructive pulmonary disease, interstitial pulmonary fibrosis, and chronic lung transplant rejection.
- **Laryngeal carcinoma**
- **Esophageal adenocarcinoma**
- **Laryngeal papillomas**
- **Paradoxical vocal-fold motion disorder**
- **Recurrent croup**
- **Reinke edema**
- **Ulceration**
- **Vocal fold granulomas**
- **Dental caries**

Differences between LPR and GERD

- hoarseness was the most prevalent symptom of patients with LPR (100%), but none of the patients with GERD
- Heartburn: GERD (89%); LPR (6%)
- majority of LPR patients have normal Esophageal acid clearance mechanisms
- LPR patients typically have upright (daytime) reflux with good esophageal motor function and no esophagitis, whereas GERD patients have supine (nocturnal) reflux and esophageal dysmotility

DIAGNOSIS

- **The Reflux Symptom Index (RSI)**
 - consists of 9 questions related to symptoms
 - total score >13 (of 45) suggests of LPR

**TABLE
66.1**

THE REFLUX SYMPTOM INDEX

Within the last month, how did the following problems affect you?
Circle the appropriate response.

		0	1	2	3	4	5
		0 = No Problem 5 = Severe Problem					
dysphonia	1. Hoarseness or a problem with your voice	0	1	2	3	4	5
	2. Clearing your throat	0	1	2	3	4	5
	3. Excess throat mucus or postnasal drip	0	1	2	3	4	5
Dysphagia	4. Difficulty swallowing food, liquids, or pills	0	1	2	3	4	5
	5. Coughing after you ate or after lying down	0	1	2	3	4	5
	6. Breathing difficulties or choking episodes	0	1	2	3	4	5
globus	7. Troublesome or annoying cough	0	1	2	3	4	5
pharyngeus	8. Sensations of something sticking in your throat or a lump in your throat	0	1	2	3	4	5
GERD Sx	9. Heartburn, chest pain, indigestion, or stomach acid coming up	0	1	2	3	4	5
	TOTAL						

A score >13 suggests the presence of LPR.

From Belafsky PC, Postma GN, Koufman JA. Validity and reliability of the reflux symptom index (RSI). *J Voice* 2002;16(2):274–277, with permission.

- **Laryngopharyngoscopy**
 - Posterior laryngitis with thickening and pachyderma of the laryngeal posterior commissure and postcricoid mucosa has been classically associated with LPR
 - The reflux finding score (RFS)
 - A score of > 7 (of 26) suggests LPR

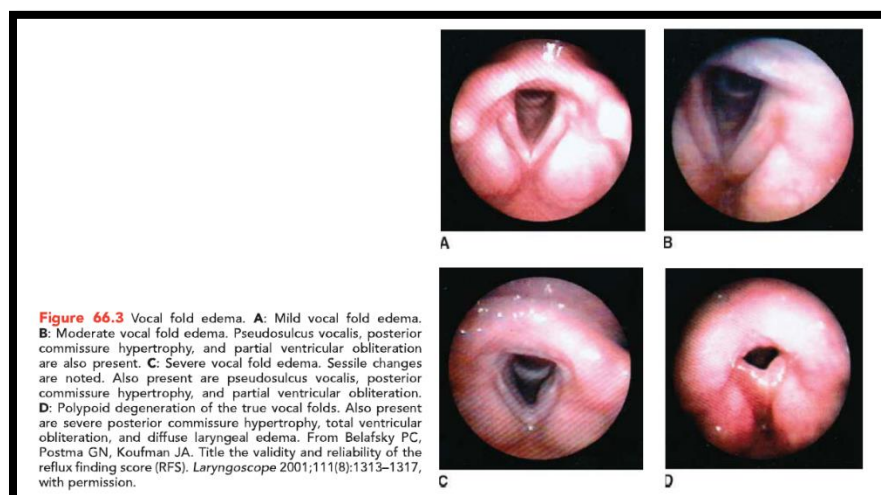
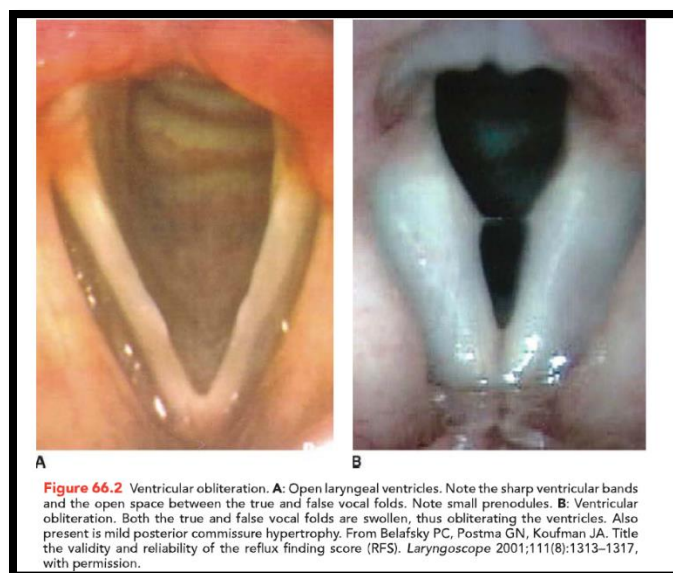
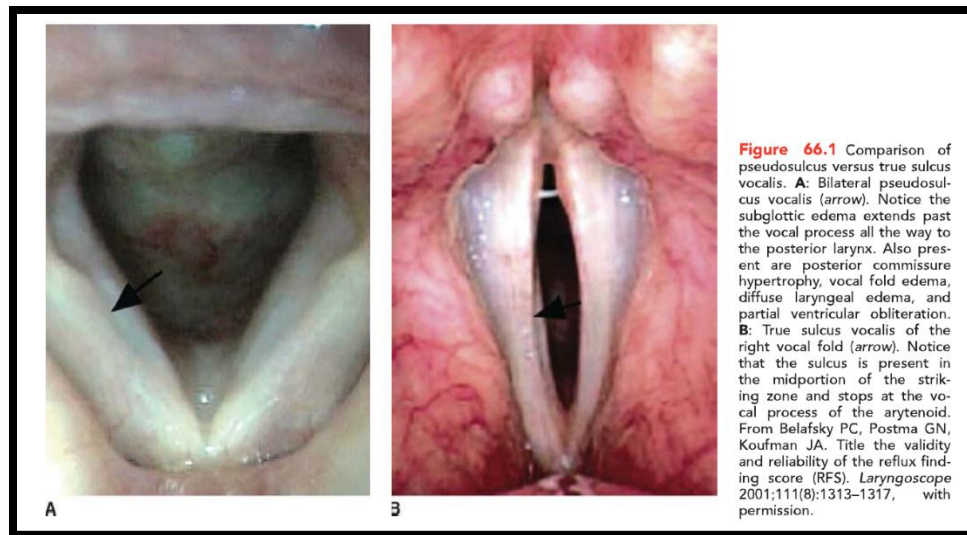
**TABLE
66.2**

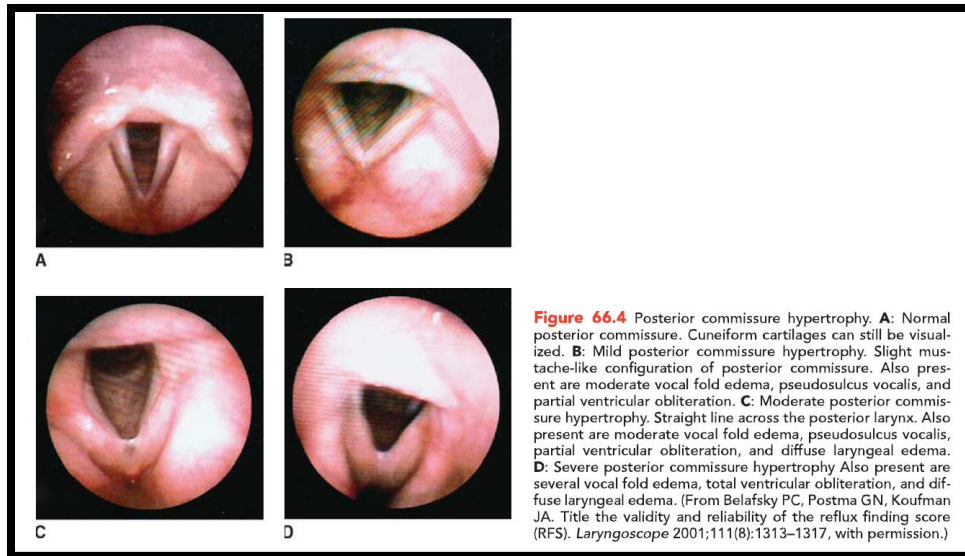
THE REFLUX FINDING SCORE

pseudosulcus	Subglottic edema	0 = absent 2 = present
	Ventricular obliteration	2 = partial 4 = complete
	Erythema/hyperemia	2 = arytenoids only 4 = diffuse
	Vocal fold edema	1 = mild 2 = moderate 3 = severe 4 = polypoid
	Diffuse laryngeal edema	1 = mild 2 = moderate 3 = severe 4 = obstructing
pachyderma	Posterior commissure hypertrophy	1 = mild 2 = moderate 3 = severe 4 = obstructing
	Granuloma/granulation tissue	0 = absent 2 = present
	Thick endolaryngeal mucus	0 = absent 2 = present

A score of >7 suggests LPR. of 26

From Belafsky PC, Postma GN, Koufman JA. Title the validity and reliability of the reflux finding score (RFS). *Laryngoscope* 2001;111(8):1313–1317, with permission.



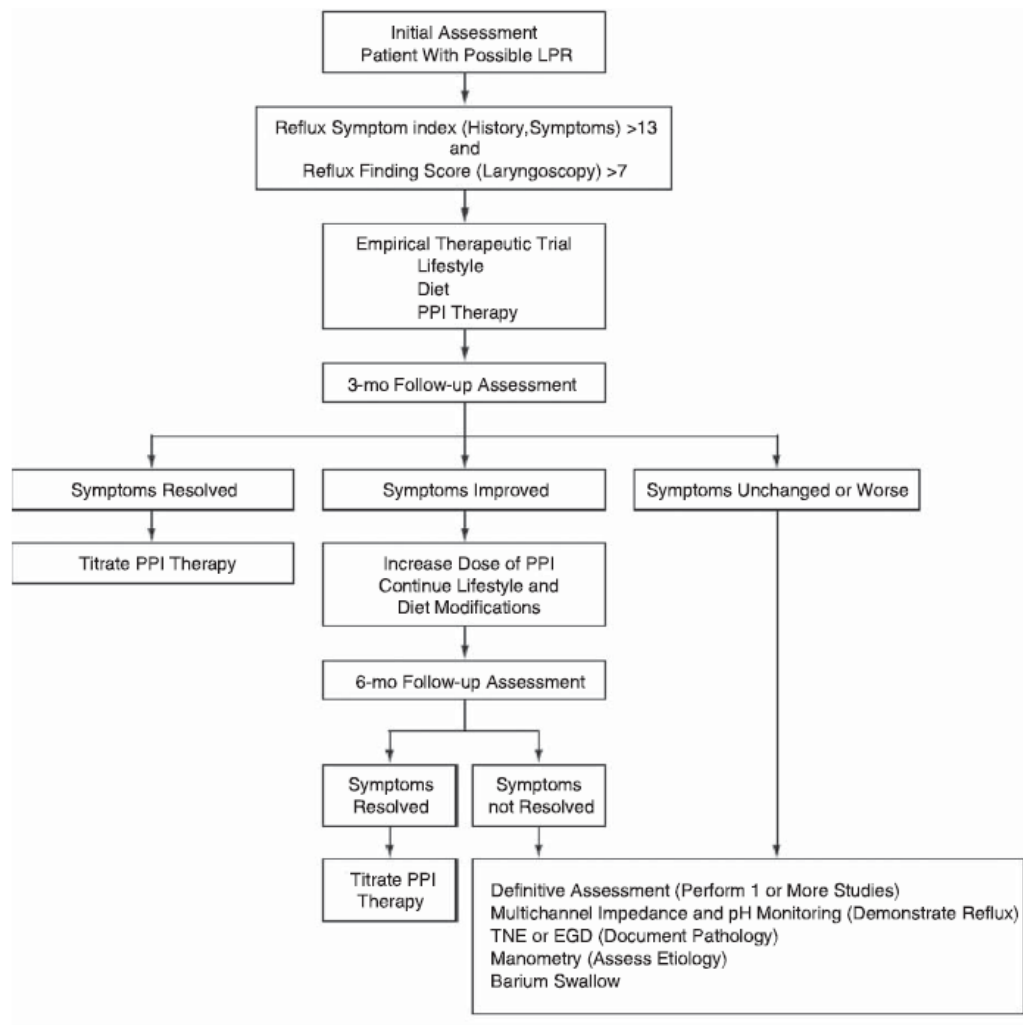


- Esophagoscopy
 - rule out important phenomena such as Barrett's intestinal metaplasia
 - flexible large bore transoral or flexible small bore transnasal telescropy
- **Reflux Detection**
 - The most widely used is a pH monitor
 - single (esophageal)- for GER
 - dual (esophageal and pharyngeal) – for LPR
 - **24-hour ambulatory dual-probe pH monitoring:**
 - is most commonly used (currently **Gold Standard**)
 - placed for 24 hrs while the patient is ambulatory
 - upper probe can be placed just below, at or just above the cricopharyngeus
 - as little as one reflux event to the pharynx is significant enough to diagnose LPR
 - Multichannel Intraluminal Impedance (**MII-pH**) monitoring
 - Capability of detecting acid, weakly acidic, and nonacid events
 - multiple stations (esophagus, stomach, and pharynx)
 - liquid or gas events
 - anterograde (swallow) and retrograde (reflux) direction
- Radiologic Imaging
 - barium swallow, modified barium swallow
 - sensitivity of reflux detection only 25%-33%
- Tissue and Fluids Assay-Pepsin Detection
 - acid or bile salts may or may not be present in Reflux
 - all refluxate contains pepsin
 - Now confined to research

TREATMENT

- **Lifestyle Modification:**
 - Weight loss (≤ 25 kg/m²)
 - Small meal size
 - Avoid lying down within 3 h of eating a meal
 - Eat a low fat, low acid diet
 - Avoid carbonate beverages
 - Avoid spicy food and chocolate
 - Stop smoking
 - Reduce alcohol intake
 - Avoid tight-fitting clothes
- **Medical Treatment:**
 - Bicarbonate Gum (Sugar free)
 - Prokinetic Agents
 - Increase lower esophageal sphincter pressure
 - Increase esophageal motility
 - Promote gastric emptying
 - Metoclopramide, bethanacol, domperidone, bromopride
 - Their use has fallen out of favor in recent years
 - Sucralfate
 - reacts with HCl to form a cross-linking viscous material that acts as an acid buffer for up to 8 hrs
 - mucosal protection
 - Alginate
 - Gaviscon Advance
 - react with stomach acid to form a gel raft that floats on top of the stomach, helping to keep gastric contents in the stomach and preventing GER
 - licensed in the UK for the treatment of LPR
 - improving symptoms, laryngeal findings, and patient QOL
 - **H₂ Receptor Antagonists**
 - effective in up to 80% GERD patients
 - only effective in approximately 50% LPR
 - **Proton Pump Inhibitors**
 - irreversibly inhibit the H⁺ /K⁺ ATPase enzyme
 - Treats acid refluxate only
 - very effective for the treatment of GERD
 - treatment of LPR have shown variable results
 - **empiric therapy with PPIs remains the backbone of medical management of LPR**
 - omeprazole, lansoprazole, esomeprazole, pantoprazole, rabeprazole, and dexlansoprazole
 - twice daily dosage is recommended for LPR
 - side effects: bloating, constipation, or diarrhea
 - Long-term sequelae
 - malabsorption of vitamin B12 and calcium
 - increased rate of hip fracture

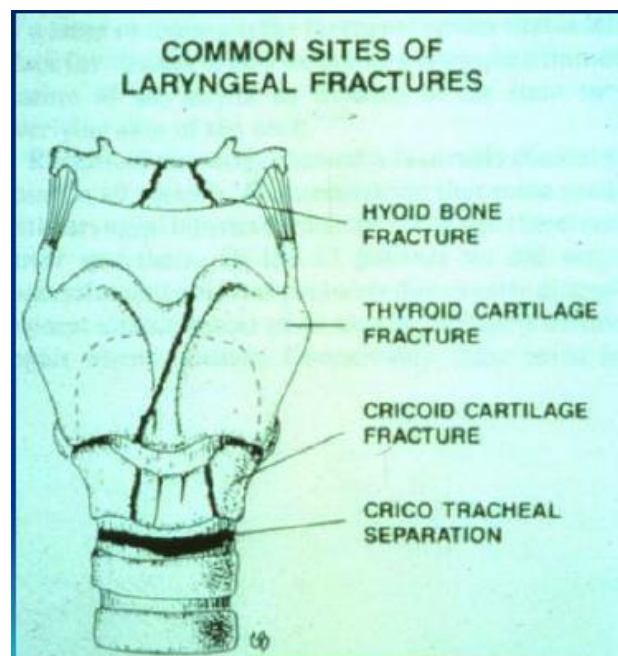
- alteration of the gastric milieu and bacterial flora
 - increased risk of pneumonia
 - decrease anticoagulant activity of clopidogrel (Plavix)
- **Surgical Management**
 - **A favorable outcome:**
 - extra esophageal symptoms are nocturnal
 - esophageal and laryngeal acid exposure is identified by pH monitoring
 - there is an association between extraesophageal symptoms and reflux (both acidic and nonacidic)
 - there is a positive response to medical therapy
 - **Fundoplication**
 - **Complete** (360°) wrap, as in the **Nissen** or Rossetti procedures
 - **Partial** (270°) wrap, as in the Toupet or Bore procedures
 - The most common reported GE junction operation for EER is the laparoscopic Nissen fundoplication
 - 80% improvement
 - up to 20% of patients may need to return to PPI treatment of their condition after 10 years
 - **Reserved for:**
 - medically refractory EER
 - the more severe sequelae of EER such as airway stenosis, reflux-associated asthma, dysplasia or cancer



لا تنسوني من دعواتكم... أخوكم يحيى فقيه

Laryngeal Trauma

- Larynx contains the narrowest region of the ventilatory conduit, houses the phonatory apparatus, and functions to protect the airway during swallowing
- Laryngeal trauma is a relatively uncommon
- Considered life threatening
- If not promptly recognized and treated, it can cause significant long-term morbidity
- Each case of external laryngeal trauma presents a unique set of problems, but despite the diversity of injuries, specific management guidelines can be applied.
- **Severity of injury** and **delay in treatment** correlate with poor outcome
- Laryngeal trauma is often associated with concomitant **cervical** and intracranial injuries and frequently occurs as part of multisystem poly-trauma. (Multidisciplinary team)
- The larynx is structurally protected by
 - The mandible superiorly
 - The sternum inferiorly
 - The sternocleidomastoid muscles laterally
 - The spinal column posteriorly
- Further protection is afforded through its mobility
- It is susceptible to crushing **anterior** cervical trauma, which compresses it against the spinal column (mainly with extended neck)
- Laryngeal calcification increases the likelihood of fractures following trauma (more in men)



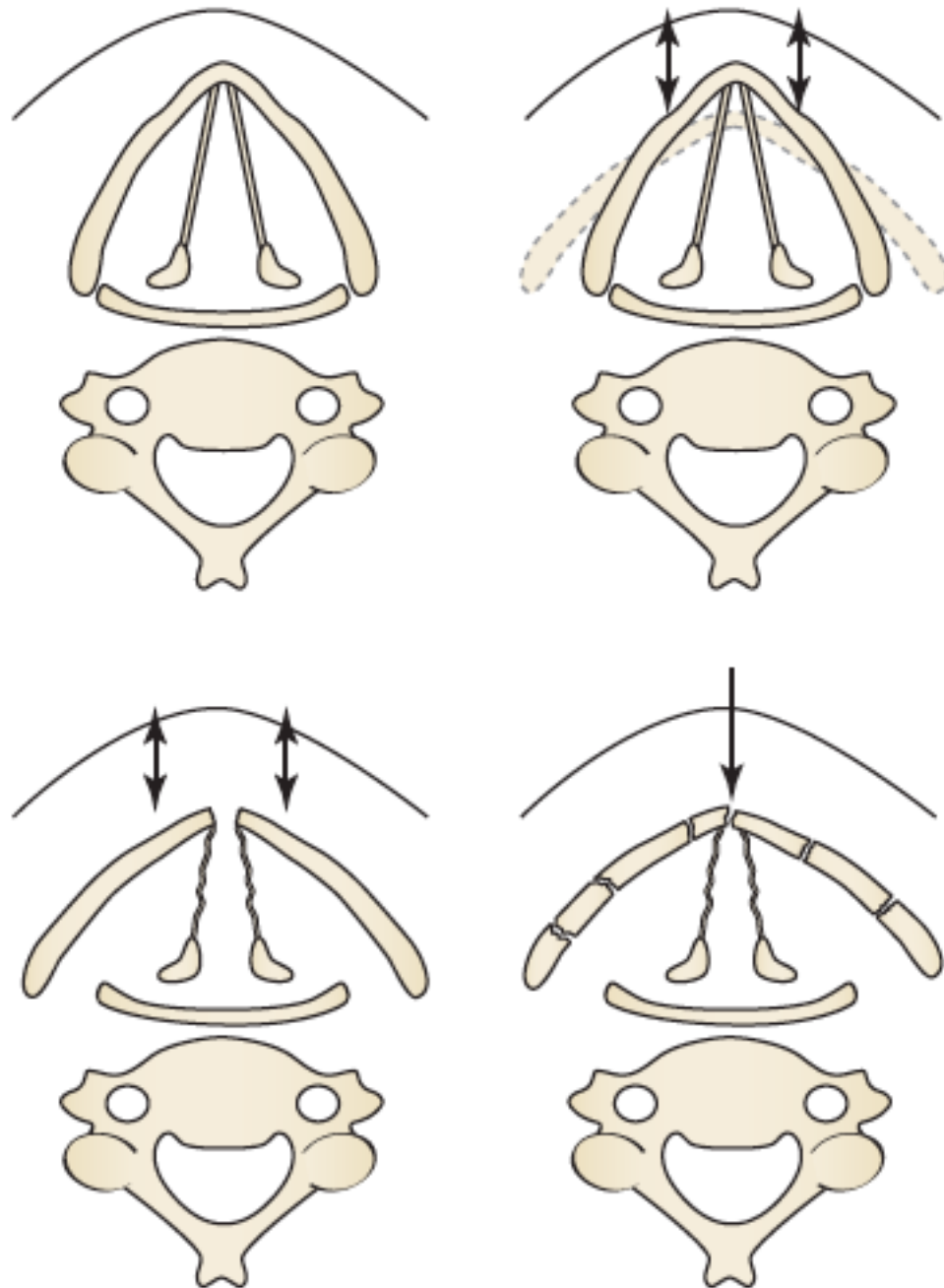


FIGURE 67-2. Impact of anterior cervical trauma on the larynx. *Top:* Uncalcified thyroid cartilage may fracture but is more likely to spring back. *Bottom:* Calcified cartilage may shatter, and the larynx appears flattened.

MECHANISMS OF LARYNGEAL TRAUMA

Broadly divided into:

- External trauma, (blunt or penetrating)
- Internal trauma, (iatrogenic, thermal, caustic)

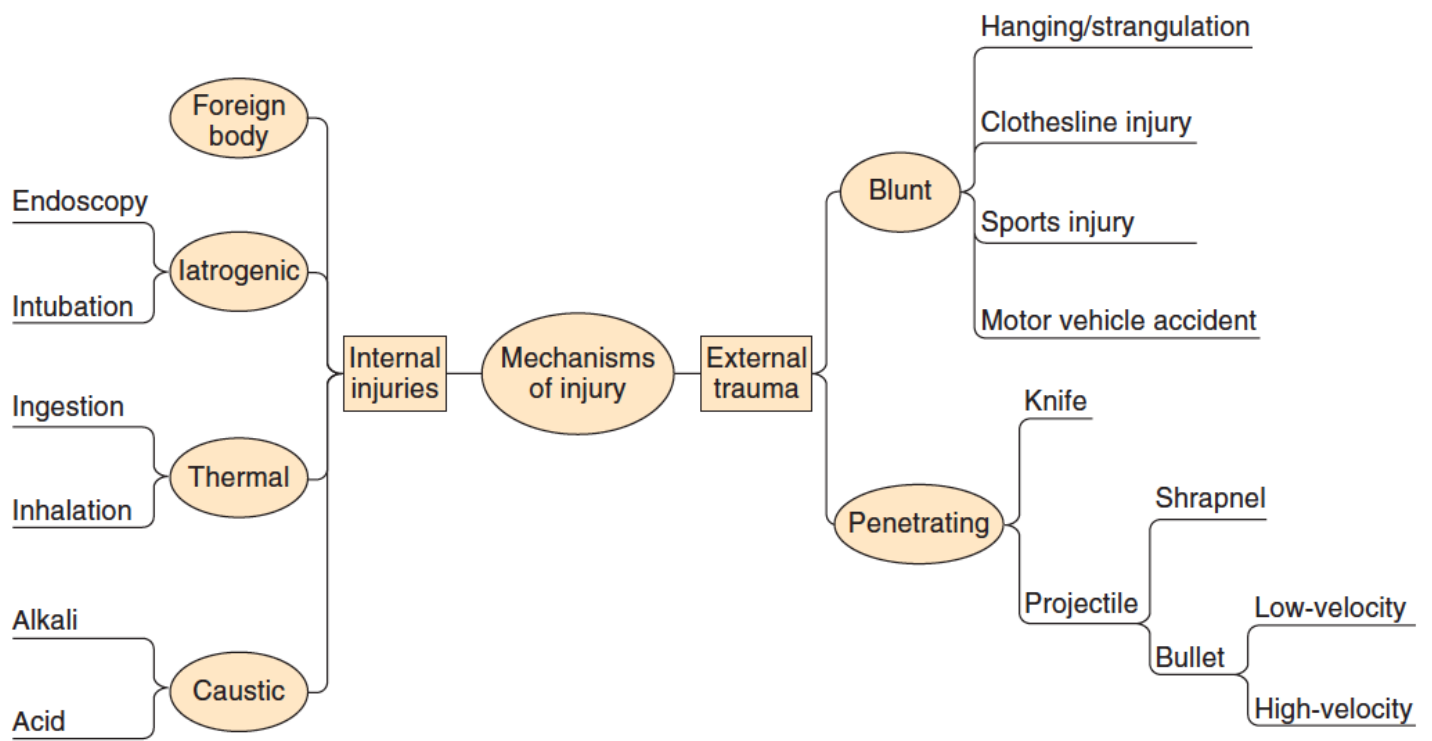


FIGURE 67-1. An overview of mechanisms of laryngeal and esophageal injury.

EXTERNAL TRAUMA

Incidence

- External laryngeal trauma is rare.
- It has a population incidence of 1 in 137,000 in adults
- Accounts for 0.5% of trauma admissions in children.

Blunt Trauma:

- Motor vehicle accidents (most common)
- Clothesline injury (encounters fixed horizontal object at neck level)
(Commonly cause laryngotracheal separation)
- Personal assaults
- Sports injuries
- Manual strangulation or hanging-type

Moderate blow to the larynx => the momentum of the vocal folds causes a shearing effect between the vocalis muscle and the internal perichondrium
=>This results in injuries such as endolaryngeal mucosal tears, edema, or hematoma

More severe trauma produces fractures of the laryngeal cartilages and disruption of the laryngeal ligaments.

Subluxation or dislocation of the arytenoid cartilage can produce an immobile vocal fold.

Unilateral injury to the recurrent laryngeal nerve often is associated with cricoarytenoid joint injuries owing to the proximity of the recurrent laryngeal nerve to the cricoid cartilage.

Bilateral injury often associated with cricotracheal separation

Structural integrity of the cricoid cartilage is essential in airway maintenance.
(The only complete ring of the airway)

Fractures of the hyoid bone associated with epiglottic injuries
The greater or lesser cornu of the thyroid cartilage can lacerate the pharyngeal mucosa

Penetrating Trauma

- Knife and gunshot wounds, blast
- Injuries vary from minor lacerations to severe disruption of the cartilage, mucosa, soft tissue, nerves, and adjacent structures.
- Penetrating trauma is more likely to damage additional cervical structures
- Gunshot wounds are more associated with severe tissue damage
- High-velocity projectiles cause greater tissue destruction and wound contamination than low-velocity
- Knife wounds cause less peripheral soft tissue damage
- Death from penetrating trauma may be caused by complete disruption of the larynx, massive soft tissue edema, or associated neurovascular injuries.

DIAGNOSIS AND EVALUATION

- **Mechanism of Injury.**
- **History**
- Any patient with anterior neck trauma is considered to have an upper airway injury.
- No single symptom seems to correlate well with the severity of injury

TABLE 67-1. Symptoms and Signs of Laryngeal Trauma

	Patients (N = 33)
Hoarseness	28 (85%)
Dysphagia	17 (52%)
Pain	14 (42%)
Dyspnea	7 (21%)
Hemoptysis	6 (18%)

- When the laryngeal lumen is severely compromised, aphonia and apnea occur
- Cardiopulmonary arrest may result because of airway obstruction
- Other Sx: dysphagea, odenophagea, can't tolerat supine position

- **Physical Examination**
- Examination and initial management of laryngeal trauma should be part of primary and secondary surveys
- **Priority** is to establish a safe airway with cervical spine protection (c-spine injury in 10% of cases)
- Breathing: Resp rate, saturation, Type of strider if present (inspir-expir-combined)
- Pt may need emergent **trach under local anesthesia**
- Endotracheal intubation is **not a preferred** method of airway control in laryngeal trauma, because it can exacerbate a laryngeal injury and precipitate total airway obstruction
- If endotracheal intubation has been performed, it should be converted at the earliest opportunity—no longer than 24—to a tracheotomy to prevent long-term laryngeal injury
- In selected cases, transtracheal jet ventilation or cricothyroidotomy may be helpful temporizing measures

1. Signs of vascular injury

- Active bleeding
- Expanding hematoma
- Bruits
- Loss of pulses
- The neck is **inspected** for evidence of injury—such as skin abrasions, bruising, emphysema and entry and exit wounds in penetrating trauma, trachea deviation
- Look for active bleed, exposed cartilage, bubbles from the wound
- **Palpated** for crepitations, laryngeal tenderness, and any obvious changes in laryngeal anatomy such as loss of the prominence
- **Open wounds** should not be explored at this stage, because instrumentation may restart hemorrhage
- o **Flexible nasoendoscopy** is then performed
- Oropharynx and hypopharynx are examined for injuries.
- The laryngeal mucosa is examined for lacerations and hematomas
- Arytenoids and vocal fold mobility
- Any exposed or protruded cartilage or submucosal distortion of the framework.
- A sign of cartilaginous injury is failure of the vocal cords to meet in the same horizontal plane.
- In patients with minor laryngeal injuries, stroboscopy should be performed to better assess vocal fold function

- **Radiologic Evaluation**
- Computed tomography (**CT**) is the most useful radiologic examination
- **Assess** larynx **framework**, **C-spine** and **adjacent** structures and vessels
- CT scan can be **avoided** in Patients with minimal anterior neck trauma and normal physical findings, stable airway

Box 67-1. NATIONAL EMERGENCY X-RADIOGRAPHY UTILIZATION STUDY (NEXUS) CRITERIA FOR CERVICAL SPINE IMAGING IN TRAUMA PATIENTS*

1. No posterior midline cervical tenderness
2. No evidence of intoxication
3. Normal level of alertness
4. No focal neurologic deficit
5. No painful distracting injuries

*According to NEXUS low-risk criteria, cervical spine radiography is indicated for trauma patients unless they exhibit *all* the listed criteria (sensitivity 99%).

- CT useful to assess poorly visualized areas, such as the subglottic and anterior commissure regions, and to identify associated cervical injuries
- Chest X-ray helpful to rule out pneumothorax
- **Arteriography**, CT or MRI angiography, color Doppler as indicated if suspecting vascular injury
- Pharyngeal and esophageal mucosal penetration can be identified with the use of a Gastrografin followed by barium swallow examination.

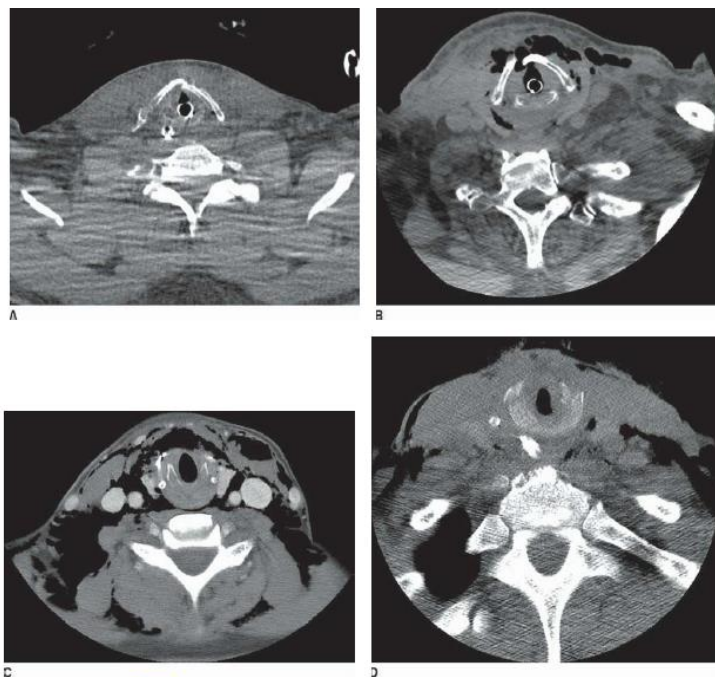


Figure 77.1 A: Minimally displaced laryngeal fracture. B: Moderately displaced laryngeal fracture. C: Severely displaced laryngeal fracture. D: Displaced laryngeal fracture.

**TABLE
77.1**



**DIAGNOSIS
LARYNGEAL TRAUMA**

Symptoms

- Hoarseness
- Pain
- Dyspnea
- Dysphagia

Signs

- Stridor
- Hemoptysis
- Subcutaneous emphysema
- Laryngeal tenderness
- Loss of thyroid cartilage prominence
- Vocal fold immobility
- Laryngeal hematoma
- Laryngeal edema
- Laryngeal lacerations

Radiology

- Computed tomography
- Arteriography
- Cervical spine radiography
- Contrast esophagography

Classification of Laryngeal Trauma.

Box 67-2. SCHAEFER-FUHRMAN CLASSIFICATION OF LARYNGEAL TRAUMA

Group 1: Minor endolaryngeal hematomas or lacerations; no detectable fracture

Group 2: Edema, hematoma, minor mucosal disruption without exposed cartilage; nondisplaced fracture; varying degrees of airway compromise

Group 3: Massive edema, large mucosal lacerations, exposed cartilage; displaced fracture(s); vocal cord immobility

Group 4: Same as group 3 but more severe with:

Severe mucosal disruption

Disruption of the anterior commissure

Unstable fracture, two or more fracture lines

Group 5: Complete laryngotracheal separation

Lee-Eliashar system:

Table 1. Classification of laryngotracheal trauma according to the degree of the injury

Type	Degree	Symptoms	Signs (in order of their incidence)
1	Mild	Mild voice change, mild dyspnea, cough	Minor hematomas, small lacerations, no fractures or dislocations
2	Moderate	Compromised airway, hemoptysis	Obstructing hematoma, edema, minor mucosal disruption, nondisplaced fractures
3	Severe	Severe airway compromise, stridor	Massive edema and hematoma, deep mucosal tears, exposed cartilage, aspiration, displaced fractures, unilateral vocal fold immobility
4	Profound	Impending airway obstruction	Massive edema, mucosal avulsion, fragmented cartilage, aspiration, displaced arytenoids, bilateral vocal fold immobility
5	Critical	Complete airway obstruction	Skeletal collapse, structural disruption and breakdown, complete laryngotracheal separation

MANAGEMENT

Goal is to restore function (airway, protection, phonation)

1- **Airway Management.** While protecting the cervical spine

- Respiratory distress at presentation should immediately proceed to **tracheotomy under local anesthesia.**
- If pt stable but flexible showed unexpected airway-encroaching injuries are identified at this stage, and the patient proceeds to tracheotomy
- More subtle endolaryngeal injuries are further assessed with CT and again, if airway-encroaching injuries are identified, the patient proceeds to tracheotomy
- **Do not** attempt to intubate, might make things worse

2- **Surgical Versus Nonsurgical Therapy.**

- Decision regarding the most appropriate therapy for laryngeal trauma rests on
 - 1) The stability of, and the extent of injury to, the laryngeal framework
 - 2) The extent of mucosal injuries
 - 3) Injury to the vibratory apparatus
 - 4) The integrity of the laryngotracheal junction

Aim to repair all laryngeal injuries within 12 hours of presentation and not to delay beyond 24 hours

Delays in treatment can lead to granulation and scar tissue formation that can progress to laryngeal stenosis

Nonsurgical (conservative):

- Patients with a normal endolarynx on nasoendoscopy and patients with minimal endolaryngeal abnormalities and a stable laryngeal framework whose airway patency has been confirmed by CT
- Minimum of a 24-hour admission to a high-dependency unit
- Regular observations
- Head is elevated to reduce further edema
- Voice rest (48-72 hrs)
- Serial flexible nasoendoscopy
- Use of humidified oxygen (mixture of oxygen and helium should be on standby in case respiratory distress develops)
- Corticosteroids (within 24hrs of injury)
- All patients are prescribed a proton pump inhibitor
- If the laryngeal mucosa has been breached, patients are also given prophylactic broad-spectrum antibiotics

Surgical options:

- Endoscopy alone
- Endoscopy with exploration
- Endoscopy with exploration and stenting

Surgical goal:

- Homeostasis
- Evacuation of hematoma
- Reconstruction of laryngeal framework
- Coverage of de-epithelialized surfaces

• **Diagnostic/therapeutic endoscopy:**

- All patients who are not managed conservatively should undergo a microlaryngoscopy and tracheoscopy, direct pharyngoscopy, and direct esophagoscopy.
- Hematomas can be drained
- Mucosal and selected vocal cord lacerations can be repaired
- Dislocated cricoarytenoid joints can be reduced.
- If injuries to opposing laryngeal mucosal surfaces are extensive, stents or keels can also be deployed endoscopically to prevent adhesions

• **Open laryngeal repair.**

- **Indications:**

1. Displaced, unstable, multiple or comminuted laryngeal fractures
 2. Cricotracheal separation
 3. Detachment of the anterior commissure
 4. Extensive mucosal disruption and lacerations
 5. Exposed cartilage
 6. Complete Vocal fold immobility or joint CA joint disruption
- Transverse neck incision is placed over the cricoid=>and subplatysmal flaps are elevated=>Strap muscles are separated=> anterior vertical laryngofissure.

Box 67-3. FACTORS THAT DETERMINE THE NEED FOR AND THE NATURE OF SURGICAL INTERVENTION

Laryngeal Framework

Stable

- No fractures
- A single undisplaced fracture

Unstable

- A single displaced fracture
- More than one fracture line
- Cricoid fracture

Potentially nonviable

- Framework comminution with devitalized cartilage fragments

Laryngeal Mucosa

Intact/minimally injured

- No mucosal injuries
- Small submucosal hematoma
- Linear laceration with no exposed cartilage

Injured

- Jagged/multiple linear lacerations
- Large hematoma(s)

Exposed Cartilage

Massively injured

- Significant loss of mucosa
- Devitalized mucosal tissue

Vibratory Apparatus

Intact

Injured

- Anterior commissure
- Vibrating edge of the vocal cord(s)
- Arytenoid dislocation

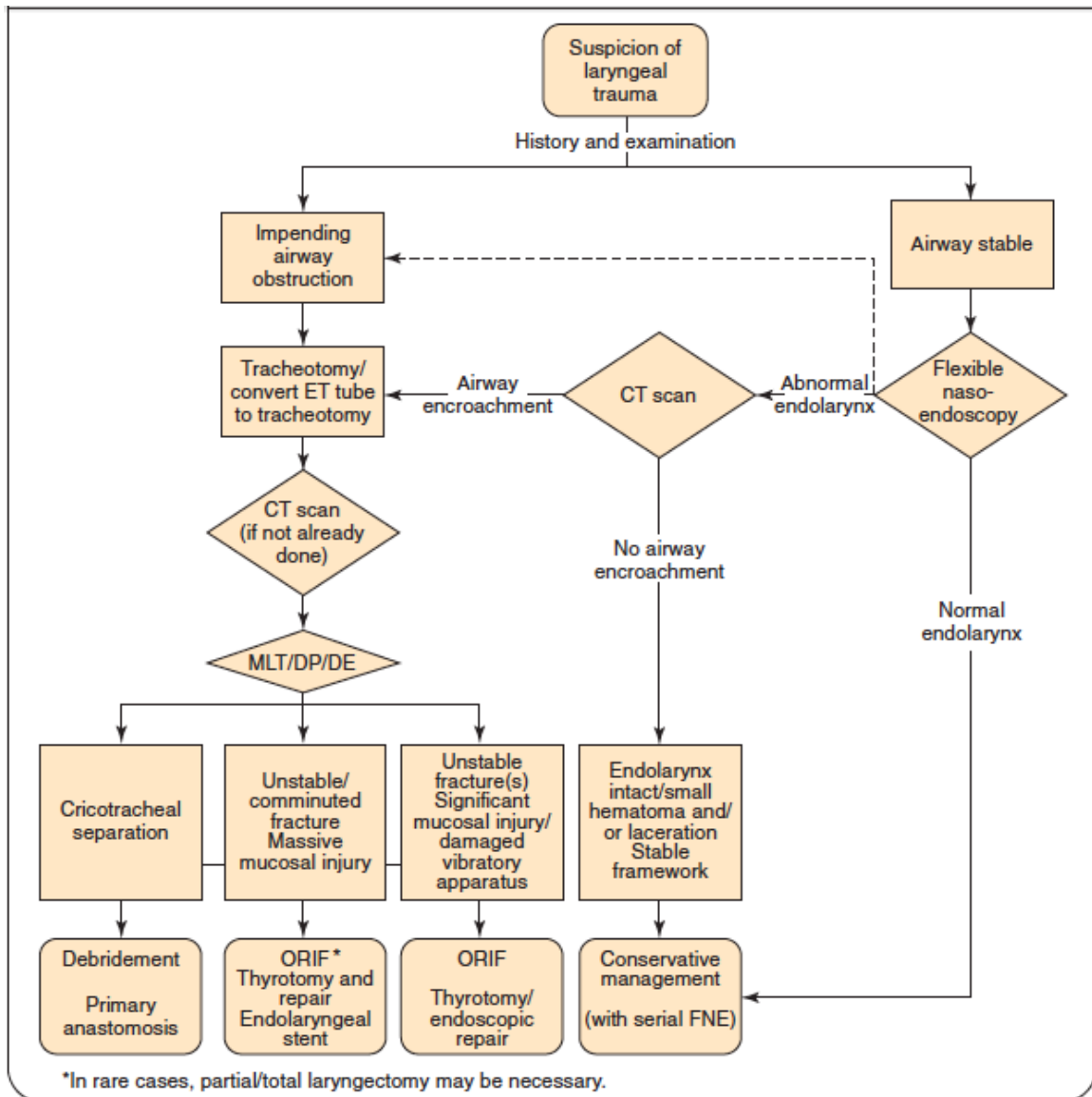
Laryngotracheal Junction

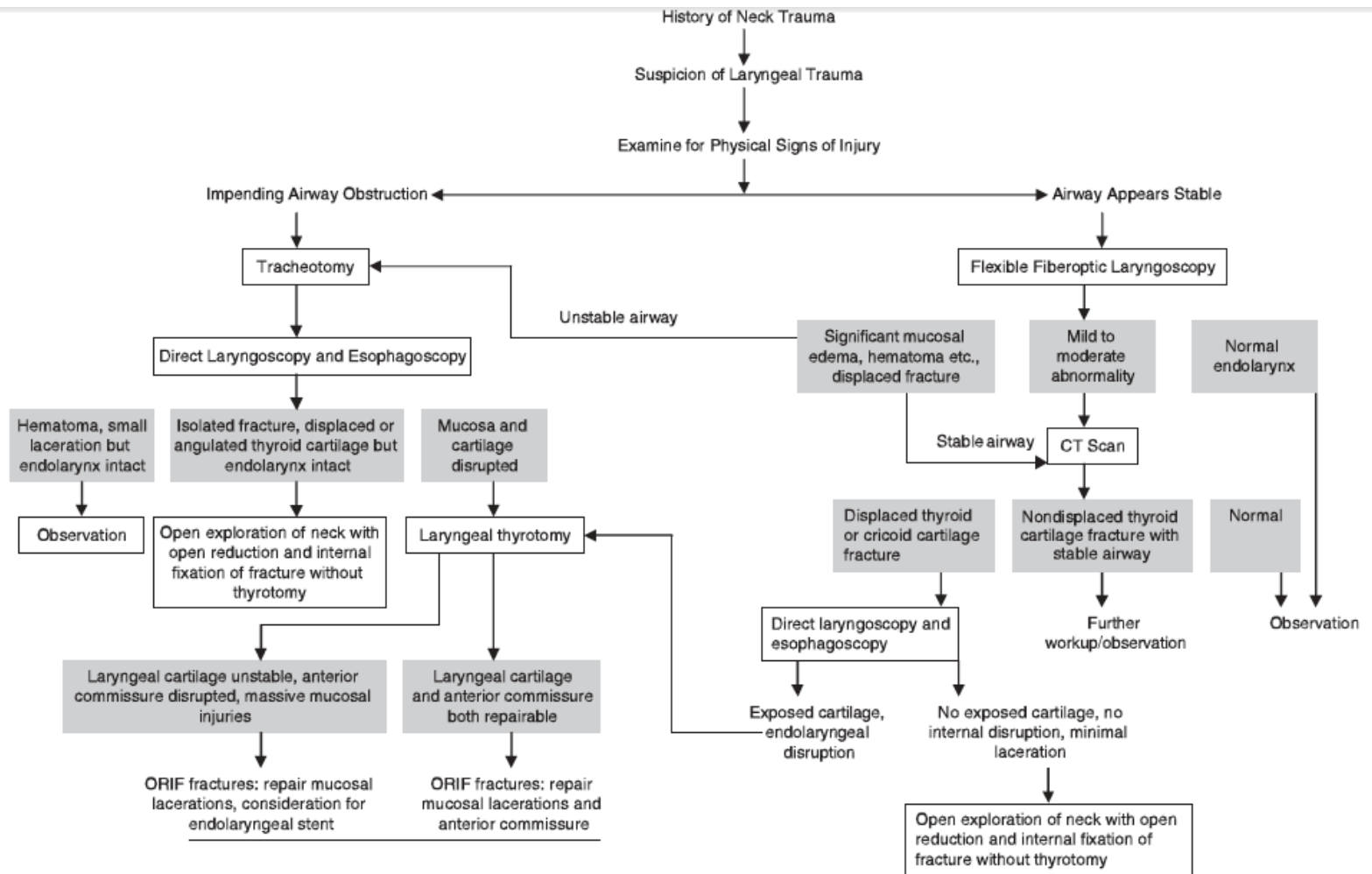
Intact

- Any degree of laryngotracheal separation

- **Endolaryngeal stenting:**

- Primary indication for stenting is significant **framework comminution**.
- Stents can also be used to prevent **anterior commissure** webbing in cases of bilateral vocal fold epithelial loss
- **Massive mucosal injury**
- The stent is removed endoscopically 10 to 14 days later





Management table

Group	Management
1	-Conservative: head elevation Steroids. Antibiotics. Anti-reflux medications. Humidification. Voice rest
2	-Should be serially examined -Injuries may worsen or progress with time. -Occasionally, these injuries may require a tracheotomy. -Medical adjuncts may also be helpful (Steroids, anti-reflux medications, humidification, voice rest, antibiotics).
3	-Tracheotomy is often required. -Exploration and surgical repair -The following injuries will require surgical repair: • Disruption of anterior commissure. • Major endolaryngeal lacerations. • Tear involving vocal cord. • Immobile vocal cord. • Cartilage exposure. • Displaced cartilage fractures. • Arytenoid subluxation or dislocation.
4	-Tracheotomy is always required. -Surgical repair of these injuries will require stent placement to maintain integrity of the larynx
5	-Urgent airway evaluation and management. -Tracheotomy is necessary to secure the airway, but can be very difficult due to the altered anatomy. -Complex laryngotracheal repair must be performed through a low cervical incision (see below) after the airway is secured.

**TABLE
77.3**



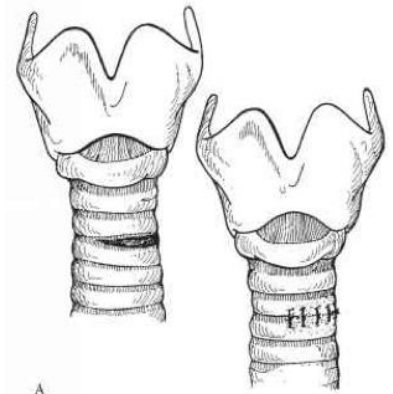
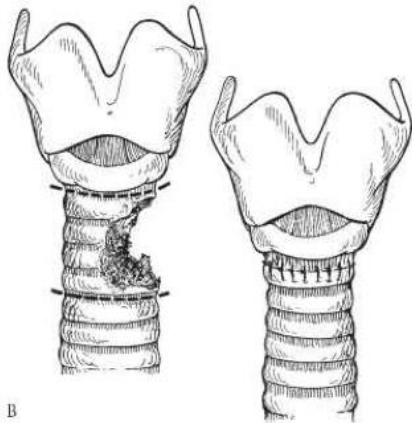
**TREATMENT
LARYNGEAL TRAUMA**

Medical

Voice rest
Systemic steroids
Elevate head
Humidified air
Antibiotics
Antireflux measures

Surgical

Tracheotomy
Endoscopy
Exploration
Thyrotomy
Closure of lacerations
Insertion of stents for disrupted anterior commissure
Grafting for severe mucosal loss only
Fixation of fractures

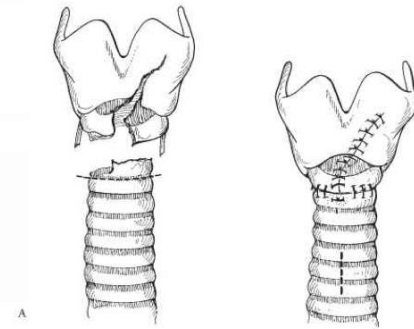


Special Considerations:

- In fracture injury, the cartilage reduction and fixation should be performed prior to endolaryngeal soft tissue repair (Suture, wiring or miniplates)
- All major endolaryngeal lacerations should be repaired with 5-0 or 6-0 absorbable suture.

- Vocal Cord Immobility.
 - Assessed by scope, strob, microlaryngoscopy and instrumentation of joint, EMG
 - CA joint dislocation=>endoscopic manipulation and reduction
 - Once reduced, the arytenoid tends to be held in place by the remaining muscles and ligaments but also by the fact that the cricoid facet has some depth, into which the arytenoid sits
- RLN injury:
 - Only if complete palsy then exploration is indicated
 - If anatomically intact nerves should be allowed to regenerate
 - Primary anastomosis with no tension
 - Cable graft from greater auricular or sural
 - The other option is to consider ansa cervicalis to recurrent laryngeal repair (Nerve repairs do not restore the intricate motor function of the larynx but may provide sufficient muscle tone to improve vocalization)

- Management of Cricotracheal Separation
- Following surgical exposure, a tracheotomy is fashioned or moved to a healthy part of the lower trachea prior to reanastomosis
- The repair begins with the posterior anastomosis using a combination of 3-0 absorbable and nonabsorbable sutures, working toward the anterior trachea (knot is out side the lumen to avoid granulation)
- Avascular and damaged tissues are resected
- The neck is kept in flexion for 7 days postoperatively to prevent traction on the anastomosis
- A large monofilament suture from the submental skin to the anterior chest skin, placed at the time of surgery, may be necessary for the first few postoperative days
- Partial/Total Laryngectomy.



In cases of massive laryngeal injury with significant tissue loss

- **Sequel of laryngeal Trauma**
- All patients with laryngeal trauma should be regularly followed up for a minimum of 12 months
- Long-term morbidity may be due to any combination of laryngeal stenosis, dysphonia, and aspiration as a consequence of both structural and neurovascular injuries
- In patients with a normal vibratory apparatus structure and unilateral vocal cord paralysis, definitive medialization procedures should only be attempted after 6 months of observation for functional recovery.
- In cases of bilateral vocal cord palsy that causes laryngeal inlet obstruction, consideration should be given to retaining the tracheotomy in preference to performing an early laser arytenoidectomy in anticipation of return of vocal cord function.
- Laryngeal stenosis as a result of posterior commissure or tracheal granulation tissue should be promptly treated with intralesional steroid injections and reduction of granulation tissue.
- The granulation tissue should not be allowed to mature into a scar
- Outcome is dependent both on the nature and severity of the original injury and on whether the injury was promptly recognized and adequately treated

Special Considerations in Pediatric External Laryngeal Trauma

- The neonatal larynx lies at the level of C3 and descends during the first 3 years of life to its adult position at the level of C6. (in pediatric larynx better shielded)
- The pediatric larynx is smaller in absolute and relative dimensions compared with the adult larynx.
- Laryngeal mucosa is less firmly adherent to the cartilaginous framework in infants and children (easy edema and hematoma)
- It is usually not possible to perform tracheotomies under local anaesthetic in children
- so start with endotracheal tube or ventilation through bronchoscope then Trach

IATROGENIC LARYNGEAL TRAUMA

- Endotracheal intubation remains by far the most common cause of laryngeal trauma.
- Approximately 10% of patients have demonstrable laryngeal pathologies 1 day after short-term intubation for surgery.
- With longer term intubation and mechanical ventilation for critical illness, the incidence of laryngotracheal injuries approaches 90%
- The incidence of postintubation laryngotracheal stenosis that requires surgical correction is 1 in 204,000 Adults and 4.9 in 100,000 in children per year
- Larynx can also be injured by a poorly situated tracheotomy, either through or adjacent to the cricoid
- Another iatrogenic cause of laryngeal injury is aggressive treatment of benign laryngeal diseases

Etiology

- Pharyngeal or laryngeal lacerations
- Cricoarytenoid joint dislocation
- Neurapraxia of the lingual, hypoglossal, and laryngeal nerves
- No period of endotracheal intubation exists below which no injury occurs and above which injury is inevitable
- In pediatric => subglottic
- In adult => posterior commissure

Prevention is the best approach in relation to this injury

Performing early tracheotomies can significantly reduce occurrence

CAUSTIC/THERMAL LARYNGEAL TRAUMA

- Majority of injuries are caused by ingestion of alkaline substances.
- Thermal injury to the larynx is a sequela of inhaling superheated air. Inhalation burns occur in 30% of all burn patients
- 20% of patients with inhalation injury have extensive laryngeal injury.
- 7% of patients with inhalation injury have both laryngeal and tracheobronchial injuries

Etiology

- Alkali causes liquefaction necrosis of muscle, collagen, and lipids and creates an injury that worsens with time.
- Acids cause coagulation necrosis of the superficial tissues
- In children=> usually its an accident=>small amount
- In adult=> usually attempting suicide=> large amount
- Inhalation injury is typically a consequence of fires in closed spaces and causes a sixfold increase in the mortality rate associated with the burn

Management

- All pts should be admitted for a minimum of 24 hours for airway observation.
- The first priority is the establishment of a safe airway
- Followed by cardiovascular resuscitation as per standard burn-care protocols
- Presence of facial or body burns, soot in the oral cavity, and an endoscopic finding of laryngeal edema predict the need for airway intervention
- If patients are to undergo a microlaryngoscopy, tracheobronchoscopy, or esophagoscopy, these should be within 24 hours of injury.
- Beyond this time, edema and ulceration are more marked, and instrumentation may exacerbate subsequent problems.
- In cases of caustic injury, the upper aerodigestive tract should be irrigated to remove any residual substances

Sequel of castic and thermal injury (stricture, dysphonia 70% of inhalation injury)

**TABLE
77.4**



**COMPLICATIONS
LARYNGEAL TRAUMA^a**

Granulation tissue

- Prevent by covering all exposed cartilage
- Avoid stents when possible
- Careful excision

Laryngeal stenosis

- Excision with mucosal coverage
- Stenting selected cases
- Laryngotracheoplasty
- Tracheal resection with reanastomosis

Vocal fold immobility

- Observe
 - Vocal fold injection
 - Thyroplasty-type vocal fold medialization
 - Arytenoidectomy and vocal fold lateralization for bilateral paralysis
-

Pediatric:

- 1- Clinical Evaluation of Airway Obstruction**
- 2- Subglottic Stenosis**
- 3- Laryngeal Web, Atresia, PGS, Clefts, Fistula**
- 4- Congenital Vascular Anomalies**
- 5- Tracheal Anomalies and Vascular Rings**
- 6- Neonatal Nasal Obstruction**
- 7- Aerodigestive Foreign Bodies**
- 8- Caustic and Corrosive Ingestions**
- 9- Congenital Pediatric Neck Lesions**
- 10- Pediatric Syndromes**

Clinical Evaluation of Airway Obstruction (Stridor, Aspiration, Cough):

- Any upper or lower airway obstruction generates a pathological respiratory sound resulting from the rapid, turbulent flow of air through the narrowed segment of the respiratory tract.
- Other symptoms and signs of respiratory distress may occur such as:
 - Dyspnea
 - Nasal flaring
 - Supraclavicular, sternal, intercostal and subcostal retractions
 - Grunting (short deep hoarse voice due to sudden glottic obstruction)
 - Life-threatening cyanotic episodes
 - Somnolence and apnea
- **Degree of Respiratory Distress:**
 - In clinical practice, the symptoms of respiratory obstruction can be graded as follows:
 - 1. Grade I (Mild):**
 - Noisy breathing with mild to moderate dyspnea and chest retractions.
 - No anxiety or restlessness.
 - Normal intake of food and drink
 - Interest in playing
 - 1. Grade II (Moderate):**
 - Noisy breathing with severe dyspnea and chest retractions.
 - Anxiety and restlessness.
 - Refusal of food and drink.
 - No interest in playing.
 - 1. Grade III (Severe):**
 - Exhausted child with decreased stridor and intercostal muscle retractions.
 - Slowing down of respiratory rate and heart rate.
 - Ashy-grey color and perspiration.
 - Spells of somnolence.
- If the brief clinical examination reveal **SEVERE** (Grade III) or **PROGRESSIVE** respiratory distress in a child with an impending airway compromise:
 - Immediate airway stabilization in the operating room is mandatory (Endotracheal intubation +/- Tracheostomy).
- Otherwise, in patients with STABLE Mild (Grade I) +/- Moderate (Grade II) respiratory distress:
 - Complete history, physical examination and radiological investigation should be obtained.

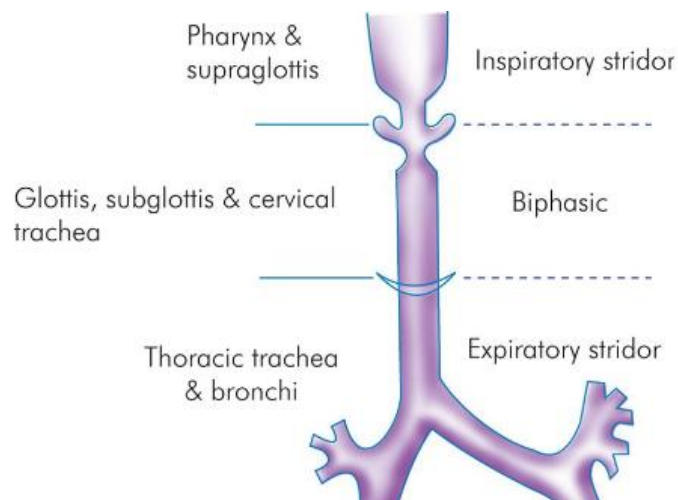
Site and Cause of Airway Obstructions:

- The assessment of a dyspnoeic infant or child is based on the following factors:
 1. Quality of pathological respiratory sounds.
 2. Phase of the respiratory cycle during which the pathological sounds are produced.
 3. Assessment of the three main functions of the larynx.
 4. Medical history.

• Pathological Respiratory Sounds:

1. Stridor:

- Harsh musical respiratory sound caused by turbulent airflow through a restricted (narrow) area.
- Phases of Stridor:
 1. **Inspiratory:** Supra-glottic or pharyngeal obstruction.
 2. **Biphasic:** Glottic, subglottic or cervical trachea obstruction.
 3. **Expiratory:** Thoracic trachea and bronchi obstruction.



2. Stertor:

- Low-pitched **inspiratory** fluttering and snoring sound produced by nasal, nasopharyngeal or oropharyngeal obstruction.

3. Wheezing:

- **Expiratory** whistling sound produced by turbulent airflow passing through constricted small airways (bronchioles).

- Variable airway obstruction is influenced by atmospheric, intratracheal and intrapleural pressures during inspiration and expiration.

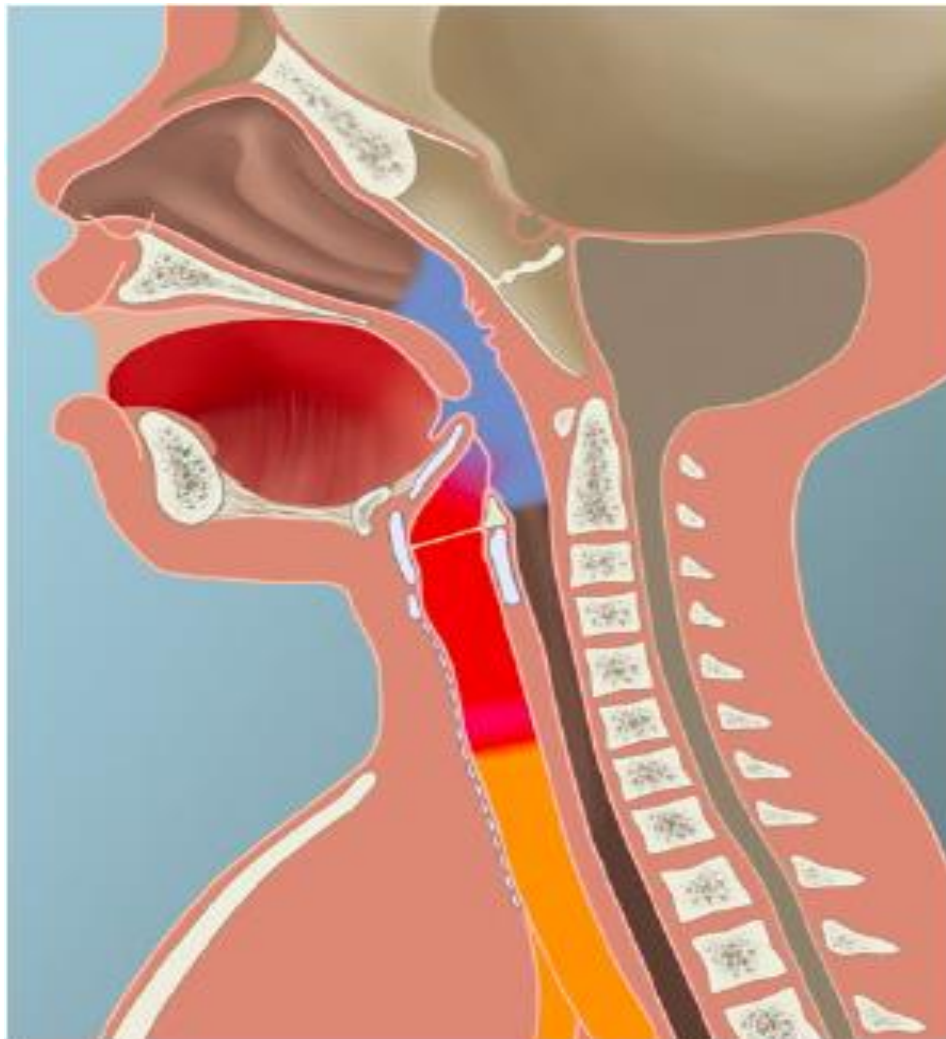


Fig. 3.1 Pathological respiratory sounds

Stertor (blue): Naso-oropharyngeal obstructions: –Adenoid hyperplasia –Tonsillar hyperplasia –Base of tongue mass –Glossoptosis, retrognathia –Pharyngeal mass

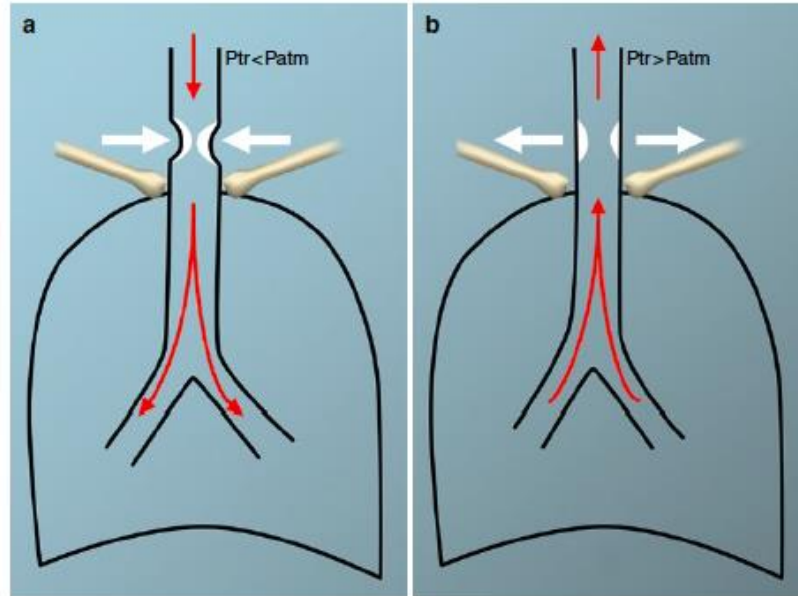
Stridor (red): Laryngotracheal, variable extra-thoracic tracheal obstructions: –Laryngomalacia –BVCP bilateral vocal cord palsy –Soft tissue SGS –Cysts and laryngoceles –Cleft larynx –Glottic web

Wheezing (orange): Variable intrathoracic obstruction of the tracheobronchial tree: –Tracheobronchomalacia –Extrinsic vascular compressions –Non-circumferential tracheal stenosis –Mediastinal masses

- **Variable Extrathoracic Obstruction:**

- Based on Bernoulli Effect.
- Variable extrathoracic obstruction of the airway produces:
 - An inspiratory pathological sound (stridor or stertor).
 - A prolonged inspiratory phase.
 - A hindered inspiratory phase dependant on the severity of the obstruction.

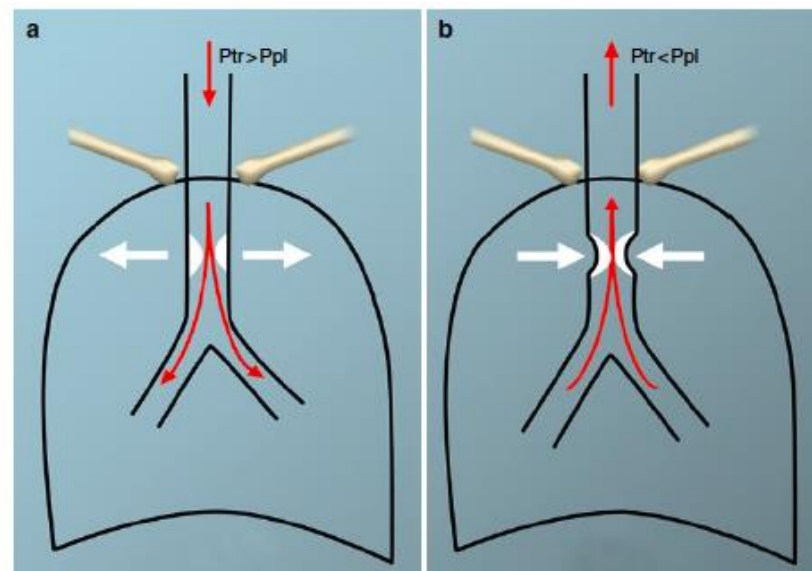
Fig. 3.2 Variable extrathoracic obstruction (Reproduced from Kryger et al. 1976 [24]. With permission). (a) During inspiration, the intratracheal pressure (P_{tr}) is lower than the atmospheric pressure (P_{atm}). This generates a prolonged inspiratory phase with an inspiratory pathological sound (stridor or stertor). (b) During expiration, the intratracheal pressure (P_{tr}) is higher than the atmospheric pressure (P_{atm}). This maintains an almost normal expiratory flow rate, depending on the severity of the obstruction



- **Variable Intrathoracic Obstruction:**

- Based on Bernoulli Effect.
- Variable intrathoracic obstruction of the airway produces:
 - An expiratory pathological sound (wheezing).
 - A prolonged expiratory phase.
 - A hindered expiratory phase dependant on the severity of the obstruction.

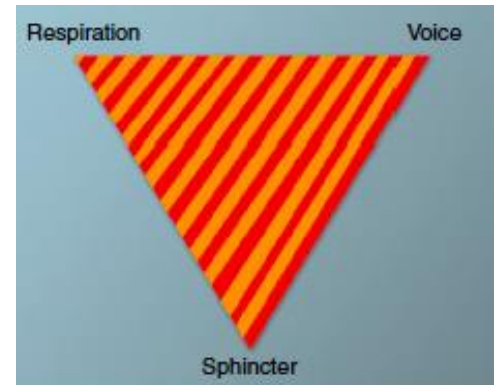
Fig. 3.3 Variable intrathoracic obstruction (Reproduced from Kryger et al. 1976 [24]. With permission). (a) During inspiration, the intratracheal pressure (P_{tr}) is higher than the pleural pressure (P_{pl}). This maintains an almost normal inspiratory flow rate, depending on the severity of the obstruction. (b) During expiration, the intrapleural pressure (P_{pl}) is higher than the intratracheal pressure (P_{tr}). This generates a prolonged expiratory phase and pathological expiratory sound (wheezing)



- **Fixed Airway Obstruction:**
- Intra- and extraluminal pressures do not affect the size of the narrowed segment of the airway..
- Typically the case in congenital cartilaginous or acquired cicatricial subglottic stenosis.
- A fixed obstruction of the airway produces:
 - A typical biphasic stridor of equal intensity during both phases of the respiratory cycle.
 - Variability in the inspiratory and expiratory phases, depending on the severity of the condition.

- **Assessment of Laryngeal Functions:**

- Feeding difficulties with **aspiration** (coughing, choking, gagging, regurgitation or aspiration pneumonia) should be evaluated:
 - Observed in laryngotracheo-oesophageal clefts (LTOC), oesophageal atresia with tracheo-oesophageal fistula (TOF), nerve palsies of the pharyngolarynx and to a lesser degree in unilateral vocal cord paralysis.



- The quality and amplitude of the **voice or cry** should also be evaluated:
 - **Weak and breathy cry:**
 - Corresponds to **incomplete glottic closure**, as seen in unilateral vocal cord paralysis.
 - **Hoarse voice or aphonia:**
 - Indicative of a **vocal cord pathology**, which may be due to inflammation, oedema, tumour or scarring.
 - **Muffled sound:**
 - Suggestive of a **supraglottic pathology** (cyst, lymphovascular malformation).
 - **High-pitched cry:**
 - Seen in conditions such as **bilateral vocal cord palsy, synechia or web** of the vocal cords.
 - Unless it is very severe, tracheal stenosis does not influence voice quality and cry.

- **Complete Medical History:**

- Performed only in the absence of severe or progressive respiratory distress.

1. **Age of onset:**

- Immediately at birth or during the neonatal period warrants questions regarding the pregnancy (fetal distress and prematurity) and delivery (difficult labor, asphyxia, apnoeic episodes, low Apgar scores, and birth injury).

Table 3.1 Pathological conditions relating to the age of onset in the case of respiratory symptoms

• Birth
– Bilateral vocal cord paralysis
– Congenital cysts
– Laryngeal webs and atresia
– Choanal atresia
– Vascular mediastinal anomalies
– Severe subglottic stenosis
• Age: first few weeks of life
– Laryngomalacia
• Age: 1–4 months
– Moderate subglottic stenosis
– Subglottic haemangioma
• Age: 1–3 years
– Croup
– Bronchiolitis
• Age: 3–6 years
– Membranous laryngo-tracheobronchitis
– Epiglottitis

Sign	Score = 0	Score = 1	Score = 2
Heart Rate	Absent	Below 100 per minute	Above 100 per minute
Respiratory Effort	Absent	Weak, irregular, or gasping	Good, crying
Muscle Tone	Flaccid	Some flexion of arms and legs	Well flexed, or active movements of extremities
Reflex/Irritability	No response	Grimace or weak cry	Good cry
Color	Blue all over, or pale	Body pink, hands and feet blue	Pink all over

2. **Mode of onset:**

- Sudden (foreign body or airway edema).
- Gradual progressive (laryngomalacia, subglottic hemangioma, papillomas).

3. **Duration:**

- Short (Foreign body, edema, and infection).
- Long (laryngomalacia, stenosis, subglottic hemangioma, anomalies tongue and jaw).

4. **Aggravating factors:**

- Dyspnea and stridor worsen during feeding, crying or in the case of agitation, due to increased airway demands.
- Dyspnoea and stertor worsen during sleep in nasopharyngeal or oropharyngeal obstructions, as a result of muscular tone loss.
- Coughing and choking episodes increase during meals in tracheo-oesophageal fistulae and clefts.

Table 3.2 Factors worsening airway obstruction in infants and children

• Worsening with crying, feeding or straining (laryngeal anomalies) – Laryngomalacia – Bilateral vocal cord paralysis – Subglottic stenosis – Subglottic haemangioma • Worsening during sleep (naso-oro-pharyngeal obstruction) – Retrognathia with tongue base prolapse – Epiglottic prolapse – Adenotonsillar hypertrophy • Worsening with deglutition (deficient sphincteric function of the larynx) – Unilateral vocal cord paralysis – Discoordinated pharyngolarynx (neurological disorder) – Tracheoesophageal fistula and cleft

5. Influence of body position:

Table 3.3 Influence of the body position in the case of airway obstruction

• Decreased noisy breathing in prone position – Laryngomalacia – Vallecular cyst – Micro-retrognathia – Macroglossia – Innominate artery compression • Decreased or worsened noisy breathing in lateral position* – Unilateral vocal cord paralysis – Lateral pharyngolaryngeal cyst, mass
--

6. History of intubation, endoscopy, surgery or laryngeal trauma.

7. Cyanotic spells.

Table 3.4 Mnemonic history of airway obstruction [17]

S=severity: parents' subjective impression regarding the severity of the obstruction

P=progression: evolution of the obstruction over time

E=eating: feeding difficulties, aspiration and failure to thrive

C=cyanosis: cyanotic episodes, apparent life-threatening events

S=sleep: disturbed sleep, suprasternal or chest retractions during sleep



- **Assessment of the Patient's General Condition**

- **Prematurity:**

- Possibility of poor pulmonary reserve as a sequela to hyaline membrane disease as well as neurological deficits secondary to periventricular leucodystrophy caused by brain hemorrhage.

- **Congenital anomaly:**

- May need genetic evaluation and karyotyping.

- **Neurological evaluation:**

- May need Brain MRI.

- **Cardiological evaluation:**

- May need ECG or ECHO.

- **Pulmonology evaluation:**

- May need PFT or spirometry.
- Pulmonary reserve may be obtained based on intentional episodes of apnea during endoscopy under general anesthesia:
 - The rapidity with which the child recovers from temporary oxygen desaturation provides useful information on the quality of the gas exchanges in the lung parenchyma.

- **GI evaluation:**

- May need pH monitoring or MBS.

- **Physical Examination:**

- **Performed only in the absence of severe or progressive respiratory distress.**
- Careful non-invasive inspection.
- Evaluation of dysmorphic facial features.
- Retractions of the accessory respiratory muscles (suprasternal, intercostal, subcostal and abdominal movements).
- Flaring of ala nasi.
- Mouth breathing
- Size and position of the mandible and tongue.
- The patency of nasal airways.
- Attention should be paid to the intensity of the pathological sound and its relation to the respiration phase.
 - In case of old child you may ask them to breath rapidly through open mouth and observe for stridor.
 - Examine infants in different positions.
 - Auscultation is very useful when the pathological respiratory sound is not clearly audible.
- **In-Office Trans-nasal Flexible Laryngoscopy (TNFL)**
 - Done for infants and cooperative older children.
 - Toddlers often refuse this examination and should not be forced to undergo it.
 - Patency of the nasal cavities, choanae, nasopharynx and pharyngolarynx should be carefully assessed.
 - The degree of dynamic functional narrowing of the airway at the naso-oro-pharyngeal junction and larynx during inspiration should also be determined.
 - Inspection of the larynx is aimed at documenting the abnormalities of the supraglottis (e.g., laryngomalacia, lymphovascular malformations and cysts) & VF mobility.
 - If restricted abduction or true immobility of both vocal cords is perceived:
 - Complete examination under general anesthesia is justified in order to differentiate neurogenic bilateral vocal cord paralysis from posterior glottic stenosis.
- **Indications for Endoscopy Under General Anesthesia:**
 - In the case of symptom progression, associated abnormalities or atypical clinical presentations.
 - In the case of chronic stridor, clinical indicators such as feeding difficulties, failure to thrive, obstructive sleep apneas and cor pulmonale.

- ALL of the following endoscopies should be done under GA for complete assessment of the airway:
 1. TNFL for dynamic evaluation of the upper and lower airways.
 2. Direct laryngotracheoscopy with a bare rod-lens telescope.
 3. Suspension MicroLaryngoscopy.
 4. Broncho-esophagoscopy, when possible and depending on the type of airway obstruction.

Table 5.1 Indications for spontaneous respiration anaesthesia in paediatric airway endoscopy

- Dynamic evaluation of the upper airways by TNFL
- Predictable difficult intubation
- Endoscopic therapeutic procedure requiring a free laryngeal field:
 - Endoscopic LTOC repair
 - Laryngotracheal stenosis
 - Microlaryngeal laser surgery for various indications

- **Radiological Evaluation:**
- Performed only in the absence of severe or progressive respiratory distress.
- Types:
 - High kilo-voltage X-ray:
 - No extra-radiation.
 - AP and Lateral Cervical and Chest films.
 - CT scan
 - MRI
 - Barium swallow
 - US



**TABLE
89.1****DIFFERENTIAL DIAGNOSIS:
STRIDOR IN A CHILD****Congenital**

Laryngomalacia, laryngocele, saccular cyst, laryngeal web/atresia, subglottic cyst, subglottic stenosis, tracheomalacia, tracheal web/stenosis, vascular anomalies, complete tracheal rings, thymic cysts

Infections

Retropharyngeal abscess, epiglottitis, angioedema, viral laryngotracheobronchitis, bacterial tracheitis

Neoplasms

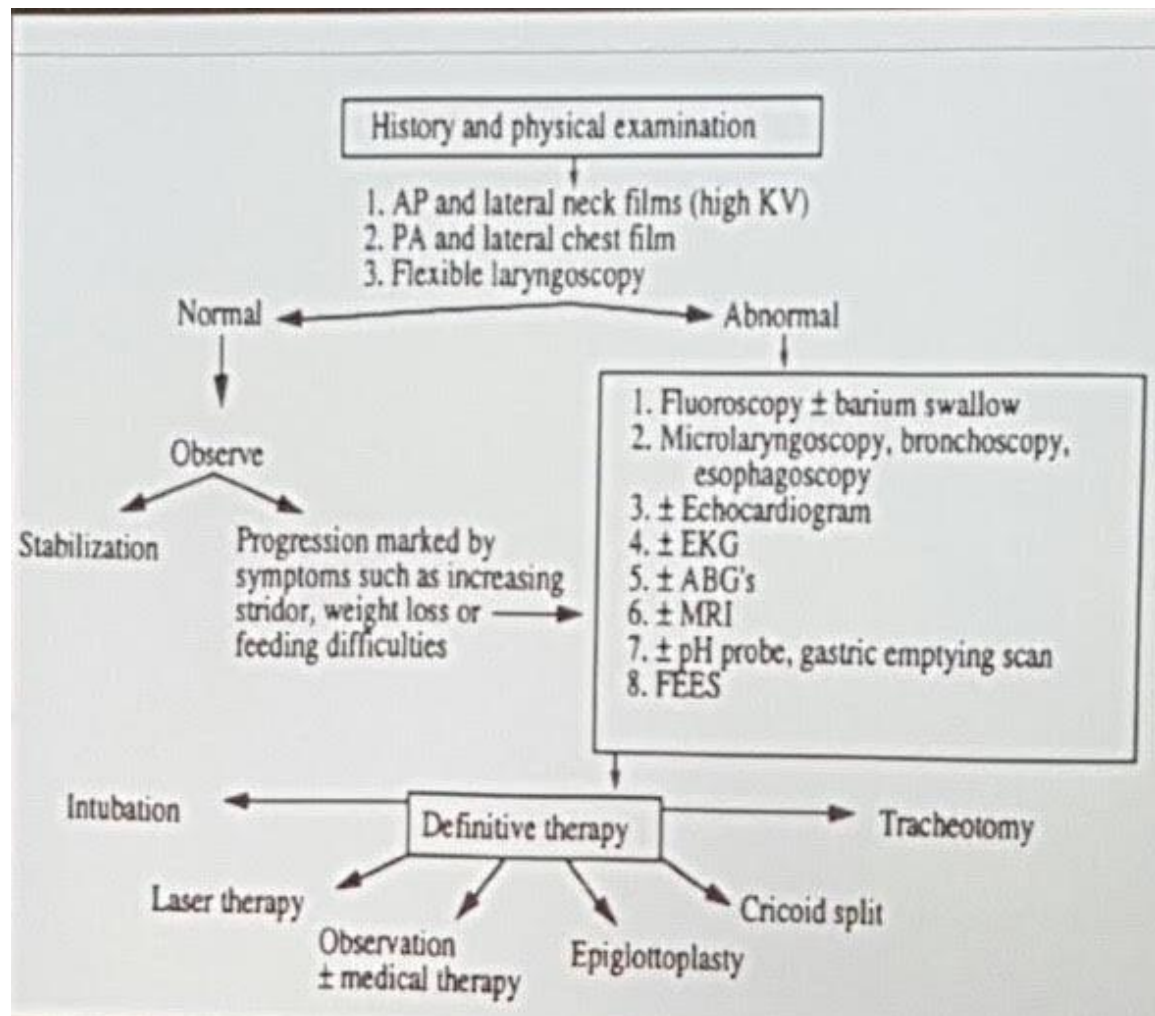
Vascular tumors—hemangiomas, lymphatic/venous/arterial malformations, recurrent respiratory papillomas, thyroid tumors, mediastinal tumors

Other

Vocal cord immobility, foreign body, laryngeal fracture, intubation trauma-cricoid dislocation

**TABLE
89.2****DIFFERENTIAL DIAGNOSIS:
ASPIRATION IN A CHILD**

Congenital	Neurologic	Other
Laryngeal cleft	Hypotonia	Vocal cord immobility
Tracheoesophageal fistula	Encephalopathy	Cleft palate
Esophageal atresia/webs/duplications, vascular rings, achalasia	Anoxic brain injury Arnold-Chiari malformation Muscular dystrophies Cerebral palsy	Neoplasm GERD Foreign body Postsurgical



Evaluation of Aspiration:

- Aspiration is considered pathologic if pulmonary complications ensue.
- Barriers of aspiration:
 - o Swallow
 - o Epiglottis
 - o A-E folds
 - o Vocal cords
 - o cough reflex
- Predisposing factors:
 - o Low Level of consciousness (LOC).
 - o Swallowing problems
 - o Aero-digestive tract abnormalities.
- Sequels of aspiration:
 - o Chronic cough
 - o Hoarseness
 - o Pneumonia (Recurrent)
 - o Pulmonary fibrosis
- Findings (not specific):
 - o Fever
 - o Tachypnea
 - o Wheezing
 - o Rales
- Findings of Upper motor neuron pathology:
 - o Spastic
 - o Straining
 - o Strangling
- Findings of Lower motor neuron pathology:
 - o Oral/pharyngeal weakness
 - o Worse with liquids
- Sources of aspirate:
 - o Oral contents
 - o Oral secretions
 - o Reflux
- With anti-reflux medications:
 - o Higher risk of bacterial contamination
 - o Lower risk of pneumonitis
 - o All from higher pH
- FB aspiration:
 - o Usually in kids < 5 years old.
 - o Presents with sudden stridor.

Diagnostic Studies of Aspiration:

1. Video-fluoroscopic swallowing study (Modified Barium swallow/MBS):
 - Upper Aero-digestive examination.
 - Consistencies: liquid; paste; solid.
 - Good study; time consuming; radiation exposure.
2. Functional endoscopic evaluation of swallowing (FEES):
 - Flexible scope placed behind the palate to observe swallowing using dye.
 - Evaluate for:
 - Pharyngeal pooling; premature spillage; laryngeal penetration; aspiration; residue.
 - Invasive; only see pharyngeal phase; no radiation exposure.
3. Endoscopy:
 - Look for structural abnormalities; biopsies if needed.
 - Lipid-laden macrophages on bronchial lavage = aspiration.
4. Imaging:
 - Upper GI can show abnormal anatomy if present.
 - CT and MR for possible intracranial lesions.
 - US (real time) good for oral phase; tongue movement seen.
 - Radionuclide scintigraphy: quantify aspiration; emptying.
5. EMG
 - Advantage: can study individual muscles.

Management of Aspiration:

- **Medical management and swallowing rehab:**
 - Alter food consistencies, posture, exhale while swallowing.
 - Palatal appliance – stimulus to trigger involuntary swallowing
 - Antibiotics, intubation and toilet for severe pneumonia.
 - Sialorrhea:
 - Antihistamines; anticholinergics, Botox
- **Surgical management:**
 - Feeding tube (G or J): Doesn't prevent aspiration of secretions.
 - Fundoplication: 90% resolution if caused by reflux.
 - CP myotomy: good only for CP dysfunction (Doesn't relax).
 - Salivary glands:
 - Duct ligation; gland excision; rerouting; only good for aspiration of secretions; rerouting for drooling only.
 - Bilateral SMG excision with Stenson's ligation: former for resting salivary flow; latter for food induced spit.
 - Tracheostomy: not good for chronic; limits laryngeal elevation.
 - VC medialization: good for failure of glottic closure; 50% cure.
 - Laryngeal closure: needs trach; small post opening for voice; 50-80% success; usually reversible.
 - Laryngotracheal separation: >90% success; stoma with blind pouch; only used in severely impaired who won't use voice.

Evaluation of Cough:

- History and physical:
 - If chronic, recurrent or serves no useful function, look further.
 - Duration and character; PND or GER; asthma/LRT disease.
 - Smoking; toxin exposure; allergies; sinusitis.
 - H&N, respiratory and CV exam; examine upper A/W and ears.
 - Voluntary cough to witness and characterize it.
- Investigations:
 - CBC, CXR and sputum culture; PFTs if suspicious
 - Endoscopy if all else non-diagnostic

• Differential Diagnosis of Cough:

- Pharyngeal, laryngeal or bronchial receptors:
 - Environmental: tobacco; pollution; allergens.
 - Inflammatory: pharyngitis; laryngitis; PND; pneumonia; GER.
 - Neoplastic: pharyngeal or laryngeal Ca; bronchogenic Ca.
 - Cough-variant asthma.
- Afferent neurons of the cough reflex (rare):
 - Vagus neurilemmomas; cervical osteophytes.
- Other receptor sites:
 - Ear; pleura; pericardium; stomach.
- Central: psychogenic.

• Management of Cough:

- Treat cause; resolution in 80% of cases.
- Cough reflex suppressants: narcotics only ones proven to work.
 - **Codeine** best, others too addictive.
 - Dextromethorphan used frequently; no data to prove it works.
- Peripheral suppressants:
 - Block receptors directly; topical lido; limited duration.
- Expectorants and mucolytics:
 - Increase volume and decrease viscosity of secretions.
 - Humidity, lozenges and syrups examples.
 - Avoid combination preparations – side effects and not proven.



King Abdulaziz Medical City
National Guard Health Affairs

Pediatric Otolaryngology Sheet

Patient's name: _____ Age: _____ MRN: _____

1. Patient's status:

Stridor:

- Mild or Severe (Feeding / Distress / Cyanosis / Apnea)
- Duration: _____
- Onset / Progression: _____
- Aggravating Factors: _____
- Relieving Factors: _____
- Voice Changes: _____

2. Background:

- Prenatal / postnatal history / B.W. : _____
- Pre / post term: _____
- ICU admission / Intubation/ size ETT: _____
- Vaccination: _____
- Developmental history: _____
- Past surgical / cardiac / neurological history: _____
- Family history: _____
- Social history: _____
- Medication: _____
- Allergy: _____
- Patient was fine until: _____

3. Assessment *V/S*

- Patient looks: well / ill / distressed / Growth chart _____
- Voice / cry: _____
- Feeding: oral / OGT / NG / PEG tube _____
- Auscultation: Stridor: inspiratory / expiratory / mixed _____
- Ear: _____
- Nose: _____
- Throat: _____
- Neck: _____
- Chest: _____

4. Differential Diagnosis:

5. Final Diagnosis:

6. Plan / Recommendation:

Recommended tubes and scopes based on patient age

Patient age	Bronchoscope		Oesophagoscope	Tracheostomy tube ISO	ETT ID
	size	OD (mm)			
Premature	2.5	4.2	4	2.0/2.5	2.5
Term newborn	3.0	5.0	4–5	3.0/3.5	3.0/3.5
6–12 months	3.5	5.7	5–6	3.5/4.0	3.5/4.0
1–2 years	3.7	6.4	6	4.0	4.0/4.5
2–3 years	4.0	6.7	6–7	4.0/4.5	$\left. \begin{array}{l} \\ \\ \end{array} \right\} \frac{\text{Age} + 16}{4}$
3–4 years	4.5	7.3	7	5.0	
4–5 years	5.0	7.8	8	5.0/5.5	

Description

Endotracheal Tube, 2.5 mm ID x 3.3 mm OD

Endotracheal Tube, 3.0 mm ID x 4.0 mm OD

Endotracheal Tube, 3.5 mm ID x 4.7 mm OD

Endotracheal Tube, 4.0 mm ID x 5.3 mm OD

Endotracheal Tube, 4.5 mm ID x 6.0 mm OD

Endotracheal Tube, 5.0 mm ID x 6.7 mm OD

Endotracheal Tube, 5.5 mm ID x 7.3 mm OD

Endotracheal Tube, 6.0 mm ID x 8.0 mm OD

Endotracheal Tube, 6.5 mm ID x 8.7 mm OD

Subglottic Stenosis (SGS):

- Subglottic airway is the narrowest area of the pediatric airway.
- Cricoid is a complete, non-expandable and non-pliable ring.
- The upper half of infant cricoid is V-shaped and becomes rounded at its lower level.
- Normal subglottic lumen diameter is:
 - 3.5 mm in premature neonates.
 - 4.5 mm in full term neonates.
- Cartilages of infant larynx are softer and more pliable than adults.
- Laryngeal mucosa are lax in infants and hence more prone to edema when inflamed or injured causing significant airway obstruction in the subglottic area.
- In infants:
 - 1 mm of subglottic edema in will decrease subglottic luminal diameter by 75% and a 16-fold increase in airway resistance.
- In adults:
 - 1 mm of subglottic edema in will decrease subglottic luminal diameter by 30% and a 2-fold increase in airway resistance.

Fig. 2.15 Infant subglottis and risk of dyspnea: The size of the infant subglottis has a maximum diameter of 5–6 mm and a cross-sectional area of 28 mm². One millimetre of mucosal oedema reduces the diameter by 2 mm and the cross-sectional area (12.6 mm²) by nearly 50% (Reproduced with permission of Holinger, Chicago [31])

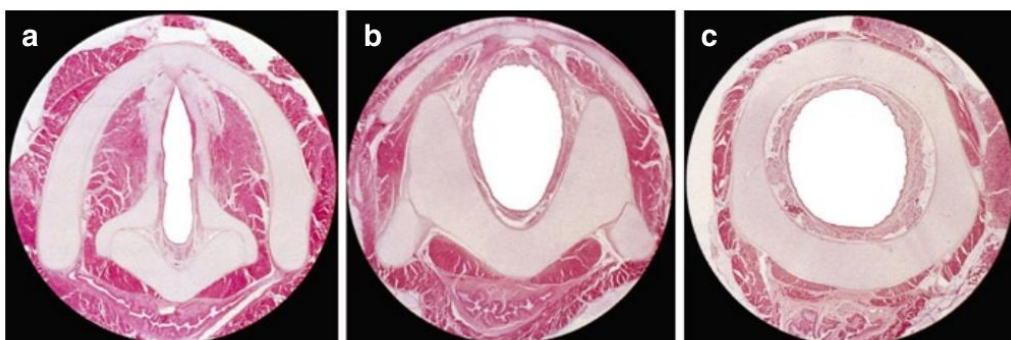
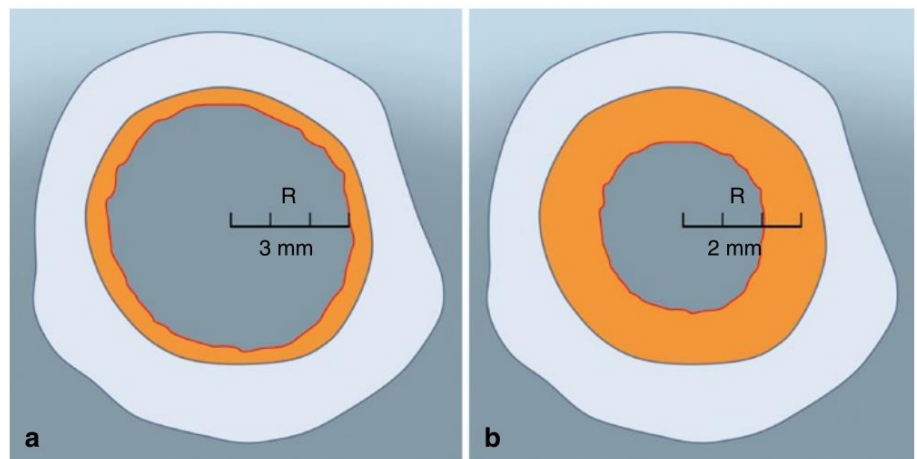


Fig. 2.5 Horizontal histological sections of the infant larynx. (Reproduced from Holinger, Chicago [32]. With permission.) (a) The thyroid cartilage is round and not V-shaped as in adults; the arytenoids are long, contributing to one-half of the glottic length; the cricoid plate is slightly V-shaped (section at the

glottic level). (b) Due to the V-shaped configuration of the upper cricoid, the subglottic lumen is elliptical (section at the mid-portion of the cricoid cartilage). (c) At the lower level of the cricoid cartilage, the lumen is round-shaped

- Classification of SGS:

1. Congenital / C-SGS (5%):

- Less common cause of SGS.
- 3rd most common congenital laryngeal anomaly (10–15%).
- Most common laryngeal anomaly requiring tracheotomy in children under 1 year of age.
- C-SGS is defined as:
 - Subglottic diameter **< 3.0 mm** in a pre-term.
 - Subglottic diameter **< 4.0 mm** in a full-term.
 - **No history of previous ETT or laryngeal trauma.**
- Results from failure of the laryngeal lumen to recanalise completely during 10th week of gestation.
- Closely related to laryngeal webs and atresia, which also result from a laryngeal recanalization failure.
- Histopathology:
 - Mainly **cartilaginous stenosis**.
 - For this reason, endoscopic laser resection or incisions and dilatation will be ineffective because of troublesome granulation tissue formation with subsequent severe restenosis.
 - Types:
 - 1. Small cricoid with normal shape:**
 - 2. Abnormal cricoid shape:**
 - Most common form of C-SGS.
 - Common forms are:
 - Elliptical-shaped cricoid.
 - Thick anterior lamina
 - Generalized cricoid thickening
 - Elliptical-shaped cricoid cannot accommodate a round-shaped ET tube and emergency tracheostomy is necessary in this case to secure the airway.
 - 3. Trapped first-tracheal ring:**
 - Less frequent cause of C-SGS.
 - Associated with flattened cricoid.

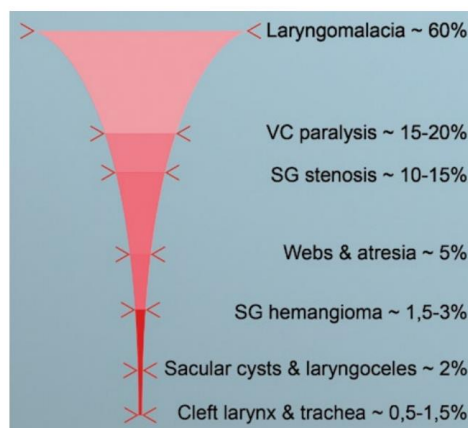


Table 1 Prevalence of congenital laryngeal anomalies

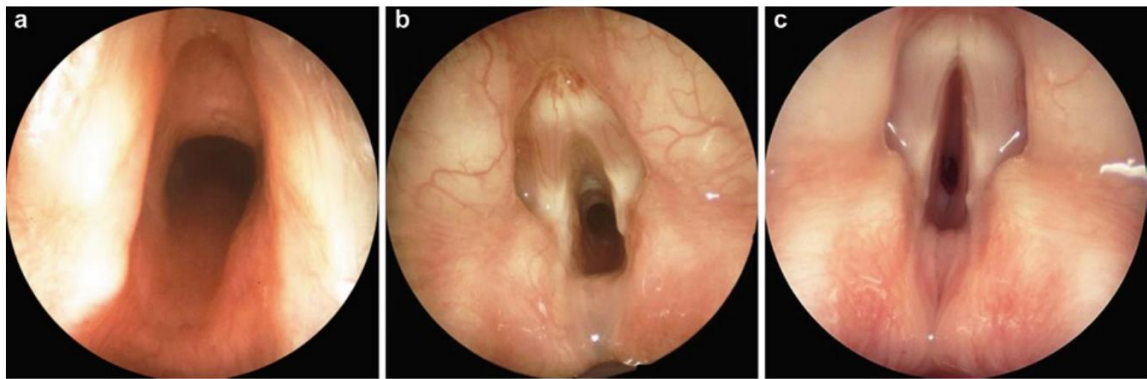


Fig. 8.2 Endoscopic views of congenital cartilaginous SGSs: (a) Thick anterior lamina of the cricoid ring: asymptomatic <50% SGS. (b) Generalised thickening of the cricoid ring: mild grade III C-SGS requiring surgery. (c) Elliptical cricoid: grade III C-SGS requiring surgery



Fig. 8.1 Flattened cricoid with hyperplasia of the submucosal glands: association of a cricoid ring deformity with increased soft tissue within the lumen (Reproduced with the permission of Holinger [4])



Fig. 8.3 Elliptical cricoid: This abnormally shaped cricoid does not accommodate any of the ET tubes without severe mucosal damage. Once diagnosed, tracheostomy or immediate single-stage laryngeal reconstruction with postoperative intubation must be performed (Reproduced with the permission of Holinger [4])

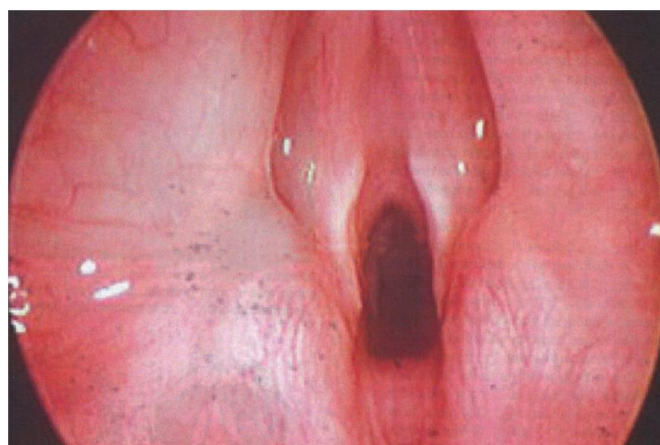


FIGURE 205-2. Endoscopic view of a congenital glottic web with subglottic extension.

2. Acquired / A-SGS (90%):

- Most common cause of SGS.
- More severe, requiring aggressive and long-term management compared to C-SGS.
- Causes:
 - **Trauma:**
 - Intubation (90% of A-SGS).
 - Blunt or penetrating laryngeal trauma
 - Caustic or thermal laryngeal injuries
 - **Surgical interventions**
 - Inappropriate use of lasers and dilation.
 - High tracheotomy, cricothyrotomy.
 - Failed laryngotracheoplasty
 - Common causes of failure:
 - Surgical factors
 - Presence of GERD
 - Secondary infection
 - Granulation tissue formation
 - **Neoplasms**
 - Benign or malignant
 - Chondroma, fibroma, hemangioma or carcinoma.
 - **Infections:**
 - Bacterial or fungal infections.
 - **Inflammatory/Autoimmune disorders**
 - Wegner granulomatosis.
 - Sarcoidosis
 - SLE
 - **GERD and Eosinophilic esophagitis:**
 - Exacerbating factor in SGS.
 - Should be treated medically or surgically prior to reconstructive surgery.
 - **Post-radiation therapy:**
 - Secondary to chondroradionecrosis.
 - **Idiopathic causes**
- Histopathology:
 - Mainly **Soft tissue stenosis.**
 - Mixed stenosis (soft tissue and cartilaginous) occurs in cases of emergency ET intubation leading to acquired on top of congenital subglottic stenosis.
 - Types:
 1. **Submucosal gland hyperplasia**
 2. **Ductal cysts**
 3. **Submucosal fibrosis**
 4. **Granulation tissue**

Table 8.1 Histopathologic classification of subglottic stenosis
(Adapted with permission from Holinger [6])

Cartilaginous stenosis (usually congenital)

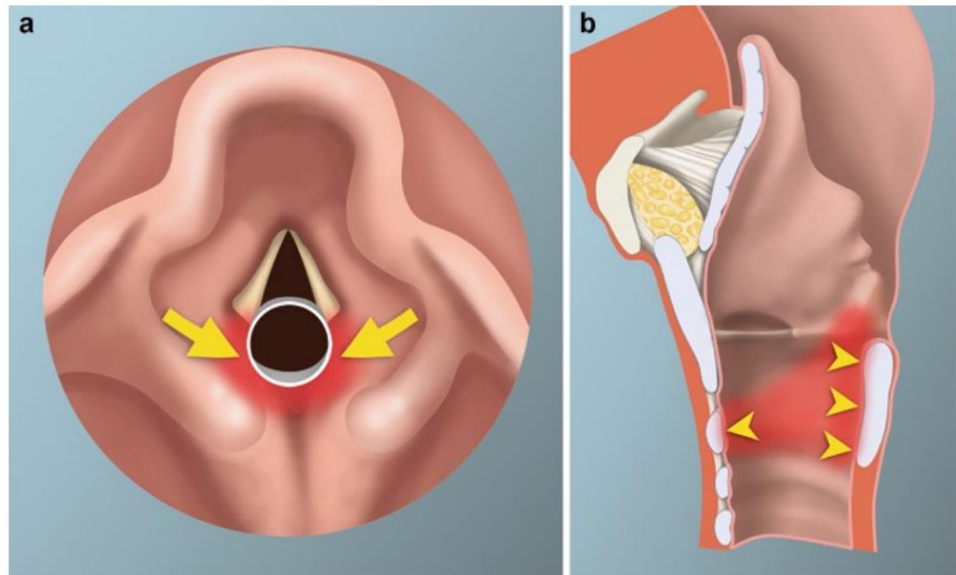
- Cricoid cartilage deformity
 - Normal shape, small size
 - Abnormal shape
 - Elliptical
 - Cleft (partial, submucosal)
 - Flattened
 - Generalized thickening
- Trapped first tracheal ring

Soft-tissue stenosis (usually acquired)

- Submucosal gland hyperplasia
- Ductal cysts
- Submucosal fibrosis
 - ±Distorted cartilage
 - ±Cricoid ossification
- Granulation tissue

- **Intubation-related SGS:**
- Most common cause of laryngotracheal stenosis.
- 90% of all causes of acquired SGS.
- Incidence of post-intubation stenosis in children dropped from previously (20%) to currently **1-8%** due to the following reasons:
 - Improvement of neonatal care.
 - Red rubber ETT have been abandoned.
 - Current clinical belief that the ideal ETT is:
 - Smallest ETT permitting adequate ventilation
 - Should have an air leak around it with subglottic pressure <25 cm H₂O.
- **Most important factors in development of laryngotracheal stenosis:**
 - **Size of ETT:**
 - Most important factor.
 - In children, ETT size should allow an air leak at 20 cm water pressure if possible.
 - Polymeric silicone and polyvinyl chloride are considered the safest materials for prolonged intubation.
 - **Duration of intubation:**
 - No definite safe time limit for ET intubation has been established.
 - Studies in adults shown that **7-10 days** period is acceptable, after which risk of laryngotracheal complications increases (15%).
 - Premature infants tolerate more prolonged intubation (weeks), but beyond **4 weeks** of intubation, the risk of laryngotracheal stenosis increase.
 - Good tolerance of infant larynges to long-term ET intubation due to the soft and pliable cartilaginous framework of the cricoid ring.
 - With growth, laryngeal framework becomes more rigid and partially ossifies at adulthood or after chronic injuries caused by ET tubes.
- **Most common site of injury post-intubation is:**
 - **Posterior endolarynx in adults.**
 - Most vulnerable structures are medial aspect of both arytenoids where the cartilage is protected by the mucoperichondrium only.
 - **Subglottic region in children, due to:**
 1. Subglottic region is narrowest portion of pediatric airway.
 2. Cricoid cartilage is the only complete ring cartilage in upper airway which prevents outward extension of traumatic edema.
 3. Subglottic mucosa is delicate and tends to deteriorate under the stress of ETT.
 4. Subglottic submucosa is made up of loose areolar tissue that allows edema to develop easily.

Fig. 14.2 Diagram of the potential sites of pressure-induced endotracheal tube injuries in the larynx: **(a)** Maximum pressure is exerted on the medial aspect of the arytenoid cartilages (*arrows*). **(b)** Other sites of predilection for pressure-induced necrosis include the posterior laryngeal commissure, and the posterolateral and circumferential subglottis (*arrows*)



- Pathophysiology of intubation-related SGS:
 - Endotracheal tube (ETT) causes pressure necrosis at the point of contact with subglottic mucosa.
 - Mucosal edema and ulceration
 - Exposed perichondrium and cartilage
 - Interruption of normal ciliary flow
 - Mucociliary stasis
 - Secondary infection and perichondritis.
 - Chondritis and cartilaginous necrosis
 - Healing by secondary intention
 - Granulation tissue formation
 - Deposition of fibrous tissue in submucosa.

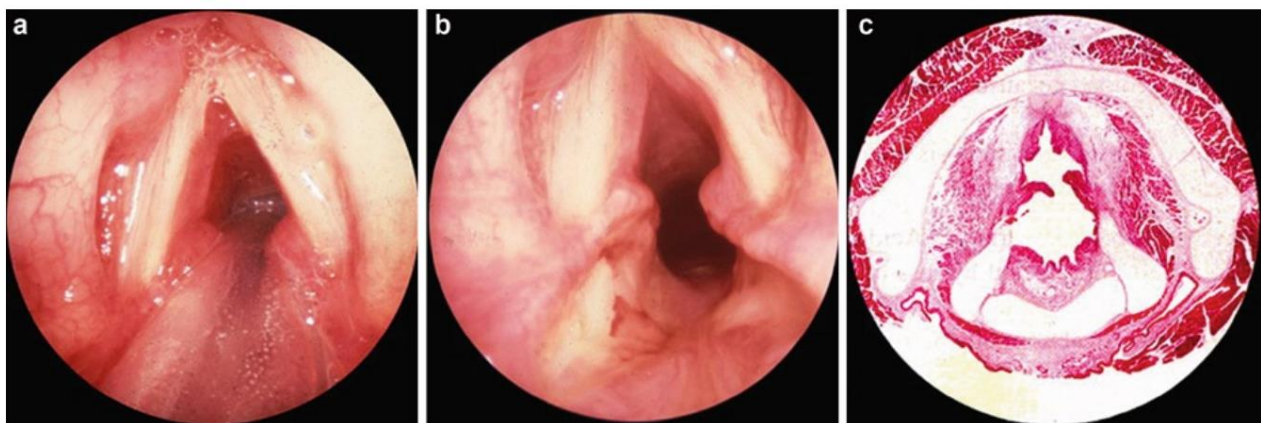


Fig. 14.3 Acute lesions of intubation at the posterior glottis: **(a)** Granulation tissue at the vocal processes of the arytenoids in response to pressure necrosis due to the endotracheal tube. **(b)** Exposed cartilage at the medial aspect of both arytenoid cartilages. **(c)** Corresponding histology for the same type of lesions (Reproduced from Holinger [14]. With permission)

- Local factors contributing to post-intubation stenosis:

1. Intubation trauma:

- Due to anatomical differences or a lack of experience.
- May lead to development of severe endolaryngeal lesions or aggravating prolonged ETT injuries.

2. Pressure-induced ET tube injury:

- ET tube always lies in the posterior glottis because of its oro-pharyngeal curvature.
- When pressure of ETT exceeds capillary mucosal perfusion pressure (20–40 mmHg), ischemic necrosis ensues.
- Degree of ischemic necrosis is more significant than actual duration of intubation.
- Major changes are seen within 48–72 hours of intubation.
- Superinfection due to prolonged intubation increases severity of perichondritis and cartilaginous necrosis.
- Tracheotomy performed at this stage worsens the condition by increasing bacteriologic contamination of the airway.
- Process of repair begins slowly with granulation tissue formation creeping from the edges of denuded cartilage to provide a vascularised bed for re-epithelialization.
- As granulation tissue grows significantly faster than the epithelium, it leads to excess tissue that may cause airway obstruction and subsequent scarring.

Fig. 14.1 Faulty technique of intubation: **(a)** Diagram: a rotating motion should never be imparted to the anaesthesiology laryngoscope. In infants, this manoeuvre exposes the epiglottic petiole in a frontal plane, thereby preventing easy insertion of the endotracheal tube into the subglottis. **(b)** Endoscopic view: acute traumatic lesion at the anterior laryngeal commissure resulting from a faulty intubation technique

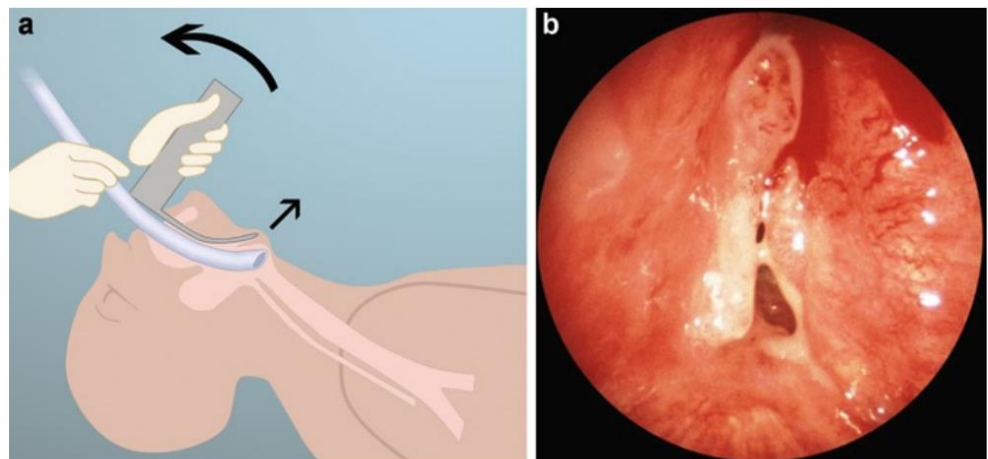


Table 14.2 Predisposing factors of acquired laryngotracheal stenosis

• Patient – Age: infant < adolescent – Larynx: small size, abnormal shape, infection – Abnormal wound healing (keloid formation) – Systemic diseases: GOR, shock (infectious, cardiogenic), immuno-suppression, diabetes, anaemia, hypotension, bronchopulmonary dysplasia
• ET tube – Oversized – Excessive hardness – Poor biocompatibility
• Intubation – Traumatic – Multiple – Prolonged – Followed by tracheotomy
• Nursing – Inadequate patient's sedation – Traumatic and repeated suction – Presence of nasogastric tube – Ventilator-driven tube motion

Table 14.1 Acute post-intubation injuries of the larynx

• Supraglottis – Absent to minor lesions – Oedematous protrusion of ventricular mucosa – Erythema or oedematous swelling of the ventricular bands
• Glottis – Non-specific swelling in Reinke's space of the vocal cords – Pressure-induced ischemic necrosis : ◦ Ulcers at the medial aspect of the arytenoids with flanges of granulation tissue at vocal processes ◦ Medial exposure of the cricoarytenoid joint ◦ Posterior interarytenoid ulcer
• Subglottis – Non-specific oedematous swelling – Posterolateral cricoid ulcers – Concentric subglottic ulceration

- Stenotic laryngeal sequelae:
 1. Interarytenoid adhesions at the posterior laryngeal commissure or subglottic level
 2. Posterior glottic stenosis (PGS) with or without cricoarytenoid joint fixation
 3. Concentric subglottic cicatricial narrowing
 4. Mucous retention cysts.
- Combined glottis-subglottic stenosis are most challenging to treat.
- They are encountered in very severe cases or when prolonged intubation has been proceeded by trauma.

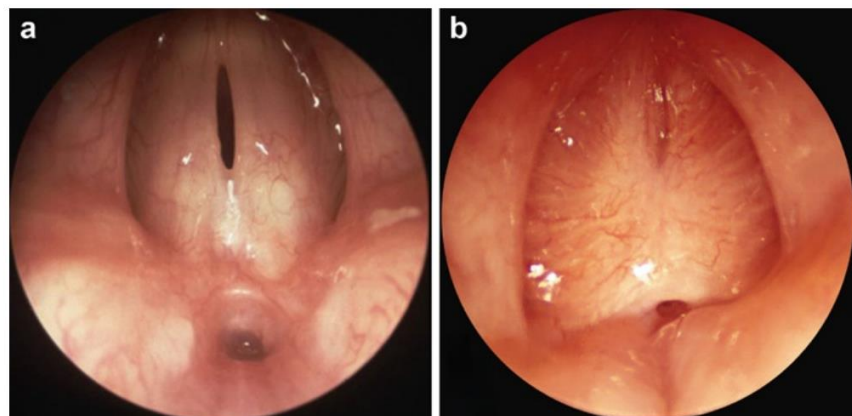


Fig. 14.7 Combined glotto-subglottic stenoses due to prolonged intubation: (a) Posterior glottic stenosis associated with subglottic stenosis. (b) Severe vocal cord synechia associated with subglottic stenosis

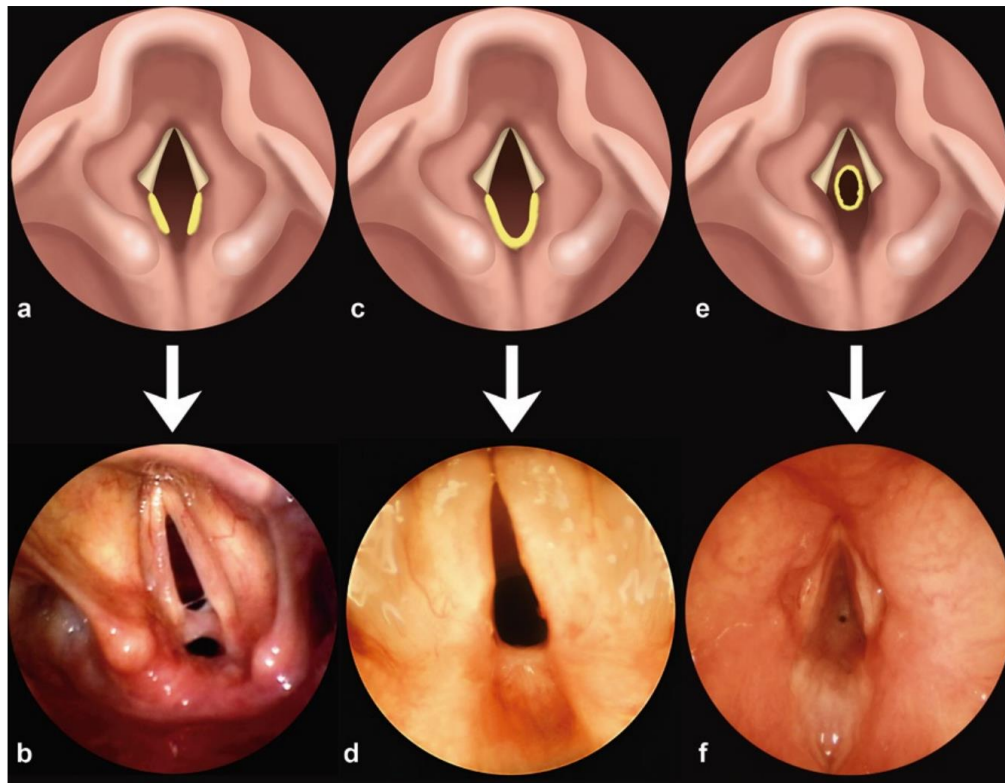


Fig. 14.6 Stenotic cicatricial sequelae due to prolonged endotracheal tube intubation: (a) Diagram of ulcerated troughs with an intact median strip of mucosa (b) Interarytenoid adhesion (c) Diagram of annular posterior ulceration with no residual mucosal strip (d) Posterior glottic stenosis, with or without cricoarytenoid joint fixation (e) Concentric subglottic ulceration (f) Cicatricial subglottic stenosis

- Prevention of acquired laryngotracheal stenosis:
 1. Intubation and endoscopy should be performed gently on relaxed patients.
 - At a rule, ETT must never be forcibly inserted and it should be changed to smaller size with the slightest resistance during introducing the ETT into the larynx.
 - The smallest ETT provides adequate ventilation for infant and child should always be chosen.
 2. Predisposing factors contributing to laryngeal trauma secondary to intubation should be recognized and avoided.
 3. Early exploration of laryngeal fractures should be done to minimize serious sequelae.
 4. During tracheotomy, avoid extensive resection of tracheal wall and smallest size tracheotomy tube compatible with ventilation and suctioning should be used.
 5. High tracheotomy and cricothyroidotomy should be avoided except in extreme emergencies.
 - If it was done high then neck should be explored and tracheotomy site should be relocated to lower level to prevent SGS.
 - If PCTR is planned for the patient, high tracheotomy is preferred because it reduces length of the resection.
 6. Aggressive endoscopy for benign laryngeal lesions should be avoided especially in anterior commissure area to prevent formation of anterior glottis web.

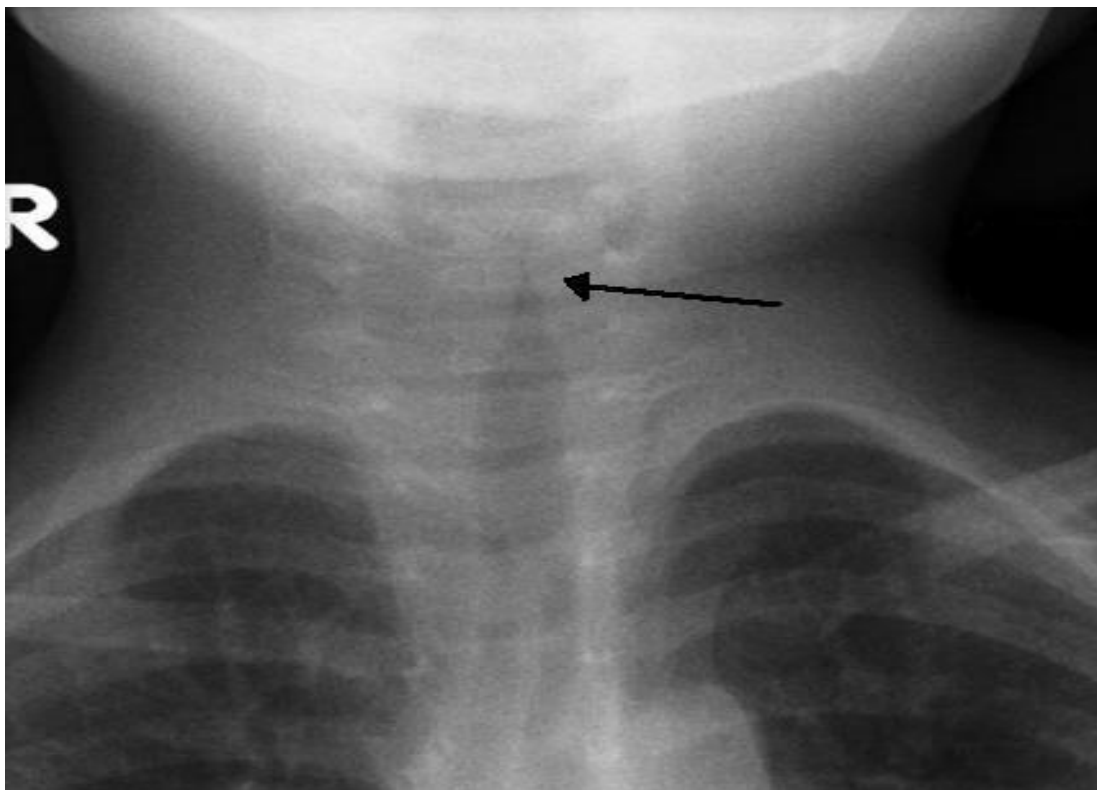
- Clinical picture of SGS:
 - Mild to moderate SGS (Grade I-II) may be asymptomatic and become symptomatic only during upper airway infection and present as recurrent episodes of croup with a barking cough.
 - Significant airway findings on DLB were seen in **10%** only of patients presenting with recurrent croup.
 - Progressive respiratory difficulty:
 - Biphaseic stridor.
 - Severe respiratory distress with more severe SGS:
 - Severe dyspnoea
 - Suprasternal or chest retractions
 - Abnormal cry or hoarseness occurs when vocal cords are affected.
 - Feeding abnormality.
 - Congenital SGS may present at or shortly after birth with respiratory distress and difficult to pass size 3 (in full term) or even size 2.5 ETT during intubation.
 - Size 2.5 ETT has outer diameter of 3.6 mm.
 - Size 3.0 ETT has outer diameter of 4.2 mm.
 - History of prolonged ET intubation or laryngeal trauma in case of acquired SGS.
 - Symptoms occur 2-4 weeks after the original insult, although latent period can occasionally be longer.

**TABLE
90.1**
**SIGNS AND SYMPTOMS
OF LARYNGEAL STENOSIS**

Laryngeal Region	Signs and Symptoms
Subglottis	Hoarse or normal voice, biphasic stridor, normal feeding (except with severe obstruction), barking cough
Glottis	Hoarse voice or aphonia, inspiratory (early) or biphasic (late) stridor, normal feeding (with or without severe obstruction), no cough
Supraglottis	Muffled voice, fluttering inspiratory stridor, severe feeding problems, no cough

Adapted from Reichert TJ. *Diseases of the Larynx in Infants and Children*. Washington, DC: American Academy of Otolaryngology-Head and Neck Surgery, 1987.

- Diagnosis of SGS:
 - **Flexible scope:**
 - Performed in stable patient.
 - Evaluate presence of supraglottic and glottic anomalies or lesions and assessing the mobility of the vocal folds.
 - Associated anomalies are observed in > 50% of the cases of C-SGS.
 - **High-Kilovoltage airway X-ray:**
 - Performed in stable patient.
 - Lateral and AP soft tissue x-rays of neck and chest during full inspiration.
 - Identify the classic steeple seen with subglottic edema.



- **CT scan:**
 - Performed in stable patient.
 - Evaluate site and length of involved segment.
 - Complementary techniques to rigid endoscopy in preoperative evaluation and follow-up of laryngotracheal stenosis.

- **Direct Laryngobronchoscopy (DLB):**
 - Gold standard investigation.
 - Done under GA in OR.
 - Rigid telescope is used initially for better visualization of airway followed by suspension microlaryngoscopy.
 - Should assess:
 - **SGS:**
 1. Extent:
 - Isolated SGS or glotto-subglottic stenosis.
 2. Length:
 - Length of SGS.
 - Length between SGS and vocal folds.
 - Length between SGS and tracheostoma.
 - Length of tracheostoma.
 - Length between tracheostoma and carina.
 3. Nature:
 - Cartilaginous or soft tissue.
 4. Size of residual lumen:
 - Measured by passing bronchoscope or known size ETT.
 - Mayer-cotton grading system.
 - **Vocal folds mobility:**
 - By palpation of the cricoarynenoid joints.
 - **Associated anomalies:**
 - Supraglottis, glottis, trachea and lower airway.

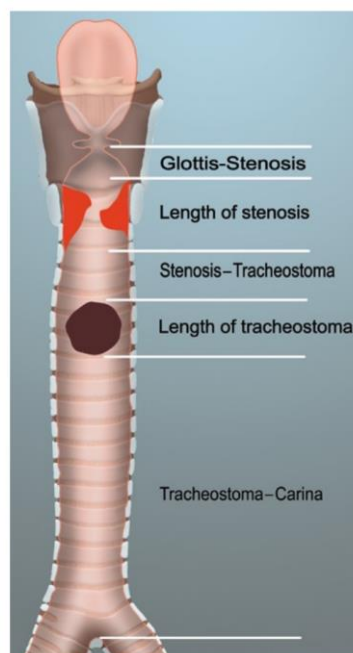


Fig. 5.8 Diagram of the endoscopy report: the length and precise location of the subglottic stenosis with respect to the vocal cords and tracheostoma, as well as the length of the tracheostoma and residual normal trachea (in centimetres and number of normal tracheal rings) must be recorded



Fig. 4.15 Paediatric rigid bronchoscopes:(a) Set of eight Karl Storz bronchoscopes from size 2.5–6.0. (1) Light-cable connection. (2) Straight-forward telescope. (3) Prismatic light deflector for illumination of the open-tube. (4) Working/jet ventilation channel. (5) Standard 15 mm side port with connected rubber

pipe for anaesthesia. (b) Lateral and frontal views of a variety of optical forceps mounted on straight-forward telescopes. From top to bottom: scissors, biopsy, foreign body, cup-biopsy and grasping forceps

Fig. 4.8 Proposed set of paediatric laryngoscopes to manage the most common situations: (a) Anaesthetist laryngoscope. (b) Kleinsasser laryngoscope. (c) Benjamin–Lindholm laryngoscopes. (d) Parson laryngoscope. (e) Benjamin–Haves light-clip. Only one size per laryngoscope is shown, with the exception of (c)

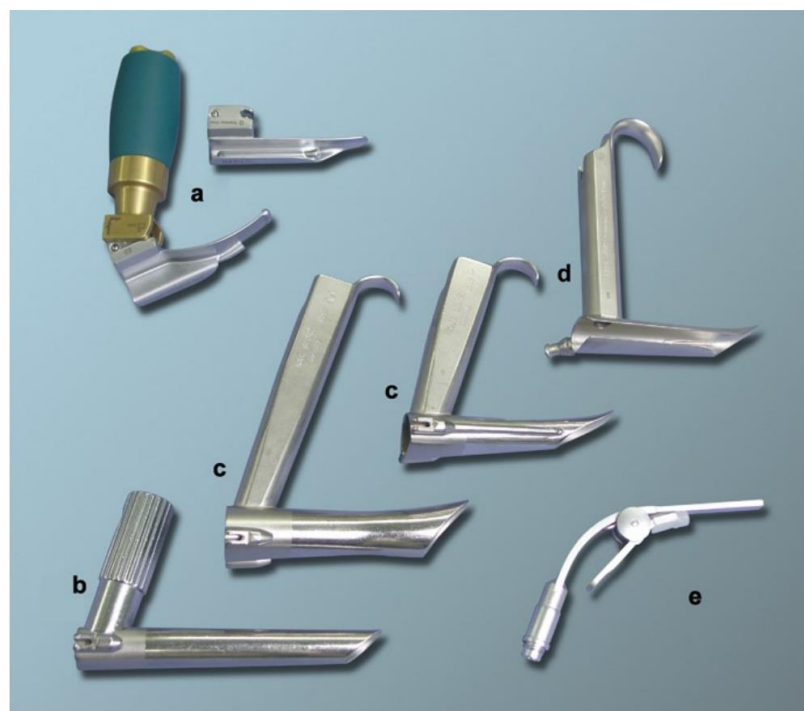


Fig 4.3 Set of Parsons laryngoscopes: (a) Infant and child sizes. (b) Adolescent and adult sizes

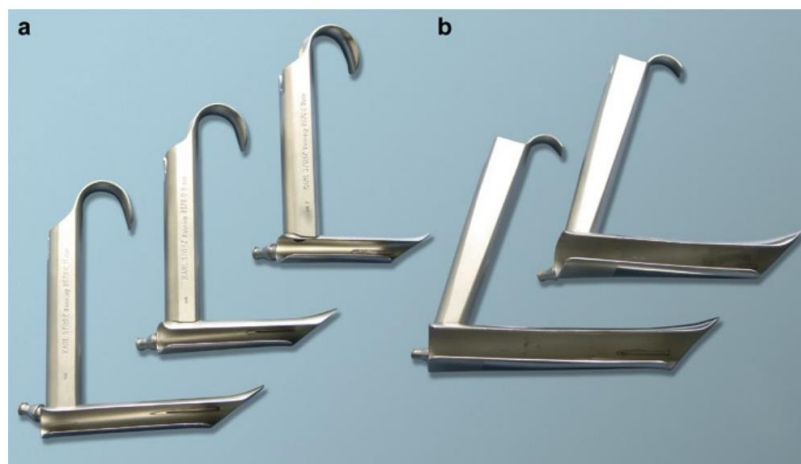


Fig. 4.4 Benjamin-Lindholm laryngoscopes: They are available in different sizes for infants and children, providing a panoramic view of the pharyngolarynx



Fig. 4.5 Kleinsasser laryngoscopes: These straight scopes are suitable for laser work in the paediatric larynx

Recommended tubes and scopes based on patient age

Patient age	Bronchoscope		Oesophagoscope	Tracheostomy tube ISO	ETT ID
	size	OD (mm)			
Premature	2.5	4.2	4	2.0/2.5	2.5
Term newborn	3.0	5.0	4–5	3.0/3.5	3.0/3.5
6–12 months	3.5	5.7	5–6	3.5/4.0	3.5/4.0
1–2 years	3.7	6.4	6	4.0	4.0/4.5
2–3 years	4.0	6.7	6–7	4.0/4.5	$\left. \begin{array}{c} \text{Age} + 16 \\ 4 \end{array} \right\}$
3–4 years	4.5	7.3	7	5.0	
4–5 years	5.0	7.8	8	5.0/5.5	

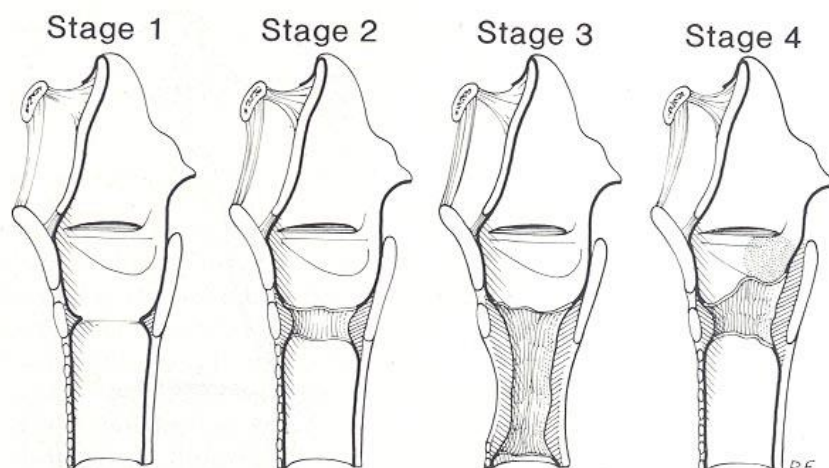
- Grading systems of SGS:

1. Lano grading system:

- Based on the subsites involved.
- Does not take into account length of stenosis or lumen diameter.
- Helpful in predicting the rate of success decannulation after resection and anastomosis.
- Stages:
 - **Stage I:**
 - One subsite involved.
 - 94% successful decanulation rate.
 - **Stage II:**
 - Two subsites involved.
 - 80% successful decanulation rate.
 - **Stage III:**
 - Three subsites involved.
 - 20% successful decanulation rate.

2. McCaffrey's grading system:

- Used for adult laryngotracheal stenosis.
- Based on subsites involved and length of stenosis.
- Does not include lumen diameter
- Helpful in predicting the rate of success decannulation after resection and anastomosis.
- Stages:
 - **Stage I:**
 - Isolated subglottic stenosis.
 - < 1cm in length.
 - **Stage II:**
 - Isolated subglottic stenosis.
 - > 1cm in length.
 - **Stage III:**
 - Subglottic and tracheal stenosis not involving the glottis.
 - **Stage IV:**
 - Subglottic and tracheal stenosis with glottic involvement.



3. Cotton-Myer grading system:

- Most widely used.
- Based on relative reduction of subglottic cross-sectional area.
- Good for mature, firm, circumferential lesions.
- Does not take into account extension to other subsites or length of stenosis.
- Exact diameter of stenosis can be difficult to measure.
- **NOT helpful in predicting the rate of success decannulation after resection and anastomosis.**
- Grades:
 - **Grade I:**
 - Lumen obstruction <50%.
 - **Grade II:**
 - Lumen obstruction 51-70%.
 - **Grade III:**
 - Lumen obstruction 71-99%.
 - Pinpoint hole.
 - **Grade IV:**
 - Complete stenosis.
 - No detectable stenosis.

Classification	From	To
Grade I	 No Obstruction	 50% Obstruction
Grade II	 51% Obstruction	 70% Obstruction
Grade III	 71% Obstruction	 99% Obstruction
Grade IV	No Detectable Lumen	

Fig. 5.12 Myer-Cotton airway grading system [36]

4. Modified Cotton-Myer grading system:

- New grading system.
- Based on relative reduction of subglottic cross-sectional area in addition to associated comorbidities and glottic involvement.
- **Helpful in predicting the rate of success decannulation after resection and anastomosis.**

Table 5.2 New airway grading system [35]

Myer–Cotton grade		Isolated SGS	Isolated SGS + comorbidities	SGS + glottic involvement	SGS + glottic involvement + comorbidities
		(a)	(b)	(c)	(d)
I	0–50%	I a	I b	I c	I d
II	51–70%	II a	II b	II c	II d
III	71–99%	III a	III b	III c	III d
IV	No lumen	IV a	IV b	IV c	IV d

Table 5.3 Overall decannulation rates of 100 PCTRs for grades III and IV SGS according to the new airway grading system [35]

Types of grade III–IV SGS		Nb	Overall decannulation (follow-up from 6 months to 21 years)
(a)	Isolated SGS	36	97%
(b)	Isolated SGS + comorbidities	31	93%
(c)	SGS + glottic involvement	19	89%
(d)	SGS + comorbidities + glottic involvement	14	64%

**TABLE
90.4**

**COMMON CONDITIONS THAT
MIMIC LARYNGEAL STENOSIS**

Neonate

Laryngomalacia
 Vascular compression (e.g., innominate artery, aberrant subclavian artery, vascular ring)
 Vocal fold paralysis
 Bilateral (e.g., Chiari deformity)
 Unilateral (e.g., traumatic post ductus ligation)
 Hemangioma, subglottic
 Complete tracheal rings
 Laryngeal cleft
 Cri du chat syndrome
 Laryngeal saccules, laryngoceles

Child

Dermoid, saccular, epiglottic cysts
 Lingual thyroid
 Infections (e.g., epiglottitis, croup, bacterial, laryngotracheitis)
 Foreign body
 Papillomatosis

Adult

Vocal fold paralysis (status post thyroid surgery, laryngeal carcinoma)
 Viral, granulomatous, fungal laryngitis
 Reinke edema
 Laryngeal trauma (blunt or penetrating)
 Postintubation
 Hemangioma, supraglottic
 Idiopathic (e.g., spastic dysphonia)
 Neurologic (e.g., amyotrophic lateral sclerosis, multiple sclerosis)
 Caustic ingestion
 Systemic disease (e.g., rheumatoid arthritis, polychondritis)
 Tracheal tumors or obstruction

- **Treatment plans of SGS:**

- **Newborn baby with SGS and respiratory distress with failure of intubation:**

○ **DLB:**

- Endoscopic evaluation should precede intubation or tracheotomy if possible in order to establish the site and cause of the airway obstruction.
- Patient should be deeply sedated on spontaneous breathing (no muscle relaxants) to avoid potential laryngospasm when bronchoscope is situated just above the vocal cords.
- Larynx is exposed using laryngoscope with blade inserted into vallecula.
- Examination endolarynx, subglottis and trachea all the way down to carina with rigid 2.7-mm-telescope (4mm telescope is preferred if possible).
- Smallest available ETT can be loaded over the telescope for trial of intubation.

○ **Emergency Tracheostomy:**

- Performed once the intubation is failed with ETT, especially in newborns with C-SGS with elliptical cricoid shape.
- If PCTR is planned for the patient (SGS grade 3 or 4), tracheotomy is preferred be done either:
 - High tracheotomy immediately below cricoid ring to reduces length of the resection and preserve maximally the distal normal trachea.
 - Low tracheotomy between 6-7th tracheal rings to spare a sufficient amount of normal tracheal rings between subglottic stenosis and tracheostomy site.
- In neonate with congenital cardiac anomalies, trial should be made to avoid tracheostomy as possible, as it may delay crucial cardiac surgeries in order to avoid spreading of the infection from nearby tracheostomy site to the fresh mediastinal wound.
- After securing the airway with tracheostomy, complete work-up should be done to prepare the patient for further elective airway surgery (endoscopic or open).

- **Intubated baby with failure of extubation:**

○ **DLB:**

- Endoscopic evaluation should precede tracheotomy if possible in order to establish the site and cause of the airway obstruction.
- Examination of airway should be done while patient is deeply sedated on spontaneous breathing (no muscle relaxants).
- **Findings:**
- **Granulation tissue:**
 - Suspension MLS + removal of exophytic granulation tissue using biopsy forceps followed by Mitomycin C application.
 - Using CO₂ laser is contraindicated as it will carbonise the highly vascularized granulation tissue leading to heat diffusion into surrounding tissues with subsequent scarring.
 - Atraumatic re-intubation is done with a one-size smaller ETT covered with Gentamycin-corticosteroid ointment (Diprogenta).
 - Patient is covered with IV dexamethasone (2 mg/kg) and antibiotics.
 - DLB is done after 4 days to assess the airway followed by trial of extubation in OR.
 - Post-extubation medical care:
 - Racemic epinephrine (50 mg/kg in 4 ml of NaCl 0.9%).
 - IV Dexamethason (2 mg/kg)
 - CPAP
 - Heliox through face mask
- **Laryngeal edema without mucosal necrosis:**
 - Re-intubation with one-size smaller ETT covered with Diprogenta.
 - Patient is covered with IV dexamethasone (2 mg/kg) and antibiotics.
 - Extubation after 4 days with good post-extubation medical care.
 - If extubation failed, then plan for Anterior cricoid split (ACS) if the patient met the required criteria to avoid tracheostomy.
- **Acquired soft-tissue mild SGS:**
 - Endoscopic balloon dilation with Mitomycin C application.
- **Severe SGS:**
 - Open surgical approach as single stage or double stage (depending of patient comorbidities).

Fig. 14.12 Exuberant granulation tissue filling the posterior glottis after endotracheal tube removal: (a) Severe dyspnoea resulting from airway obstruction of the posterior glottis. (b) Status 5 days after endoscopic treatment with removal of granulation tissue and topical application of mitomycin C. Uneventful extubation

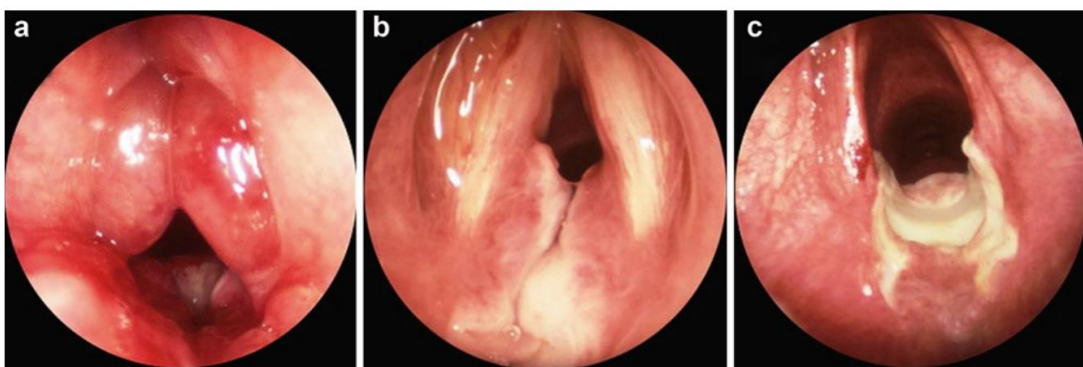
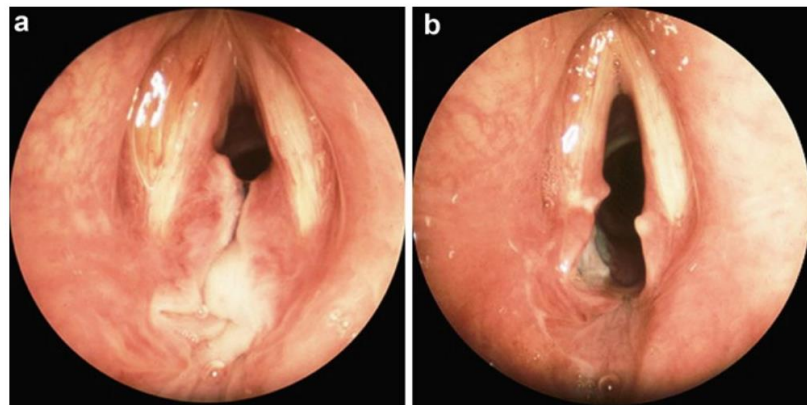


Fig. 14.8 Acute intubation lesions with different symptoms and outcomes: (a) Prominent oedematous protrusion of ventricular mucosa. Post-extubation dyspnoea, with complete resolution following medical treatment alone. (b) Ulcerated troughs with exuberant granulation tissue filling the posterior glottis. Post-extu-

bation dyspnoea, with complete resolution following combined endoscopic and medical treatments. (c) Annular interarytenoid ulceration with exposed cartilage but no granulation tissue in an immuno-compromised adolescent. Post-extubation dysphonia but no dyspnoea. Progression to posterior glottic stenosis

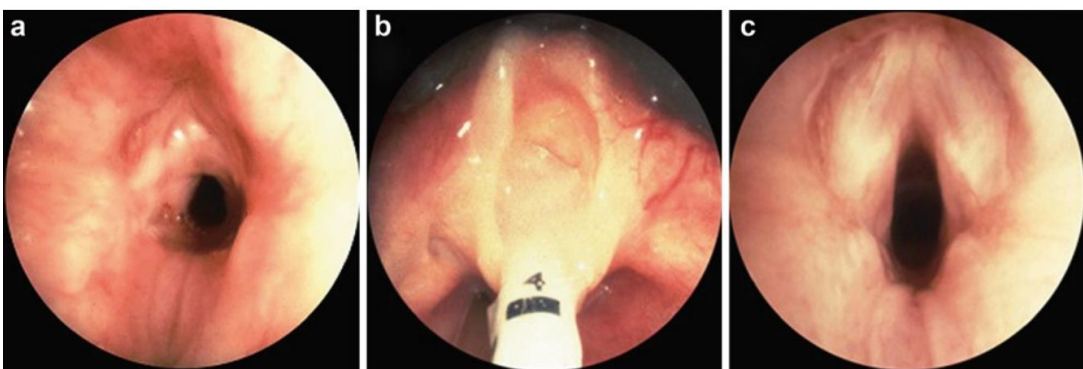


Fig. 14.9 Post-intubation soft-tissue stenosis in a newborn baby: (a) Diffuse submucosal swelling of the glottis and subglottis, narrowing the lumen to 70%. (b) Re-intubation with a

one-size smaller soft Portex Blue Line® tube and topical application of a gentamicin-corticosteroid ointment (Diprogenta®) plug. (c) Subnormal airway at extubation 4 days later

○ **Elective Tracheostomy:**

- Endoscopic evaluation should precede tracheostomy if possible in order to establish the site and cause of the airway obstruction.
- If PCTR is planned, high or low tracheostomy is done.
- Residual normal trachea must be maximally preserved.
- Try to avoid tracheostomy as possible if the patient is going for open cardiac surgery.
- After securing the airway with tracheostomy, complete work-up should be done to prepare the patient for further elective airway surgery (endoscopic or open).

Fig. 14.18 Correct placement of tracheostomy with impending laryngotracheal stenosis: (a) Tracheostoma situated immediately below the cricoid ring to maximally preserve normal trachea. (b) Tracheostoma situated at the sixth–seventh or eighth tracheal ring to spare sufficient length of normal trachea between the stenosis and the tracheostomy

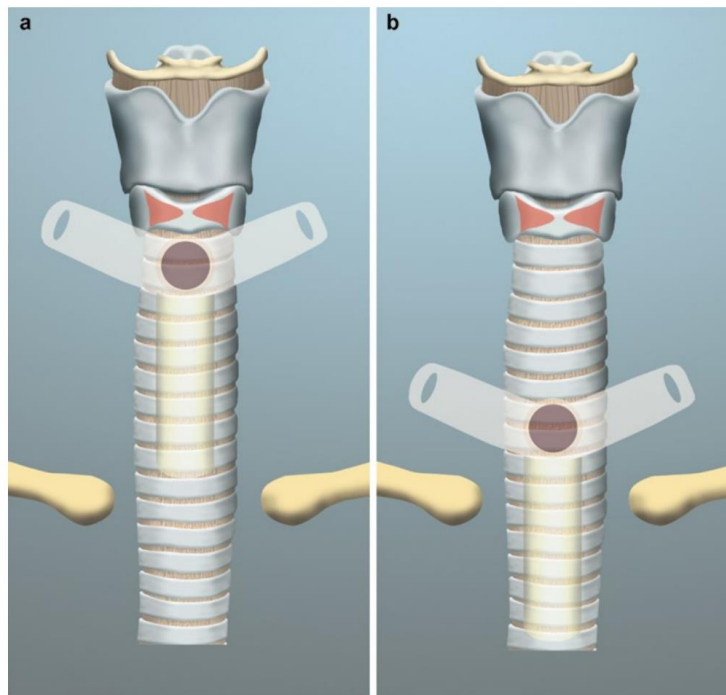
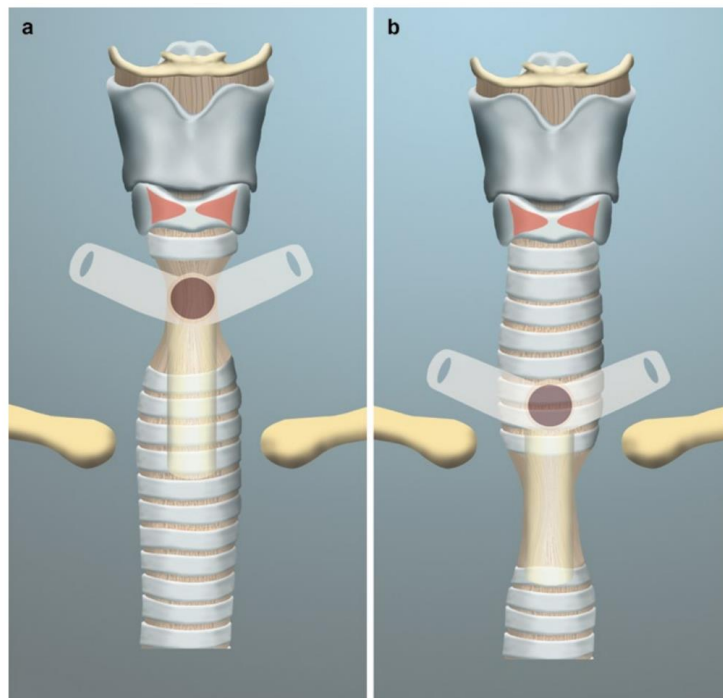


Fig. 14.19 Correct placement of tracheostomy when subglottic stenosis is associated with tracheal stenosis: (a) Cervical cuff or post-tracheostomy stenosis: the new tracheostomy must be placed in the stenotic segment. (b) Substernal cuff or cannula tip stenosis: the new tracheostomy must be placed just above the stenosis, which is stented by the tracheostomy cannula



- **Stable child with known airway obstruction:**
 - **Diagnostic DLB:**
 - Confirm the diagnosis of SGS.
 - Done with rigid telescope followed by suspension microlaryngoscopy.
 - Complete assessment of:
 1. SGS
 2. Associated airway anomalies
 3. Vocal folds mobility
 - **Counseling:**
 - Thorough discussion with child's parents is essential before any surgical intervention.
 - Parents expect the outcome to be a normally breathing and speaking child after surgery, which is not realistic, especially in terms of voice.
 - In difficult cases in children with comorbidities, multidisciplinary approach should be applied.
 - **Consultations:**
 - Gastroenterology:
 - GERD can induce chronic irritation of the pharyngolarynx which can cause or aggravate an upper airway stenosis.
 - **24-hour dual pH monitoring** is indicated prior to surgical management to evaluate the presence and severity of GERD.
 - All patients should be kept on oral PPI (**1mg/kg**) prior and after any surgical intervention to control the acid reflux.
 - Patients with uncontrolled reflux on PPI should be considered for fundoplication prior to open airway surgery.
 - Uncontrolled reflux is considered a cause of failure of open airway surgery.
 - Swallowing:
 - Aspiration should be ruled out before considering any open airway surgery.
 - Swallowing assessment is done either by fiberoptic endoscopic evaluation of swallowing (**FEES**) or modified barium swallow (**MBS**).
 - Cardiology:
 - Cardiac patients should undergo thorough evaluation with ECHO and ECG.
 - If the patient is planned for surgical intervention to correct any cardiac anomaly, tracheostomy should be avoided as possible and single-stage LTR or PCTR can be considered instead.

▪ Pulmonology:

- Pulmonary function tests (**PFT**) provide precise information regarding pulmonary reserve, which can't be done in tracheostomized patients.
- Pulmonary reserve can be estimated in tracheostomized patients from the rapidity of recovering from temporary O₂ desaturation after intentional apnea episodes during DLB.
- History of prematurity raise possibility of poor pulmonary reserve as a sequela to hyaline membrane disease.
- Pulmonary reserve can help decide if a single-stage or a double-stage surgery should be considered for SGS.

Table 17.2 Patient's general condition

• Overall appearance
• Nutritional aspect
– Bodyweight and height for age
• Dysmorphic features
– Syndromic or non-syndromic
• Pulmonary status
– Chest deformity
– Oxygen requirement, need for bronchodilators
• Cardiac anomalies
– Pulmonary hypertension
– Shunts
• Neurologic impairments
– Swallowing disorders
– Mental retardation
• Gastro-oesophageal reflux, eosinophilic oesophagitis

Table 17.10 Optimization of patient's status prior to airway reconstruction

• Address and treat all comorbidities amenable to improvement
• Consider potential contraindication to surgery in severely handicapped children who cannot be improved by any means
• Start medical treatment of MRSA and pseudomonas aeruginosa airway colonisation 5 days prior to surgery
• Treat all patients for GOR, starting in the preoperative period and continuing possibly for 6 months to a year in overt GOR
• Postpone surgery and treat for eosinophilic oesophagitis if it has been biopsy-proven

- **Timing of surgery:**

- Observation:
 - Indicated for patients with mild stenosis (Grade I and mild Grade II), especially in C-SGS, as the airway is expected to enlarge with growth.
 - Follow-up endoscopies should be done every 6 months until complete resolution of respiratory symptoms.
- Early surgical intervention:
 - In children with no significant comorbidities, early intervention during the first months of life is advisable.
 - Important for speech and language development and eliminating morbidity and mortality of tracheotomy.
 - No need to wait until the child reaches 10 kg before doing any surgical intervention.
- Prerequisites before any airway surgery for SGS:
 - In C-SGS, there should be no other relevant airway obstruction.
 - In A-SGS, original reason for intubation should no longer exist.
 - Patient should be in good respiratory and neurological condition.
 - Stenosis must be composed of mature scar tissue.
 - Associated medical conditions should be treated.

Table 17.9 Temporary contraindications to surgery for LTS

- | |
|--|
| <ul style="list-style-type: none"> • Local factors <ul style="list-style-type: none"> – Highly reactive larynx, severe reflux laryngitis – Immature LTS – Other sites of significant airway obstruction – Infected airway – Pharyngolaryngeal discoordination with aspiration |
| <ul style="list-style-type: none"> • Systemic factors <ul style="list-style-type: none"> – Persistent pulmonary disease – Uncontrolled GORD – Severe neurological impairment – Severe mental disability – Severe cardiovascular anomalies |

- **Preoperative Planning Questions:**

- Have all comorbidities been addressed prior to surgery?
 - Gastro-esophageal reflux disease?
 - Neurologic, pulmonary and cardiac diseases?
 - Nutrition?
 - Airway infection/contamination?
- Is surgical timing appropriate?
 - Maturity of subglottic stenosis?
 - Patient's age in relation to potential comorbidities?
- What type of surgery should be performed?
 - Endoscopic
 - LTR
 - PCTR
 - Extended PCTR
- Type of costal grafting?
 - No graft
 - Anterior graft
 - Posterior graft
 - Combined grafts
- How many stage?
 - Single-stage
 - Double-stage
- Need for stenting?
 - With
 - Without
- Need for post-op PICU and for how long?
- Risks involved with the different types of surgery?
 - LTR: significant recurrence rate in grade III–IV SGS
 - PCTR: anastomotic dehiscence and RLN injury.
- Expected results regarding airway, voice and swallowing?

- **General surgical approach based on SGS grading:**

- **Grade I (<50% luminal obstruction):**
 - Mostly, it does not require any surgical intervention.
 - **Normal glottis:**
 - Managed using **endoscopic approach**.
 - **Abnormal glottis:**
 - Managed using **LTR with anterior or posterior graft + stenting** (depending on the location of glottic involvement).

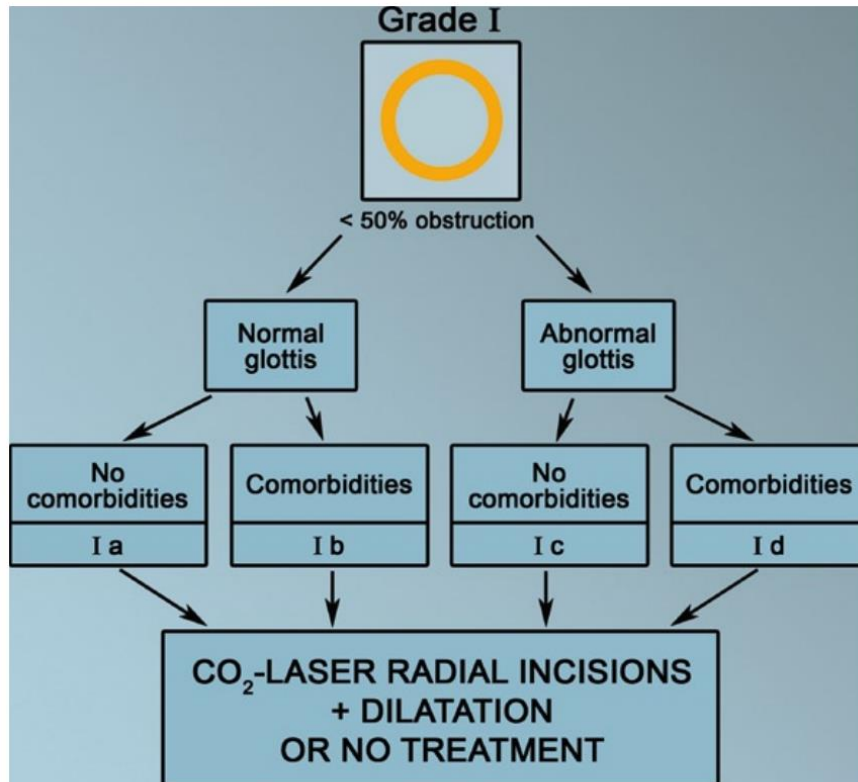


Table 17.5 Treatment algorithm for Grade I SGS

- **Grade II (51-70% luminal obstruction):**
 - **Normal glottis:**
 - Managed using **endoscopic approach**.
 - Extensive SGS in cranio-caudal axis is managed using **LTR with anterior graft + stenting**
 - **Abnormal glottis:**
 - Managed using **LTR with anterior or posterior graft + stenting** (depending on the location of glottic involvement).

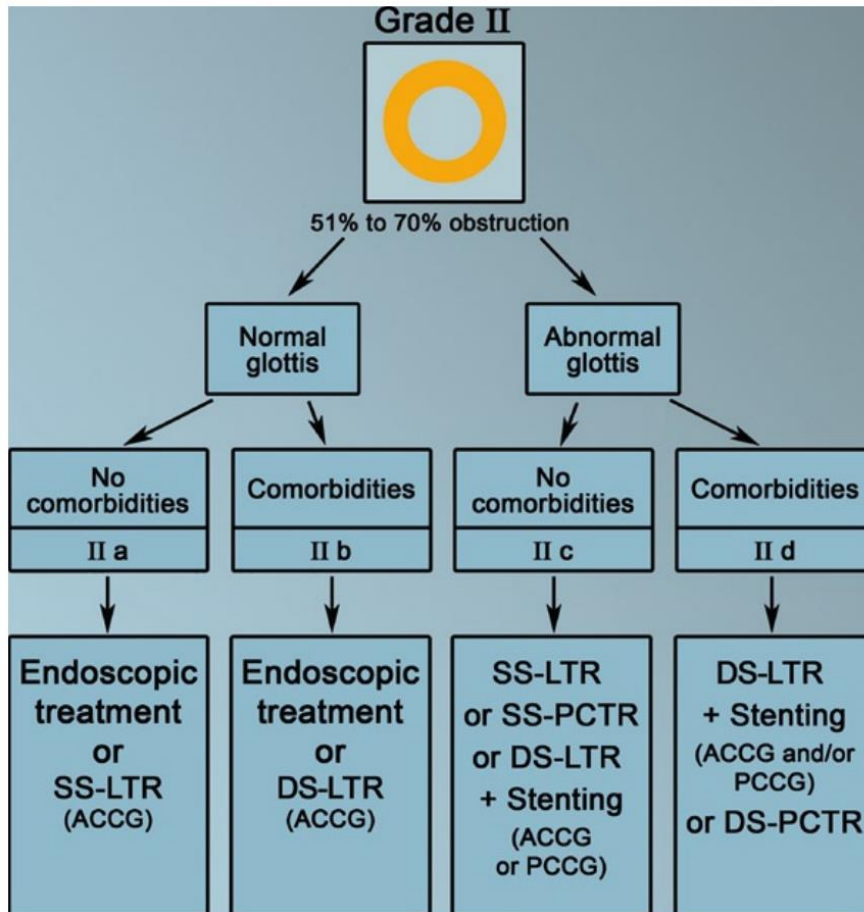


Table 17.6 Treatment algorithm for Grade II SGS

- **Grade III (71-99% luminal obstruction):**
 - **Normal glottis:**
 - Mild Grade III (71-80%):
 - **<5mm cranio-caudal axis:**
 - Managed using **endoscopic approach**.
 - If stenosis reoccurs to its original grade, then managed using open approach.
 - **>5mm cranio-caudal axis:**
 - Managed using **LTR with anterior graft + stenting**.
 - Severe Grade III (81-99%):
 - Managed using:
 - **PCTR (Surgery of choice)**
 - **LTR with anterior and posterior grafts and long-term stenting.**
 - **Abnormal glottis:**
 - Managed using:
 - **Extended PCTR (Surgery of choice)**
 - **LTR with anterior and posterior grafts and long-term stenting.**
- **Grade IV (100% luminal obstruction):**
 - **Normal glottis:**
 - Managed using:
 - **PCTR (Surgery of choice)**
 - **LTR with anterior and posterior grafts and long-term stenting.**
 - **Abnormal glottis:**
 - Managed using:
 - **Extended PCTR (Surgery of choice)**
 - **LTR with anterior and posterior grafts and long-term stenting.**

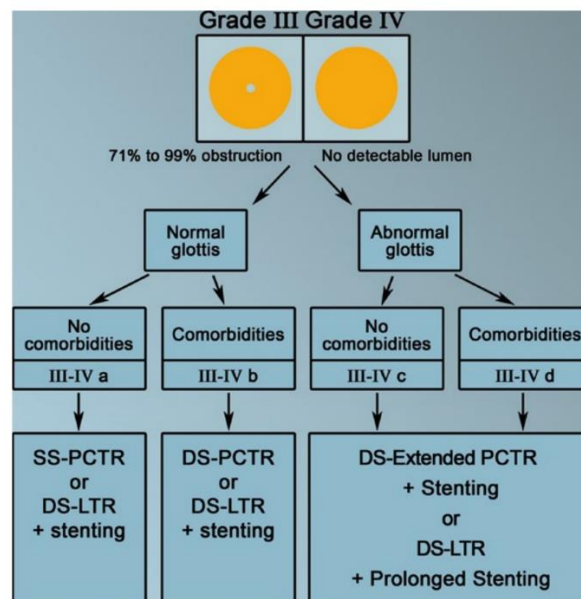


Table 17.7 Treatment algorithm for Grade III and IV SGS

- **Endoscopic approach for treatment of SGS:**
- Composed of:
 1. CO2 laser
 2. Dilations
 3. Mitomycin C application
- Indications of endoscopic approach:
 1. Grade 1 acquired SGS
 2. Grade 2 acquired SGS
 3. Mild Grade 3 acquired SGS $\leq 5\text{mm}$ in cranio-caudal extension
 4. Additional treatment to open surgery to optimize the final surgical results.
- Contraindications of endoscopic approach:
 1. Congenital cartilaginous SGS
- Risk factors for failure of endoscopic approach:
 1. Failure of previous endoscopic procedures
 2. Glottic involvement with interarytenoid fibrosis
 3. Malacia and collapse due to loss of cartilaginous support
 4. Circumferential cicatricial scarring $>1\text{ cm}$ in vertical dimension
 5. Exposure of perichondrium or cartilage during Co2 laser excision predisposing to perichondritis and chondritis
 6. Severe bacterial infection of trachea after tracheotomy
 7. Concomitant tracheal stenosis.
- **Endoscopic CO2 laser:**
 - Success rates 90% in grade I, 50% in grade II and 15% in grade III SGSs.
 - Allows a successful outcome without a tracheotomy.
 - Preferred for:
 - Thin web-like SGSs $\leq 5\text{mm}$ in cranio-caudal extension in pediatrics.
 - Advantages:
 - Precise surgical excision
 - Low surgical morbidity
 - Minimal damage to underlying or surrounding tissues
 - Shapshay's technique for concentric web SGS:
 - Laser radial incisions at the four cardinal points followed by dilatation.
 - Prevent circumferential restenosis.
 - Technique for asymmetrical web SGS:
 - Laser resection of $< 50\%$ of the subglottic circumference.
 - Preserve the mucosa of the contralateral side for the re-epithelialization process.

Fig. 18.2 Diagram of Shapsay's endoscopic technique for treating a concentric web-like subglottic stenosis: (a) CO₂ laser radial incisions at the four cardinal points. (b) Result after dilation: residual mucosal bridges between laser incisions facilitate the reepithelialisation process

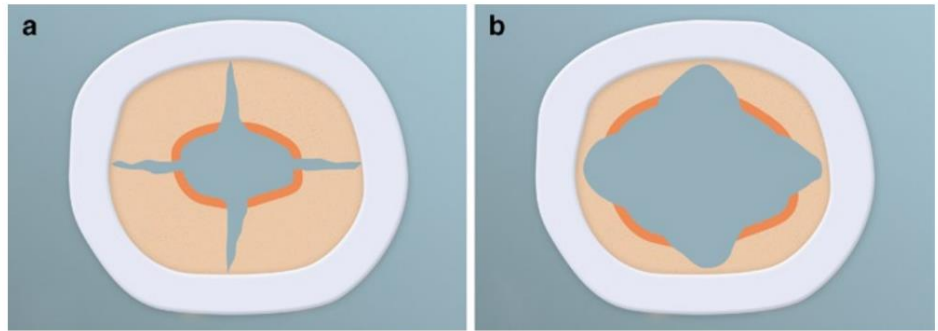
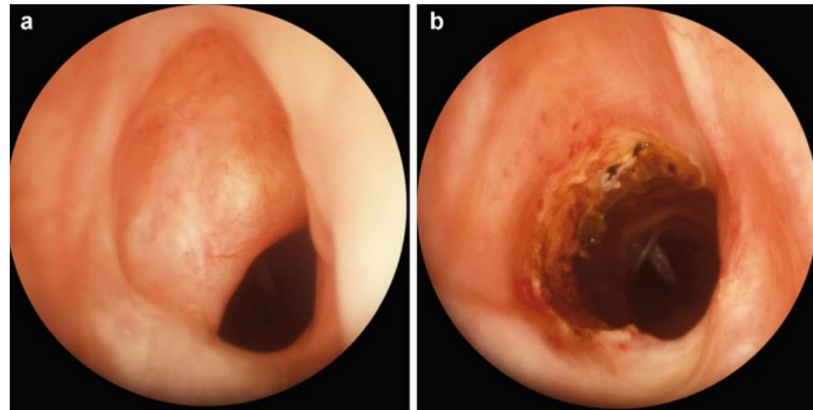


Fig. 18.1 Web-like, asymmetrical subglottic stenosis: (a) Preoperative view: the thin, web-like diaphragm occupies the left subglottic space. (b) Immediate postoperative view: the web has been fully resected using the CO₂ laser. Note the absence of charring. The late outcome was a subnormal airway after a single endoscopic treatment



- **Dilation:**
 - Preferred for:
 - Early fresh SGS.
 - Not preferred for:
 - Mature firm stenosis
 - Cartilaginous stenosis.
 - Introduced into the airway during intermittent apneas.
 - Types:
 1. **Savary-Gilliard dilators:**
 - Consist of flexible, tapered, incompressible bougies.
 - Ranging from 5-15 mm in diameter.
 - Advantage:
 - The resistance felt during dilation is an excellent indicator of maximal size that should be used.



Fig. 4.40 Savary-Gilliard tracheal dilators: The well-tapered nose allows for smooth and progressive dilation

2. Balloon Dilators:

- Angioplasty high-pressure noncompliant balloons are inflated with water to increase their inside pressure.
- Promote an outward expansion, even in the presence of very reduced lumen.
- Less risk of tissue injury and scar formation.
- Lack of tactile feedback.
- Comes in different sizes.



Figure 1. Stylet and balloon port location

Airway Balloon Size (diameter x length)	Maximum Pressure
5x24 mm	16 atm
7x24 mm	16 atm
8.5x24 mm	12 atm
10x40 mm	12 atm
12x40 mm	10 atm
14x40 mm	10 atm
16x40 mm	8 atm

- **Mitomycin C (MMC) application:**
 - Anti-neoplastic antibiotic acts as alkylating agent by inhibiting DNA synthesis.
 - Isolated from streptomyces caespitosus strain of actinomyces.
 - Modulate wound healing response by:
 1. Inhibiting neo-angiogenesis
 2. Inhibiting fibroblast proliferation
 - Results in:
 - Prevention of scar tissue and granulation formation.
 - Usage in airway surgery:
 - Topical application of pledgets soaked in 4ml of MMC (4mg/ml) for 4 mins over raw mucosal surfaces.
 - Complications:
 - Excessive fibrinous exudate production and potential airway obstruction 24–48 h after surgery **(5%)**.
 - Prevented by application of pledgets soaked in saline over the wound.
 - Contraindications:
 - Topical applications over denuded cartilage due to risk of cartilage necrosis from preventing granulation tissue formation over the exposed cartilage.
 - Topical applications within 6 weeks following PCTR due to risk of anastomotic dehiscence from reducing scar formation.



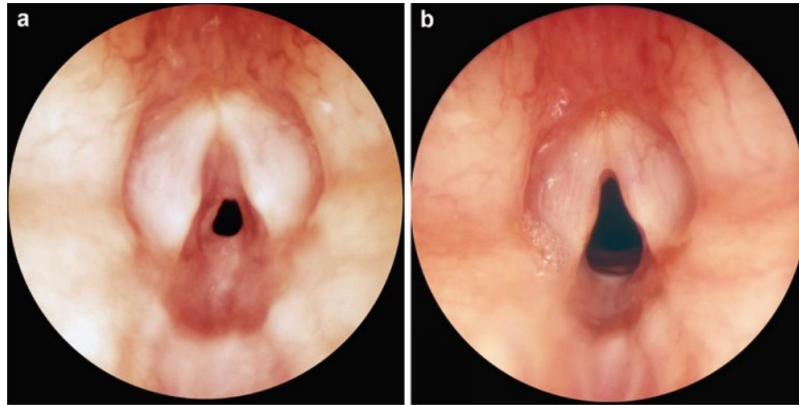


Fig. 18.4 Recurrent web-like concentric glotto-subglottic stenosis after partial cricotracheal resection: **(a)** Preoperative view: mild grade III subglottic stenosis treated by CO₂ laser incisions, dilation and mitomycin C (2 mg/ml for 2 min). **(b)** Postoperative view at 6 months showing the result after a

single endoscopic treatment: The airway is adequate, albeit with an anterior synechia of the vocal cords. Parents refused any further treatment over the next 5 years. Initially, the endoscopic treatment was performed solely to “buy time” before considering open revision surgery

- **Local corticosteroids injection:**
 - Controversial.
 - Local injection of long-acting steroid (Triamcinolon acetone/Kenacort)
 - Modulate wound healing response by:
 1. Delaying synthesis of collagen in early stages of wound healing.
 2. Increasing collagen lysis in late stages.
 - Results in:
 - Decrease scar formation
 - Complications:
 - Increasing scar formation and predisposing to infection of the exposed cartilage by delaying epithelial migration necessary to resurface the denuded area.
 - Resorption of cartilage secondary to presence of local corticosteroids is a serious complication.

Table 2. Treatment results

Procedure	Total procedures	# With successful outcome	% With successful outcome
CO ₂ laser	20	3	15%
CO ₂ with steroid injection	11	2	18.2%
CO ₂ with mitomycin-C	16	12	75%

- **Open approach for treatment of SGS:**
- Composed of:
 1. Anterior Cricoid Split (ACS)
 2. Laryngotracheal Reconstruction (LTR)
 3. Partial Cricotracheal Resection (PCTR)
 4. Extended Cricotracheal Resection (Extended-PCTR)
- Aim of open approach:
 - o Early decannulation with minimal effect on the voice.
- Indications of open approach:
 1. Congenital cartilaginous SGS
 2. Advance grade 3 and 4 acquired SGS
 3. Glottic involvement
- Contraindications of open approach:
 - o **Local factors:**
 1. Immature SGS
 2. Presence of other sites of significant airway obstruction
 3. infected airway
 4. Swallowing incoordination with aspiration
 - o **Systemic factors:**
 1. Persistent pulmonary disease requiring tracheostomy
 2. Uncontrolled GERD
 3. Severe neurological or mental impairment
 4. Severe cardiovascular anomalies
 - o **Anterior cricoid split (ACS):**
 - Success rate reaches 80%
 - Indication:
 - Alternative to tracheostomy in intubated babies with acquired soft-tissue SGS following several failed extubation attempts.
 - Principle:
 - Anterior vertical midline transection of cricoid ring allows the cartilage to spring open.
 - Followed by mucosal section which results in leakage of submucosal edema fluid through the wound.
 - The overall outcome is reducing subglottic edema which allows for extubation without the need for tracheostomy.
 - Strict criteria should be met:
 1. ≥ 2 failed extubation trials due to SGS.
 2. Weight > 1.5 kg
 3. Off ventilator for ≥ 10 days
 4. Oxygen requirement $< 30\%$
 5. No current acute respiratory infection
 6. Non-congestive heart failure for ≥ 1 month
 7. No hypertensive drug intake for ≥ 10 days

▪ Approach:

- Lower anterior laryngofissure done from lower 1/3 of thyroid cartilage, anterior arch of cricoid and first two tracheal rings.
- Incision should be below anterior commissure cranially to preserve the voice.
- Thyroid alar cartilage graft is placed over split site to prevent superinfection of the wound and diminish subcutaneous emphysema.
- Patient is kept intubated for 5 days before extubation.
- Corticosteroids are administered before extubation and continued for 5 days.

Fig. 14.10 Anterior cricoid split modified into an anterior laryngotracheal reconstruction: (a) Anterior laryngotracheal incision: the cricoid ring springs partially open. Harvesting of the thyroid cartilage from the upper alar portion. (b) Suturing the thyroid cartilage graft into position

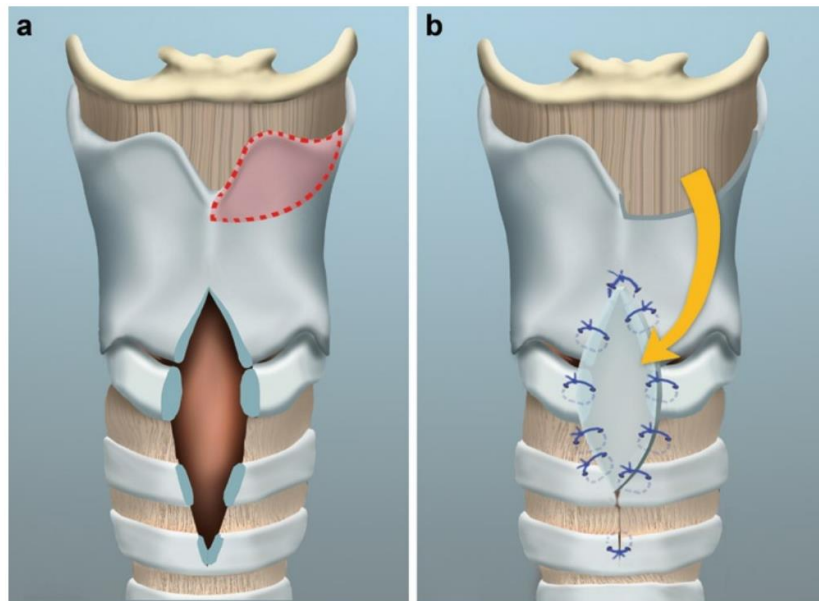
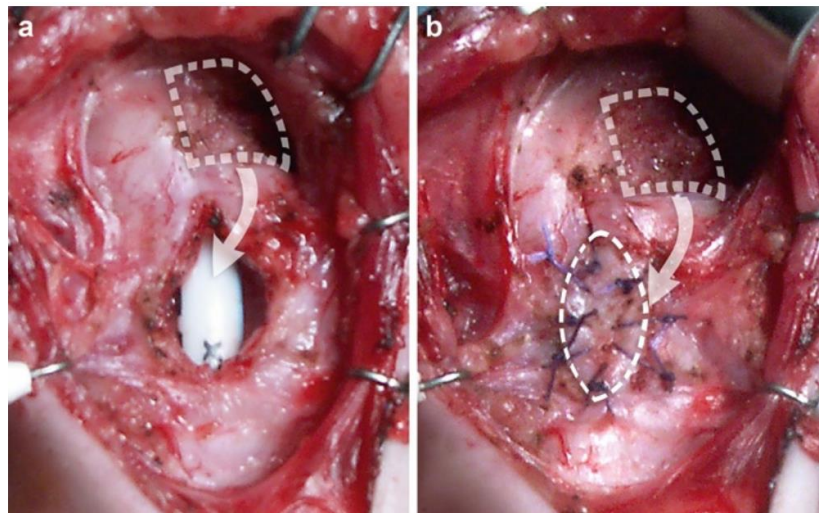


Fig. 14.11 Anterior cricoid split with thyroid cartilage graft in a premature baby intubated for 1 month: (a) The anterior cricoid split extends from the lower third of the thyroid cartilage to the second tracheal ring. The white dashed line shows the site of harvesting of the left upper thyroid ala. (b) The oval-shaped thyroid cartilage graft is sutured in position



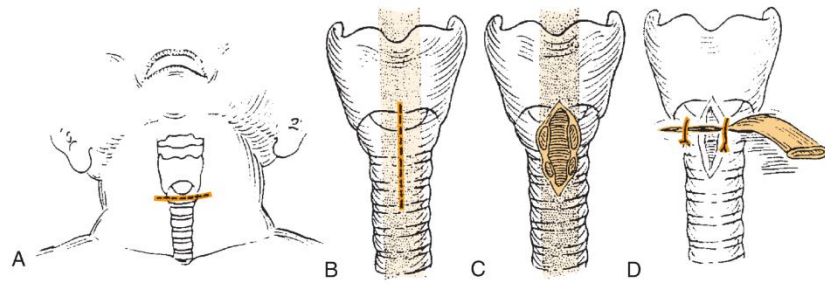


FIGURE 205-5. Anterior cricoid split operation. **A**, Skin incision. **B**, Midline laryngeal incision through cartilage and mucosa. **C**, With the cricoid cartilage decompressed, the endotracheal tube is ghosted in **B** and **C**. **D**, Loose skin closure with a drain inserted.

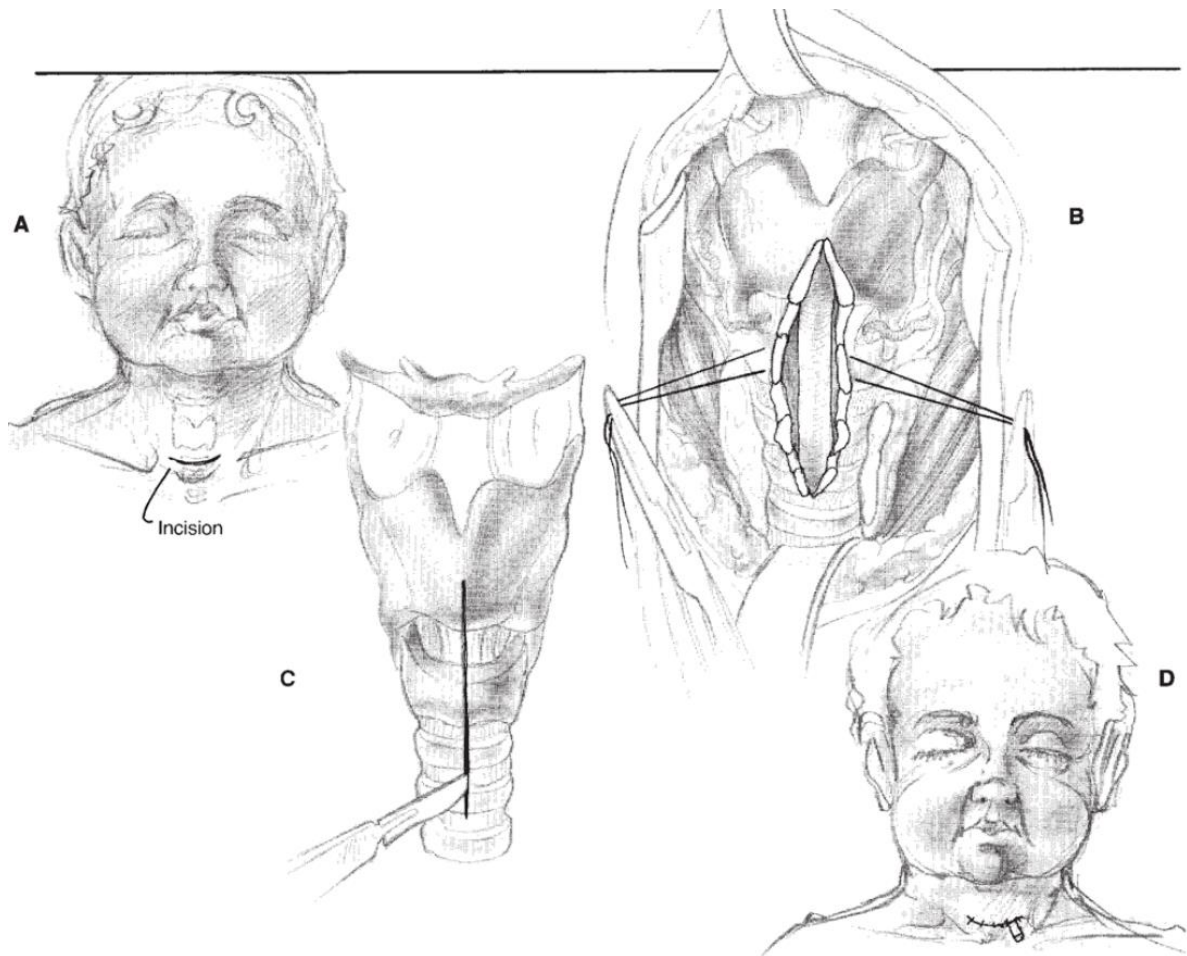


Figure 90.2 Anterior cricoid split. **A**: Skin incision (cricoid level). **B**: Vertical incision through the upper two tracheal rings, cricoid, and lower thyroid cartilage. **C**: Stay sutures. **D**: Loose skin closure with drain.

- **Laryngotracheal Reconstruction (LTR):**
 - Success rate reaches 90%
 - Indications:
 1. Congenital cartilaginous SGS
 2. Any SGS with glottic involvement
 3. Failed endoscopic approach for mild SGS
 4. Grade 3 and 4 SGS
 - Principle:
 - Augmentation of laryngotracheal complex by anterior or posterior midline incision of cricoid cartilage with insertion of cartilage grafts to expand the airway.
 - Scar tissue is not resected to preserve residual mucosa in the stenotic tract.
 - Types of graft:
 - **Costal cartilage:**
 - Best grafting material due to:
 - Good rigidity.
 - Availability in large quantities.
 - Easily trimmed and shaped.
 - Lower 3 medial costal cartilages (7,8,9th):
 - Flat and fused together to provide adequate width for the graft.
 - Steps:
 - Horizontal skin incision below mammary gland.
 - Desired length costal cartilage graft is harvested after preservation of inner surface perichondrium.
 - Surgical wound is filled with saline to rule out pneumothorax.
 - Drain is inserted and wound is closed.
 - **Other graft materials:**
 - Thyroid cartilage
 - Septal cartilage
 - Auricular cartilage
 - Hyoid bone

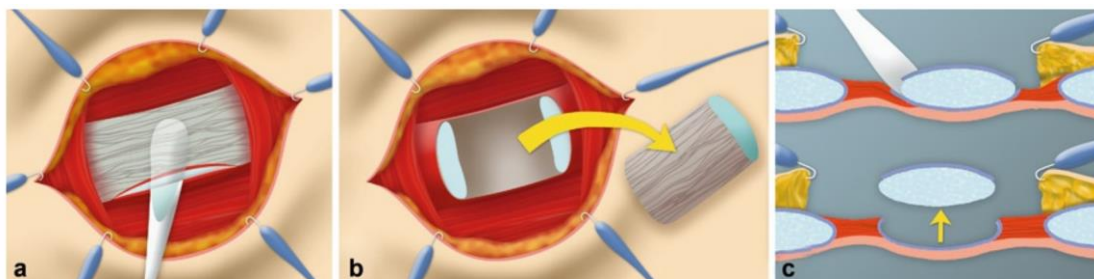


Fig. 19.4 Technique for obtaining a costal cartilage graft: (a) Incision at mid-distance of the rib thickness on the superior and inferior borders of the costal perichondrium. (b) Dissection in a subperichondrial plane on the deep chest surface and transsec-

tion of the cartilage at both extremities. A 2–4 cm long costal graft can usually be obtained. (c) Axial view of subperichondrial dissection on the deep chest surface

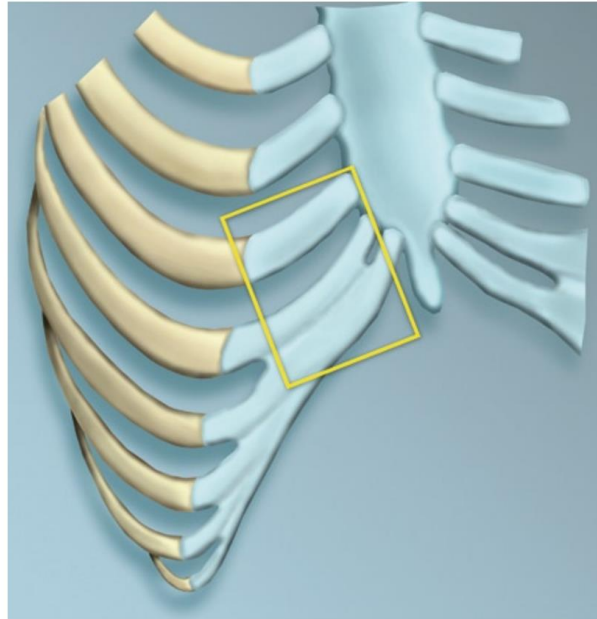


Fig. 19.3 Costal cartilage graft harvesting: the seventh, eighth and ninth rib cartilages frequently present a flat and slightly concave surface at the bony-cartilaginous junction to the sternum. As they are often fused together, they provide adequate width for the graft

- Location of cartilage graft:
 - **Anterior cartilage graft:**
 - Indications:
 - Congenital cartilaginous mild SGS.
 - Failed endoscopic approach for grade 1-2 and mild grade 3.
 - Steps:
 - Lower anterior laryngofissure is done from lower 1/3 of thyroid cartilage, anterior arch of cricoid and first two tracheal rings.
 - Incision should be below anterior commissure cranially to preserve the voice.
 - Carving the harvested anterior costal cartilage graft to a boat-shaped graft with flanges to prevent prolapse of the graft into the airway.
 - Placement carved anterior costal cartilage graft into the site of anterior laryngofissure, with the perichondrium facing the lumen followed by meticulous absorbable suturing.
 - Placement of Penrose drain and closure of the wound.

Fig. 19.9 Carving of the anterior cartilage graft: (a) On the perichondrial side, a boat-shaped template is drawn to match exactly the anterior laryngotracheal defect, preserving superior, inferior and lateral flanges of the cartilage. (b) Results after proper carving of the anterior costal cartilage graft

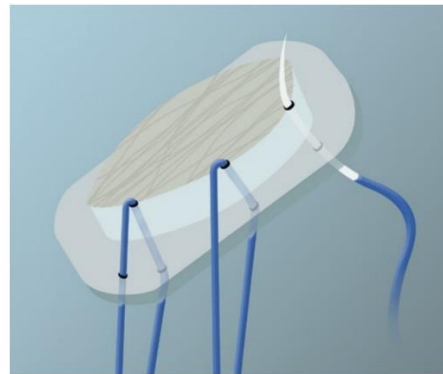
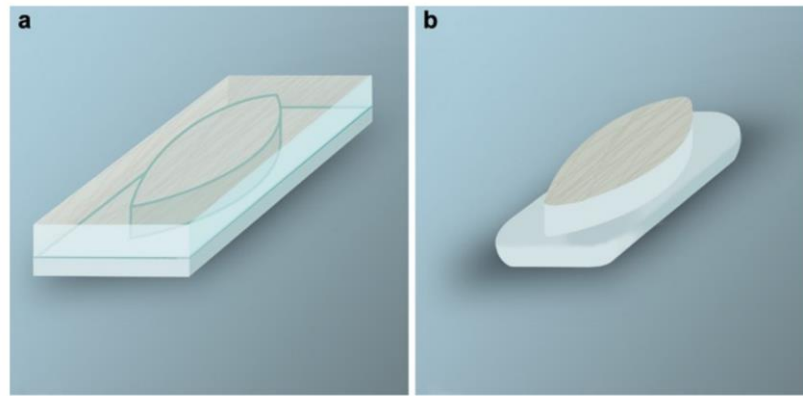


Fig. 19.10 Correct placement of stitches through the boat-shaped anterior costal cartilage graft for laryngotracheal reconstruction: the stitch, inserted through the dorsal portion of the cartilage, must emerge exactly at the edge of the perichondrium on the boat-shaped portion of the costal cartilage graft

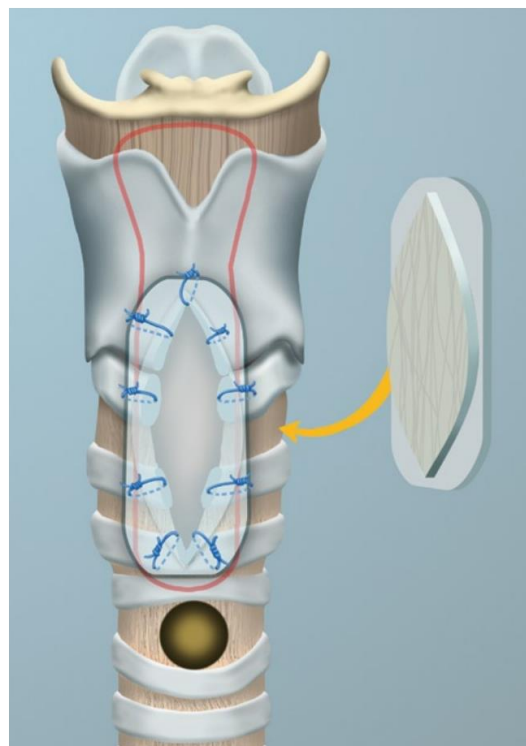


Fig. 19.11 Double-stage laryngotracheal reconstruction with anterior costal cartilage graft: the costal cartilage graft is sewn into position, with the perichondrium facing the lumen. Large cartilage flanges prevent prolapse of the costal cartilage graft into the airway. For details on suturing in cross section, refer to Fig. 19.7c)

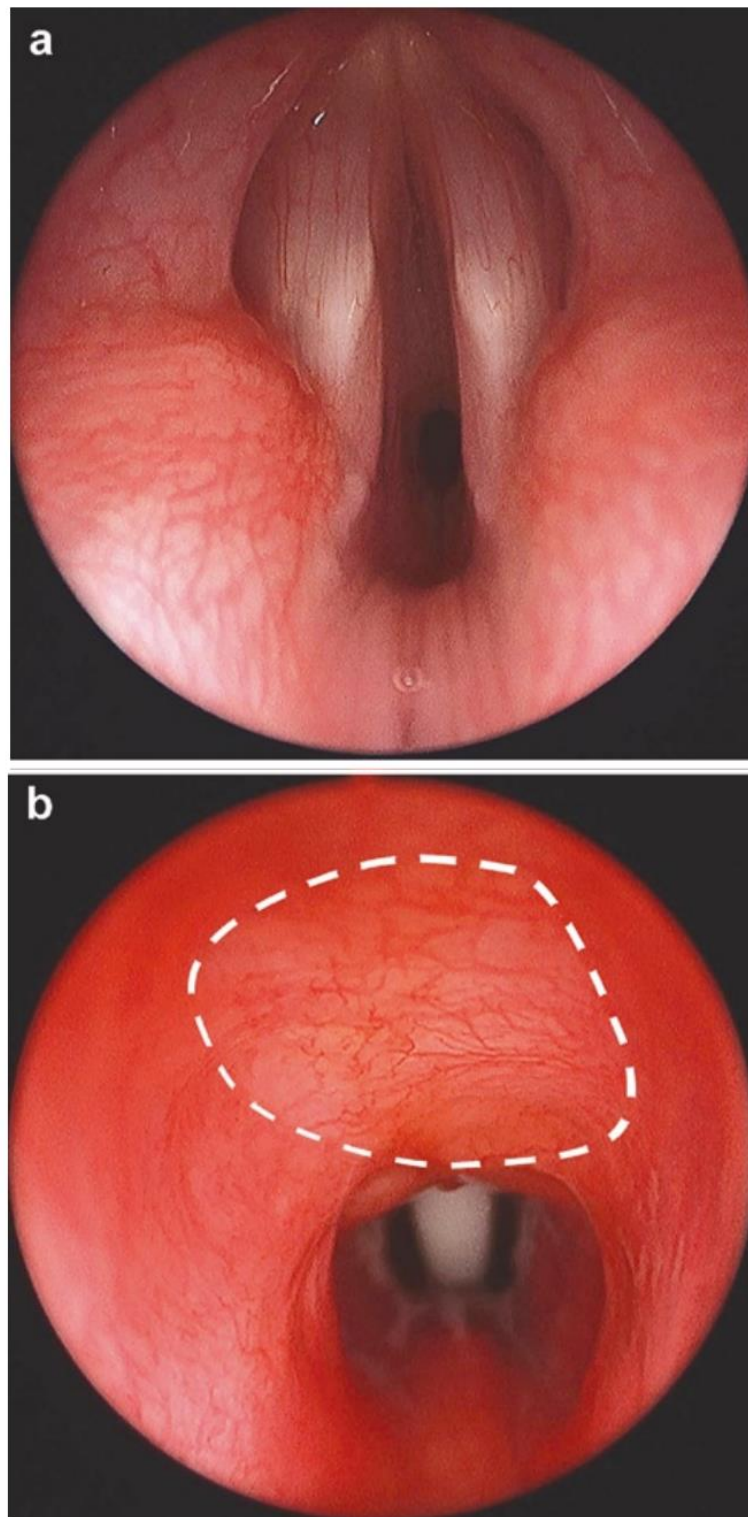


Fig. 19.12 Laryngotracheal reconstruction with anterior costal cartilage graft for Grade II congenital subglottic stenosis: (a) Preoperative view: elliptical cricoid. (b) Postoperative view at 3 months: large, round-shaped subglottis with perfect integration of the anterior costal cartilage graft (*dotted white line*)

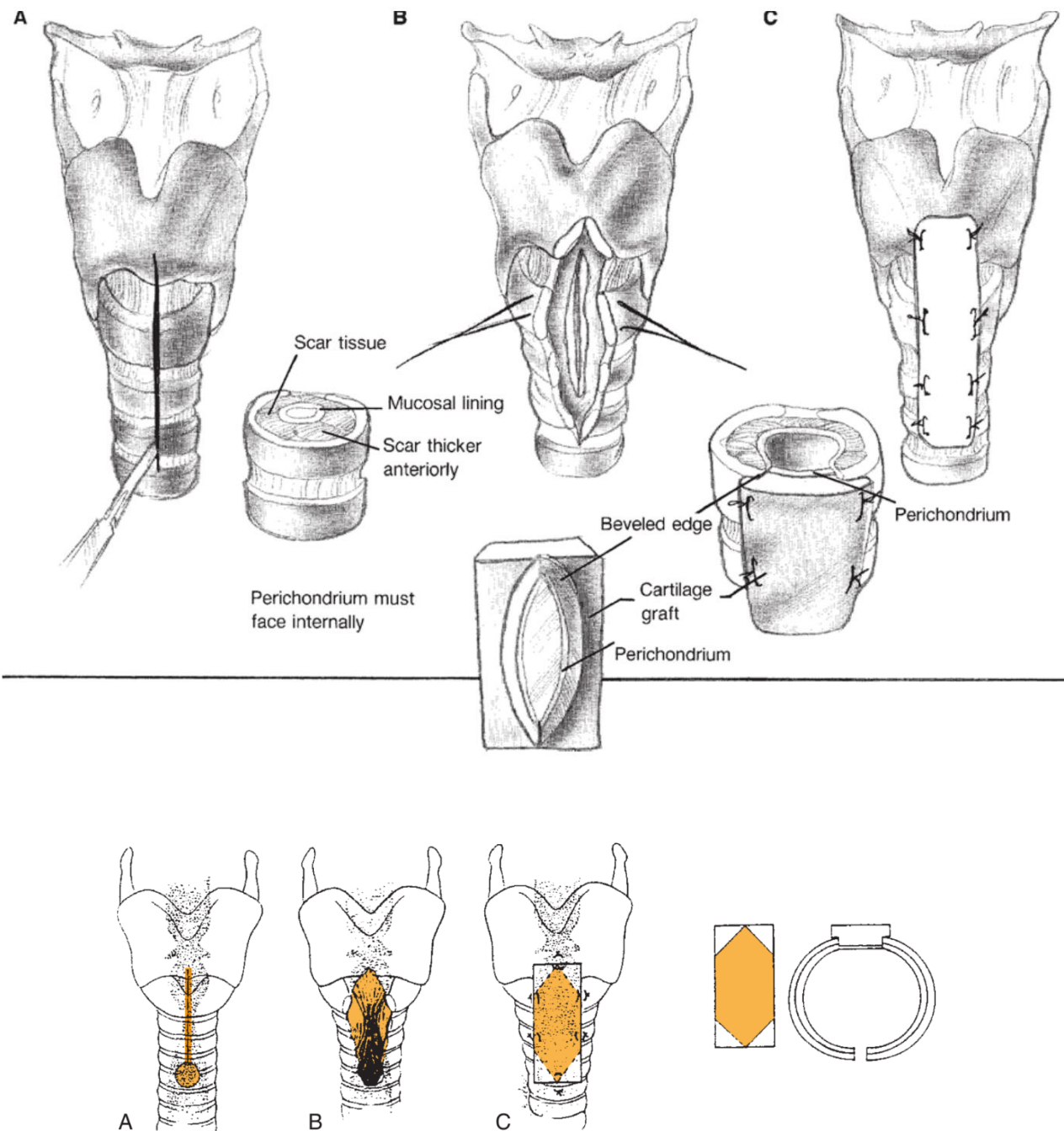
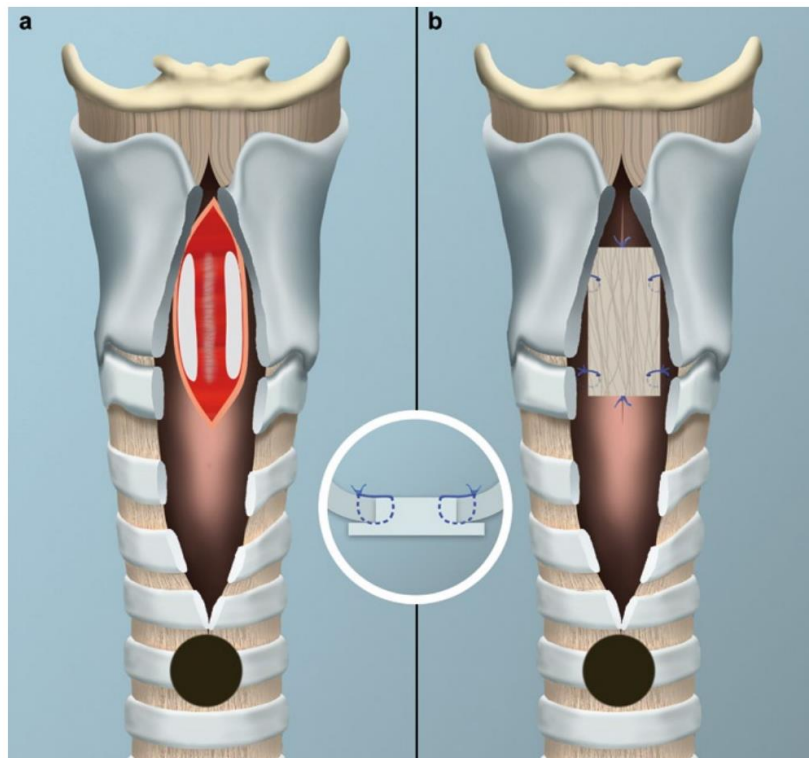


FIGURE 205-9. Anterior cartilage graft. **A**, Vertical incision is made into the thyroid cartilage from a point immediately below the anterior commissure through the upper tracheal rings, with care taken to remain in the midline. **B**, Intraluminal scar and lining mucosa are incised along the length of the stenotic segment. **C**, Costal cartilage is shaped into a modified boat (*inset*) and is placed in position with the lining of the perichondrium facing internally.

- **Posterior cartilage graft:**

- Indications:
 - Mild SGS (1 or 2 or mild 3) with posterior glottic involvement.
- Steps:
 - Full anterior laryngofissure is done from thyroid notch of thyroid cartilage, anterior arch of cricoid and first two tracheal rings (lower anterior laryngofissure is preferred if possible as not to disrupt the anterior commissure).
 - Provide optimal exposure of cricoid plate for the posterior cricoid split.
 - Posterior cricoid splint is done from inter-arytenoid region cranially to membranous tracheal wall.
 - Carving the harvested posterior costal cartilage graft to a rectangular-shaped graft with flanges.
 - Placement carved anterior costal cartilage graft between the two posterior cricoid laminae, with the perichondrium facing the lumen followed by meticulous absorbable suturing.
 - Anterior laryngofissure is sutured without grafting.

Fig. 19.2 Full laryngotracheofissure for subglottic stenosis combined with glottic involvement: **(a)** The incision is first made just above the thyroid notch in order to provide complete visualisation of the glottis during the midline laryngofissure. The posterior cricoid split must be placed precisely in the midline. The cuts should be perpendicular to the plane of the cricoid plate. **(b)** Results after posterior costal cartilage grafting: The interarytenoid and subglottic spaces have been enlarged



- **Anterior and Posterior cartilage grafts:**
 - Indication:
 - Severe Grade 3 or 4 SGS.
 - Steps:
 - Combined anterior and posterior cartilage grafts utilizing full anterior laryngofissure.

Fig. 8.4 Diagram of LTR for elliptical cricoid cartilage stenosis: (a) Severe SGS with normal anteroposterior distance and preserved mucosa. Dotted arrows show anterior and posterior midline cricoidotomies. (b) Situation after subglottic enlargement with posterior and anterior costal cartilage grafting. The normal mucosa on the divided portions of the elliptical cricoid facilitates rapid reepithelialisation of the graft's periosteum

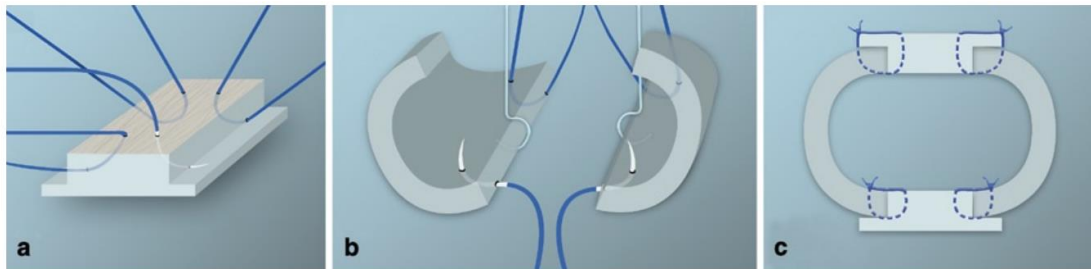
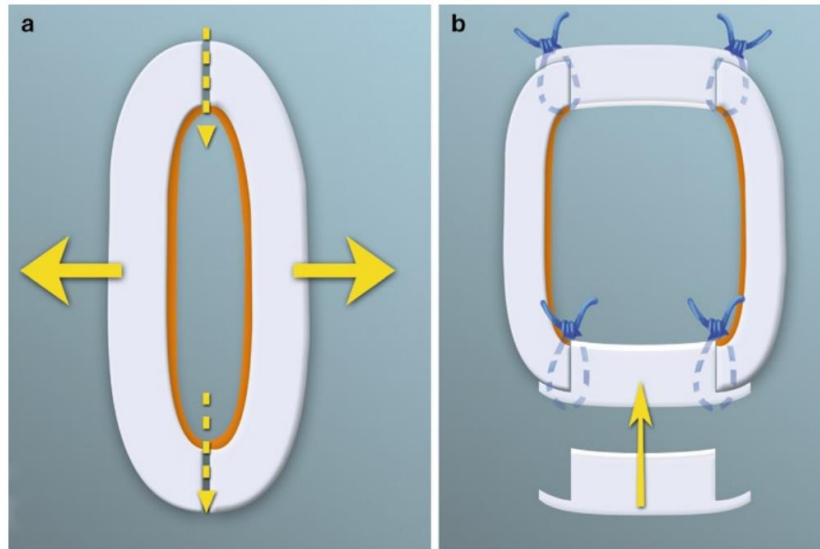


Fig. 19.7 Diagrammatic representation of suturing the posterior graft into position: (a) The needle must be inserted through the perichondrium and emerge exactly at the angle created by the lateral flanges of the cartilage. Four stitches are sufficient to stabilise the graft. (b) Correct placement of the stitches through the cricoid plate: The edge of the cricoid lamina is lifted using a skin

hook. The needle is inserted obliquely from the postero-inferior crest of the cricoid plate so as to fit exactly the height of the costal cartilage graft. (c) Final result in cross section with additional anterior graft. Note the exact placement of the stitches, which is different for anterior and posterior costal cartilage grafts

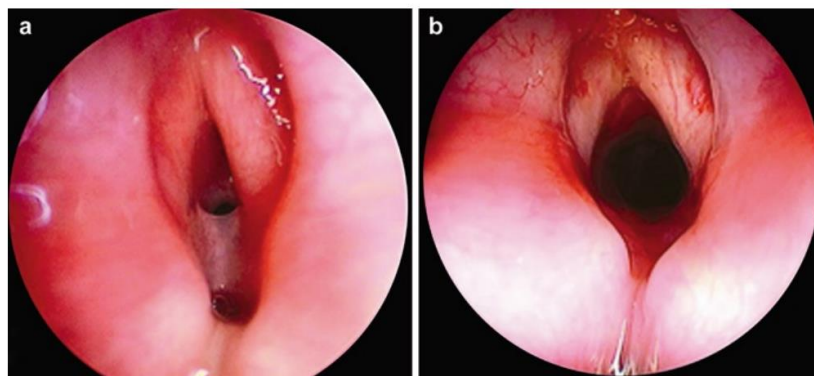


Fig. 19.13 Laryngotracheal reconstruction for Grade III subglottic stenosis combined with posterior glottic stenosis: (a) Preoperative view: the posterior glottic stenosis is thin, and the cricoarytenoid joints are passively mobile during palpation.

(b) Postoperative view at 3 months: restoration of an adequate airway following laryngotracheal reconstruction with anterior and posterior costal cartilage grafts, and 1-month stenting using an LT-Mold

- Stages of LTR:
 - **Single-stage LTR:**
 - Reconstruction the airway and resection of tracheostoma (if present) in a single surgery.
 - Indications:
 - No significant comorbidities
 - $SGS \leq$ mild grade 3
 - No glottic involvement
 - Single anterior or posterior graft
 - Close tracheostoma (if present) from SGS.
 - Advantages:
 - Facilitates healing process due to absence of tracheostoma (risk of bacterial contamination).
 - Shorten tracheostomy dependency.
 - Stenting:
 - Post-operative ET intubation should be kept in place as stenting for a duration of :
 - **7 days** with anterior graft.
 - **14 days** with posterior graft.

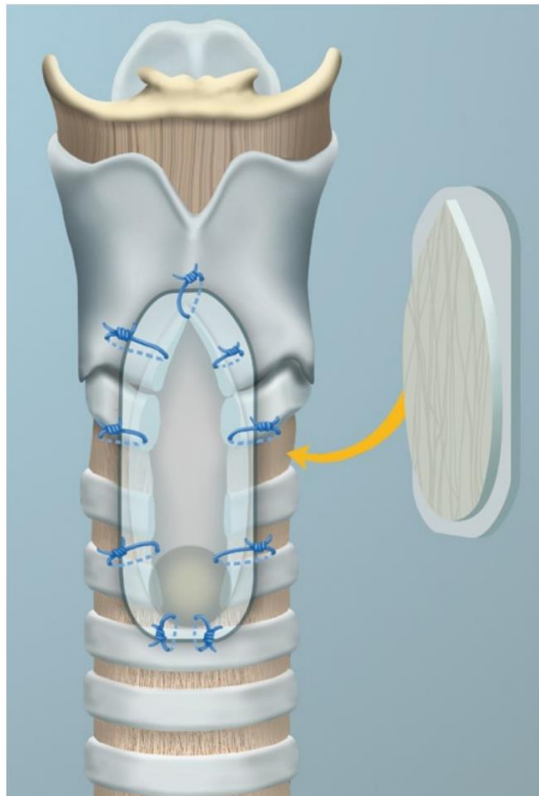


Fig. 19.14 Single-stage laryngotracheal reconstruction with anterior costal cartilage graft: the shape of the cartilage inlet is more triangular in order to accommodate the round opening of the tracheostoma. The anterior costal cartilage graft is sutured into position, including the tracheostomy site. The suturing technique is identical to that used for double-stage surgery

- **Double-stage LTR:**

- Reconstruction the airway in a single surgery while keeping the tracheostomy for future decannulation.
- Indications:
 - Significant comorbidities
 - Severe grade 3 and 4 SGS
 - Glottic involvement
 - Combined anterior and posterior grafts
 - Far tracheostoma (if present) from SGS.
- Stenting:
 - Kept for 2-6 weeks.
 - Types:
 - **T-Tube:**
 - Montgomery (without inner cannula) or Healy (with inner cannula).
 - Silicone tube.
 - Soft and pliable.
 - Commonly used in adults.
 - Disadvantages:
 - Does not conform to the complex inner contours of larynx.
 - Risk of granulation tissue formation.
 - Migration
 - Plugging in no inner cannula.



Fig. 2.18 Montgomery T-tube: Simple open silicone T-tube suitable for tracheal stenting but not for glotto-subglottic airway reconstructions



Fig. 2.20 Healy paediatric T-tube: This prosthesis comprises an inner cannula that can quickly be removed and changed in case of clogging by dried secretions. This inner cannula further diminishes the inner size of the prosthesis required for the passage of air

▪ **Aboulker Stent:**

- Cigar-shaped Teflon stent.
- Commonly used in children.
- Disadvantages:
 - Too hard
 - Does not conform to the complex inner contours of larynx.
 - Risk of granulation tissue formation.
 - Risk of infection
 - Stent migration
 - Pressure necrosis.
 - Cannot restore a sharp anterior commissure which has negative impact on voice quality.

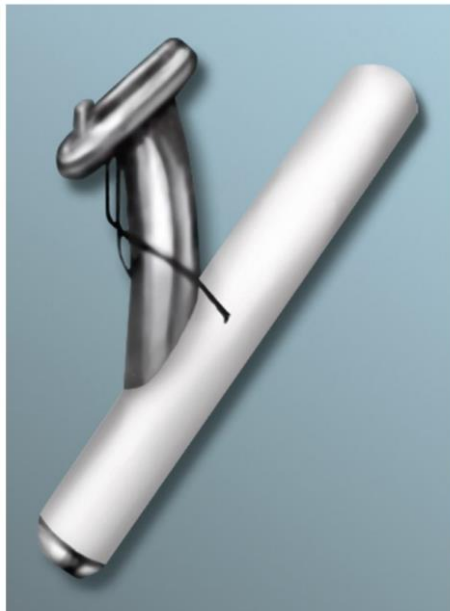


Fig. 2.17 Aboulker stent: Cigar-shaped, hard-Teflon prosthesis, unsuitable for stenting glotto-subglottic stenoses

- Currently recommend antibiotics for 1 week post-op with coverage for Staph and Pseudomonas as it shown to be correlated with granulation tissue formation.
- Patients must be evaluated for **granulation tissue** prior to stent removal with fiberoptic exam.

- Post-op care of LTR:
 - **PICU:**
 - Duration depends on child's age and the type of surgery (SS-LTR vs. DS-LTR) performed.
 - **CXR:**
 - Rule out pneumothorax after rib cartilage harvesting.
 - Rule out atelectasis from plugged bronchial secretions or blood.
 - **Medications:**
 - Antibiotics
 - PPI
 - **Frequent daily evaluation for early complications**
 - **Drain removal:**
 - After 48 h, when dressing is dry.
- Complications of LTR:
 - **Intra-op:**
 - Hypoxia
 - Pneumothorax
 - Pneumomediastinum.
 - Bleeding
 - **Early post-op:**
 - Wound infections
 - Subcutaneous emphysema
 - Haematoma
 - Graft displacement
 - Stent dislodgement and aspiration
 - **Airway complications:**
 - Accidental extubation:
 - In single-stage LTR.
 - Avoided with deep sedation (risk of narcotic withdrawal) and use of muscle relaxants (risk of losing the airway after accidental extubation).
 - Blockage of ET or tracheostomy tube and the need for replacement:
 - Avoided with proper humidification and saline instillation.
 - Granulation tissue formation:
 - Treated with systemic steroid or with topical or aerosolized steroid/antibiotic drops through tracheostomy tube.
 - **Late post-op:**
 - Re-stenosis
 - Failure of reconstruction
 - Distortion of laryngeal anatomy
 - Voice impairment
 - Aspiration

- Outcome of LTR:
 - 10-20% failure rate, mainly after:
 - Primary surgery for Grade III/IV SGS
 - PCTR has a superior outcome.
 - SGS with glottic involvement.
 - Extended PCTR has a superior outcome.
 - Factors decrease success rate:
 - Severe grade
 - Glottic involvement
 - Revision surgery
 - Double stage
 - Presence of significant comorbidities

Table 19.3 Results of the Cincinnati experience in 199 LTRs for a sole diagnosis of SGS[43]

Myer-cotton Grade	Double-stage LTR (n = 101)		Single-stage LTR (n = 98)	
	OP specific DR	Overall DR	OP specific DR	Overall DR
II	85% (18/21)	95% (20/21)	82% (37/45)	100% (45/45)
III	37% (23/61)	74% (45/61)	79% (34/43)	86% (37/43)
IV	50% (7/14)*	86% (12/14)*	67% (2/3)*	100% (3/3)*
II, III, IV	~ 50% (48/96)	~ 80% (77/96)	~ 80% (73/91)	~ 93% (85/91)
Revision surgery	48% of all cases~ 1.6 per child		18% of all cases~ 1.3 per child	

OP=Operation DR=Decannulation rate

*=Please note the small numbers

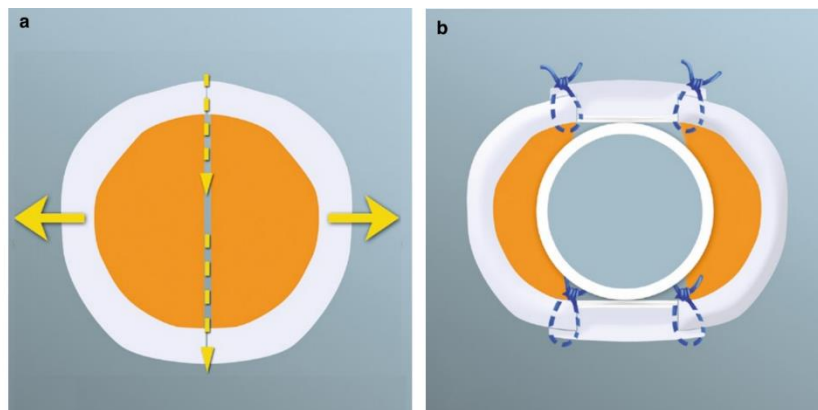


Fig. 20.1 Diagram of laryngotracheal reconstruction with anterior and posterior costal cartilage grafts for grade IV laryngotracheal stenosis: (a) Prior to surgery, the subglottic airway is fully obstructed by cicatricial tissue. (b) Diagram of airway expansion with anterior and posterior costal cartilage grafts: The reconstructed airway is fully devoid of any mucosal lining. A circumferential (up to 1.5 cm in length) airway segment must heal by secondary intention around the stent (displayed in white), which leads to difficult wound healing, granulation tissue formation, and restenosis

Box 19.3 and 19.4

Surgical Highlights for LTR

- Do not perform a full laryngofissure for anterior or posterior CCG alone
- Extend the midline thyrotracheal incision from the lower third of the thyroid cartilage down to the first two tracheal rings for a simple LTR with ACCG alone
- Avoid incising the thyroid cartilage cranially to the anterior commissure of the vocal cords (mid-distance from the thyroid notch to the inferior border of the thyroid cartilage) in order to preserve good voice quality
- Extend the vertical midline thyrotomy into a full laryngofissure for a combined ACCG and PCCG
- Carefully divide the thyroid cartilage in the midline in order to preserve an intact anterior laryngeal commissure
- Extend the posterior cricoid split through the transverse interarytenoid muscle in the case of posterior glottic scarring
- Harvest the rib cartilage with the perichondrium on its flat or slightly concave surface
- Systematically check for pneumothorax after harvesting a CCG
- Carve the PCCG in a rectangular fashion with the perichondrium facing the lumen and preserve lateral flanges of cartilage in order to wedge the graft between the two divided cricoid laminae
- Carve the ACCG in an oval shape on the perichondrial side and preserve flanges of cartilage on the opposite side in order to avoid prolapse of the ACCG into the airway
- Always place the perichondrial side of the CCG intraluminally
- Select an appropriate stent shape, diameter and length in order to splint the reconstructed airway
- For single-stage LTR, avoid full laryngofissure in order to maintain optimal laryngeal stability
- Carve and suture the ACCG into position after reintubation with a normal size ET tube, corresponding to the patient's age, in the case of SS-LTR

- **Partial Cricotracheal Resection (PCTR):**
 - Success rate reaching 90%
 - Technically challenging surgery.
 - Indications of PCTR:
 1. Surgery of choice for management of severe isolated grade 3 and 4 SGS.
 2. Failed previous LTR
 3. Tracheostomal malacia (Suprastomal collapse)
 - Relative contraindication of PCTR:
 - SGS < 3mm distance from vocal folds (Extended PCTR should be done instead).
 - Principle:
 - Resection of diseased segment of the airway with reconstruction of a normal, rounded mucosalised subglottic airway.
 - No graft is needed.
 - Risk factor for failure of PCTR:
 - Unilateral or bilateral vocal fold paralysis.
 - Steps:
 - Tracheal and laryngeal dissection up to lower edge of the cricoid cartilage without identifying the RNLs.
 - Resection of subglottic stenosis leaving the posterior cricoid plate and anterior rectangular wedge of tracheal wall intact for anastomosis.
 - Tracheoesophageal separation
 - Reshaping of the subglottic space to easily accommodate the distal tracheal stump.
 - Posterior cricotracheal and thyrotracheal and anastomosis with absorbable sutures
 - Integrity of the anastomosis is checked by pouring normal saline into surgical field, while the patient is ventilated using positive pressure to detect any leakage of air.
 - Penrose drain is placed
 - Neck is maintained in a flexed position with chin-to-chest sutures to limit the extension of the neck during the postoperative period.

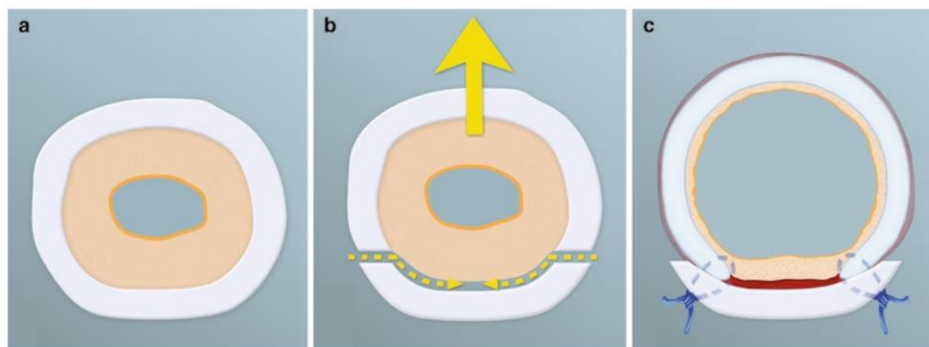


Fig. 8.5 Diagram of PCTR for flattened cricoid ring with mucosal hyperplasia and fibrosis (axial views of the subglottis): (a) Initial situation requiring entire removal of the abnormally thick subglottic mucosa. (b) During PCTR, the anterior arch of the cricoid ring is completely removed with the thick abnormal mucosa. The flattened cricoid ring, with its wide cricoid plate, represents a favourable situation for this type of surgery. (c) Reconstruction of the subglottis with a normal tracheal ring

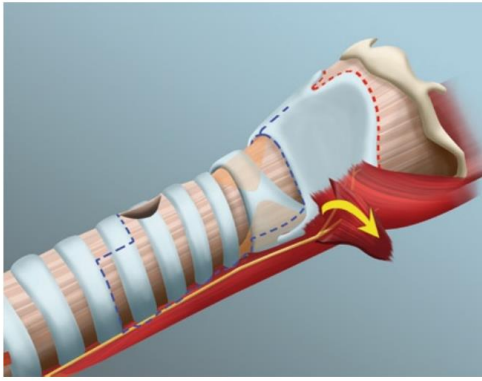


Fig. 20.6 Laryngeal dissection for partial cricotracheal resection: The sternothyroid and thyrohyoid muscles are sectioned from their thyroid insertion (not shown on the diagram). The cricothyroid muscles are reflected laterally in order to protect the recurrent laryngeal nerves (yellow arrow). The thyrohyoid membrane is incised to provide a mini-laryngeal drop (dotted red line). The resection lines (dotted blue lines) are shown for single-stage partial cricotracheal resection

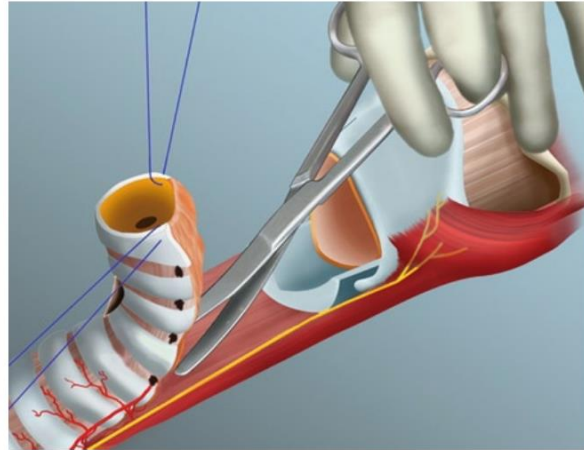


Fig. 20.7 Tracheoesophageal separation: The dissection is extended caudally to the tracheostoma level, while avoiding a compromise of the vascular supply to the distal tracheal stump

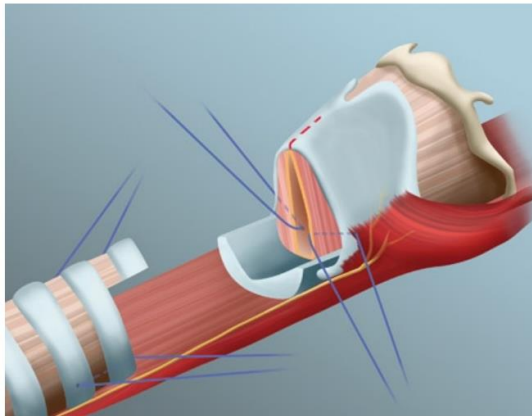


Fig. 20.8 Reshaping of the subglottic space after partial cricotracheal resection: The slit-like subglottis is enlarged by suturing the lateral subglottic mucosa to the inferior edge of the thyroid cartilage. The red dotted line shows the cranial extent of the inferior midline thyrotomy that must not extend cranially beyond the mid-distance from the thyroid notch to the inferior border of the thyroid cartilage

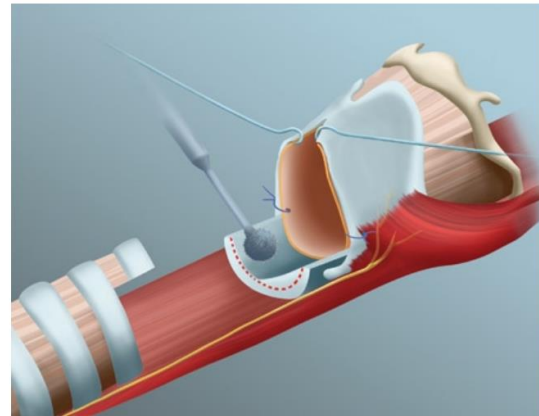


Fig. 20.10 Reshaping of the subglottic space after partial cricotracheal resection: The inferior borders of the thyroid alae are spread apart using skin hooks to enlarge the subglottic lumen without affecting voice quality. The V-shaped cricoid is flattened and widened with a diamond burr to easily accommodate the distal tracheal stump (red dotted line)

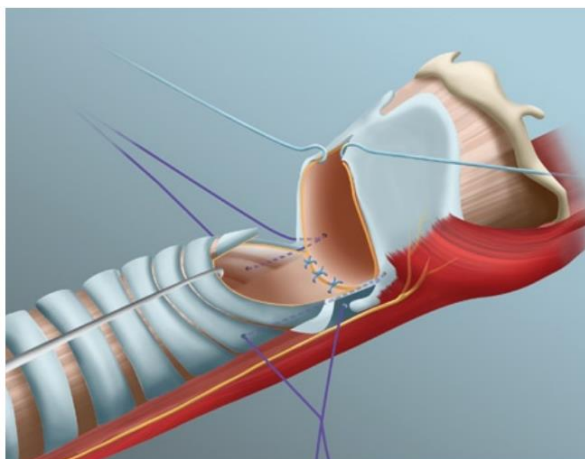


Fig. 20.14 Completion of posterior cricotracheal anastomosis: Great care should be taken to achieve perfect mucosal approximation, the only guarantee for primary healing without scar tissue formation

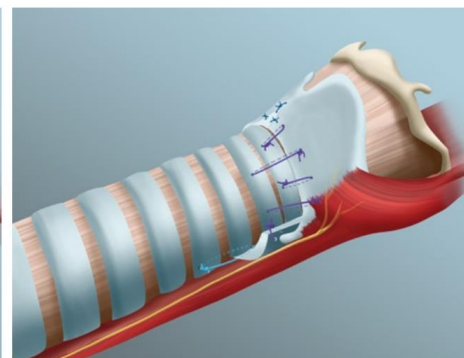


Fig. 20.15 Completion of thyrotracheal anastomosis: Note the alternate position of the stitches through the first and second tracheal rings so as to distribute the anastomotic tension onto different levels. An additional tension-releasing suture is placed between the posterolateral aspect of the cricoid plate and the trachea (displayed in turquoise). Staying in a subperichondrial plane at the cricoid level is essential to avoid injury to the recurrent laryngeal nerves. The triangular wedge of pedicled trachea is trimmed to the size of the corresponding subcommissural defect and sutured in place with two or three 5.0 vicryl sutures

	LTR	PCTR
Advantages	1. Easier technically. 2. Good result in Grade 1 and 2 or minor grade 3 SGS.	1. Higher rate of decannulation in severe grade 3 and 4 SGS 2. Glottic sparing. 3. Avoidance of donor site morbidity. 4. Minimizes wound-healing problems from costal cartilage grafts and stenting in LTR. 5. Resecting the diseased airway segment with fully mucosalized reconstructed airway.
Disadvantages	1. Less optimal results with severe grade 3 and 4 SGS. 2. Higher risk of distortion of laryngeal framework. 3. Reconstructed airway is fully devoid of any mucosal epithelium. 4. Higher risk of granulation tissue formation. 5. Higher risk of delayed wound healing. 6. Higher risk of re-stenosis.	1. Technically challenging surgery 2. Higher risk of RLN injury 3. Higher risk of anastomotic dehiscence 4. Require laryngeal release procedure and tracheal mobilization when resecting > 5 tracheal rings to achieve tension-free anastomosis.

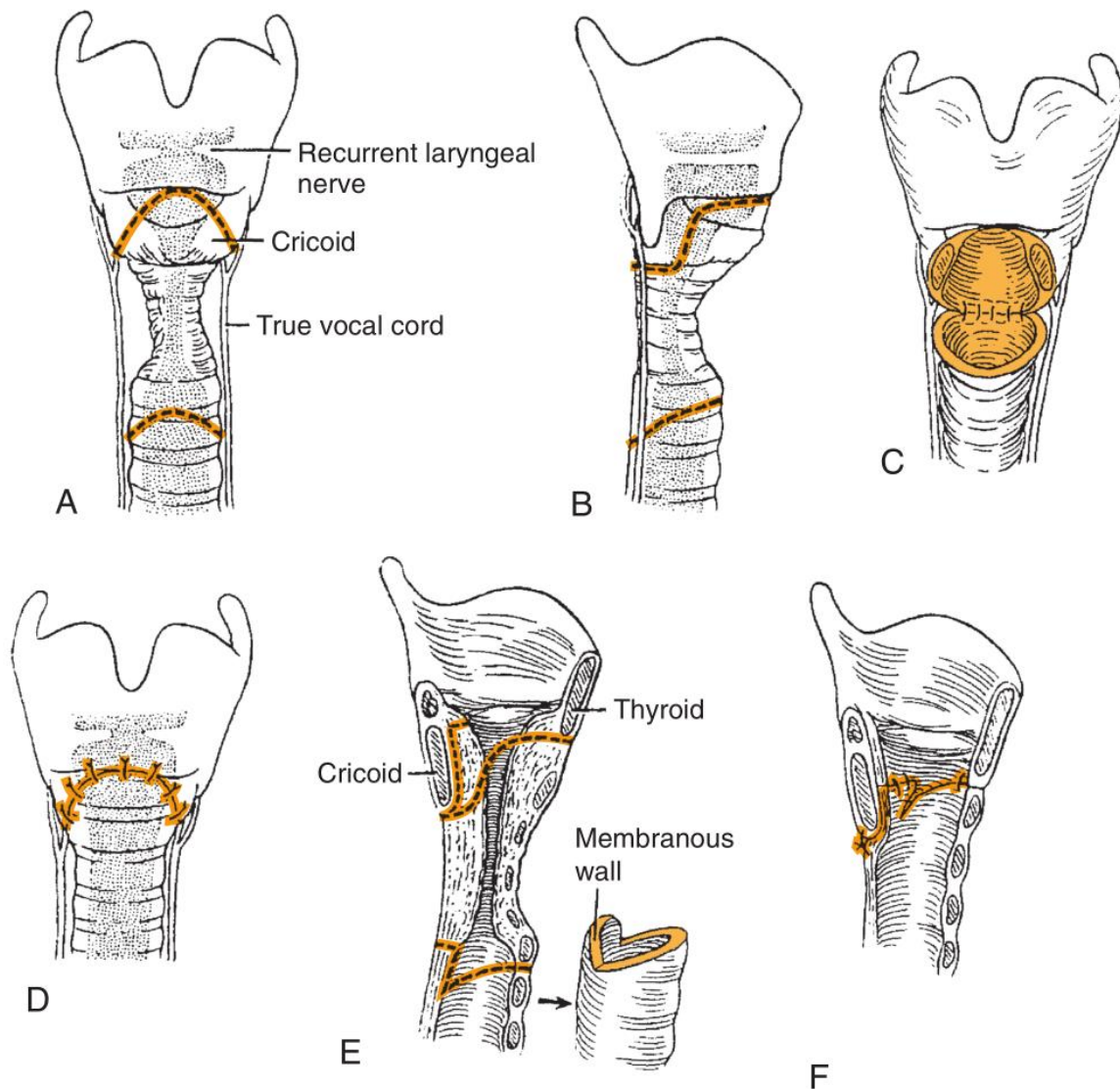


FIGURE 205-10. Partial cricoid resection with thyrotracheal anastomosis. **A**, Anterior view of the stenotic area to be resected, including the anterior cricoid lamina. **B**, Lateral view of the same area. Note the preservation of the posterior cricoid lamina and the location of the recurrent laryngeal nerve. **C**, After resection. The trachea is beveled and approximated to the subglottis. **D**, Suturing is completed (3-0 polyglactin 910). **E**, If a posterior subglottic thick scar is present, it is resected with preservation of the posterior cricoid cartilage, and a mucosal flap is developed from the posterior tracheal wall. **F**, Suturing of the mucosal flap is performed, and coverage of raw areas is achieved.

- Stages of PCTR:
 - **Single-stage PCTR:**
 - Reconstruction the airway and resection of tracheostoma (if present) in a single surgery.
 - No stenting is required.
 - Indications:
 - Grade 3 and 4 SGS:
 - No significant comorbidities
 - Intact vocal cords/glottis
 - Resection of ≤ 5 tracheal rings with the SGS
 - Complications:
 - Longer tracheal resections carry greater risks of anastomotic dehiscence.
 - **Double-stage PCTR:**
 - Reconstruction the airway in a single surgery while keeping the tracheostomy for future decannulation.
 - No stenting is required.
 - Indications:
 - Grade 3 and 4 SGS:
 - Significant comorbidities
 - Glottis involvement
 - Resection of > 5 tracheal rings with the SGS
- Tracheostomy situations:
 - **High tracheostomy:**
 - Level of 1st tracheal ring.
 - Ideal for SGS patients' requiring PCTR.
 - PCTR is done as a single-stage.
 - Tracheostoma is included in the resected SGS and tracheal rings and anastomosis is done with the distal tracheal stump.
 - **Usual location tracheostomy:**
 - Level of 3-4th tracheal ring.
 - PCTR is done as a double-stage.
 - Tracheostoma is included in the resected SGS and tracheal rings resulting in resection of > 5 tracheal rings.
 - Anastomosis is done with the distal tracheal stump.
 - New tracheostoma is placed distally.

- **Low tracheostomy with steady tracheal rings between SGS and tracheostoma:**
 - Level of > 5th tracheal ring.
 - PCTR is done as a double-stage.
 - Tracheostoma is not included in the tracheal resection.
 - Resection of SGS and tracheal rings is done and the anastomosis is performed with a steady tracheal ring.
- **Low tracheostomy with unsteady tracheal rings between SGS and tracheostoma:**
 - Level of > 5th tracheal ring.
 - PCTR is done as a double-stage.
 - Resection of SGS and tracheal rings with tracheostoma (>5 rings) is done and the anastomosis is performed with a steady tracheal ring in the distal stump.
 - Requires laryngeal release and tracheal mobilization.

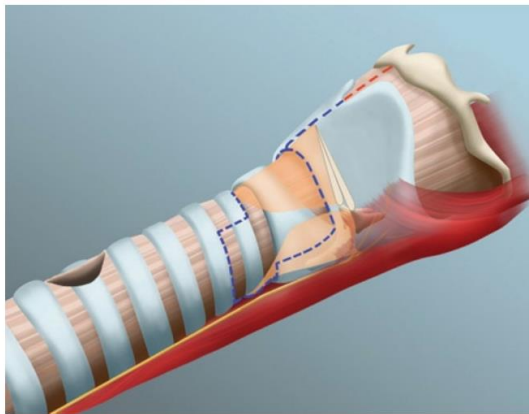


Fig. 20.17 Severe glotto-subglottic stenosis with distally placed tracheostoma: Subglottic resection is limited to the anterior cricoid ring and first or second tracheal ring laterally to keep a wedge of cartilage pedicled to the anterior tracheal wall

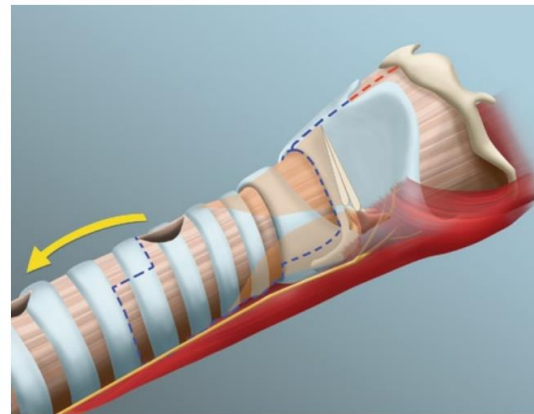
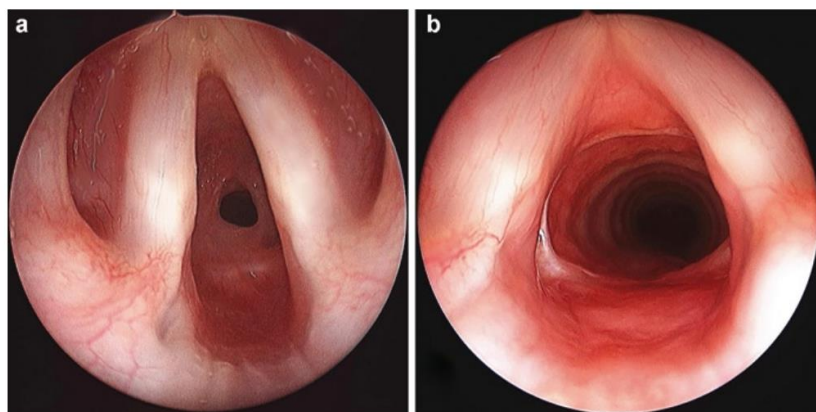


Fig. 20.18 Severe glotto-subglottic stenosis with tracheostoma placed at the third tracheal ring: The segment of residual trachea between the subglottic stenosis and the tracheostoma is too short to be used for the anastomosis. The original tracheostoma must be excised as part of the resected specimen, and a new tracheostoma must be repositioned more distally (yellow arrow)

Fig. 20.16 Single-stage partial cricotracheal resection for isolated grade III subglottic stenosis: (a) Preoperative view: The grade III subglottic stenosis is away from the normal vocal cords. (b) Postoperative view: patent subglottic airway 2 years after single-stage partial cricotracheal resection. The anastomotic line is barely visible posterolaterally under the left vocal cord



20.1 Box 20.1 Surgical Highlights for Simple PCTR

- Place a haemostat for traction on either side of the ellipse of skin left around the stoma in order to facilitate tracheal dissection and mobilisation.
- Use a retractor ring with elastic stay hooks to optimise the exposure throughout the surgical procedure.
- Do not attempt to visualise or dissect the RLNs.
- Tracheal dissection is performed by staying over the outer perichondrium of the tracheal rings.
- Carefully preserve the vascular supply to the trachea from the tracheo-oesophageal grooves, except for the segment that is to be resected.
- Coagulate before dividing the feeding vessels of the trachea using a bipolar forceps to avoid retraction of the vessels into the peri-tracheal fat pad and prevent injury to the RLNs during coagulation.
- Do not dissect the larynx and trachea using a monopolar coagulation probe.
- Avoid dissection above the posterolateral border of the cricoid plate in order to avoid injury to the RLNs.
- Dissection is safe up to the cricoid ring, short of the trachea.
- Reflect the cricothyroid muscles over the cricothyroid joints by sharp dissection off the cricoid ring from the midline so as to protect the RLNs.
- Carry out infrahyoid laryngeal release and extensive intrathoracic tracheal mobilisation if five or more tracheal rings must be resected for a single-stage PCTR.
- • Open the airway first at the lower edge of the cricoid ring in order to determine the distal extent of the SGS.
- Perform the upper resection line along the inferior edge of the thyroid cartilage and stay anterior to the cricothyroid joint laterally in order to avoid injury to the RLNs.
- Remove any cicatricial tissue from the cricoid plate and flatten it down with a diamond burr to optimise adaptation of the tracheal ring used for the anastomosis.
- Keep an anterior cartilaginous wedge pedicled to the tracheal stump used for the anastomosis, perform an inferior midline thyrotomy to enlarge the subglottic lumen, and suture the anterior pedicled wedge of the trachea into the subcommissural defect upon completion of the anastomosis.
- Meticulous surgical technique is required throughout the entire surgical procedure, particularly for the thyrotracheal anastomosis.
- Perfect mucosal approximation is the only way of preventing granulation tissue formation and subsequent restenosis at the anastomotic level.
- Use magnifying (3x) glasses when operating on infants and small children.

- **Extended Partial Cricotracheal Resection (Extended PCTR):**
 - Technically challenging surgery.
 - Indications of Extended PCTR:
 1. Surgery of choice for management of severe grade 3 and 4 SGS with glottic involvement.
 2. Distorted laryngeal framework due to previous failed LTRs.
 - Principle:
 - Resection of diseased segment of the airway in addition to posterior cricoid split and posterior costal cartilage graft.
 - Need stenting for 4–6 weeks.
 - Performed only as a double-stage procedure and a tracheostomy must be left in situ until the airway is fully healed and stable.

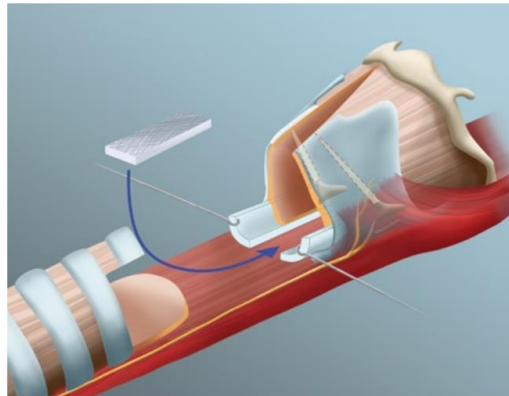


Fig. 20.20 Posterior cricoid split: Through the anterior laryngofissure, the cricoid plate is divided exactly in the midline. This transection is fully extended through the posterior commissure scarring and interarytenoid muscle. A blunt haemostat is used to spread apart the divided portions of the cricoid plate, and a costal cartilage is harvested

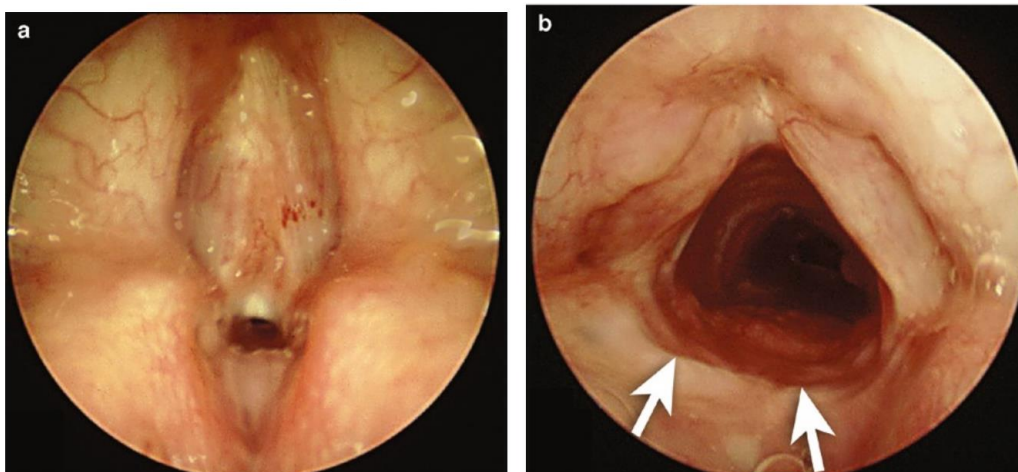


Fig. 20.26 Extended partial cricotracheal resection for glotto-subglottic stenosis with cicatricial fusion of the vocal cords: (a) Preoperative view: acquired on congenital glotto-subglottic stenosis with fusion of the vocal cords and pinhole residual posterior opening. (b) Postoperative view: patent glotto-subglottic airway, albeit with an overexpanded interarytenoid space. The posterior mucosal flap was sutured above the glottic level (white arrows)

Box 20.2 Surgical Highlights for Extended PCTR

- Keep an ellipse of skin around the initial tracheostoma so as to facilitate tracheal dissection and mobilisation.
- Create a new tracheostoma at the end of the surgery.
- Perform a full laryngofissure under visual control in order to preserve the integrity of the vocal cords and anterior laryngeal commissure, in case of a tight posterior glottic stenosis or vocal cord fusion combined with a subglottic stenosis.
- Expand the posterior cricoid plate using the same technique as for LTR with PCCG.
- On the tracheal stump, create a pedicled flap of membranous trachea in order to resurface the posterior subglottis, and use a cartilage wedge anteriorly so as to increase the size of the reconstructed subglottic lumen.
- Select an appropriately sized LT-Mold and recreate a perfect anterior laryngeal commissure by proper realignment of the vocal cords during closure of the supraglottic portion of the laryngofissure.
- Reposition the tracheostoma more distally, if deemed appropriate.

- Post-op care of PCTR and Extended PCTR:
 - **PICU:**
 - In SS-PCTR, patient is kept intubated in PICU.
 - Neck is kept in flexed position to avoid tension over the anastomosis.
 - Routine chest physiotherapy.
 - **CXR:**
 - Rule out pneumothorax after rib cartilage harvesting in Extended PCTR.
 - Rule out atelectasis and bronchopneumonia.
 - **Medications:**
 - Antibiotics
 - PPI
 - **Frequent daily evaluation for early complications**
 - **Drain removal:**
 - After 48 h, when dressing is dry.

- Complications of PCTR and Extended PCRTR:
 - **Intra-op:**
 - Hypoxia
 - Pneumothorax
 - Pneumomediastinum.
 - Bleeding
 - **Early post-op:**
 - Wound infections
 - Subcutaneous emphysema
 - Haematoma
 - Bronchopneumonia
 - Graft displacement (In Extended PCTR)
 - Anastomotic dehiscence (5.6%).
 - RLNs injury.
 - If extubation is not tolerated despite a patent subglottic airway, then a distally placed tracheostomy is required.
 - **Late post-op:**
 - Re-stenosis:
 - Endoscopic approach can apply to improve the mild stenosis post PCRTR.
 - Topical applications Mitomycin C should not be done within 6 weeks following PCTR due to risk of anastomotic dehiscence from reducing scar formation.

Fig. 20.39 CO₂ laser resection of a left subglottic stenosis 3 months after single-stage partial cricotracheal resection: (a) Significant cicatricial tissue at the anastomotic level below the left vocal cord. (b) Status after endoscopic resection using the CO₂ laser

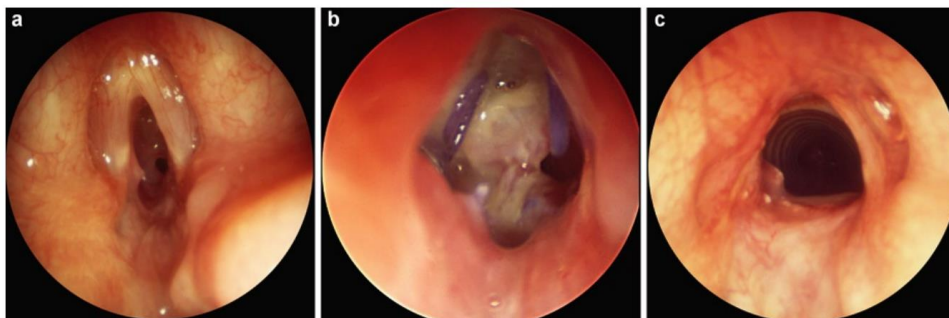
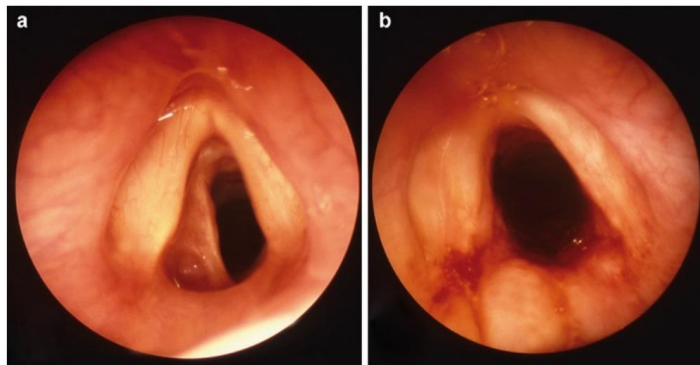


Fig. 20.42 Anastomotic dehiscence 3 weeks after single-stage partial cricotracheal resection for grade III subglottic stenosis: (a) Preoperative view: grade III subglottic stenosis. (b) Anastomotic dehiscence 3 weeks after single-stage partial cricotracheal resection due to respiratory syncytial virus infection with severe coughing. (c) Final outcome 3 weeks after revision surgery: patent glotto-subglottic airway, albeit with thinned vocal cords

Table 20.5 Results of PCTR for severe Grade III and IV LTS

Patients' characteristics	Cincinnati [69] <i>n</i> = 93	Lausanne [22] <i>n</i> = 100
Stenosis Grade		
II	5%	4%
III	60%	64%
IV	35%	32%
Glottic involvement	23%	33%
Comorbidities	NR	45%
Tracheostomy at surgery	85%	82%
Primary PCTR	46%	62%
Salvage PCTR	59%	38%
Extended PCTR	27%	23%
Revision surgery	29%	14%
Results		
Operation-specific success rate	71%	76%
Overall success rate	94%	90%
Anastomotic dehiscence	2%	4%
RNL injury	2%	0%

NR = not reported

Laryngeal Web/Atresia:

- Classification:

1. Congenital Web/Atresia:

- Uncommon congenital anomalies (5%).
- Resulting from **failure of recanalization** of the primitive larynx during 10th week of gestation.
- Since recanalization of larynx proceeds from caudal to cranial, the web's extent and its cartilaginous SG component are directly related to the time at which this recanalization process ceased.
- Pathophysiology:
 - **Laryngeal Atresia:**
 - Results due to failure of recanalization at an early stage of embryogenesis.
 - Incompatible with life unless associated with an esophageal atresia and tracheoesophageal fistula (TOF).
 - **Laryngeal Web:**
 - Results due to failure of recanalization at a late stage of embryogenesis.
 - Results in a canalized posterior glottic airway with anterior glottic web.
 - Laryngeal webs and atresia share the same pathogenesis as C-SGS which explains why most severe types of glottic webs are associated with a cartilaginous SGS.
 - Types:
 - **Supraglottic (2%)**
 - **Glottic (75%):**
 - Mainly anteriorly based.
 - Mainly membranous.
 - **Subglottic (7%)**

2. Acquired Web/Atresia:

- More common than congenital type.
- Causes:
 - **Iatrogenic:**
 - Most common cause.
 - Traumatic intubation.
 - Intra-laryngeal surgery.
 - **Inflammatory process** (TB , Diphtheria)
 - **Trauma**
 - **Reflux**

TABLE 202-4. Glottic Web Classification by Cohen			
Severity	Extent of Glottic Obstruction	Subglottic Involvement	Symptoms
Type 1	<35%	None or little	Mild hoarseness
Type 2	35%-50%	Thin anterior web with little subglottic extension	Hoarse, weak cry, stridor with exertion
Type 3	50%-75%	Thin-thick web, extends to lower border of cricoid	Severe hoarseness, moderate airway obstruction
Type 4	75%-90%	Thick web, extends to lower cricoid	Aphonic, severe airway obstruction, requires tracheotomy

From Cohen SR, Congenital glottic webs in children. A retrospective review of 51 patients. *Ann Otol Rhinol Laryngol Suppl* 1985;121:2-16.

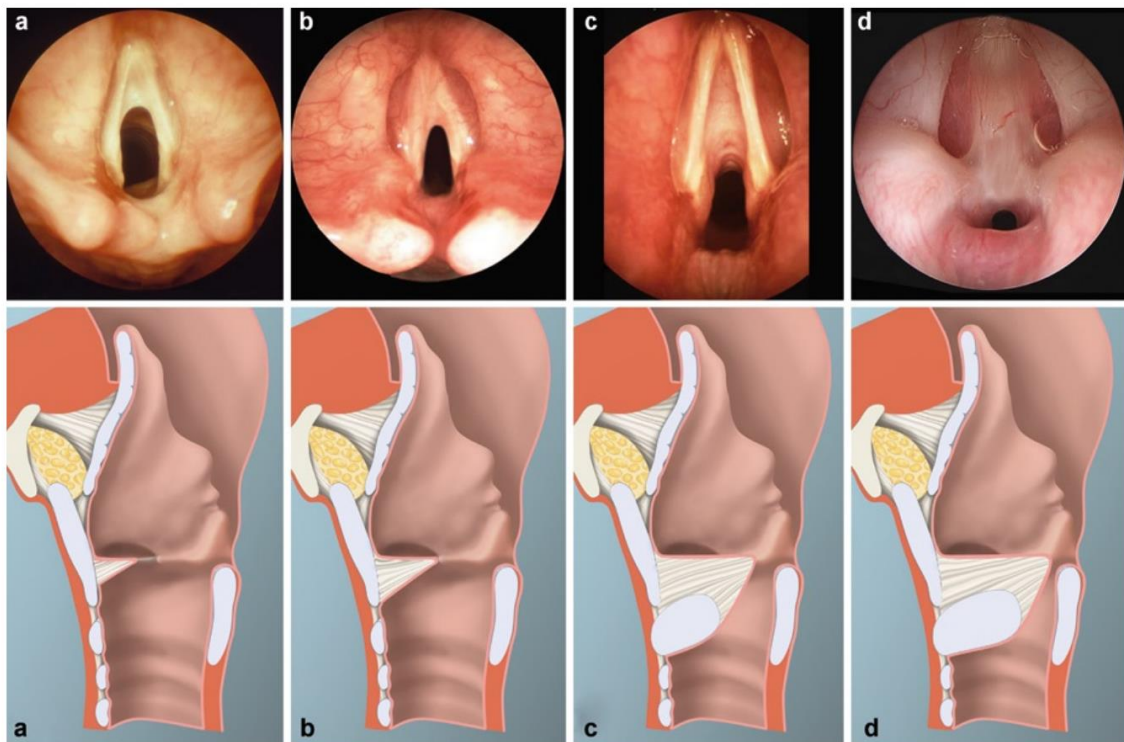


Fig. 9.1 Cohen's classification of glottic webs: endoscopic views and sagittal diagrams: (a) Type I: <35% of glottic length, no subglottic extension. (b) Type II: 35–50% of glottic length, minimal subglottic extension. (c) Type III: 50–75% of glottic

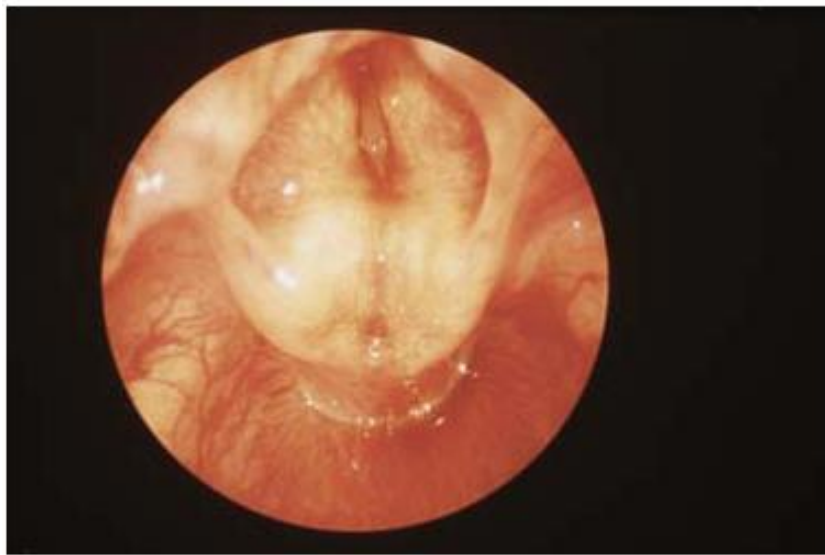
length, significant anterior subglottic extension, possibly with cartilaginous component. (d) Type IV: 75–90% of glottic length, severe subglottic extension with cartilaginous component

- Association with congenital laryngeal webs:
 - **Velo-cardio-facial/ DiGeorge syndrome:**
 - **Deletion of chromosome 22q11**
 - Found in 65% of patients with congenital laryngeal webs.
 - CATCH-22:
 - Cardiac abnormality (Tetralogy of Fallot)
 - Abnormal facies
 - Thymic aplasia
 - Cleft palate
 - Hypocalcemia/Hypoparathyroidism

- Clinical picture:

○ **Congenital laryngeal atresia:**

- Present immediately after delivery.
- Incompatible with life unless associated with an esophageal atresia and tracheoesophageal fistula (TOF).
- Can be diagnosed prenatally by Fetal MRI.
- Delivery by ex utero intrapartum treatment (EXIT procedure) is necessary to secure the airway by immediate tracheotomy, while the neonate remains on placental support to avoid death from airway obstruction.
- Following successful tracheostomy at birth, reconstructive surgery with eventual decannulation is sometimes possible.



○ **Laryngeal web:**

- Dysphonia:
 - All patients present some degree of dysphonia.
 - Ranging from mild dysphonic cry to aphonia.
- Stridor:
 - Inspiratory or biphasic stridor.
 - Airway symptoms increase with the extension of the web.
 - Most severe cases warranting a tracheotomy to secure the airway.
- Feeding difficulty:
 - Due to respiratory limitation.

- Diagnosis:
 - **Flexible laryngoscopy:**
 - Detect a web if a good view of anterior commissure is achieved.
 - **Rigid Microlaryngoscopy and bronchoscopy:**
 - Gold standard.
 - Allows for definitive diagnosis of laryngeal web.
 - Evaluate:
 - Nature of the web
 - Location of the web
 - Extension of the web
 - Arytenoids mobility
 - Detection for any concomitant airway pathology
 - **Genetic screening and cardiovascular evaluation:**
 - Should be done for all patients with congenital laryngeal web to rule out associated chromosome 22 deletion.
- Treatment:
 - **Observation:**
 - Indication:
 - Infants with anterior membranous web (Type I-II) and voice problems only.
 - Anti-reflux medications.
 - Endoscopic repair can be delayed until pre-school age.
 - Surgical correction is easier as the larynx is larger.
 - Silicone keel is not well tolerated in an infant larynx without a tracheotomy.
 - **Endoscopic division of the web:**
 - Indication:
 - For more symptomatic webs (Type I-II and membranous type III).
 - Timing of repair is based on respiratory symptoms.
 - Methods of division:
 - Cold instruments
 - CO2 laser
 - Methods to reduce the recurrence:
 - Mitomycin
 - Mucosal flap rotation
 - Silastic keel (removed after 4 weeks)

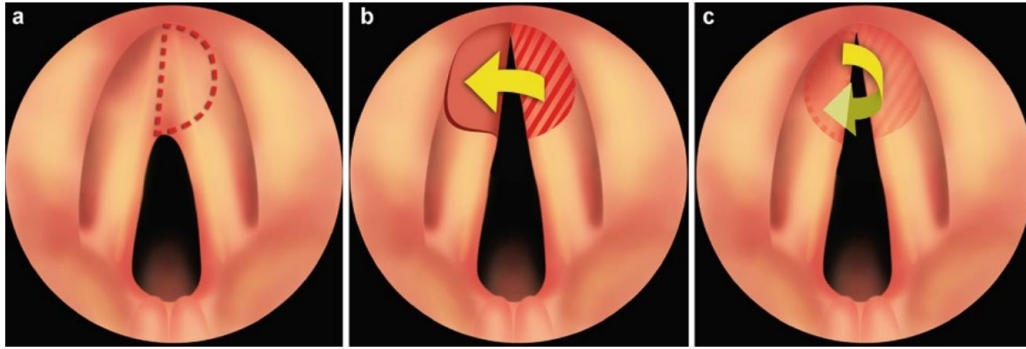


Fig. 9.2 Diagram of endoscopic treatment for type I membranous web of the vocal cords: (a) Flap design. (b) Elevation of flap and web section. (c) Reflexion of mucosal flap on the homolateral vocal cord

Fig. 9.3 Endoscopic treatment of Type II–III vocal cord webs: (a) CO₂ laser incision (ultrapulse mode). (b) Design of the keel

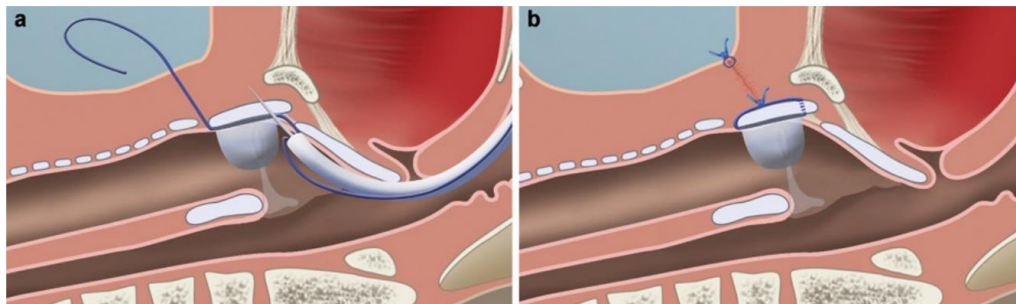
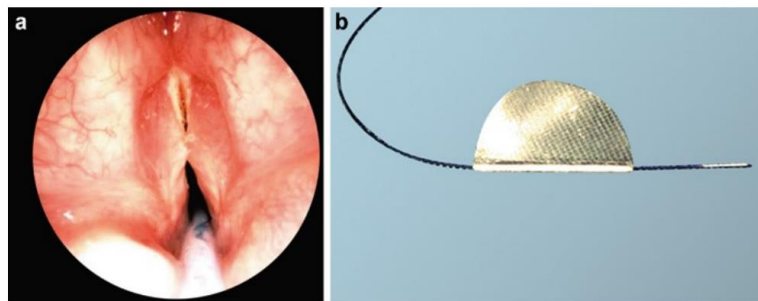
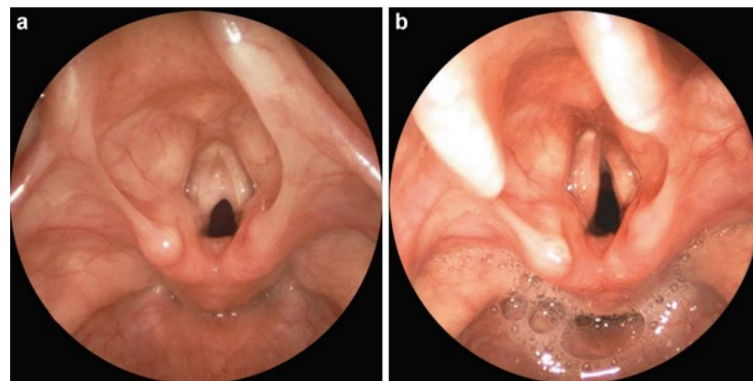


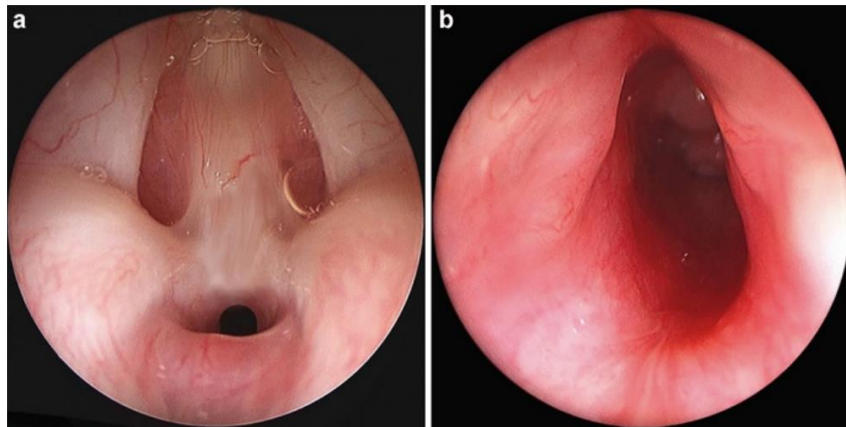
Fig. 9.4 Diagram of endoscopic placement of the silicone keel: (a) A first inside-out stitch using the Lichtenberger needle-carrier is made in the subglottic region. (b) A second inside-out stitch is made above the anterior laryngeal commissure, and the knots are tied under the skin to hold the silicone keel in place at the anterior laryngeal commissure

Fig. 9.5 Treatment result of a type II web after endoscopic splinting with a keel: (a) Preoperative view: Type II web. (b) Postoperative view: sharp anterior laryngeal commissure



- **Open Repair:**
 - Indications:
 - Cartilaginous type III and IV laryngeal web.
 - If tracheotomy is necessary to secure the airway open surgical repair can be performed early, during the first 2 years of life.
 - Methods:
 - LTR with anterior graft (For Type III)
 - LTR with anterior and posterior graft (For Type IV)
 - Extended PCTR (For Type IV)

Fig. 9.6 A 5-month-old child presenting with velo-cardio-facial syndrome: (a) Cohen's Type IV webbing of the vocal cords with Grade III congenital cartilaginous subglottic stenosis. (b) Result at 3 months after extended partial cricotracheal resection and 2 weeks after LT-Mold removal: patent glotto-subglottic airway with sharp anterior commissure



Posterior glottic web/ Posterior glottic Stenosis (PGS):

- Presence of inter-arytenoid web or fibrosis will cause limited vocal fold abduction which may mimic bilateral vocal folds paralysis.
- Types:
 - **Congenital:**
 - Rare (1-4% of all webs)
 - Often necessitates a tracheotomy in early years of life.
 - **Acquired:**
 - More common.
 - Secondary to previous intubation trauma.
- Diagnosis:
 - **Rigid Microlaryngoscopy and bronchoscopy:**
 - Gold standard.
 - Differentiate PGS from BVFP by:
 - Limited mobility during arytenoids palpation
 - Presence of inter-arytenoid fibrosis

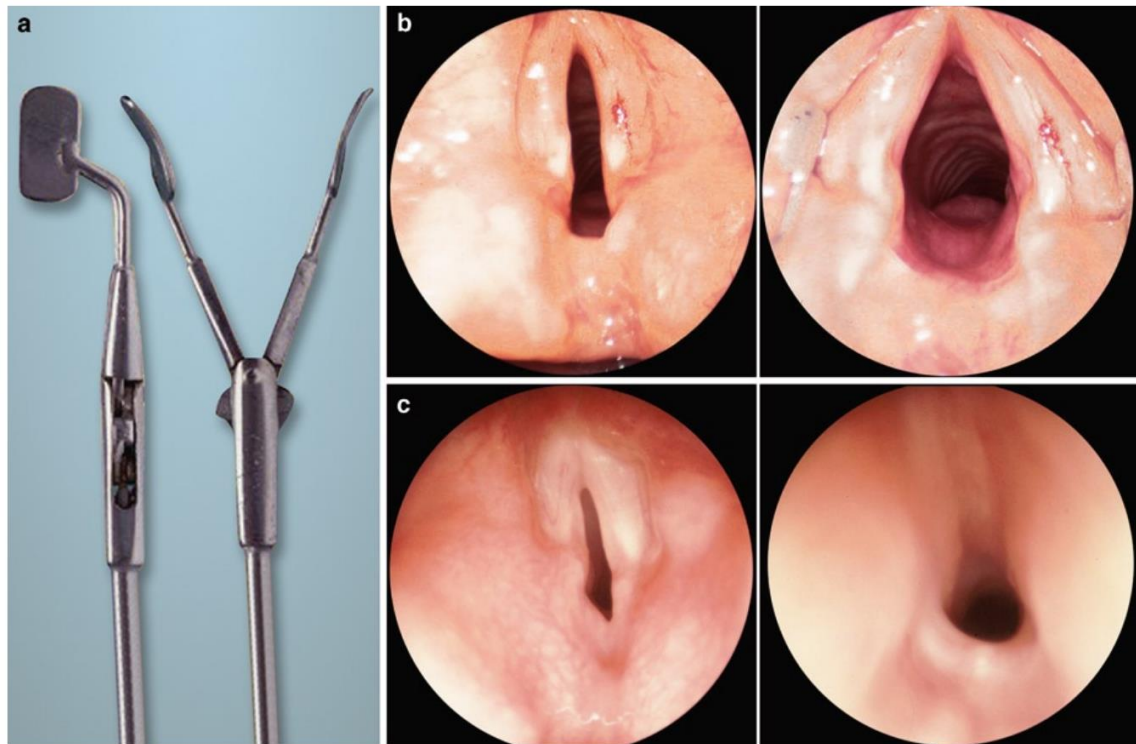


Fig. 5.9 Contribution of the Lindholm false vocal cord retractor in bilateral immobility of the vocal cords: **(a)** The Lindholm self-retaining false vocal cord retractor. **(b)** Bilateral vocal cord paralysis: the paramedian position of both vocal cords (*left*) is easily spread apart with the false vocal cord retractor (*right*). **(c)** Scarring of the posterior commissure: the paramedian position of both vocal cords (*left*) is not improved by the false vocal cord retractor, but a band of scar tissue is conspicuous (*right*)

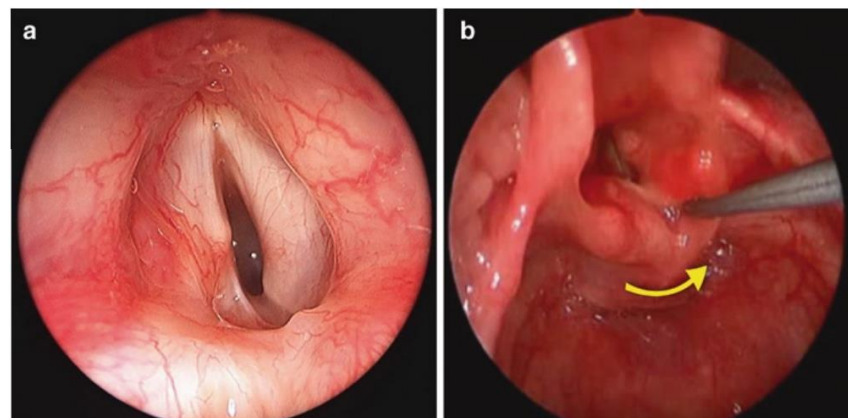
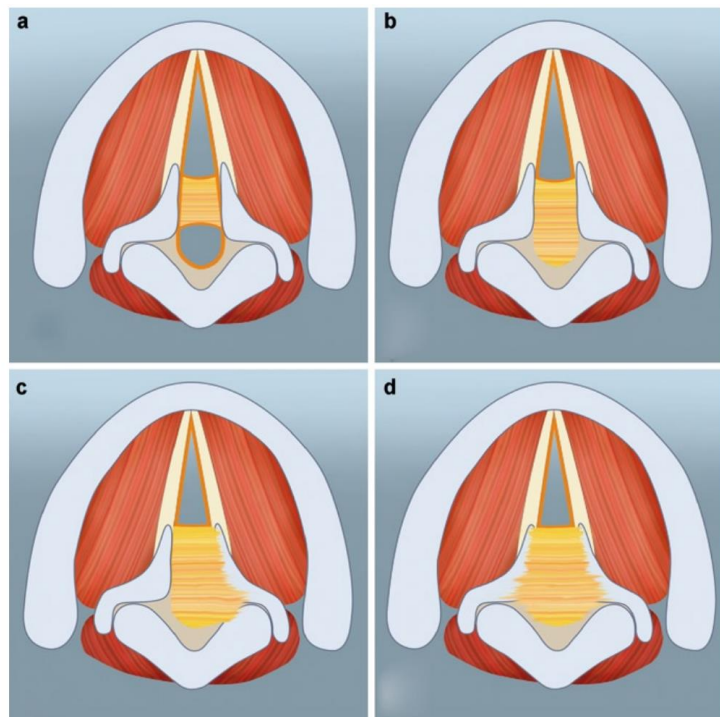


Fig. 5.10 Posterior glottic stenosis without cricoarytenoid joint fixation: **(a)** Endoscopic aspect of a posterior glottic stenosis. **(b)** Mobilisation of the right arytenoid to the right attracts the left arytenoid towards the right side. The reverse phenomenon is observed on the opposite side

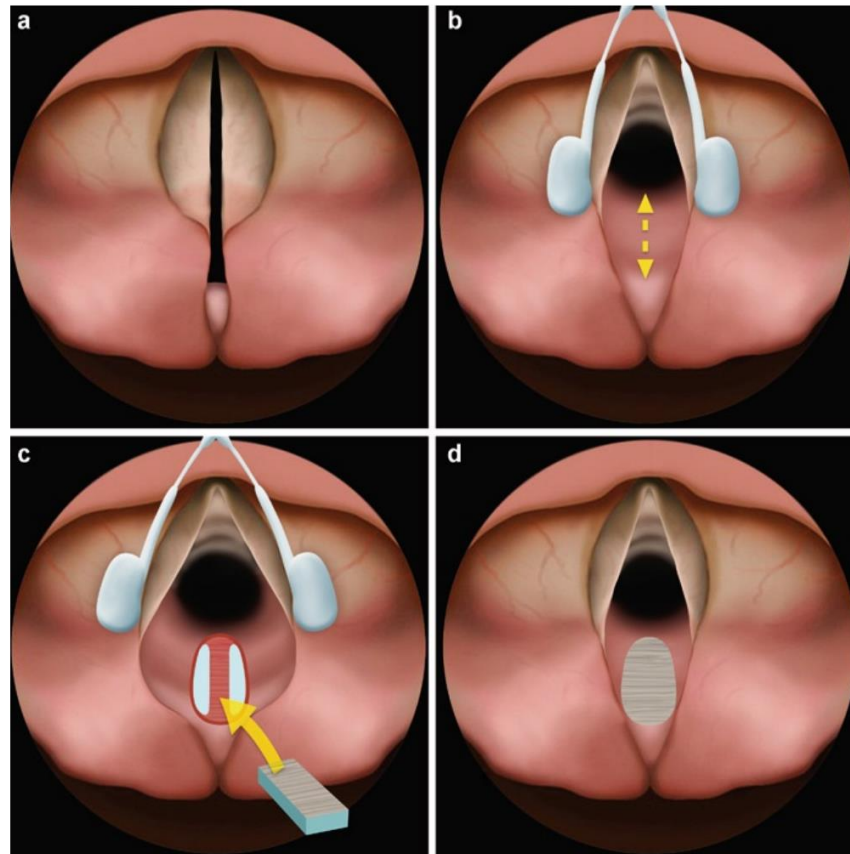
- Bogdasarian's classification of PGS:
 - **Type I: Interarytenoid Adhesion:**
 - Band of scar tissue tethering the vocal cords in midline.
 - Small residual posterior opening with intact interarytenoid mucosa.
 - **Type II: Inter-arytenoid and Posterior commissure scarring:**
 - Fibrous tissue fills the posterior glottis without any residual opening.
 - There is no cricoarytenoid joint fixation.
 - Lateral mobilization of a single arytenoid pulls the contralateral arytenoid towards the same side.
 - **Type III: Posterior commissure scarring with unilateral cricoarytenoid joint fixation:**
 - Fibrous tissue fills the posterior glottis without any residual opening.
 - Mobilization of the fixed arytenoid is impossible.
 - Contralateral arytenoid can be moved slightly laterally.
 - **Type IV: Posterior commissure scarring with bilateral cricoarytenoid joint fixation:**
 - Fibrous tissue fills the posterior glottis without any residual opening.
 - Mobilization of the both arytenoid is impossible.
 - False cord retractor cannot spread apart the vocal cords.

Fig. 5.11 Bogdasarian's classification of posterior glottic stenosis [3]:
(a) Interarytenoid adhesion: residual normal mucosal bridge between the arytenoids. **(b)** Interarytenoid and posterior commissure scarring: no residual normal interarytenoid mucosa.
(c) Scarring of posterior commissure with unilateral cricoarytenoid joint fixation. **(d)** Scarring of posterior commissure with bilateral cricoarytenoid joint fixation



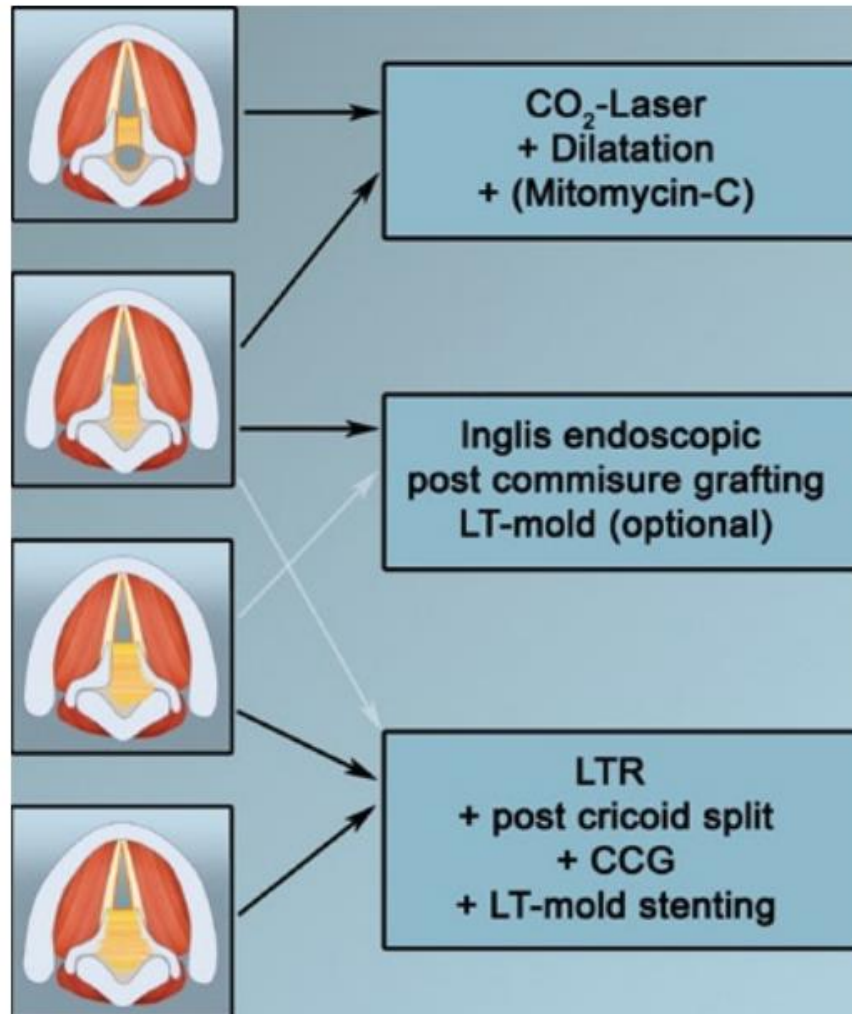
- Treatment of isolated PGS:
 - **Endoscopic division with CO2 laser:**
 - Indications:
 - Type I (interarytenoid adhesion)
 - Type II (PGS with preserved active mobility of both arytenoids).
 - Followed by dilatation +/- mitomycin application.
 - **Endoscopic posterior cricoid split and cartilage graft**
(Inglis procedure):
 - Indications:
 - Failed Co2 laser for Type II (PGS with preserved active mobility of both arytenoids).
 - Minor Type III (PGS with incomplete unilateral cricoarytenoid joint fixation).
 - Bilateral VFP.
 - Contraindications:
 - Adults (Calcified cartilages).
 - Type IV (PGS with bilateral cricoarytenoid joint fixation).
 - Subglottic involvement.

Fig. 7.8 Endoscopic posterior cricoid split with costal cartilage grafting (Inglis technique [30]): (a) Bilateral vocal cord paralysis. (b) Exposure of the posterior glottis with Lindholm false cord retractor. The *arrow* shows the extent of the cricoid split. (c) Complete posterior midline cricoid split with the CO₂ laser and costal cartilage ready to be placed between the two cricoid laminae. (d) End result: the interarytenoid distance is improved as compared to the initial condition



- **Open surgery:**
 - Indications:
 - Type III and IV PGS.
 - Method:
 - LTR with posterior cricoid split.

Table 17.8 Treatment algorithm for PGS



Laryngeal Cleft:

- Very uncommon.
- Accounts for only 0.5–1.5% of all congenital laryngeal anomalies.
- A slight male predominance.
- Majority of cases are sporadic, with only few families presenting multiple cases.
- Pathophysiology:
 - Failure of complete formation of trachea-esophageal septum.
 - Minor cases are frequently overlooked and undiagnosed.
 - Laryngeal cleft may be limited to the interarytenoid region or involve the cricoid lamina.
 - When the cleft is more extensive and reaches the cervical or intrathoracic trachea, it is termed a laryngo-tracheo-esophageal cleft (LTOC).
- Associated Anomalies:
 - Tracheobroncho malacia
 - Tracheo-esophageal fistulae (30%)
- Associated Syndromes:
 - **VACTERL Association:**
 - **V**ertebral anomalies
 - **A**nal Atresia
 - **C**ardiac anomalies
 - **T**racheo-esophageal fistula
 - **E**ar anomalies
 - **R**enal anomalies
 - **L**imb anomalies
 - **CHARGE Association:**
 - **C**oloboma
 - **H**ear disease
 - **C**hoanal **A**tresia
 - **G**rowth and mental **R**etardation
 - **G**enital anomalies
 - **E**ar anomalies
 - **G (Opitz-Frias) syndrome:**
 - Includes other defects of the median line (Cleft lip and palate, Hypospadias, Abnormal implantation of ears, Hypertelorism).
 - **Pallister–Hall syndrome:**
 - CNS, Cardiac , Pulmonary and Renal anomalies
 - Distal extremity anomalies (syndactyly and postaxial polydactyly).
 - Imperforate anus
 - **Down syndrome:**
 - Trisomy 21.

- Classification:

TABLE 202-5. Laryngeal and Laryngotracheoesophageal Cleft Classification					
Cleft Location	Peterson	Armitage	Evans	Benjamin-Inglis	Myer-Cotton
Laryngeal					
Interarytenoid	Type I	Type IA	Type I	Type I	LI
Partial cricoid	Type I	Type IB	Type II	Type II	LII
Complete cricoid	Type I	Type IC	Type II	Type II	LIII
Laryngotracheoesophageal					
Into cervical trachea	Type II	Type II	Type II	Type III	LTEI
To intrathoracic trachea/ carina	Type III	Type III	Type III	Type IV	LTEII

The Benjamin-Inglis and Myer-Cotton classification systems are the most useful.

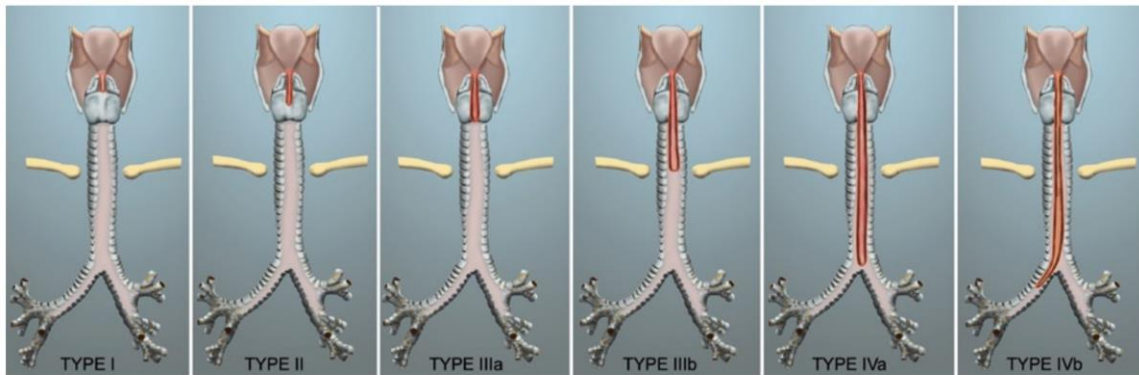
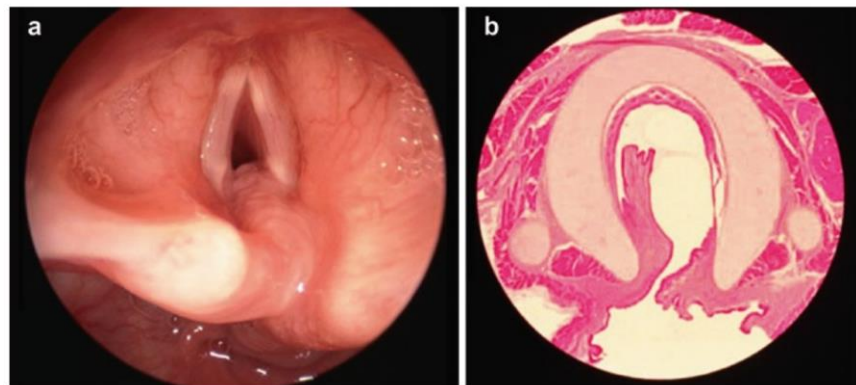


Fig. 12.1 Modified Benjamin-Inglis classification of laryngotracheo-oesophageal clefts (LTOC): The subdivision of type III into type IIIa and IIIb allows the surgeon to better define the limits of endoscopic repair: Type 0: submucosal cleft, Type I: interarytenoid cleft, Type II: partial cricoid cleft, Type IIIa: total cricoid cleft, Type IIIb: LTOC extending into the extrathoracic portion of the trachea, Type IVa: LTOC extending to the carina Type IVb: LTOC extending into one main-stem bronchus

- **Modified Benjamin-Inglis classification** (better assess the limits of endoscopic repair):
 - **Type I:**
 - Interarytenoid cleft
 - **Type II:**
 - Partial cricoid cleft
 - **Type IIIa:**
 - Complete cricoid cleft
 - **Type IIIb:**
 - LTOC extending into the cervical trachea.
 - **Type IVa:**
 - LTOC extending to the thoracic trachea/carina.
 - **Type IVb:**
 - LTOC extending into one main-stem bronchus.

- Clinical picture:
 - **Should be suspected in newborn with feeding problems and recurrent aspiration.**
 - Severity of symptoms is directly related to extent of the cleft.
 - **Asymptomatic:**
 - In mild Type I clefts.
 - **Choking and coughing during feeding:**
 - Only with thin liquids in type I clefts.
 - **Recurrent pneumonia.**
 - **Stridor:**
 - Mainly inspiratory stridor secondary to collapses of the redundant mucosa into the laryngotracheal lumen.
 - Expiratory stridor could be secondary to associated tracheomalacia

Fig. 12.2 Endoscopic view of a type III LTOC with redundant posterior mucosa herniated into the laryngeal lumen: (a) Endoscopic view (b) Corresponding histology (Reproduced from Holinger [5]. With permission)



- Diagnosis:
 - **Swallowing assessment:**
 - Modified Barium Swallow (MBS):
 - Done with water soluble contrast (Gastrografin).
 - Shows spillover into the larynx.
 - Not pathognomonic for laryngeal cleft.
 - Fiberoptic Endoscopic Evaluation of Swallowing (FEES)
 - **Flexible laryngoscopy:**
 - Done to assess vocal cord function and to rule out laryngomalacia.
 - **Rigid Laryngo-bronchoscopy and Esophagoscopy:**
 - Gold standard.
 - Done by spreading the interarytenoid mucosa laterally and probing the posterior laryngeal commissure with right-angled probe.
 - Evaluate:
 - Extent of the cleft.
 - Associated anomalies (tracheoesophageal fistula).

- Treatment:
 - **Conservative:**
 - Indication:
 - Type I laryngeal cleft (aspiration will resolve as they grow older).
 - Includes:
 - Thickening of liquids
 - Upright positioning for drinking.
 - NGT or gastrostomy feeding.
 - Anti-reflux medications.
 - Early treatment of aspiration pneumonia.
 - **Endoscopic Repair:**
 - Indication:
 - Type I and Type II.
 - Type III (a/b) with no severe concomitant congenital anomalies.
 - Pre-op airway control:
 - Non-invasive ventilation with CPAP or BiPAP:
 - Preferred treatment.
 - ET intubation may induce inflammatory reaction of the laryngotracheal mucosa and increase the risk of wound breakdown after surgical repair.
 - Method:
 - Under GA with spontaneous respiration.
 - Sharp incision of cleft mucosa with CO2 laser to separate the tracheal and pharyngo-esophageal mucosal layers.
 - Closure of the cleft in two layers from distal to proximal direction:
 - Laryngo-tracheal closure with the knots tied on the pharyngo-oesophageal side.
 - Pharyngo-esophageal closure with the knots tied on the mucosal surface of the pharyngo-esophageal repair.
 - Post-op care:
 - No need for temporary intubation or tracheotomy.
 - ICU observation.
 - In Type I–II clefts, infant resumes normal feeding the following day.
 - In Type III clefts, small soft NGT is inserted for feeding.
 - Antibiotics and steroids.
 - Complications:
 - Residual fistula resulting from partial breakdown of the suture line.

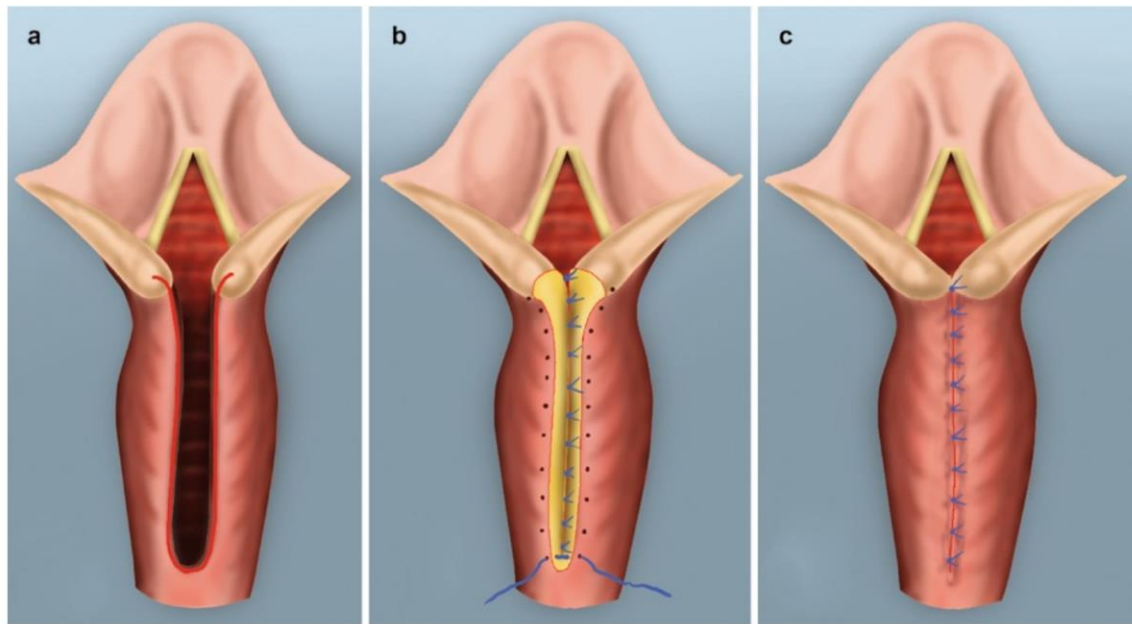


Fig. 12.3 Diagram of laryngo-tracheo-oesophageal repair: In the case of long type IIIb clefts, the CO₂ laser incisions and the two-layer closure are made in a step-wise fashion from caudal to cranial. This diagram shows the principle of endoscopic surgery:

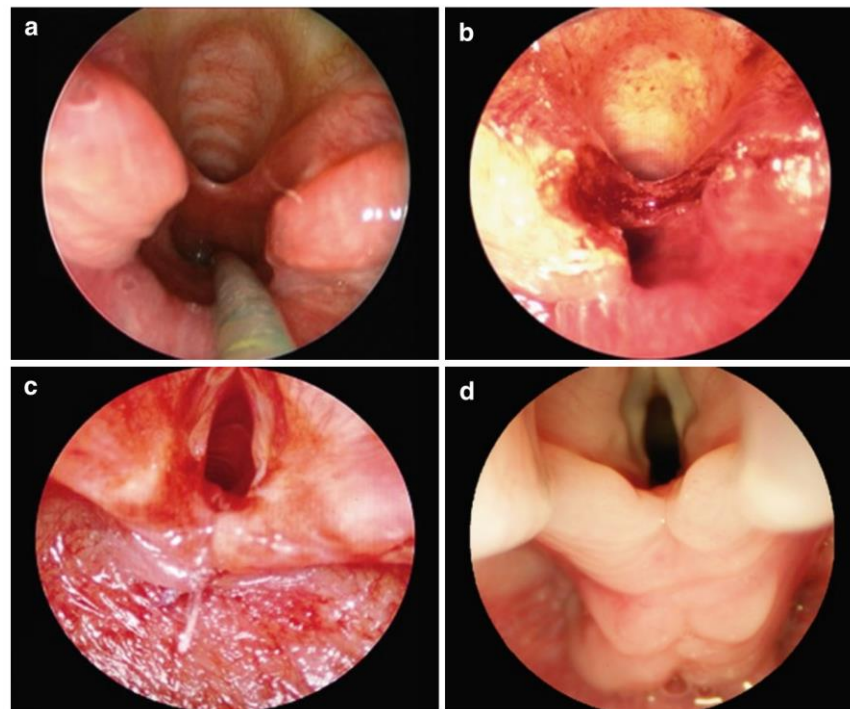


Fig. 12.4 Endoscopic views of a type IIIb laryngeal cleft repair: (a) Endoscopic exposure: the Parsons laryngoscope is inserted below the vocal cords to splay the posterior cleft. (b) CO₂ laser incisions with proper parameters: char-free incision, little bleeding. (c) End of LTOC repair. (d) Postoperative view at 2 years

- **Open Repair with Posterior Cartilage Graft:**
 - Indication:
 - Failed primary endoscopic repair.
 - Type III with severe concomitant congenital anomalies.
 - Type IV (a/b).
 - Pre-op airway control:
 - Tracheostomy:
 - If LTOC is associated with significant tracheomalacia.
 - Should be done below caudal end of the cleft.
 - Deep intubation:
 - Sufficient to provide respiratory support until open surgical repair can be conducted.
 - Method:
 - Low Tracheostomy is done first.
 - Extended laryngo-tracheofissure covering the entire length of the cleft is made.
 - Mucosa of the cleft is incised.
 - Closure of the cleft in two layers with interposition of a costal cartilage graft to restore an adequate interarytenoid distance.
 - A tracheal repair should be left without stenting (No ET tube or NG tube).

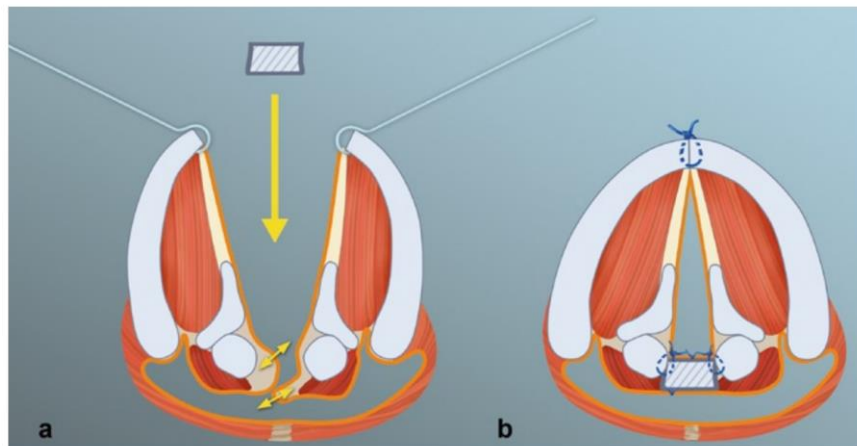


Fig. 12.7 Open surgical repair of Type IV LTOC through a combined right cervico-thoracic approach/cervical repair of LTOC: (a) through a laryngofissure, the reconstruction is made in three layers as for the repair of extrathoracic LTOC, with interposition of a costal cartilage graft to restore an adequate interarytenoid distance. The tracheal mucosa is closed with a

running 5.0 Vicryl suture. If the child does not require a tracheotomy, the ventilating ET tube is kept until a first extubation attempt is envisaged. Should the child require a tracheotomy, a short LT-Mold prosthesis that does not extend below the reconstructed cricoid plate must be used to optimise the final subglottic space. (b) final result after completion of the reconstruction

Esophageal Atresia (EA) with Tracheo-Esophageal Fistula (TEF):

- Pathophysiology
 - **Congenital:**
 - Failure of cannulation of the esophagus or developmental failure of the tracheoesophageal septum.
 - Occurs in 1:5,000 live births.
 - Most common congenital anomaly of esophagus.
 - Associated with tracheomalacia in 60% of the cases.
 - Malacic segment is usually adjacent to the fistula.
 - Due to abnormal development of tracheal muscle.
 - Should be addressed during primary fistula repair.
 - **Acquired:**
 - Secondary to:
 - Tracheostomy
 - Long-term intubation
 - Tumor
 - Inflammation
 - Trauma
- Types of EA:
 - **Type I (8%):**
 - Isolated EA.
 - **Type II (<1%):**
 - Proximal TEF and distal EA.
 - **Type III (86%):**
 - Proximal EA and distal TEF.
 - Most common type.
 - **Type IV (<1%):**
 - EA with proximal and distal TEF.
 - **Type V (<1%):**
 - Isolated TEF (H-type).
 - Located at high level of trachea.

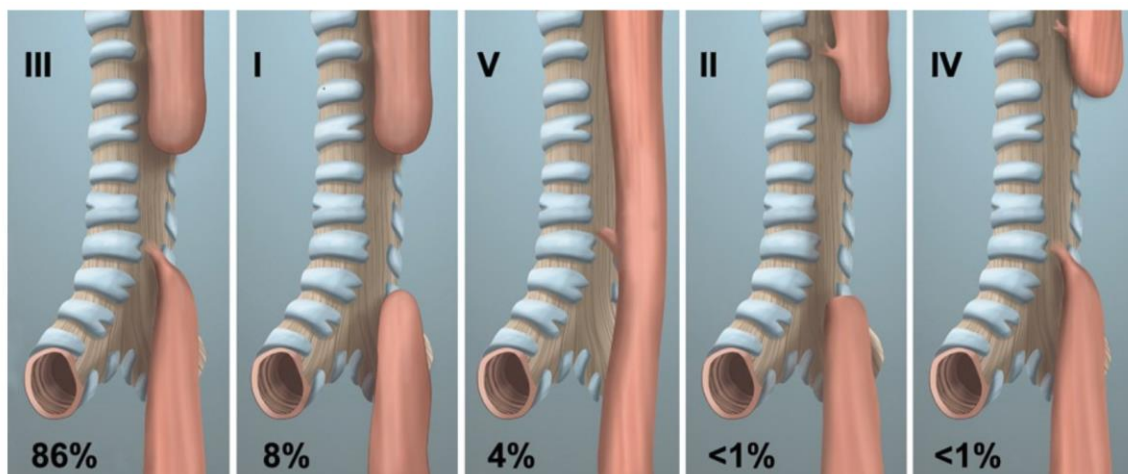


Fig. 13.8 Diagram of oesophageal atresia and tracheo-oesophageal fistula: The type III with a blind proximal oesophageal pouch and a distal fistula is by far the most common (Reproduced from Holinger [45]. With permission)

- Clinical picture of EA/TEF:
 - Excessive drooling.
 - Cyanotic episodes during feeding.
 - Coughing.
 - Vomiting.
 - Washer machine breathing
 - Respiratory distress.
 - Symptoms occurs immediately after birth **EXCEPT** for H-Type TEF without EA which present later on with recurrent pneumonia.
- Associated Syndromes (50%):
 - **VACTERL Association**
 - **CHARGE Association**
 - **G (Opitz-Frias) syndrome**
 - **Pallister–Hall syndrome**
- Diagnosis:
 - **Prenatal diagnosis:**
 - US examination:
 - Polyhydramnios
 - Absence of fluid-filled stomach
 - Fetal MRI:
 - Confirm the diagnosis
 - **Failure to pass size 10F catheter beyond 10 cm.**
 - Smaller-caliber tube is not used because it may curl up in the upper esophageal segment, giving a false impression of esophageal continuity.
 - **Chest X-ray:**
 - Gasless Abdomen suggests either:
 - Isolated EA.
 - EA with proximal TEF.
 - **Barium swallow:**
 - Should be done with small amount (0.5 to 1 mL) of water-soluble contrast.
 - **High resolution CT scan with 3D reconstruction:**
 - Clarifies the anatomy.
 - **Flexible laryngoscopy:**
 - Rules out other laryngeal pathology.
 - **Rigid Laryngobronchoscopy and Esophagoscopy:**
 - Gold standard.
 - Instillation of methylene blue may aid in the diagnosis.
 - When immediate surgical repair is planned, Fogarty or angioplasty catheter is passed through the fistula, and the balloon is inflated in the esophagus.

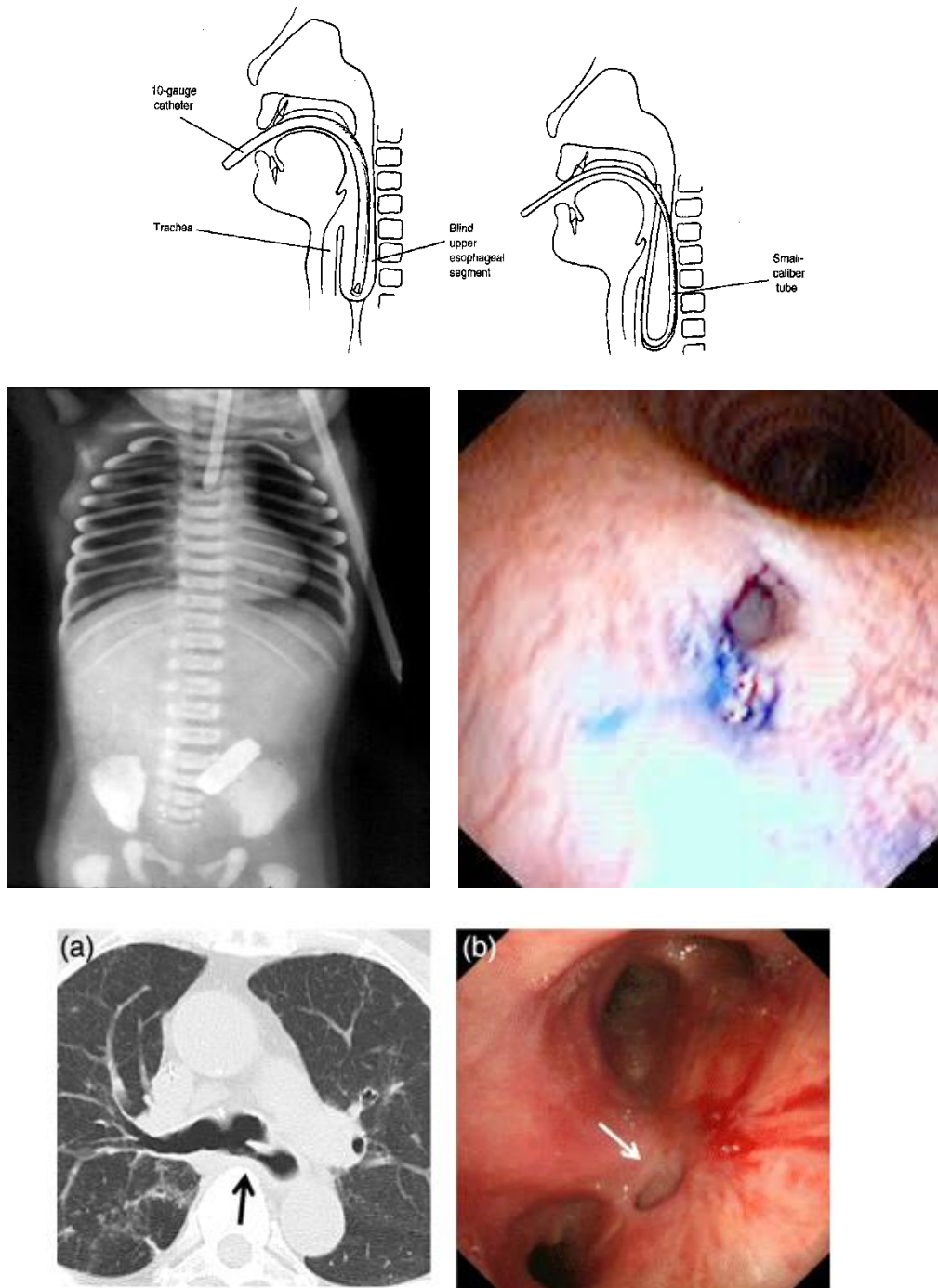
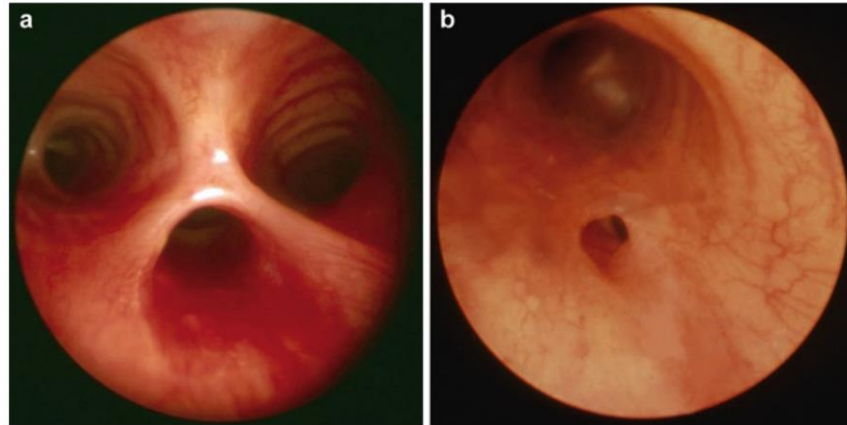


Fig. 1

Chest CT scan (a) and bronchoscopy (b) shows a TEF (size = 1 cm) at the membranous portion of the carina (arrow)

Fig. 13.9 Examples of tracheo-oesophageal fistulae (TOF) with and without oesophageal atresia (OA): (a) Type III OA with TOF: The supracarinal fistula is large. (b) H-fistula without OA (type V): The small fistula is located higher in the mid-trachea



- Treatment:
 - Simple division and closing of the fistula.
 - Restoration of esophageal continuity by single-layer end-to-end anastomosis.
 - Reinforcement of the posterior tracheal wall are indispensable in preventing residual malacia with persisting respiratory symptoms.
 - A patch of tibial periosteum is best suited to rigidify the posterior membranous wall of the trachea with mattress sutures (cartilage is less appropriate)
 - Cannot be done by endoscopic technique.
- Complications:
 - Esophageal stricture (may need dilatation)
 - Gastro esophageal reflux
 - Cranial mobilisation of the distal oesophagus for the end-to-end anastomosis may increase the incompetence of the lower oesophageal sphincter)
 - Need PPI and possibly fundoplication

CONGENITAL VASCULAR ANOMALIES

- Vascular anomalies have abnormal blood and lymphatic vasculature.
- Occur frequently in the head and neck.
- Vascular anomalies are divided into:

1. Vascular Tumors:

- Benign tumors undergo active growth phase characterized by endothelial proliferation and hypercellularity, followed by gradual tumor regression.
- Not present at birth.
- Most common type of vascular tumor is **hemangioma of infancy (HOI)**.

2. Vascular Malformations:

- Structural congenital vascular anomalies designated according to the predominant channel type present (capillaries, veins, lymphatic vessels, arteries or combination).
- Have normal levels of endothelial turnover with no evidence of cellular proliferation.
- Progressive dilation of abnormal channels with
- Present at birth and grow proportionately with the child.
- May not be clinically apparent for a few months.
- No involution phase.
- Most common type of vascular malformation that involves the head and neck is **lymphatic malformation**.

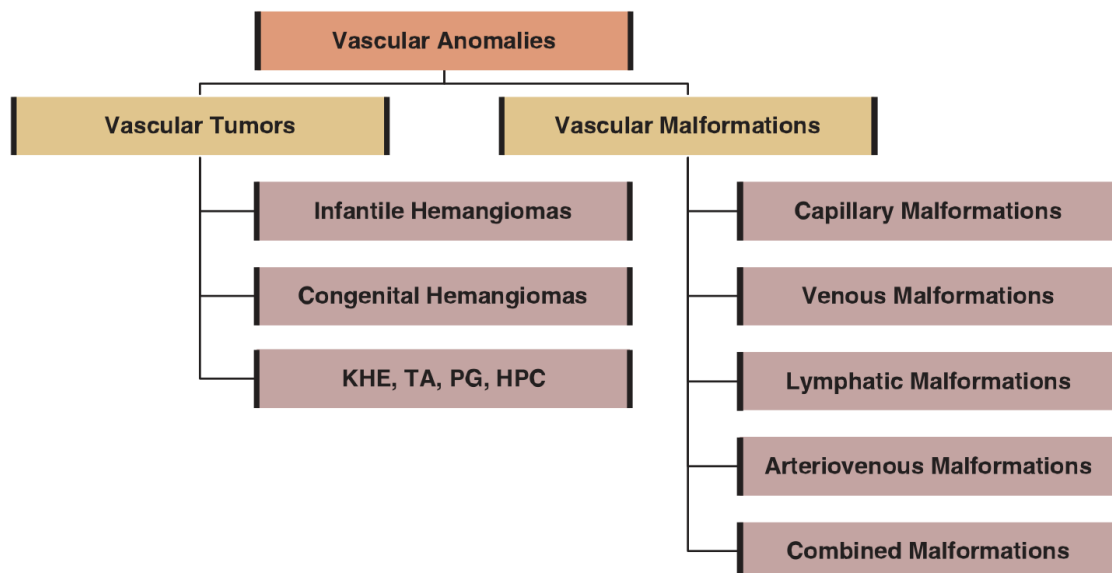


Figure 104.1 A schematic representation of the 1996 ISSVA classification of vascular anomalies. KHE, kaposiform hemangioendothelioma; TA, tufted angioma; PG, pyogenic granuloma; HPC, hemangiopericytoma.

Vascular Tumors:

- **Hemangioma of Infancy (HOI):**

- Most common vascular tumor.
- Most common tumor of infancy.
- Incidence up to 10%.
- More common:
 - o Females (3:1).
 - o Caucasians
 - o Low birth weight infants.
- Clinical picture:

1. Cutaneous Hemangioma:

- Appearance:
 - Firm/rubbery irregular erythematous lesions.
- Location:
 - Head and Neck **(60%)**
 - Trunk (25%)
 - Extremities (15%)
- Distribution:
 - Isolated lesions **(80%)**
 - Multiple lesions **(20%)**
 - o Infant with multiple cutaneous hemangiomas is likely to have visceral lesions, especially in the liver.
 - o Infant with large segmental cervicofacial hemangiomas in the "beard" distribution is at high risk of having airway lesions.
- Time of Appearance:
 - Absent at birth.
 - Appears in first 6 weeks of infancy (85%).
- Growth Phases:
 - **Proliferation:**
 - o Rapid growth of the lesion in 1st year of life.
 - o Endothelial proliferation and hypercellularity.
 - **Involution:**
 - o Gradual tumor regression with hypocellularity and fibrosis.
 - o Color of the lesion fades and become grayish or dull purple.
 - o Starts at 12-18 months of life.
 - o 50% regress by 5 years old.
 - o 70% regress by 7 years old.
 - o 90% regress by 9 years old.
 - o 100% regress by 10-12 years old.

- Types of Cutaneous Hemangioma:
 - **Superficial:**
 - Capillary Hemangioma.
 - Raised, bright strawberry-red color.
 - **Deep:**
 - Cavernous Hemangioma.
 - Dark red/blue color.
 - Present as a mass covered by skin.
 - **Mixed:**
 - Contains both types.
- Classification Cutaneous Hemangioma:
 - **Focal:**
 - Localized.
 - **Segmental:**
 - Plaque-like in contour.
 - Involve cutaneous dermatomes.
 - Typically has extended proliferative period of 18 months.
 - They do not completely regress.
 - Higher risk of ulceration and residual scarring.
 - CN-V3 distribution (Cervicofacial area/beard distribution) is associated with airway involvement (**65%**).
 - Ocular involvement can lead to blindness.
 - **Disseminated:**
 - Increased risk of visceral Hemangioma with presence of > 5 cutaneous hemangiomas.



FIGURE 199-5. Types of hemangioma of infancy. **A**, Superficial focal hemangioma; forehead. **B**, Mixed segmental hemangioma; midface, lip, forehead. **C**, Deep focal hemangioma; malar region. **D**, Mixed focal hemangioma; malar lower eyelid. **E**, Mixed segmental hemangioma; cheek, malar region.



FIGURE 199-3. Segmental hemangioma appearance. **A**, Initial appearance in infancy. **B**, Late proliferative stage. **C**, Beginning involution phase.



A



B



C

Figure 104.3 A series of photographs of children with hemangiomas are presented demonstrating variations in morphology. **A**: Example of a child with an isolated focal hemangioma. **B**: Example of a child with multiple cutaneous hemangiomas. **C**: Example of a child with a facial segmental hemangioma occupying the periorbital, temporal, cheek, and chin areas of the face. Segmental hemangiomas are prone to ulceration as can be seen in this child around the left nasolabial crease.

2. Airway Hemangioma:

- Occurs anywhere in upper and lower airway.
- Has same clinical course and histological appearance of cutaneous Hemangioma.
- Asymptomatic if limited to superficial mucosa only.
- Frequently presents during proliferation in association with URTI.
- Risk factors:
 - Large segmental hemangioma of lower face **(65%)**
 - PHACES Syndrome **(50%)**
- **Subglottic Hemangioma (SGH):**
 - Rare (1.5%) congenital laryngeal anomaly.
 - Most common location of airway hemangioma.
 - 50% of children with subglottic hemangioma will have a cutaneous lesion.
 - Clinical picture:
 - Asymptomatic during the first weeks of life.
 - Inspiratory stridor followed by biphasic stridor with barking cough and slight hoarseness start at 2–4 months and becoming manifest in all infants by age of 6 months.
 - Progression of symptoms reaches a plateau between ages of 10–12 months.
 - Symptoms then decrease slowly and finally disappear around age of 2 years.
 - Complete resolution of tumor may take as long as 5–10 years.
 - Recurrent or prolonged episodes of croup should always alert the physician to a likely congenital anomaly.
 - Symptoms of respiratory distress depend on the severity of the airway obstruction.
 - Empiric use of high-dose corticosteroids in infant with cutaneous HOI and new-onset stridor usually improves the stridor and can be used as diagnostic assessment.

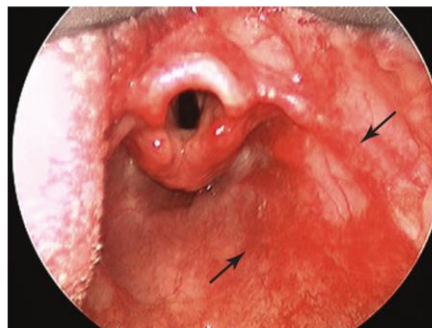


FIGURE 199-9. Endoscopic appearance of airway hemangioma of infancy showing diffuse hypopharyngeal mucosal involvement (arrows).

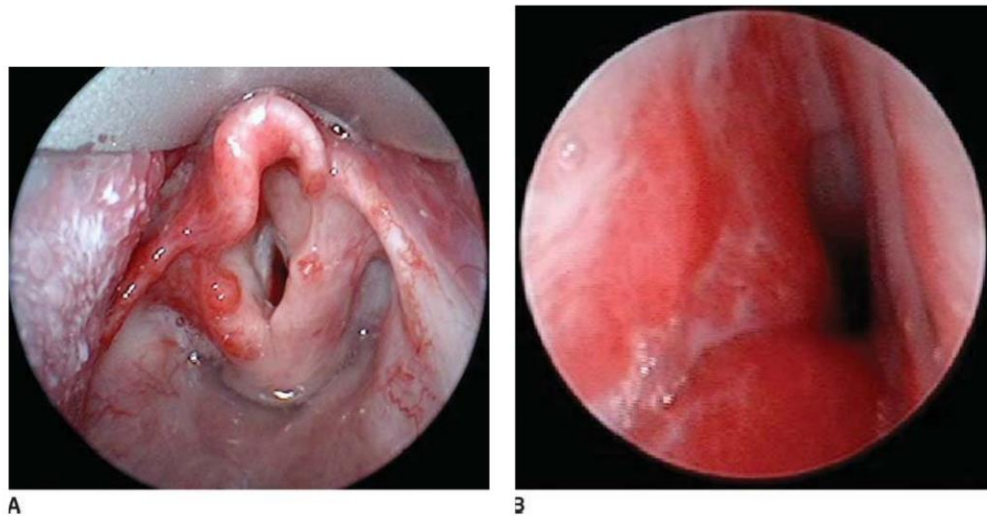


Figure 104.5 Endoscopic photographs of (A) a child with flat supraglottic hemangiomas occupying the left epiglottis and aryepiglottic fold and (B) a child with a large left subglottic hemangioma that severely narrows the subglottic lumen.

3. Parotid Hemangioma:

- Most common benign parotid tumors of childhood.
- 50% parotid hemangiomas are associated with cutaneous hemangiomas
- Parotid gland hemangiomas typically occupy entire gland and adjacent parapharyngeal space.
- Children with extensive hemangioma of parotid and pharyngeal area have chronic recurrent otitis media secondary to extrinsic compression of eustachian tube.
- They grow rapidly, causing extrinsic compression of upper pharyngeal airway.
- Frequently have a prolonged proliferative phase (12-16 months) and may require systemic therapy if airway is compromised.

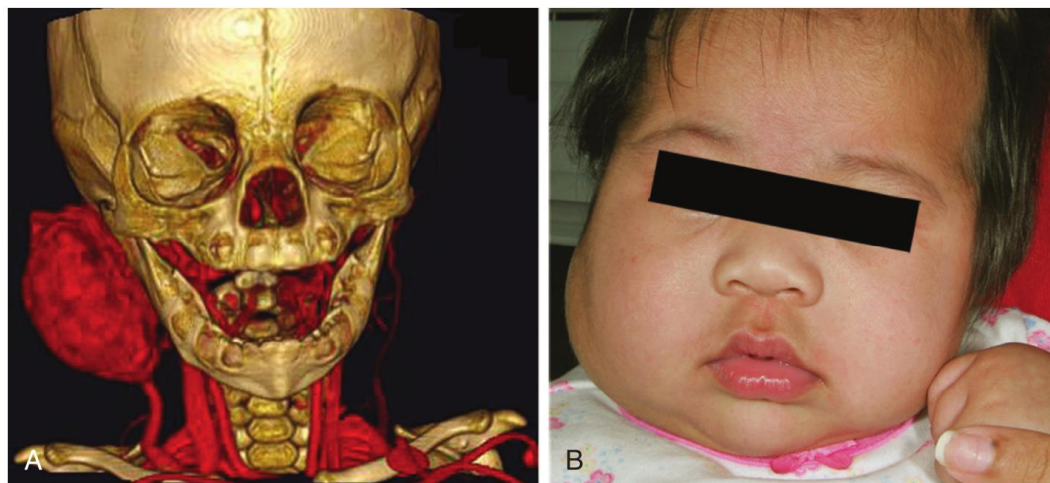


FIGURE 199-I. A, Computed tomographic angiography appearance of deep focal hemangioma of parotid neck region. B, Clinical image of same patient.

4. PHACES Syndrome:

- **P**osterior cranial fossa anomalies
- **H**emangioma (facial segmental)
- **A**rterial/carotid anomalies
- **C**ardiac anomaly/Coarctation of aorta
- **E**ye anomaly
- **S**ternal pit
- Diagnosis:
 - Consider PHACES workup **in all patients with large segmental hemangiomas.**
 - More common in females (90%)
 - MRI, Ophthalmic exam and Cardiac echo.



FIGURE 199-6. Clinical appearance of PHACES. **A**, Segmental hemangioma that involves the lips, nose, and left upper and lower face. **B**, Segmental hemangioma that involves the lips, nose, and left upper and bilateral lower face. **C**, Segmental lower-lip hemangioma with sternal cleft. **D**, Focal lower-lip hemangioma with sternal cleft.

- Complications:
 - Occur from lesion-induced functional compromise and cutaneous ulceration.
 - **Cosmetic deformity:**
 - Unpredictable cosmetic outcome.
 - In 20-40% of patients, residual changes of skin (laxity, discoloration, telangiectasia, fibrofatty masses or scarring are seen).
 - Especially with large segmental HOI.
 - The longer it takes to involute the higher the incidence of permanent cutaneous residua.
 - Proliferation of lesions nasal tip can cause nasal distortion.
 - **Ulceration and scarring:**
 - Proliferating HOI in places of repetitive skin trauma (lips and neck skin folds) can ulcerate and result in scarring.
 - Ulceration begins as brown crusts that become painful and bleed, and they require local wound care for healing.
 - Lip ulceration makes oral feeding challenging because of pain and HOI-induced lip distortion.
 - **Infection:**
 - Extensive HOI ulcerations can become infected and result in tissue loss which complicates healing and treatment.
 - **Airway obstruction:**
 - Airway HOI can compromise the airway causing biphasic stridor.
 - **Visual disturbance:**
 - Periocular hemangiomas can involve the upper lid, lower lid, and for the retrobulbar space.
 - Pressure from hemangioma on the globe may lead to astigmatism and amblyopia.
 - **High-output heart failure:**
 - Large head and neck HOIs are associated with high-output cardiac failure from increased blood flow in these vascular lesions.

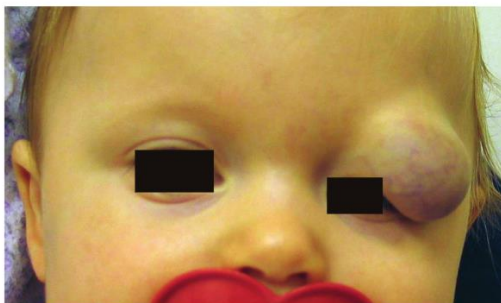


FIGURE 199-10. Deep focal hemangioma of infancy of the upper eyelid that impinges on the visual axis.



FIGURE 199-11. Ulcerated hemangioma of infancy of the ear and neck.

- Diagnosis:
 - **Ultrasound color flow Doppler :**
 - Extremely useful in differentiating hemangiomas from vascular malformations that have a similar appearance.
 - Appearance:
 - Hypoechoic
 - Well defined
 - Heterogeneous in texture
 - Small cystic and sinusoidal spaces.
 - Characteristic fast-flow pattern is visualized by Doppler US in proliferative phase.
 - **CT with contrast:**
 - Delineate extent and involvement of the hemangioma.
 - Helpful in distinguishing hemangiomas from other malformations.
 - In proliferative phase:
 - Appears as a well-circumscribed tumor with homogeneous parenchyma and intense enhancement.
 - In involutive phase:
 - Appears as a heterogeneous tumor with distinct lobular architecture and large draining veins in the center and periphery.
 - **MRI with Gad:**
 - Study of choice
 - T1 shows soft tissue mass, ISO-intense or HYPO-intense, flow voids, HYPER-intense with contrast.
 - T2 shows lobulated soft tissue mass, HYPER-intense, flow voids.
 - **CT Angiography:**
 - Rarely needed.
 - Provide information about size of the lesions and the feeding vessels.
 - Can also distinguish between hemangiomas and other vascular malformations.
 - **Biopsy:**
 - Indicated only when the diagnosis is uncertain or when the possibility of malignancy must be ruled out.
 - **GLUT-1 stain (Glucose transporter enzyme)** is strongly expressed in all phases of infantile hemangiomas.
 - Not expressed in normal surrounding vascular endothelium or in vascular malformations nor in congenital hemangiomas.
 - Very sensitive and specific for histologic confirmation of HOI.

○ **Airway endoscopy:**

- Stridor in association with segmental HOI should prompt airway evaluation.
- Mainstay of diagnosing subglottic hemangioma is rigid endoscopy under GA after laryngomalacia and vocal cord paralysis have been ruled out by fiberoptic laryngoscopy.
- Appearance of SGH:
 - Reddish smooth mass.
 - Most common location is left posterolateral subglottis and extending cranially to under surface of vocal cord.
 - Tumor mass is spongy and compressible, allowing for easy intubation with ET tube with no risk of major haemorrhage.

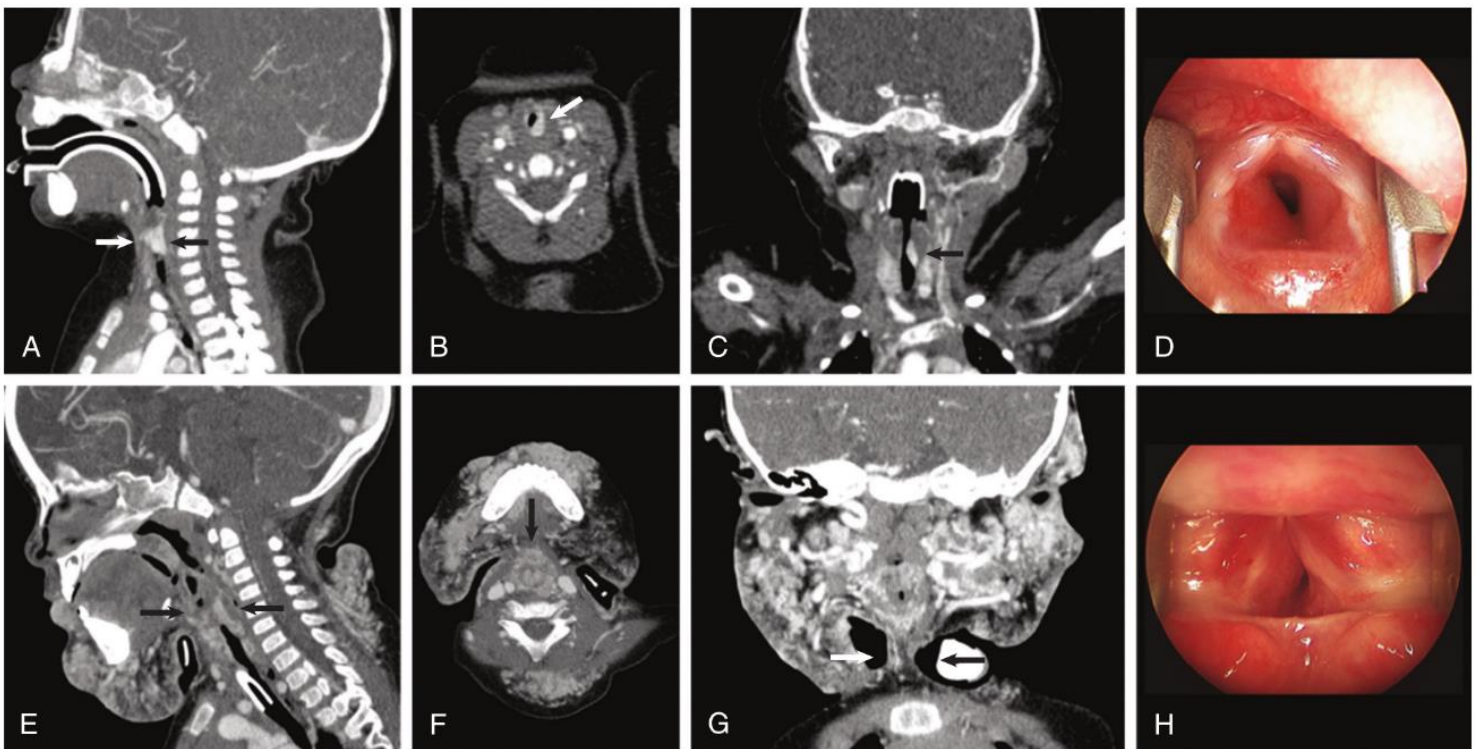


FIGURE 199-20. Computed tomography (CT) and endoscopic assessment of airway hemangioma of infancy. **A**, to **C**, CT appearance of focal airway hemangioma of infancy in medial subglottis. **A**, Sagittal. **B**, Axial. **C**, Coronal. **D**, Unilateral posterior focal hemangioma; left subglottis, same patient. **E** to **G**, CT appearance of segmental airway hemangioma of infancy. **H**, Circumferential, segmental hemangioma; subglottis, same patient.

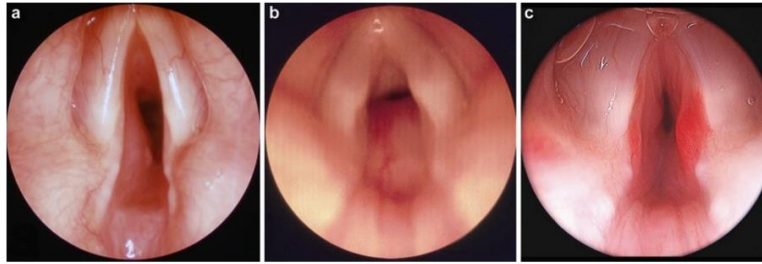


Fig. 10.2 Endoscopic views of typical aspects of subglottic haemangiomas: (a) Left posterolateral subglottic haemangioma. (b) Posterior subglottic haemangioma. (c) Bilateral subglottic haemangioma

- Management:

○ **Conservative:**

- Most uncomplicated focal hemangiomas involute spontaneously and do not require intervention.
- Rapidly proliferating lesions are sometimes problematic and can cause a wide range of complications.
- If any complications occur, the patient should be closely monitored to ensure timely intervention.
- When infant with airway HOI has severe respiratory compromise, airway protection is accomplished with ET intubation using small uncuffed endotracheal tube.
- **Indications of early intervention:**
 1. Large segmental hemangiomas.
 2. Life- threatening hemangiomas.
 3. Function-threatening hemangiomas.
 4. Presence of ulceration or secondary infection.

○ **Medical:**

- Medical therapy for proliferating HOI is directed at stopping angiogenesis.
- The aim of treatment in airway hemangioma is maintaining the airway without tracheotomy, while avoiding any long-term sequelae.

1. Propranolol (Non-selective beta blocker):

- First-line therapy for HOI impair function.
- Dose is **2mg/kg/day** for 6 months.
- Safety profile is excellent.
- Need pre-therapy cardiology evaluation.
- Can be safely initiated as outpatient therapy with appropriate education and monitoring.
- Inpatient initiation is reserved for:
 - Airway lesions
 - Young patients
 - PHACES syndrome
- Successful therapy with propranolol is associated with decreased blood vessel density in HOI.
- On average, stridor resolved within a day of initiating propranolol

**TABLE
104.2****PROTOCOL TO INITIATE PROPRANOLOL THERAPY****Inpatient propranolol algorithm**

- Preadmission diagnostic testing—echocardiogram to rule out congenital anomalies in patients with a history of murmurs, failure to thrive, or other significant medical comorbidities; EKG to rule out cardiac arrhythmia
- Admit to telemetry unit with continuous heart rate and blood pressure monitoring
- Initiate propranolol at 0.5 mg/kg PO BID for 2 doses; increase to 1 mg/kg PO BID for 2 doses; finally increase to 2 mg/kg PO BID
- Propranolol is increased in this manner as long as the patient exhibits no adverse response to the administration of the medication
- Check blood glucose 2 h after each dose of propranolol
- Check EKG 24 h after initiation of propranolol
- Discharge home after second dose of propranolol at 2 mg/kg with contact information if questions or problems arise
- Parents are instructed to maintain a strict feeding regimen. If the child is <6 mo of age, feedings should be every 2–4 h. Older children can be spaced to feedings every 6–8 h
- Parents instructed to hold medication if the child becomes ill and/or has decreased PO intake

Outpatient propranolol algorithm

- Diagnostic testing—same as for inpatient algorithm
- Monitor on telemetry unit for 6 h with continuous blood pressure and heart rate monitoring
- Initiate propranolol at 0.5 mg/kg
- Check blood glucose 2 h after each dose of propranolol
- Discharge instructions as for inpatient algorithm with contact information if questions or problems arise
- Continue propranolol at 0.5 mg/kg PO BID or 1 wk
- Return to hospital for 6-h admission on telemetry unit during which propranolol is increased to 1 mg/kg. An EKG is obtained prior to increasing the dose and blood glucose is checked 2 h after dose. Discharge home at this dose for a week with discharge instructions as previous
- Return to hospital for 6-h admission during which propranolol is increased to 2 mg/kg. Blood glucose is again checked 2 h after dose. Patient is discharged home with discharge instructions as previous with follow-up in 4 wk

TABLE 4 Contraindications to Propranolol Therapy

Cardiogenic shock
 Sinus bradycardia
 Hypotension
 Greater than first-degree heart block
 Heart failure
 Bronchial asthma
 Hypersensitivity to propranolol hydrochloride

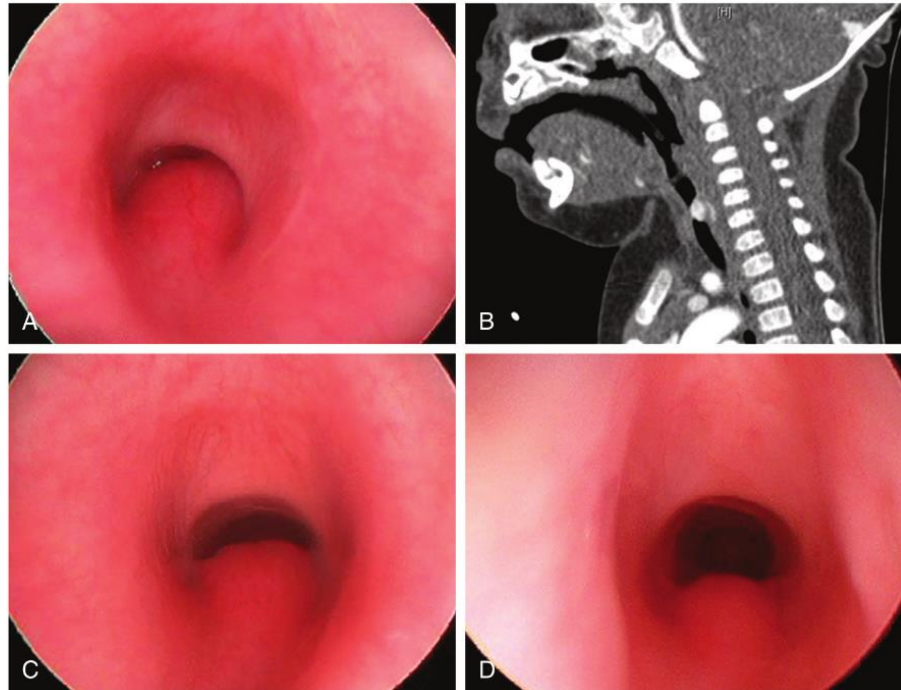
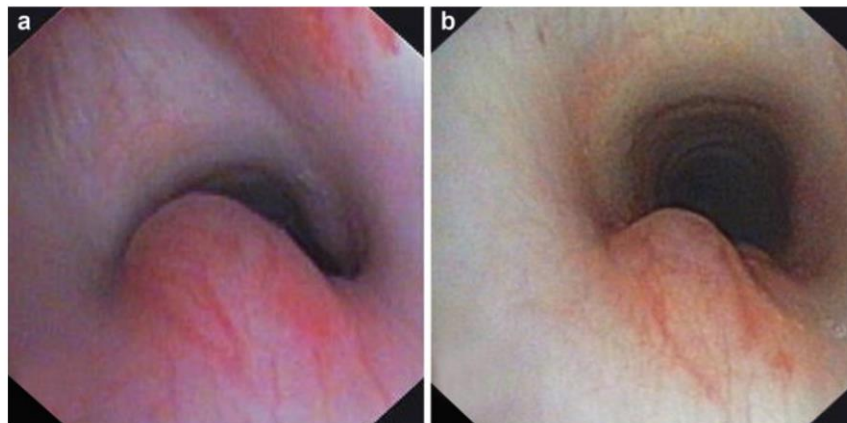


FIGURE 199-21. Endoscopic and computed tomography (CT) appearance of focal tracheal hemangioma. **A**, Before propranolol therapy. **B**, CT angiogram of tracheal hemangioma. **C**, After 1 month of propranolol therapy. **D**, After 4 months of propranolol therapy.

Fig. 10.4 Left posterolateral subglottic haemangioma treated with propranolol (2 mg/kg/day): **(a)** Initial condition: The SGH obstructs the subglottic space by 70%, causing rest dyspnoea and inspiratory stridor. **(b)** Situation 2 months after propranolol treatment: significant regression of the SGH. Symptom-free patient



2. Systemic Corticosteroids:

- Anti-angiogenic.
- Used as adjuvant therapy or as curative treatment only for a short period of time.
- Indicated mainly after failure of propranolol treatment.
- **Prednisolone 2mg/kg/day** followed by a slow tapering dose over 4-6 weeks.
- 25% of HOI will shrink after 1 month.
- 50% of HOI will stay stable.
- 25% of HOI will have poor response.
- Not recommend for longer than 3 weeks if the symptoms do not markedly improve.
- Potential long-term side effects such as failure to thrive, osteoporosis, adrenal suppression and Cushing syndrome should not be underestimated.

3. Intra-lesional Steroid Injections:

- Indicated for focal hemangiomas.
- Required repeated injections (Q6w).
- A long-acting steroid (**Triamcinolone acetonide/Kenacort**) is injected with volume not exceeding 2.5mL
- A 25-gauge needle is used and injections are made directly into hemangioma in different directions through the same needle hole.
- Direct pressure is applied for 2-10 minutes to stop bleeding.
- Risk of blindness and systemic absorption after intra-lesional injections in periorbital hemangiomas.
- **In subglottic lesions:**
 - Success rates with intra-lesional steroid injection alone exceed 75%.
 - Preferred to be used as an adjuvant treatment to CO₂ laser resection.
 - Repeated injections, temporary ET-intubations and postoperative PICU stays increase intubation-related risks as well as treatment costs.

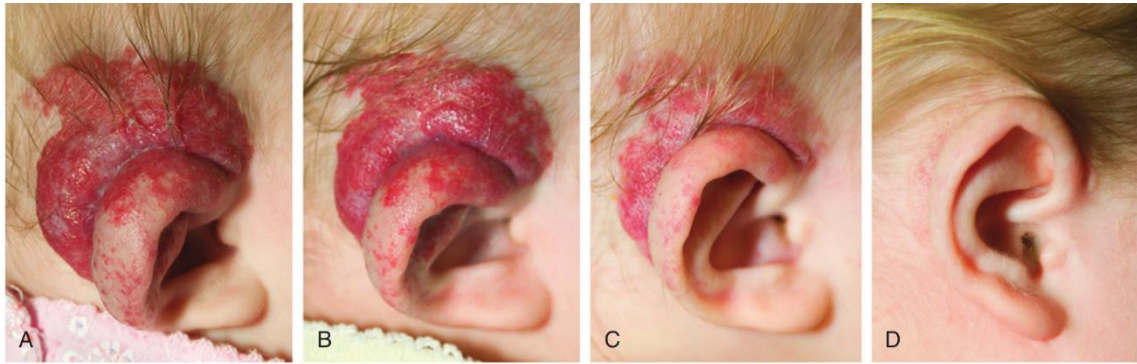


FIGURE 199-17. Superficial segmental hemangioma treated with propranolol. **A**, Pretreatment with postauricular ulceration. **B**, After 1 month of corticosteroid treatment. **C**, After 1 month of propranolol treatment. **D**, After 4 months of propranolol treatment.

4. Interferon Alpha-2a:

- Not frequently used currently.
- Effective treatment for refractory extensive HOI.
- Used as daily subcutaneous injections of 1-3 million U/m² for 6 months.
- Side effects include:
 - Low-grade fever
 - Hepatotoxicity
 - Neutropenia
 - Anemia
- Should never be used in patients less than 1 year of age due to frequent neurologic complications such as spastic diplegia.
- Although commonly used in the past, interferon a-2a has been **abandoned** owing to its severe side effects, suboptimal efficacy and the adoption of other treatment modalities.

5. Vincristine:

- Not frequently used currently.
- Effective treatment for extensive HOI.
- Can induce neurologic deficits in young children.

- **Surgical Therapy in Cutaneous Hemangiomas :**

- 1. Laser Therapy:**

- Indications:
 - Residual vascular markings of superficial HOIs at the end of involution phase.
 - Ulcerated HOI refractory to medical management.
 - Pulsed-dye laser (PDL) device reduces and removes skin redness of superficial HOIs while preserving overlying epidermis without causing scarring.
 - However, it does not reduce HOI volume.
 - Also it promote wound healing and reepithelialization of ulcerated HOI.



FIGURE 199-12. Hemangioma of infancy; lip. **A**, Ulcerated hemangioma during proliferation. **B**, Lip appearance after several pulsed-dye laser treatments and ulcer healing.

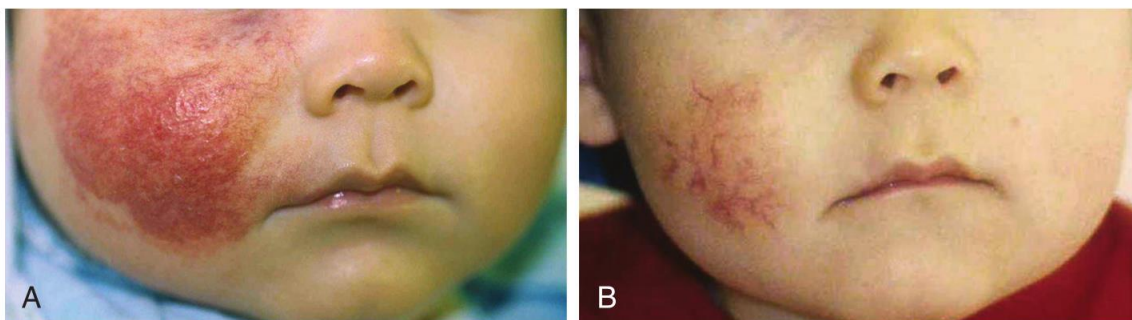


FIGURE 199-18. Laser treatment of superficial hemangioma. **A**, Pretreatment hemangioma appearance. **B**, Superficial hemangioma appearance after three pulsed-dye laser treatments.

2. Surgical Excision:

- Considered when surgery is thought to provide a better overall outcome to the patient than natural regression or medical management.
- Indicated for removal of the fibrofatty residuum or skin laxity that remains after complete regression.
- When deep facial hemangioma is mixed with large superficial HOI, staged treatment with pulsed-dye laser induces skin and dermal thickening to allow HOI excision with minimal skin loss and tension-free closure of the defect.
- Indications of early surgical excision:
 1. Hemangiomas causing significant functional compromise (vision/breathing) unresponsive to medical management.
 2. Pedunculated and ulcerated lesions causing bleeding and infection.

**TABLE
104.3**

RELATIVE INDICATIONS FOR SURGICAL MANAGEMENT OF HEMANGIOMAS

- Noninvoluting congenital hemangioma
- Ulcerative hemangioma that will likely leave a large scar
- A hemangioma that is causing significant functional compromise or symptomology and is not responding expeditiously to medical management. Examples of functional compromise include obstruction of visual axis with changes in vision or airway obstruction.
- A hemangioma that has a pedunculated appearance, and therefore will likely not involute completely.
- A hemangioma that causes significant psychosocial or appearance issues to the patient as he/she enters school can be removed with a satisfactory cosmetic outcome.



FIGURE 199-19. Combined laser and surgical treatment; small nasal tip hemangioma of infancy. **A**, Pretreatment. **B**, After pulsed-dye laser treatment. **C**, After excision of residual deep hemangioma via an external rhinoplasty approach.



FIGURE 199-16. Combined laser and surgical treatment; large upper-lip/cheek mixed segmental hemangioma of infancy. **A**, Pretreatment. **B**, After residual deep hemangioma excision.

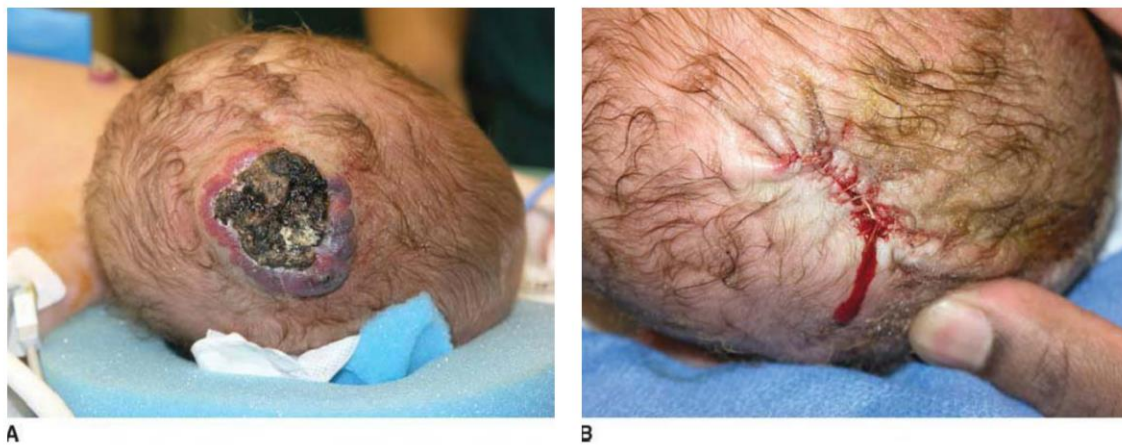


Figure 104.7 Photographs of a patient with a large ulcerative scalp hemangioma (**A**) that was surgically excised (**B**).

- **Surgical Therapy in Subglottic Hemangiomas :**
 - When infant with airway HOI has severe respiratory compromise, airway protection is accomplished with ET intubation using small uncuffed endotracheal tube.
 - This allows safe transfer to a facility capable of comprehensive HOI care and avoids using a tracheotomy for airway protection.
 - Systemic corticosteroids can be used as adjuvant postoperative therapy in airway surgery.

1. Endoscopic Laser Resection:

- Indication:
 - Slow-growing tumors that become symptomatic at age of 4–6 months.
- CO₂ laser is preferred over KTP laser in treatment of SGH due to the difficulty to control deep thermal damage with KTP laser.
- CO₂ laser resection may require 2-3 sessions to maintain a safe airway with a success rate of 90%
- Risk of cicatricial SGS (1.5%) if ablating more than 40% of subglottic circumference in a laterally situated SGH.

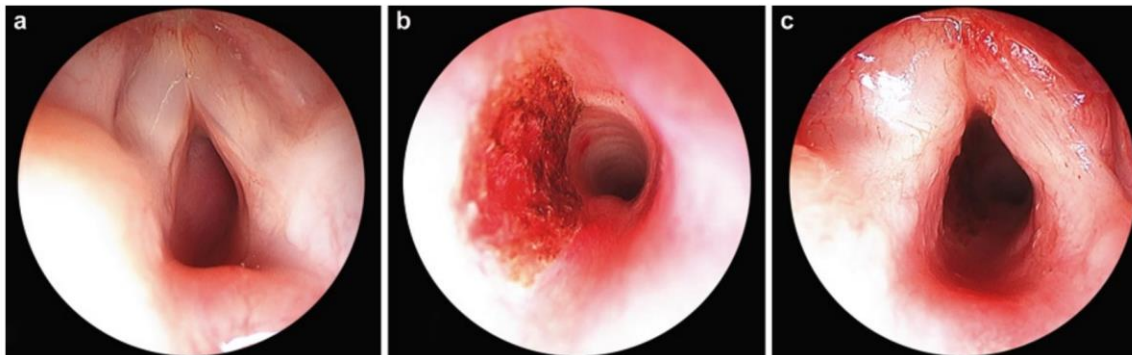


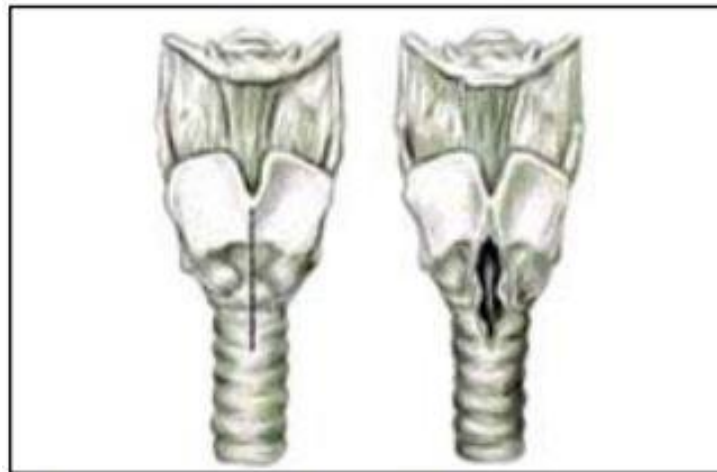
Fig. 10.3 CO₂ laser vaporisation of a left subglottic haemangioma: (a) Left-sided subglottic haemangioma. (b) CO₂ laser vaporisation with minimal charring. (c) Immediate postoperative result

2. Endoscopic Microdebrider Submucosal Resection:

- Not widely used.
- Aims at preserving the mucosa and perichondrium by inserting the microdebrider probe through a small mucosal opening.

3. Open Surgical Excision Through Laryngofissure:

- Indication:
 - Large fast-growing SGH in the proliferative phase.
 - Large SGH not involuted by age of 2 years.
 - Bilateral SGH.
 - Circumferential SGH
- Best done as a primary treatment rather than as salvage surgery after several failed laser treatments.
- Best success rate (98%) but more invasive procedure.
- Post-op care:
 - Patient is kept intubated for 24–48 h.
 - Direct laryngoscopy to check the integrity of the mucosa on the resected side is performed prior to extubation.
 - Extubation and close monitoring in the PICU are indispensable until the child can be safely transferred to a regular in-patient room.



4. Tracheostomy:

- Indication:
 - Difficult cases in centers where modern equipment and expertise are lacking.
- Safest surgical procedure.
- Has the great advantage of leaving the subglottis untouched because the high percentage of spontaneous involution of SGH (90%).
- Disadvantages:
 - Morbidity and mortality risk (1–3%)
 - Delay in speech and language acquisition.



Figure 104.2 Infantile hemangiomas undergo a process of natural regression. The following series of photographs depict a young girl at (A) 2 months, (B) 6 months, (C) 18 months, and (D) 36 months of age. As the child grows older, the hemangioma changes from a bright red color to a grayish red color, and finally to the natural skin color. The mass of the lesion also decreases, leaving little evidence of the hemangioma over time.

- **Congenital Hemangioma:**
- Uncommon variant of infantile hemangiomas.
- Comprise >3% of all hemangiomas seen in infancy.
- Rarely coexist in a patient with a typical infantile hemangioma.
- Difficult to distinguish from HOI unless careful history is taken and histologic analysis performed.
- Differ from infantile hemangiomas in:
 - **Appearance:**
 - Generally present as solitary violaceous lesions at birth.
 - Have a predilection for the head or limbs.
 - **Clinical behavior:**
 - Present at birth as fully grown lesions and do not undergo additional postnatal growth.
 - Considered as high-flow lesions and can be misdiagnosed as arteriovenous malformations (AVMs).
 - Two types:
 - **Rapidly involuting congenital hemangiomas (RICHs):**
 - Typically involute by 12-14 months of age.
 - Leaves residual patch of thin skin with prominent veins and little subcutaneous fat.
 - **Non-involuting congenital hemangiomas (NICHs):**
 - Do not undergo involution.
 - **Histological analysis:**
 - Congenital hemangiomas are **GLUT-1 negative** staining.
 - **Treatment:**
 - Medical therapy is ineffective for congenital hemangiomas.
 - Treatment for RICHs is usually observation, whereas NICHs may require laser or surgical therapy.



FIGURE 199-7. Congenital hemangioma appearance. **A**, Noninvoluting congenital hemangioma. **B**, Rapidly involuting congenital hemangioma after involution.

- **Benign non-hemangioma vascular tumors:**
 - **Kaposiform Hemangioendothelioma (KHE):**
 - Rarely occur in the head and neck.
 - Has significant lymphatic component in addition to blood vascular endothelium.
 - Appears as violaceous cutaneous nodules that extend into deep tissues.
 - Radiographic appearance shows diffuse infiltrative vascular process.
 - Careful assessment through imaging and diagnostic biopsy should guide KHE therapy.
 - **Tufted Angioma (TA):**
 - Rarely occur in the head and neck.
 - More localized and may or may not involve skin.
- Kasabach-Merritt Phenomenon:
 - Sequestration of platelets and severe thrombocytopenia, microangiopathic hemolytic anemia, and consumptive coagulopathy.
 - Occurs with KHE or TA **(Not with infantile hemangioma)**
 - Suspected with rapidly enlarging lesion.
 - Can be life-threatening
- Diagnosis:
 - Diagnosis of both KHE and TA requires incisional biopsy with possible immunohistochemical staining.
- Treatment:
 - Treatment of both KHE and TA is centered on proper diagnosis, prevention of complications, and tumor control.
 - Curative surgical resection is advisable in localized tumors.
 - For large lesions, surgery may not be curative.
 - In extensive KHE or TA, antiangiogenic chemotherapy may be necessary.



FIGURE 199-8. Kaposiform hemangioendothelioma, clinical and radiographic appearances. **A**, Cutaneous nodules. **B**, Deep lesion that extends to the skin. **C**, Coronal computed tomography of the lesion shown in **B**.

Vascular Malformations:

- **Low-flow Vascular Malformations:**
 - Lymphatic malformation
 - Venous malformations
 - Capillary malformations
- **High-flow Vascular Malformations:**
 - Arteriovenous malformations
- **Lymphatic Malformations:**
 - Low-flow vascular malformations.
 - Most common pediatric vascular malformation in head and neck.
 - 75% of all lymphatic malformations occur in the head and neck.
 - Arise from embryologic disturbances in development of the lymphatic system.
 - Associated with venous malformations (lymphatics and venous system develop concurrently).
 - Histologically:
 - Consist of abnormally dilated lymphatic channels that do not differ when compared with radiographic characterization.
 - Collections of lymphocytes are common throughout the contained connective tissue.
 - Clinical picture:
 - Present at birth (65%).
 - May not clinically apparent until after URTI or trauma.
 - Hemorrhage within the cyst spaces is common, indicating recent trauma or spontaneous intralesional bleeding.
 - 90% present in <3 years old.
 - Morphologic types:
 - **Microcystic (<2 cm):**
 - Located above level of mylohyoid muscle (**Suprahyoid**).
 - Found in oral cavity, oropharynx, tongue, parotid gland, submandibular gland and pre-epiglottic space.
 - Small soft non-compressible masses with mucosal or skin vesicles.
 - **Macrocytic (>2 cm):**
 - Located below level of mylohyoid muscle (**Infrahyoid**).
 - Found in anterior and posterior cervical triangle.
 - Large soft compressible smooth, transilluminated masses under normal or bluish skin.
 - Multilocular and consist of numerous cysts varies in size.
 - **Combined:**
 - Formerly called cystic hygroma and lymphangioma.

- Staging (Modified de Serres):
 - **Stage I:**
 - Unilateral infrahyoid lesion.
 - **Stage II:**
 - Unilateral suprahyoid lesion.
 - **Stage III:**
 - Unilateral infrahyoid and suprahyoid lesions.
 - **Stage IV:**
 - Bilateral suprahyoid lesions.
 - **Stage V:**
 - Bilateral suprahyoid and infrahyoid lesions.
 - **Stage VI:**
 - Bilateral infrahyoid lesions.
 - **Stage VII:**
 - Retropharyngeal lesion.
 - **M:**
 - Mediastinal extension.



FIGURE 199-22. Head and neck lymphatic malformation stages. **A**, Stage I, unilateral infrahyoid. **B**, Stage 2, unilateral suprahyoid. **C**, Stage 3, unilateral suprahyoid and infrahyoid. **D**, Stage 4, bilateral suprahyoid. **E**, Stage 5, bilateral suprahyoid and infrahyoid.

- Diagnosis:

○ **Prenatal US:**

- Up to 60% of all lymphatic malformations are detected by ultrasound in utero.
- Detects large cystic lymphatic lesions.
- As early as the beginning of second trimester.
- Prenatal diagnosis of large cervicofacial lymphatic malformations is important as it could be associated with airway obstruction.
 - Influence the mode, timing, and place of delivery.
 - May require performing **EXIT** (Ex Utero Intrapartum Treatment).

○ **Postnatal US:**

- Helpful in confirming the diagnosis in superficial lesions.
- Less valuable for showing extension of the lesion.

○ **CT scan:**

- Shows non-enhancing fluid density areas.
- Demonstrate the anatomic extent of cystic lesions and their relationship to soft tissues, muscle, and vascular structures.

○ **MRI:**

- Non-enhancing muscle signal in T1.
- Non-enhanced high signal without any feeding or draining vessels in T2.
- Demonstrate the anatomic extent of cystic lesions and their relationship to soft tissues, muscle, and vascular structures.

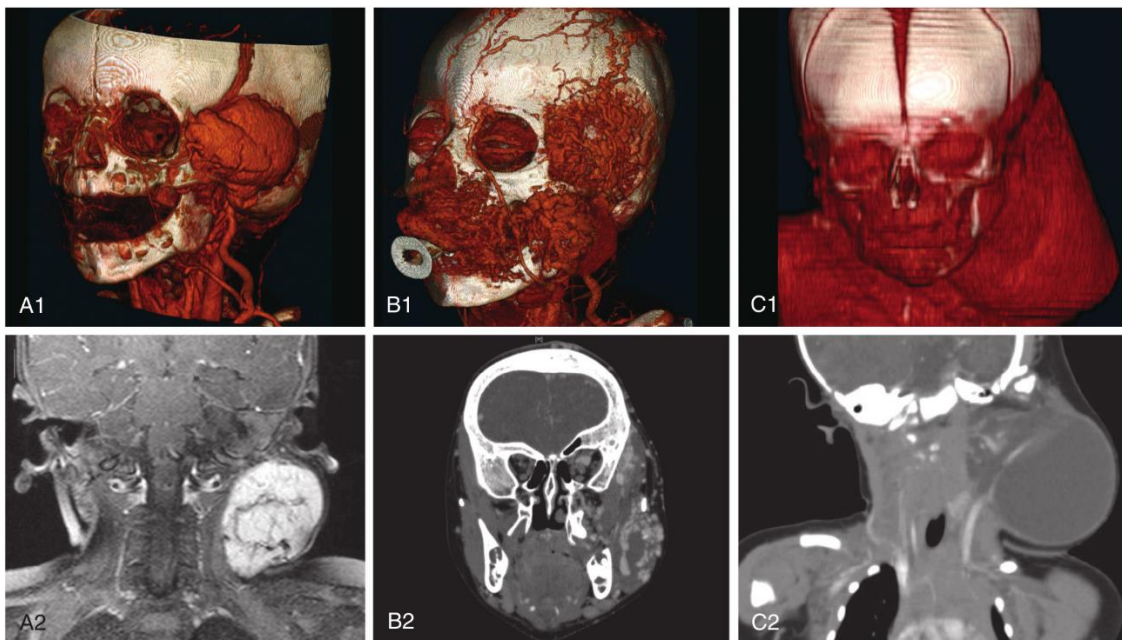


FIGURE 199-4. Computed tomographic (top row) and coronal computed tomography (bottom row) vascular anomaly imaging. **A**, Deep hemangioma of infancy. **B**, Venous malformation. **C**, Lymphatic malformation.

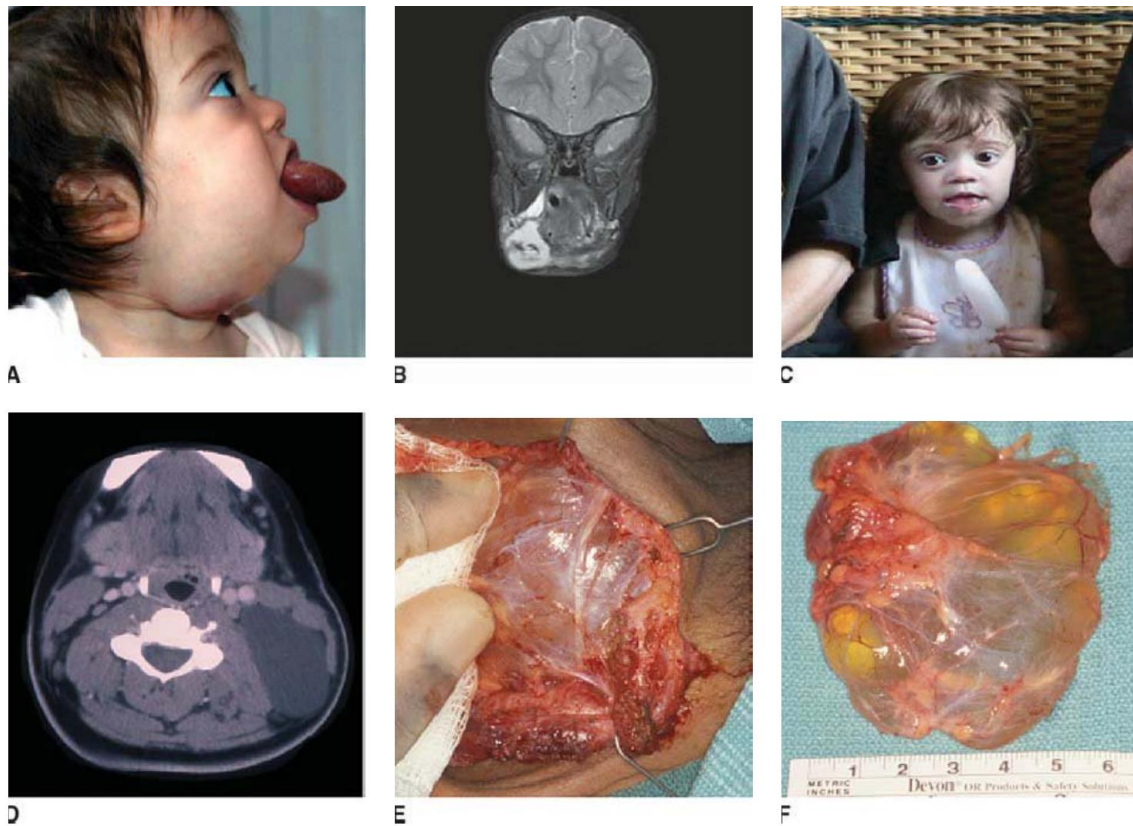


Figure 104.10 **A:** Combined microcystic and macrocystic malformation of the tongue, floor of mouth, and anterior neck. **B:** MRI with gadolinium of the same patient. **C:** Postoperative picture after excision of cervical disease and tongue reduction. **D:** CT scan of neck demonstrating macrocystic malformation of the lateral neck. **E** and **F:** intraoperative pictures demonstrating the appearance of a macrocystic lymphatic malformation.

**TABLE
104.1**

MRI CHARACTERISTICS OF VASCULAR ANOMALIES

Anomaly Type	T1-Weighted	T2-Weighted	Contrast	Gradient
Hemangioma	Soft tissue mass, isointense or hypointense, flow voids	Lobulated soft tissue mass, increased signal, flow voids	Uniform intense enhancement	High-flow vessels within and around soft tissue mass
Venous malformation	Isointense to muscle, possible high-signal thrombi	Septated soft tissue mass, high signal, signal voids (phleboliths)	Diffuse or inhomogeneous enhancement	No high-flow vessels
Lymphatic malformation	Septated soft tissue mass, low signal	Soft tissue mass, high signal, fluid levels	Rim enhancement or no enhancement	No high-flow vessels
Arteriovenous malformation	Soft tissue thickening, flow voids	Variable increased flow voids	Diffuse enhancement	High-flow vessels throughout abnormal tissue

Dubois J, Garel L. Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. *Pediatr Radiol* 1999;29(12):879–893.

- Complications:
 - **Infection:**
 - Cutaneous involvement of lymphatic malformation is associated with spontaneous lymphatic leakage from pathologic vesicles.
 - Bacteria can readily enter via cutaneous vesicles and quickly spread through the tissues affected by lymphatic lesion.
 - Results in acute and recurrent cellulitis with a rapid enlargement of the malformation.
 - Leads to serious complications such as airway obstruction and life-threatening sepsis.
 - Aggressive broad-spectrum antimicrobial therapy with or without systemic steroid is mandatory
 - **Airway obstruction:**
 - Very serious complication caused by large lymphatic malformations of head and neck obstructing larynx and trachea.
 - It might results from infection, trauma or localized hemorrhage into cysts leading to rapid enlargement of the lesion.
 - **Dysphagia:**
 - From involvement of hypopharynx and esophagus.
 - **Cosmetic disfigurement:**
 - Lymphatic malformations frequently involve and distort facial skeleton.
 - Most evident in the mandible with large bilateral suprahyoid lesions.
 - **Lymphocytopenia:**
 - Patients with large bilateral or microcystic lymphatic malformations can have significant lymphocytopenia that involves T cells.
 - Lymphocytopenia is not related to lymphocyte sequestration within the malformation.

- Treatment:

○ **Observation:**

- Indicated for favorably staged lymphatic malformations such as unilateral macrocystic posterolateral infrahyoid malformations (Stage I).
- Lesion may regress spontaneously if not complicated with infection during observation period.
- Systemic antibiotics and steroids are used to treat severe acute inflammation and infection.
- Indications of intervention:
 - Airway obstruction
 - Recurrence infection
 - Interfering with feeding
 - Interfering with speech
 - Cosmetic appearance.



FIGURE 199-25. Spontaneous resolution of macrocystic posterior cervical lymphatic malformation. **A**, Initial appearance at less than 6 months of age. **B**, Partial resolution of malformation at 9 months of age. **C**, Complete resolution by 1 year of age.

○ **Sclerotherapy:**

- Effective mainly for macrocystic malformations (>2cm).
- Ineffective for microcystic lesions.
- Involves percutaneous needle aspiration of macrocystic spaces under fluoroscopic guidance with subsequent injection of sclerosing agent.
- 40% of patients will have improvement of symptoms.
- When initial response to any sclerosing agent is unsatisfactory, treatment is repeated in 6 weeks to 3 months intervals.
- Sclerosing agents:

OK-432:

- Preferred sclerosing agent due to its low toxicity and scarring risks.

- **Doxycycline**

- **Bleomycin:**

- Toxicity lead to pulmonary fibrosis.

- **Ethanol:**

- Toxicity lead to permanent nerve injury.

- Complications of sclerotherapy:
 - Post-injection fibrosis and scarring.
 - Skin blistering and loss.
 - Erythema and pain at injection site
 - Fever
- **Surgery:**
 - Surgical excision is the most common treatment modality.
 - Goals:
 - Total excision with preservation of vital structures.
 - Subtotal excision results in more persistent post-op problems but may be necessary to preserve vital structures.
 - Best outcome with excision of localized macrocystic lesions in the posterior-inferior neck and parotid and submandibular regions.
 - Lymphatic malformations are infiltrative and involve nerves, vessels, and muscles.
 - In large facial malformations, facial nerve presents in unusual location and greater length.
 - Intra-op electromyographic nerve mapping can aid in these difficult dissections.
 - Post-op drainage of surgical site is essential following resection because significant lymphorrhea can occur.
 - Recurrence rate is 40% for incomplete excision and 20% for complete excision.
 - High risk of complications with:
 - Large mixed suprahyoid lesions (surgery should be staged).
 - Post-sclerosing agent injections due to fibrosis.
 - Complications include:
 - Infection
 - Bleeding
 - Cranial nerve injury
 - Great vessels injury
 - Fistula formation
 - Tongue edema and airway obstruction



FIGURE 199-26. Preoperative facial nerve mapping prior to lymphatic malformation excision.



FIGURE 199-27. Microcystic lymphatic malformation, mucosa and deep tongue involvement.

- **Venous Malformations:**
- Low-flow vascular malformations.
- Consist of either localized or diffuse ectatic veins with abnormal irregular venous channels.
- Types:
 - **Superficial:**
 - Intradermal or subcutaneous.
 - **Deep:**
 - Intramuscular or intraosseous.
- Clinical picture:
 - Bluish compressible mass with no palpable thrill or audible bruit.
 - Usually asymptomatic.
 - Present at birth, slowly enlarge over time, and may become inflamed and painful during puberty and trauma.
 - Enlarge when the lesion is in a dependent position, with Valsalva maneuver or high cardiac output states.
 - Intracranial and skull base extension can occur, particularly with periorbital lesions.
- Complications:
 - Congestion
 - Thrombosis and phleboliths
 - Distal emboli (large lesions)
 - Intralesional consumptive coagulopathy
- Diagnosis:
 - **Duplex US:**
 - Shows slow blood flow.
 - **CT scan:**
 - Frequently demonstrate calcified phleboliths.
 - Pathognomonic for venous malformations.
 - Assess the extent of the lesion.
 - **MRI:**
 - Bright signal on T2.
 - Assess the extent of the lesion.

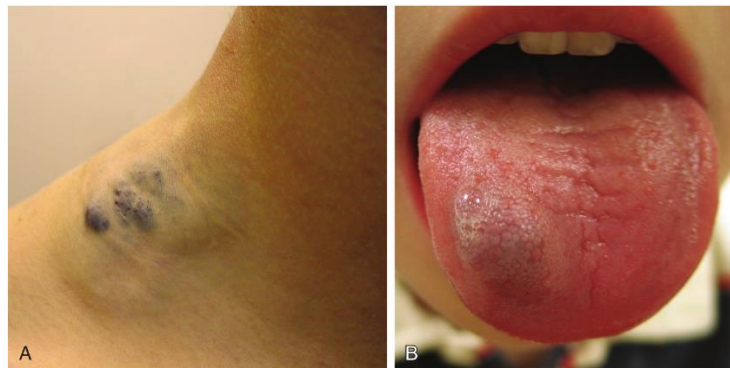


FIGURE 199-28. Venous malformation appearance. **A**, Skin. **B**, Tongue.



Figure 104.9 Photograph of a young boy with an extensive venous malformation of the right cheek and lower lip.

- Treatment:

- **Observation:**
 - For asymptomatic lesions.
 - Anti-platelet and anti-inflammatory agents can help controlling pain associated with phlebolith formation.
- **Sclerotherapy:**
 - Indicated for extensive lesions to preserve function when vital structures are involved.
 - Same sclerosing agents used for lymphatic Malformations.
- **Laser:**
 - Nd:YAG laser can be used for superficial lesions or the superficial component of deep lesions.
- **Surgery:**
 - Surgical excision results in excellent outcomes when lesion is localized and accessible.
 - Extensive lesions are not amenable to resection or can be only partially resectable.
 - Combined therapy with sclerosis and surgery is usually necessary to minimize blood loss and morbidity.

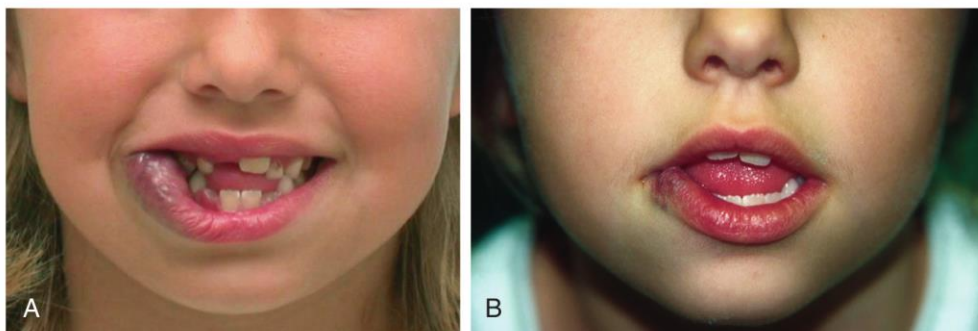


FIGURE 199-30. Venous malformation treatment with interstitial laser. **A**, Lip prior to treatment. **B**, Lip after two laser treatments.

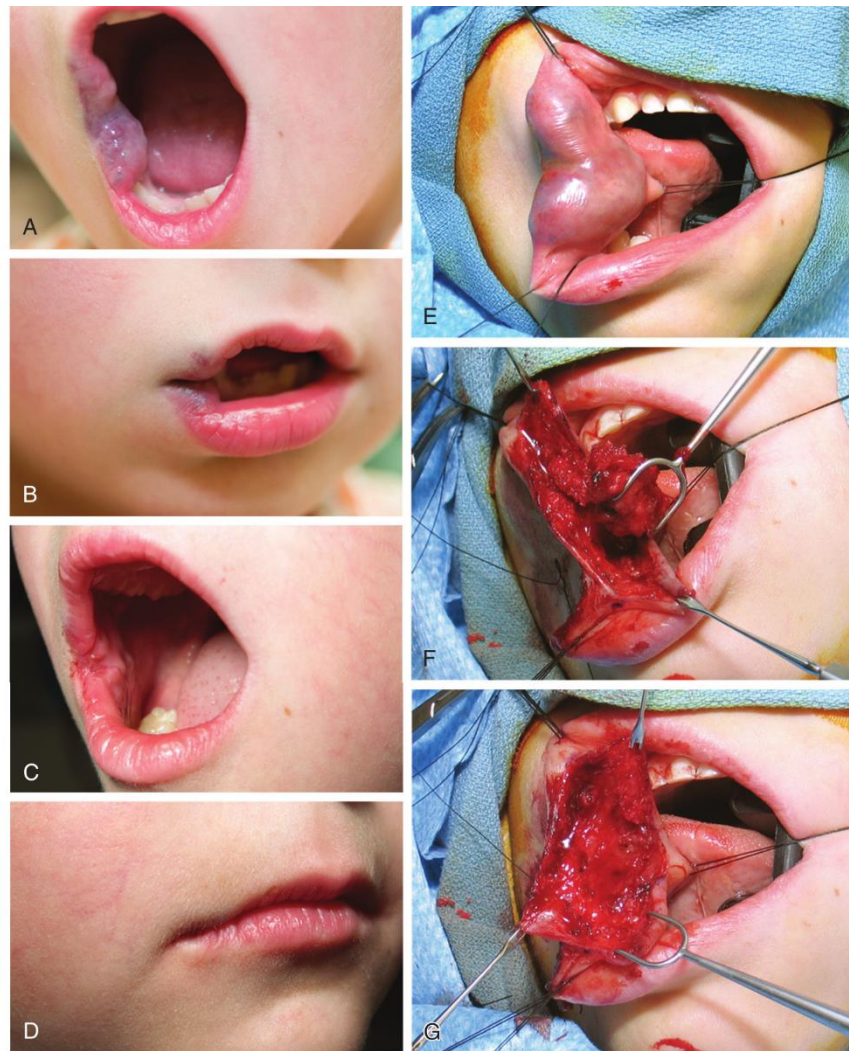


FIGURE 199-29. Excision of localized venous malformation after glue embolization. **A**, and **B**, Preoperative appearance. **E** to **G**, Intraoperative photos demonstrate the venous malformation containing glue, being excised, and being primarily reconstructed. **C**, and **D**, One-month postoperative appearance.

- **Capillary Malformations:**
- Low-flow vascular malformations.
- Superficial collection of ectatic vessels located in the dermis.
- Present at birth.
- Appear as pink macules on the face or nuchal region.
- Children with Craniofacial involvement should undergo MRI brain and ophthalmic examination to rule out associated syndromes.
- Types:
 - **Salmon patch (Nevus simplex):**
 - Fade or disappear during childhood.
 - Also called stork bite and angel kiss.
 - Appears in the face or nuchal region.
 - **Port-wine stains (Nevus flammeus):**
 - Always persists and doesn't fade.
 - Follows CN-V distribution on face.
 - Present at birth as flat, pink, macular lesions and become raised, nodular, deep red with time.
 - Association:
 - **Sturge-Weber syndrome:**
 - Present with:
 - Facial port-wine stain in ophthalmic distribution of trigeminal nerve.
 - Glaucoma
 - Vascular eye abnormalities
 - Ipsilateral intracranial vascular malformation.
 - Develop progressive neurologic problems (seizures, migraines, stroke-like episodes, learning difficulties or mental retardation, visual field impairment or hemiparesis).
 - **Wyburn-Mason syndrome:**
 - Port-wine facial stains with unilateral AVM of retina and intracranial optic pathway.



A



B

Figure 104.8 **A:** Photographs of a salmon patch on the posterior neck of a patient. **B:** Photograph of a port-wine stain covering the upper right half of the face.

- Treatment:
 - **Observation:**
 - For asymptomatic lesions.
 - **Laser:**
 - Pulsed-dye laser (PDL) device mitigate the red coloration of port-wine stains and prevent progression to more darkly colored and raised lesions.
 - Nd:YAG laser is used for capillary lesions that have thickened and become darker with age.
 - **Surgery:**
 - Surgical debulking can be done for certain lesions such as hypertrophied lip and eyelid tissue with skin grafting.
- **Arteriovenous Malformations (AVM):**
- High-flow vascular malformations.
- Congenital vascular lesions associated with arteriovenous shunting.
- Originate from arteriovenous channels that failed to regress during development.
- Most dangerous vascular anomalies.
- Always multiple and vary in diameter and length.
- Clinical picture:
 - Small and stable in childhood.
 - Rapid growth in 2nd–3rd decade of life with puberty or trauma.
 - Warm, pulsatile lesion with skin discoloration.
 - Palpable thrill and audible bruit.
- Complications:
 - Bleeding
 - Cardiac hypertrophy
 - Cardiac failure
 - Stroke
- Diagnosis:
 - Doppler US
 - CT angiography
 - MRA
- Treatment:
 - **Observation:**
 - For asymptomatic small lesions.
 - **Embolization with surgical excision:**
 - Offers the best chance for cure.
 - Complete excision may be impossible because of location and extent of the malformation.
 - When lesions cannot be excised, palliative embolization may be appropriate to control symptoms.

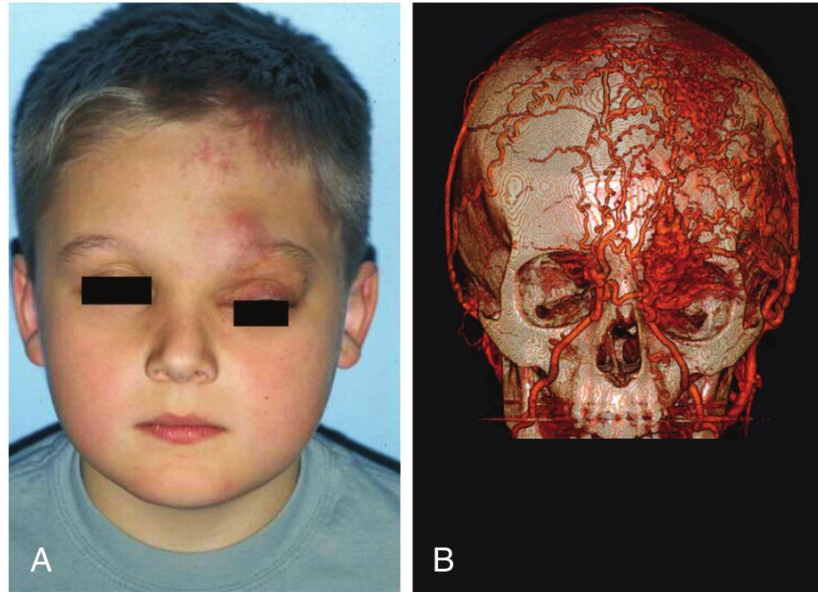


FIGURE 199-32. Extensive vasodilation associated with enlarging orbital/eyelid arteriovenous malformation. **A**, Clinical appearance. **B**, Computed tomographic angiography image with nidus in orbit and extensive vasodilatation. (**A**, Courtesy Dr. J. Gruss.)

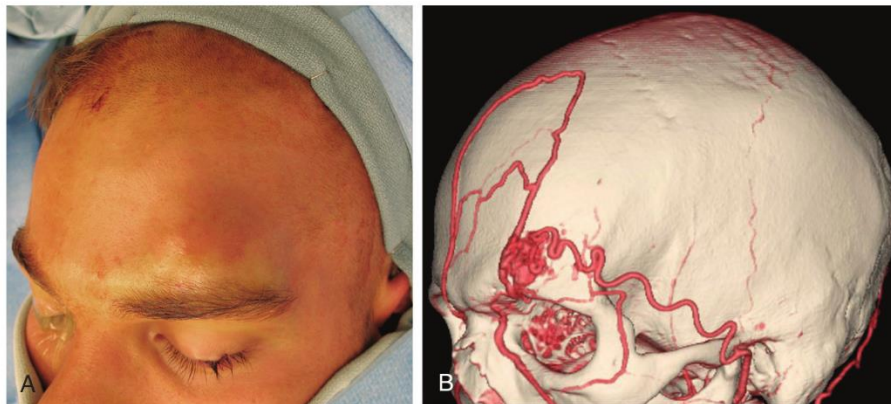


FIGURE 199-31. Computed tomographic angiography (**B**) and clinical appearance (**A**) of quiescent arteriovenous malformation nidus, left brow.

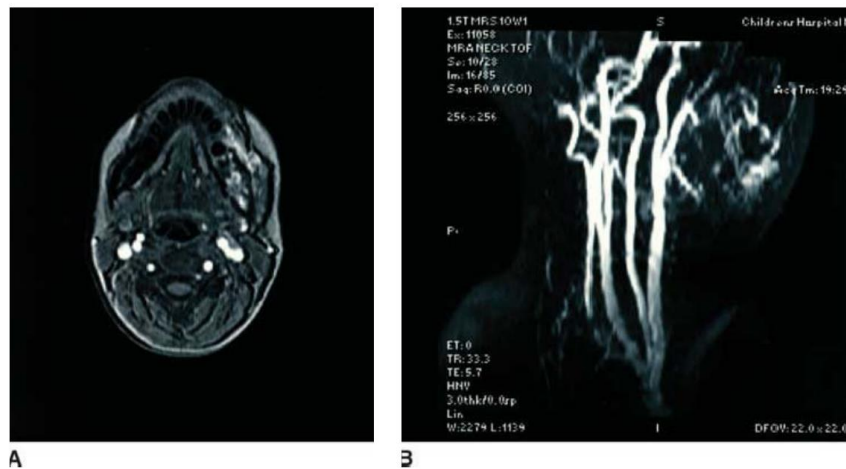


Figure 104.11 **A**: MRI and **(B)** MRA of the neck demonstrating an AVM of the left mandible.

Table 174-1 -- DISTINGUISHING CHARACTERISTICS

Hemangioma	Vascular Malformation
Usually not seen at birth	Always present at birth
Gender: Female > Male	Equal gender distribution
Race: More common in white	Equal between all races
Rapid growth and slow regression	Grows proportionately with the child
Firm and rubbery	Compressible
Rarely involves bone or cartilage	May cause significant hypertrophy and distortion of craniofacial skeleton

Table 174-2 -- MAGNETIC RESONANCE IMAGING CHARACTERISTICS OF VASCULAR ANOMALIES

	T1-Weighted	T2-Weighted	Contrast (gadolinium)	Gradient
Hemangioma	Soft-tissue mass, isointense or hypointense, flow voids	Lobulated soft-tissue mass, increased signal, flow voids	Uniform intense enhancement	High-flow vessels within and around soft-tissue mass
Venous malformation	Isointense to muscle, possible high-signal thrombi	Septated soft-tissue mass, high signal, signal voids (phleboliths)	Diffuse or inhomogeneous enhancement	No high-flow vessels
Lymphatic malformation	Septated soft-tissue mass, low signal	Soft-tissue mass, high signal, fluid/fluid levels	Rim enhancement or no enhancement	No high-flow vessels
Arteriovenous malformation	Soft-tissue thickening, flow voids	Variable increased flow voids	Diffuse enhancement	High-flow vessels throughout abnormal tissue

Tracheal Anomalies:

- **Tracheomalacia:**

- A normal framework resists collapse of the airway on expiration, when intrathoracic pressure exceeds intraluminal pressure.
- Weakness of Tracheal wall rigidity leads to dynamic collapse of the trachea during breathing, resulting in airway obstruction.
- Classification of Tracheal wall Rigidity Abnormalities:

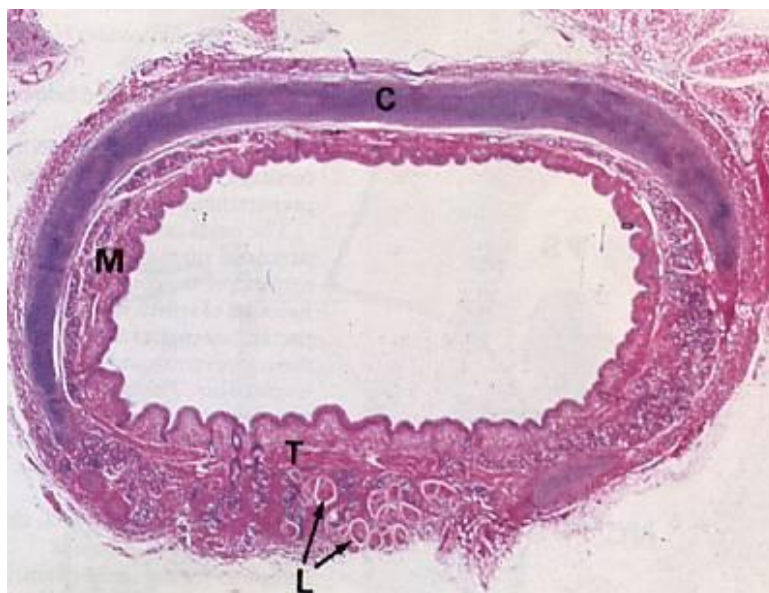
- 1. Tracheomalacia:**

- Tracheal collapse resulting from weakness of the anterior tracheal cartilaginous arc.
 - **Cartilage to muscle ratio in Tracheomalacia is reduced from normal ratio 4.5:1 to 2:1.**

- 2. Tracheal Dyskinesia:**

- Tracheal collapse resulting from dysfunction (low tone) of posterior membranous trachea.

- Tracheomalacia most commonly occurs in intra-thoracic trachea (distal 1/3) resulting in Expiratory stridor.
- Concurrent bronchomalacia is common and occurs in 30% of patients.
- Associated conditions:
 - Cardiovascular abnormalities (20-58%)
 - Bronchopulmonary dysplasia (up to 52%)
 - Gastroesophageal reflux disease (~50%)
 - Subglottic stenosis and laryngomalacia
 - Neurologic impairment (8-48%)
 - Severe developmental delay (26%)



- Types of Tracheomalacia:

- 1. **Primary (Congenital):**

- Most common form.
- Arises from intrinsic weakness in trachea itself.
- No underlying cause.
- Occurs mainly in premature babies.
- Association with TEF (Tracheoesophageal fistula):
 - High rate of symptomatic Tracheomalacia persists after repair of TEF.
- Typically present with mild to moderate symptoms.
- Resolve spontaneously with age.

- 2. **Secondary (Acquired):**

- Segmental collapse of a portion of the airway.
- Occurs secondary to known causes.
- Prolonged ET intubation.
- Tracheostomy.
- External compression (vascular, mediastinal tumors, Cardiac enlargement).

- Clinical picture:

- Most cases are mild with symptoms begin at 5-6 months of age with Expiratory stridor and cough, often described as barking or brassy.
- Inspiratory stridor is suggestive of extra-thoracic collapse.
- Moderate cases will have frequent episodes of wheezing or expiratory stridor exacerbated by exertion or upper respiratory tract infection.
- Patients with mild to moderate Tracheomalacia will have clinical improvement in 6-12 months with resolution by 2 years of age as the cartilage matures.
- Patients with severe disease will have a greater tendency toward cyanotic episodes and reflex apnea, with high mortality rates if not treated.

- Diagnosis:

- **Direct Laryngobronchoscopy:**

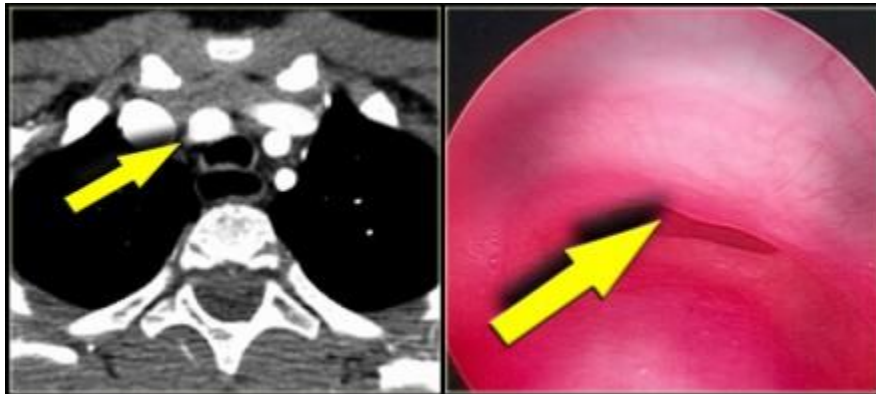
- Gold standard.
- Performed in a spontaneously ventilating patient.
- **Decrease in luminal (A-P) diameter >50% at end expiration is considered diagnostic for Tracheomalacia.**

- **CT Angiography:**

- Done to evaluate for external vascular compression of trachea in secondary Tracheomalacia.



Figure 87.11 Rigid endoscopy demonstrating tracheomalacia secondary to vascular compression imposed by the innominate artery in an infant.



- Management:
 - **Conservative:**
 - Indicated for mild to moderate cases as most cases resolved after 2 years of age.
 - Consists of:
 - Antibiotic for recurring pneumonia.
 - Humidified oxygen.
 - Intermittent steroids.
 - Proton pump inhibitors.
 - Pulmonary physiotherapy.
 - Positive pressure ventilation.
 - Prolonged intubation.

- **Surgical Intervention:**

- Indicated for severe cases not improving with conservative measures.
- Options:
 - **Tracheostomy:**
 - Long-standing option.
 - Long trach tubes is used as a stent to open the distal trachea.
 - **Intra-luminal Stenting:**
 - Counteract collapse of airway during expiration.
 - Not recommended due to its complications:
 - High rate of migration.
 - Negative effect on mucociliary clearance.
 - Granulation tissue formation.
 - Difficulty in stent removal or repositioning.
 - It should not be used when other surgical options are available.
 - Airway stents are acceptable for short-term postoperative support of tracheal lumen.
 - Tracheotomy with long distal cannula is considered a better alternative than stenting for Tracheomalacia unresponsive to aortopexy.
 - **Extra-luminal Splinting:**
 - Designed to resist external compression.
 - In contrast to intraluminal stents, extraluminal splints do not disrupt mucosa.
 - Less effective in treatment of distal tracheomalacia.
 - **Treatment of underlying cause of external tracheal compression:**
 - Repair of TEF.
 - Aortopexy.
 - **Tracheal Resection and Primary Anastomosis:**
 - Indicated in short segment tracheal collapse:
 - <30% of tracheal length in pediatrics.
 - <50% of tracheal length in adults.
 - **Slide Tracheoplasty:**
 - Indicated in long segment tracheal collapse:
 - >30% of tracheal length in pediatrics.
 - >50% of tracheal length in adults.

- **Congenital Vascular Rings:**

- Congenital anomalous configuration of the arch or associated vessels that surrounds the trachea and esophagus and forms complete or incomplete ring around them.
- Causes airway obstruction by direct tracheal compression and development of tracheomalacia.

- Types:

o **Incomplete vascular rings (60%):**

1. Aberrant High-riding Innominate Artery **(40%)**
2. Aberrant Retro-esophageal Right Subclavian Artery **(20%)**
3. Aberrant Left Pulmonary artery sling **(5%)**

o **Complete vascular rings (40%):**

1. Double Aortic Arch
2. Right Aortic Arch

- **Aberrant High-riding Innominate Artery:**

- Most common congenital vascular anomaly causing symptomatic tracheal compression.

- Innominate artery appears to originate from a more distal and leftward position on the arch causing compression on anterior wall of trachea.

- Clinical picture:

- o Stridor that improves on prone position.
- o Self-limiting most of the time.

- Diagnosis:

o **CT Angiography and MRA:**

- Confirm the diagnosis.

o **DLP:**

- Pulsatile extrinsic compression of right anterolateral mid-trachea.
- Endoscopic diagnosis is confirmed by compressing the vessel with the rigid bronchoscope tip and simultaneously palpating the right brachial or radial pulse, the intensity of the pulse will decrease.

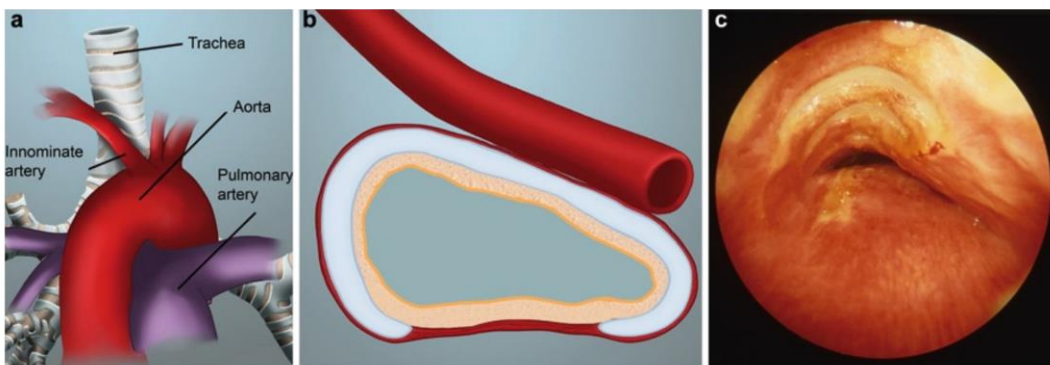


Fig. 13.2 Innominate artery compression of the right anterolateral aspect of the trachea: (a) Diagram: The anterior extrinsic compression is oblique from right to left and from cranial to caudal. (b) Transverse section: The trachea is compressed ante-

riorly by the innominate artery. (c) Endoscopic view: 15-year-old adolescent with persistent airway symptoms. Please note the right anterior extrinsic compression and the denuded cartilage resulting from intubation trauma

- **Aberrant Retro-esophageal Right Subclavian Artery:**
- Right subclavian vessel originates from descending part of aortic arch on the left side and runs posteriorly to esophagus to reach the right side of the neck.
- Associated with ipsilateral non-recurrent laryngeal nerve.
- Clinical picture:
 - o Asymptomatic mainly as it does not significantly impinge on the posterior membranous trachea.
 - o Dysphagia lusoria may occur late in life when the vessel becomes rigid from arteriosclerosis.
 - o Some patients may present with stridor.
- Diagnosis:
 - o **Barium esophagram:**
 - Reveals a filling defect in the esophagus.
 - o **CT Angiography and MRA:**
 - Confirm the diagnosis.
 - o **DLP:**
 - Pulsatile extrinsic compression of posterior wall of trachea.

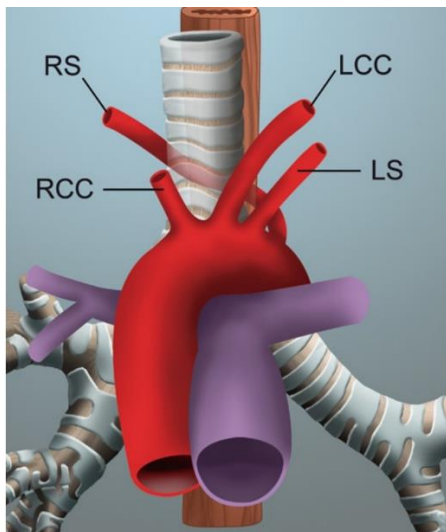
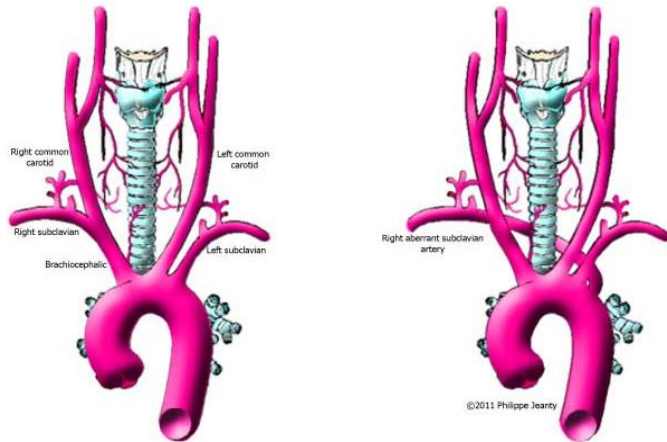


Fig. 13.3 Aberrant right subclavian artery: Diagram: The artery originates from the aortic arch on the left and passes behind the oesophagus, where it is recognised endoscopically by a pulsating extrinsic compression of the posterior wall RS, LS=Right and left subclavian arteries; RCC, LCC=Right and left common carotid arteries



- **Anomalous Left Pulmonary Artery Sling:**
- Left pulmonary artery originates from the proximal portion of right pulmonary artery (instead of from main pulmonary artery) and proceeds between the trachea and esophagus before reaching left lung.
- Complete tracheal rings and long-segment tracheal stenosis are seen in 50% of patients.
- **Clinical picture:**
 - o Stridor that worsening during lower airway infection.
- **Diagnosis:**
 - o **CT Angiography and MRA:**
 - Confirm the diagnosis.
 - o **DLP:**
 - Pulsatile extrinsic compression of lower posterior tracheal wall and right main bronchus.
 - Complete tracheal rings may be seen.

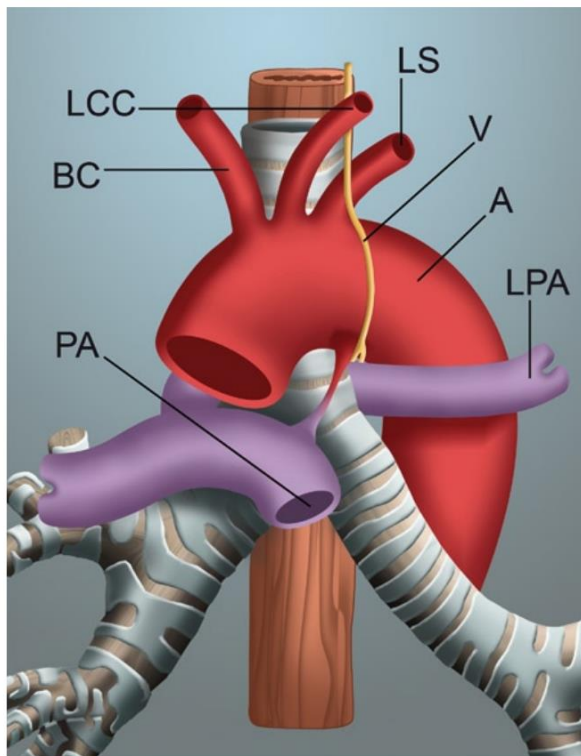


Fig. 13.4 Anomalous left pulmonary artery sling: It originates from the right pulmonary artery, encircles the lower trachea, and passes between the trachea and oesophagus. PA=Pulmonary artery; LPA=Left pulmonary artery; A=Aorta; V=Vagus nerve; BC=Brachiocephalic trunk; LCC=Left common carotid artery; LS=Left subclavian artery

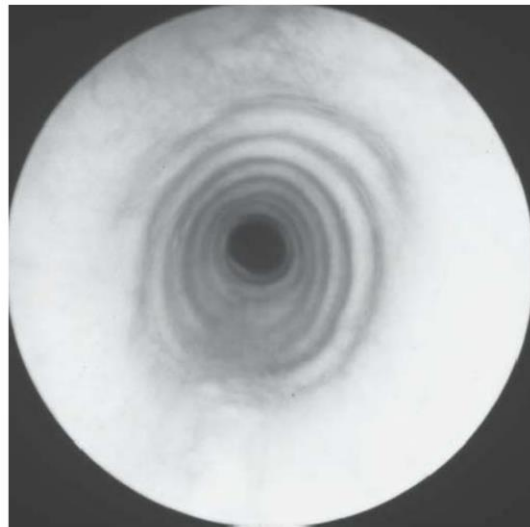


Figure 89.4 Endoscopic view of complete tracheal rings.

- **Double Aortic Arch:**
- Most common type of complete vascular ring.
- Development of paired aortas arches in which the right arch is larger and passes behind esophagus to join the left arch and form a left-sided descending aorta.
- Produces severe tracheomalacia and stridor since birth.

- **Right Aortic Arch:**
- Less common type of complete vascular ring.
- Can be either:
 - o Right aortic arch with retro-oesophageal left subclavian artery and left ligamentum arteriosum.
 - o Right aortic arch with the mirror-image branching and left ligamentum arteriosum.

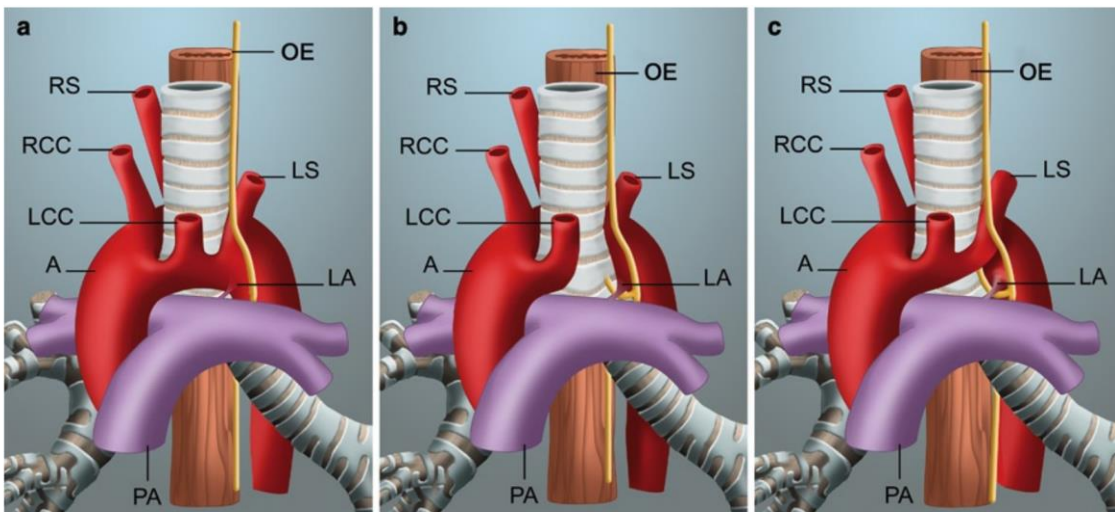


Fig. 13.5 Main vascular rings responsible for extrinsic compression of the trachea: (a) Double aortic arch: It encircles both the trachea and oesophagus. The right arch is usually predominant. (b) Right aortic arch with retro-oesophageal left subclavian artery and left ligamentum arteriosum. (c) Right aortic arch

with the mirror-image branching and left ligamentum arteriosum. OE=Oesophagus; RS, LS=Right and left subclavian arteries; RCC, LCC=Right and left common carotid arteries; A=Aorta; LA=Ligamentum arteriosum

- **Indications of surgery in vascular rings:**
- **Absolute:**
 - o Prolonged intubation
 - o Reflex apnea
 - o Failure of medical treatment
- **Relative:**
 - o Severe dysphagia
 - o Failure to thrive,
 - o Recurrent bronchopulmonary infections
 - o Associated asthma
 - o Cystic fibrosis
 - o Exercise intolerance

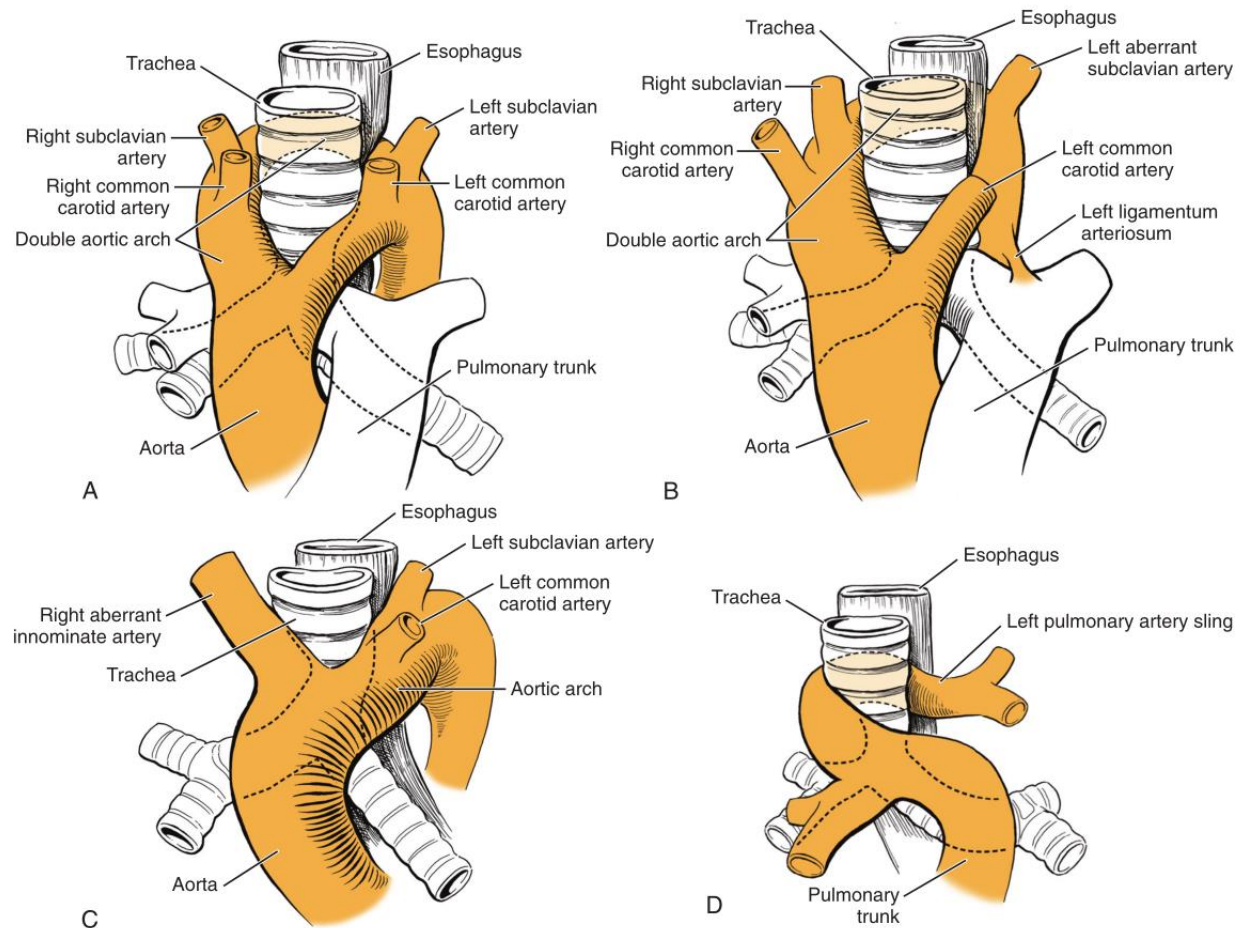


FIGURE 206-3. **A**, Double aortic arch. **B**, Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum. **C**, Aberrant innominate artery. **D**, Left pulmonary artery sling.

- **Tracheal Stenosis:**

- Classification:

- 1. Congenital:**

- The usual U-shaped tracheal cartilage is replaced by O-shaped complete cartilaginous tracheal rings.
- Patients with long-segment congenital tracheal stenosis are more likely to have early respiratory distress.
- **Most commonly associated with Pulmonary artery sling (50%).**



- 2. Acquired:**

- Results from endoluminal scarring or collapse.
- Secondary to prolonged intubation, tracheostomy, previous surgery, inhalational chemical burn injury, trauma, infection, relapsing polychondritis or Wegener granulomatosis.

Box 206-I. CANTRELL AND GUILD STRUCTURAL CLASSIFICATION

Type 1: Generalized hypoplasia of the entire trachea

Type 2: Funnel stenosis: normal proximal trachea with distal narrowing to carina

Type 3: Segmental stenosis with up to three rings involved

TABLE 206-I. Anton-Pacheco Functional Classification

Type	Symptoms/Malformations
Mild	Occasional or no symptoms
Moderate	Respiratory symptoms without respiratory compromise
Severe	Respiratory compromise
Subtype A	No other associated malformations
Subtype B	Associated malformations

- Clinical picture:
 - Symptoms correlated mainly to the diameter of the most obstructive segment.
 - Ranging from mild symptoms to severe respiratory distress.
- Diagnosis:
 - Direct Laryngobronchoscopy.
 - CT scan.
- Management (Same as SGS):
 - **Conservative:**
 - Indicated for mild symptoms.
 - Require serial follow ups with DLB or/and CT scan.
 - Catch-up phenomenon:
 - Occurs in patients with complete tracheal rings in which the normal diameter of tracheal lumen was reached by the age of 9 year.
 - **Endoscopic:**
 - Indicated for symptomatic acquired tracheal stenosis.
 - Options:
 - Laser.
 - Balloon dilation.
 - Mitomycin-C
 - **Open Repair:**
 - Indications:
 1. Congenital tracheal stenosis.
 2. Failed endoscopic approach.
 - Options:
 - **Augmentation Tracheoplasty:**
 - Vertical midline division of tracheal stenosis followed by augmentation of anterior segment with costal cartilage graft.
 - **Segmental Resection and Reanastomosis:**
 - Preferred method for patients with Short-segment tracheal stenosis:
 - <30% of tracheal length in pediatrics.
 - <50% of tracheal length in adults.
 - **Slide Tracheoplasty:**
 - Preferred method for patients with long-segment tracheal stenosis:
 - >30% of tracheal length in pediatrics.
 - >50% of tracheal length in adults.

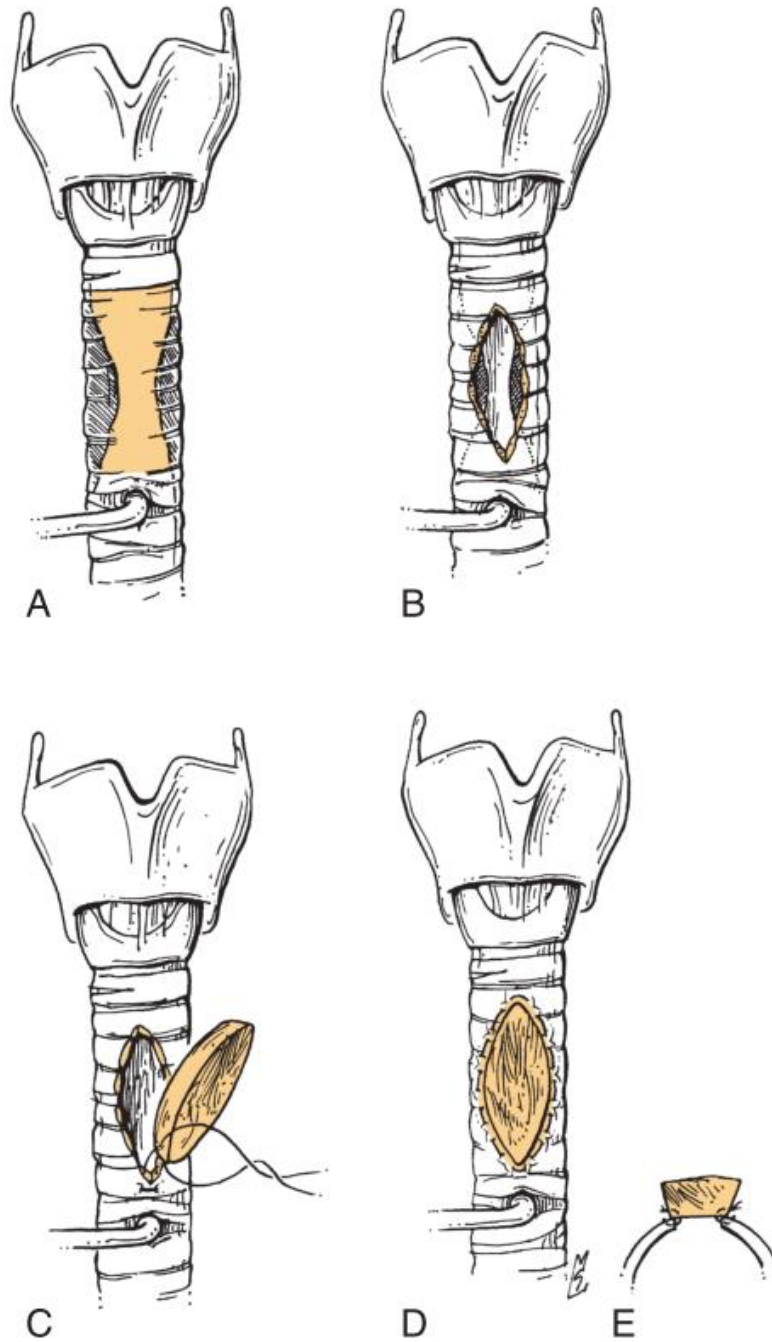


FIGURE 206-7. Cartilage graft placement for augmentation of tracheal stenosis. **A**, Area of stenosis. **B**, Stenosis excised. **C** and **D**, Placement of graft. **E**, Surgical result. Note the graft placement in the lumen.

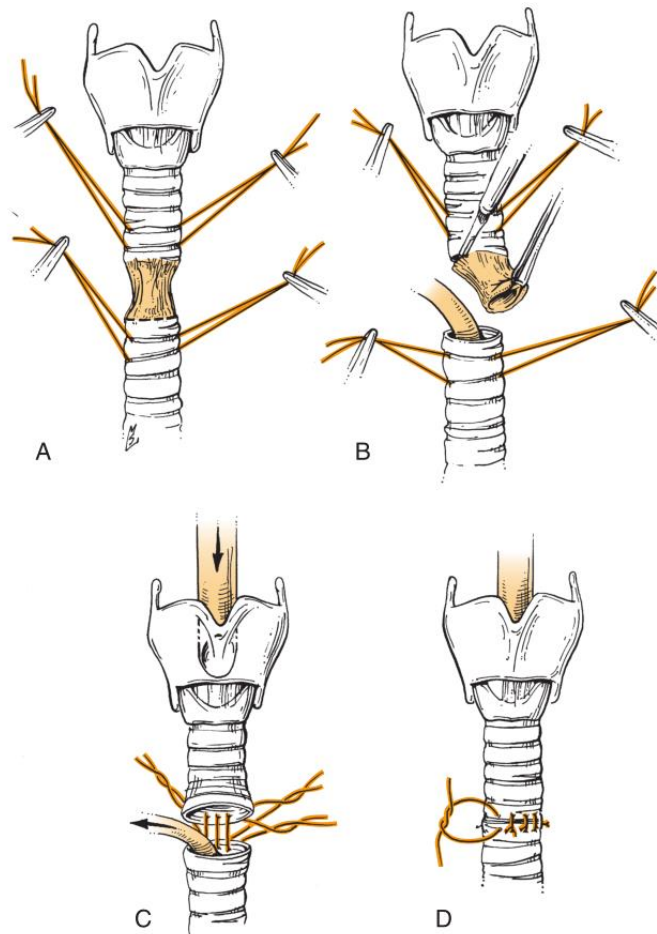


FIGURE 206-8. A, Area of stenosis. B, Resection of stenosis with intubation of trachea distally. C, Interrupted absorbable sutures placed circumferentially and reintubated from above (D).

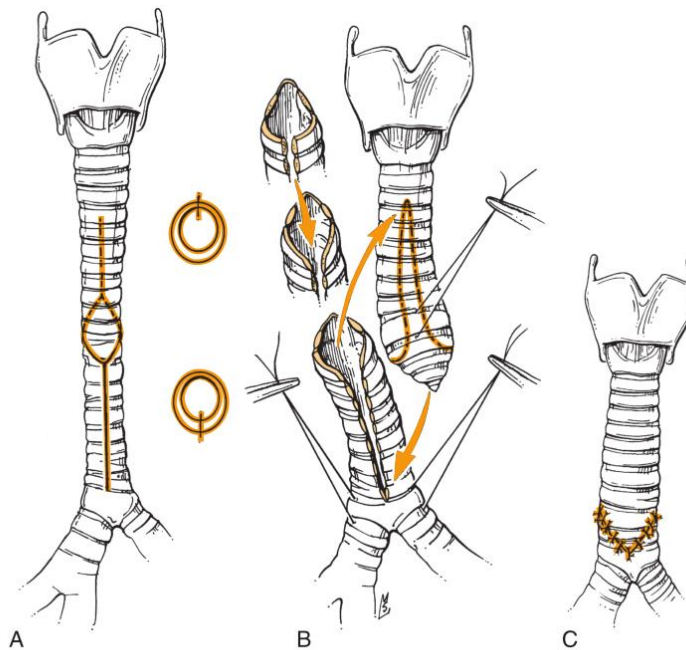


FIGURE 206-9. Slide tracheoplasty. A, Area of stenosis. B, The trachea is divided on an angle and then vertically, anteriorly, and posteriorly. C, Surgical result.

Neonatal Nasal Obstruction:

- Newborns are preferential nasal breathers for 6–20 weeks (variable).
- **Clinical picture:**
 - Stertor
 - Rhinorrhea
 - Nasal flaring
 - Cyclical/Paradoxic cyanosis (**Bilateral nasal obstruction**)
 - Worse with feeding, improve with crying
 - Tachypnea
 - Chest retractions
 - Difficulty with feeding
 - Failure to thrive
- **Diagnosis:**
 - Assessment for dysmorphisms (telecanthus, broad nasal bridge, nasal pits), ocular discharge, periocular infection/edema.
 - Trial of passing size 6 French catheter through both nostrils.
 - Anterior Rhinoscopy
 - Nasal endoscopy
 - Oral exam (palate arching or clefting)
 - Imaging:
 - CT is the preferred study for most cases of neonatal nasal obstruction.
 - MRI is indicated in midline nasal masses with potential intracranial connection.
- **Differential Diagnosis of neonatal nasal obstruction:**
 - Neonatal Rhinitis (**Most common**)
 - Septal deviation (Birth trauma)
 - Dacryocystocele
 - Congenital Nasal Pyriform Aperture Stenosis (CNPAS)
 - Midnasal stenosis
 - Choanal atresia (**Most common congenital nasal anomaly**)
 - Congenital nasal mass:
 - Dermoid (**Most common congenital nasal mass**)
 - Glioma
 - Encephalocele
 - Hamartoma
 - Hemangioma
 - Rhabdomyosarcoma
 - Neurofibroma

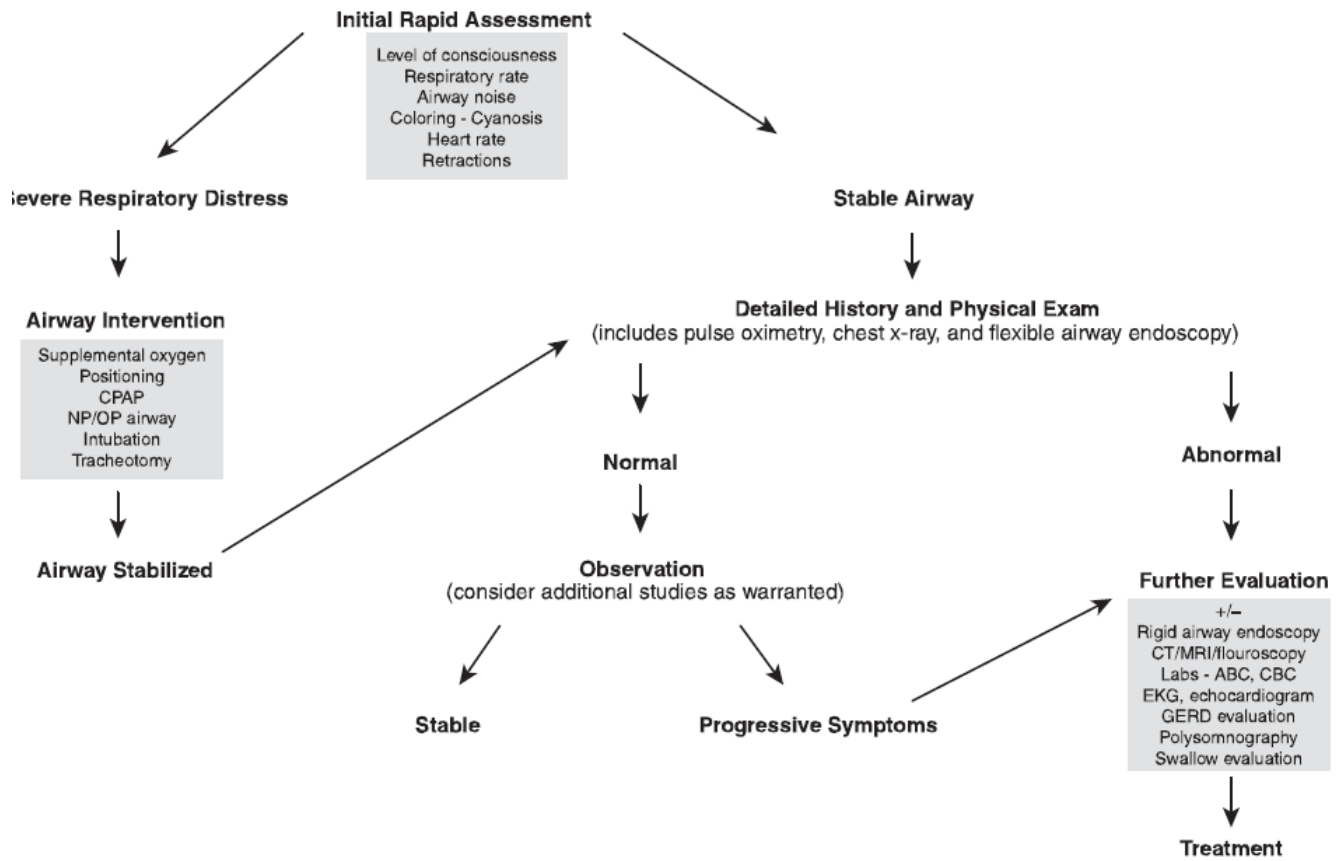


Figure 88.1 Algorithm for the evaluation and treatment of neonatal airway obstruction.

TABLE 10–1. Differential Diagnosis of a Unilateral Pediatric Nasal Mass

Vascular	Juvenile nasopharyngeal angiofibroma (JNA) Hemangioma Arteriovenous malformation (AVM)
Infectious/inflammatory	Polyp Rhinolith
Neoplastic/mass	Encephalocele Glioma Neurofibroma
Drug-related	
Idiopathic	
Congenital	Nasolacrimal duct cyst
Autoimmune/allergic	
Traumatic	Foreign body
Endocrine	

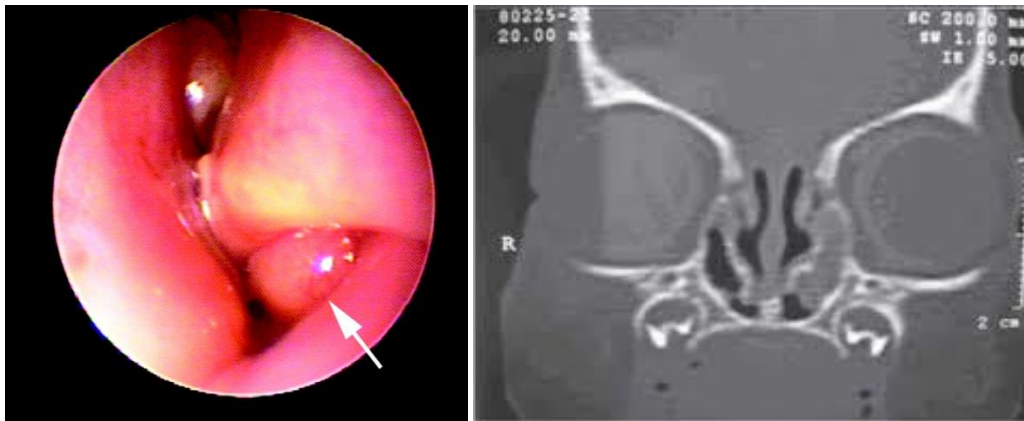
Neonatal Rhinitis:

- **Most common cause of neonatal nasal airway obstruction.**
- **Pathophysiology:**
 - Idiopathic
 - Maternal estrogen
 - Infectious (congenital syphilis, chlamydia, other),
 - Early allergic rhinitis (high prevalence of familial atopy)
 - Ciliary dyskinesia
 - Hypothyroidism
- **Clinical picture:**
 - Abundant rhinorrhea (muroid)
 - Stertor
 - Tachypnea
 - Poor feeding
 - Diffuse nasal mucosal edema
 - Turbinate hypertrophy
 - Normal pyriform aperture
 - Patent choanae
- **Diagnosis:**
 - Considered after negative full investigation for nasal obstruction.
- **Treatment:**
 - Nasal saline drops
 - 0.1% Dexamethasone ophthalmic drops
 - GERD may also contribute to neonatal rhinitis and should be treated if suspected based on the history and physical examination.



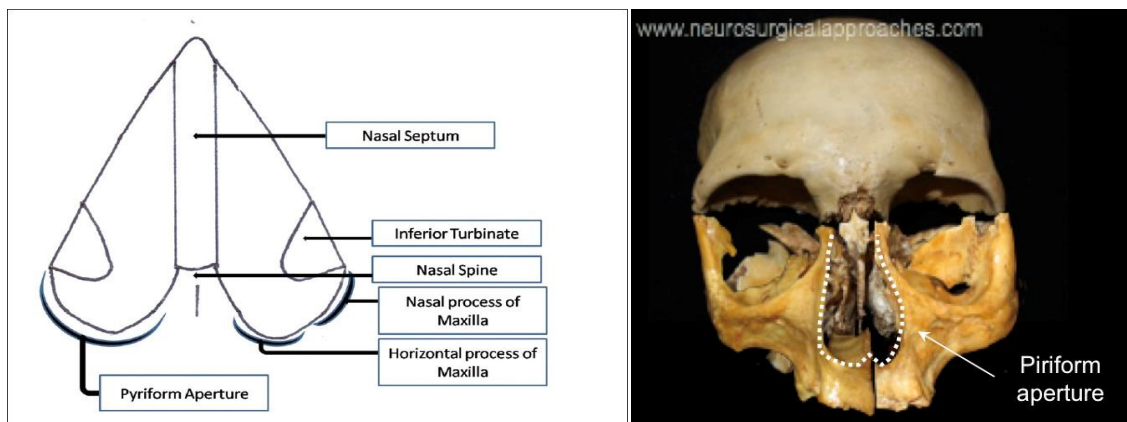
Dacryocystocele (Nasolacrimal Duct Cyst):

- **Pathophysiology:**
 - Congenital failure of opening of the distal nasolacrimal duct with cyst formation from accumulation of secretions.
- **Clinical picture:**
 - Usually asymptomatic
 - Epiphora
 - Symptoms of nasal obstruction
 - Respiratory distress and difficulty feeding in infants with large bilateral obstructive cysts.
- **Diagnosis:**
 - Examination of inferior meatus (Anteriorly)
 - CT sinuses or MRI
- **Treatment:**
 - Most (85%) spontaneously resolve by 9 months of age
 - Endoscopic marsupialization for symptomatic cysts:
 - Presence of infection
 - Respiratory obstruction
 - Feeding difficulties

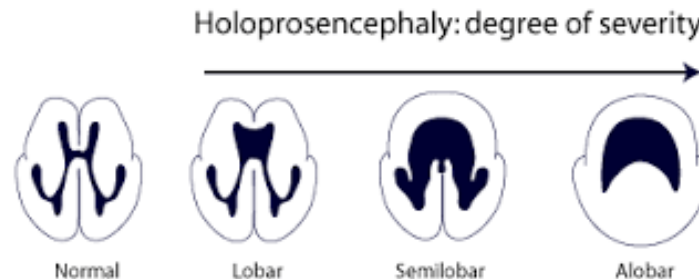


Congenital Nasal Pyriform Aperture Stenosis (CNPAS):

- Rare congenital nasal anomaly.
- 1/4 of incidence of choanal atresia.
- Occurs secondary to bony overgrowth of nasal process of the maxilla.
- **Anatomy:**
 - Pyriform aperture is the anterior most and narrowest portion of the nasal cavity.
 - Pear or heart shaped opening bounded by:
 - **Superior:** Nasal bone
 - **Lateral:** Nasal process of Maxilla.
 - **Inferior:** Horizontal process of Maxilla, Anterior nasal spine of maxilla.



- **Physiology:**
 - Very small changes in the cross-sectional area may greatly increase nasal airway resistance and produce symptoms similar to bilateral choanal atresia.
- Most commonly occurs as an isolated anomaly.
- Also could be part of holoprosencephaly which includes:
 - Solitary median maxillary central incisor (**50% Association**)
 - Brain and pituitary hypoplasia.
 - Hypertelorism
 - Craniofacial anomalies



- **Clinical picture:**

- Stertor
- Nasal flaring
- Cyclical/Paradoxical cyanosis
 - Worse with feeding, improve with crying
- Tachypnea
- Chest retractions
- Difficulty with feeding
- Failure to thrive

- **Diagnosis:**

1. Failure of passing size 6 French catheter beyond **1cm**.
2. Anterior Rhinoscopy reveals a bony obstruction at the nasal vestibule that does not allow passage of a flexible endoscope.
3. CT Sinuses:
 - Gold standard test.
 - Establish definitive diagnosis.
 - Measurement on axial CT at level of inferior meatus in full-term infant:
 - Each pyriform aperture width of **<3 mm** is diagnostic.
 - Total pyriform aperture width (between both nasal processes of maxilla) of **<8 mm** is diagnostic.
 - Normal Total pyriform aperture width is **> 11mm**.
 - CT may also show nasolacrimal ducts encased in the narrowed bony pyriform.
4. MRI Brain:
 - Done in the presence of Solitary median maxillary central incisor to rule out pituitary hypoplasia and other CNS abnormalities.



- **Treatment:**

○ **Conservative for mild cases:**

- Humidification, suctioning, decongestants.
- Feeding in upright.
- McGovern Nipple (oral airway).
- Apnea monitor at home.
- Family training in CPR.
- **If patient is able to tolerate conservative management, Nasal airway is likely to improve with growth within six months of birth.**

○ **Surgical repair for severe cases:**

▪ Endoscopic Trans-nasal approach:

- Difficult.
- Drilling the maxillary overgrowth.
- High risk of soft-tissue trauma.
- Width of the aperture should be large enough to pass a 3.5 mm ET tube.

▪ Sub-labial approach:

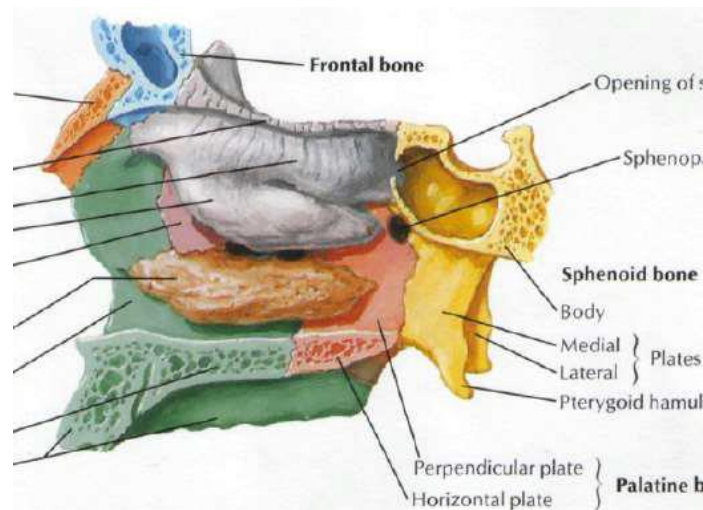
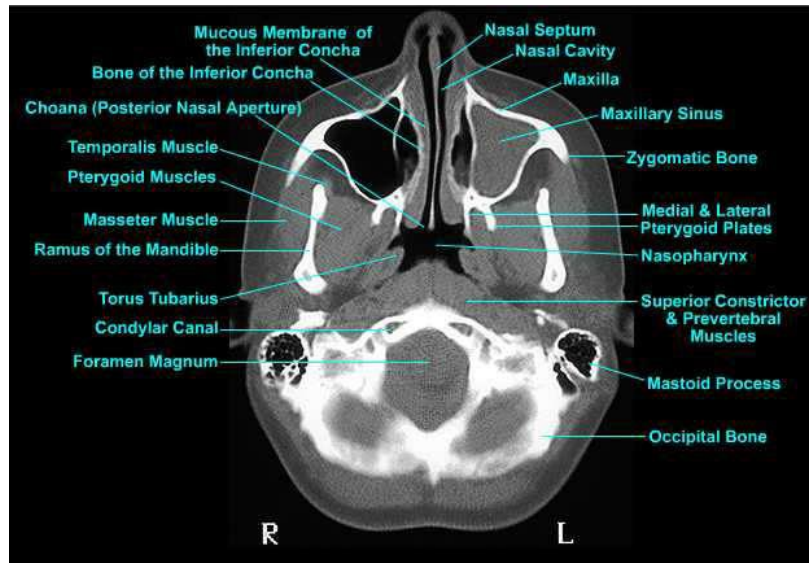
- Safe and effective
- Technically easier in the small nose of an infant.
- Provides excellent exposure without damaging the nasovestibular skin or causing scar tissue formation.
- After preservation of the mucosa, abnormal bone can be removed with drills.
- A injury to the tooth buds, contiguous soft tissues, and nasolacrimal duct.
- Width of the aperture should be large enough to pass a 3.5 mm ET tube.
- Stents are left in place no longer than 1 week postoperatively.
- Patients should be given antibiotics while the stents are in place.
- Surgery does not seem to effect nasal or facial growth, with follow-up to about 1 year.



Choanal Stenosis and Atresia:

- **Most common congenital nasal anomaly.**
- Congenital narrowing or closure of nasal airway at posterior choanae.
- Occurs in 1 per 10,000 births
- Female-to-male preponderance of 2:1.
- Two thirds (**70%**) of choanal atresia is unilateral (Right-side predominance).
- **Pathophysiology:**
 - Choanal atresia results from failure of nasobuccal membrane to recanalize in the embryonic phase of intrauterine development.
 - If the nasobuccal membrane recanalizes incompletely, choanal stenosis results.
- **Teratogenic syndromes causing bilateral Choanal Atresia:**
 - Methimazole embryopathy.
 - Carbimazole embryopathy.
- **Types:**
 1. Mixed bony and membranous (**70%**)
 2. Bony (**30%**)
 3. Membranous (**Rare**)
- **Boundaries of Atretic plate:**
 1. Undersurface of body of sphenoid bones superiorly (Below the posterior end of middle turbinate).
 2. Medial pterygoid lamina laterally (Lateral wall of nasopharynx).
 3. Vomer medially.
 4. Horizontal portion of palatine bone inferiorly (Soft palate).
- There is not just a membrane across the posterior choanae but also medialisation of the pterygoid plates and lateral wall of the nose.
 - Lateral wall is the principle challenge of choanal atresia surgery, as most surgical corrections tend to address the septum and atretic plate only.





- **Anatomic deformities associated with choanal atresia include:**
 1. Bony atretic plate is situated in front of posterior bony septum.
 2. Narrow nasal cavity.
 3. Thick vomer.
 4. Thick medial pterygoid plates.
 5. High arched palate.
 6. Narrow nasopharynx.

- **Significant association between choanal atresia and other anomalies or syndromes such as:**
 1. CHARGE
 2. VATER
 3. Crouzon,
 4. Treacher Collins.
 5. Pfeiffer,
 6. Antley-Bixler,
 7. Marshall-Smith,
 8. Schinzel-Giedion,

- **CHARGE Syndrome:**
 - Also called Hall-Hittner syndrome.
 - Most common concurrent syndrome in bilateral atresia **(50%)**.
 - Associated with unilateral atresia in **(20%)**.
 - Mutations in CHD7 gene in chromosome 8 have been identified in 60% of patients diagnosed with CHARGE.
 - Majority of mutations are sporadic.
 - Consist of:
 - **Coloboma**
 - **Heart defects**
 - **Atresia of choanae**
 - **Retarded growth and development**
 - **Genitourinary hypoplasia**
 - **Ear anomalies/deafness**

Table 1 Diagnostic criteria for choanal atresia		
Major	Minor	Diagnosis
1. Ocular coloboma	5. Cardiovascular malformations	Typical CHARGE: four majors or three majors and three minor
2. Choanal atresia	6. Genital hypoplasia	
3. Characteristic ear abnormalities	7. Cleft lip/palate	
4. Cranial nerve abnormalities including SNHL	8. Tracheoesophageal fistula	
	9. Hypothalamo-hypophyseal dysfunction	
	10. Distinctive CHARGE facies	
	11. Developmental delay	

○ **Coloboma:**

- Up to 80% of CHARGE patients
- Essentially a hole in one of structures of the eye.
 - Affects iris, retina (most common), or both
- Unilateral or bilateral
- Vision may be normal or impaired



○ **Heart Defects:**

- 70-80% of patients with CHARGE
- Most common defect is tetralogy of Fallot (33%)
- Also PDA, ASD, and VSD

○ **Atresia of Choanae:**

- 50-60% of patients with CHARGE
- Most commonly bilaterally.
- High rate of mortality with coinciding bilateral atresia and cardiac anomalies.

○ **Retarded Growth and Development:**

- Usually normal length/weight at birth.
- If growth delayed due to cardiopulmonary issues may catch up once repaired.
- Often GH deficiency in kids; obesity in adults

○ **Genitourinary Hypoplasia:**

- Easier to recognize in males.
- Common defects:
 - Microphallus, penile agenesis, hypospadias
 - Cryptorchidism, bifid scrotum
 - Solitary kidney, hydronephrosis, renal hypoplasia, horseshoe/ectopic kidney, vesicoureteral reflux.

○ **Ear Anomalies/Deafness:**

- 80-100% of patients with CHARGE.
- External Ear:
 - Cup/lop-shaped.
 - Low set with decreased vertical height.
- Middle Ear:
 - Absent stapedius.
 - Hypoplastic incus/stapes.
 - Ossicle fixation.
 - Oval window atresia.
- Inner Ear:
 - Mondini malformation.
 - Absence of semi-circular canals
- Consider CT temporal bone if already going for CT sinus.
- Often candidate for cochlear implant.

- **Clinical Picture:**
- Presentation of choanal atresia depends on whether the obstruction is unilateral or bilateral.
- - **Unilateral Choanal Atresia:**
 - Typically present later (usually 5–24 months).
 - Unilateral obstruction and persistent nasal discharge.
 - Diagnosis can be delayed to adulthood and has been recognized after unsuccessful septal surgery.
 - **Bilateral Choanal Atresia:**
 - Typically present at birth with asphyxia neonatorum.
 - Bilateral Nasal obstruction considered in newborns with:
 - Tachypnea
 - Cyclical/Paradoxic cyanosis (worse with feeding, improve with crying)
 - Difficulty with feeding
 - Failure to thrive
 - Rhinorrhea
- **Diagnosis:**
 1. Absence of condensation in cold spatula test.
 2. Failure of passing size 6 French catheter beyond **3.5 cm**.
 3. Nasal Flexible endoscopy after suctioning confirms a blind-ending nasal passageway.
 4. CT Sinuses:
 - Gold standard test.
 - Establish definitive diagnosis.
 - Should be done after stabilizing the patient condition.
 - Done after suctioning and decongestion of nasal cavity.
 - Difficult to distinguish radiologically a membranous occlusion from mucus in unprepared nose.
 - **Advantages of CT scan:**
 - Identify the type of Atresia.
 - Allows measurement of the thickness of Vomer and Medial pterygoid.
 - Estimate size of nasopharynx.
 - Rule out other differential diagnosis nasal obstruction:
 - Pyriform aperture stenosis.
 - Midnasal stenosis.
 - Nasolacrimal duct cysts.
 - Midline congenital nasal mass (Encephalocele, glioma, dermoid, teratoma, hemangioma).

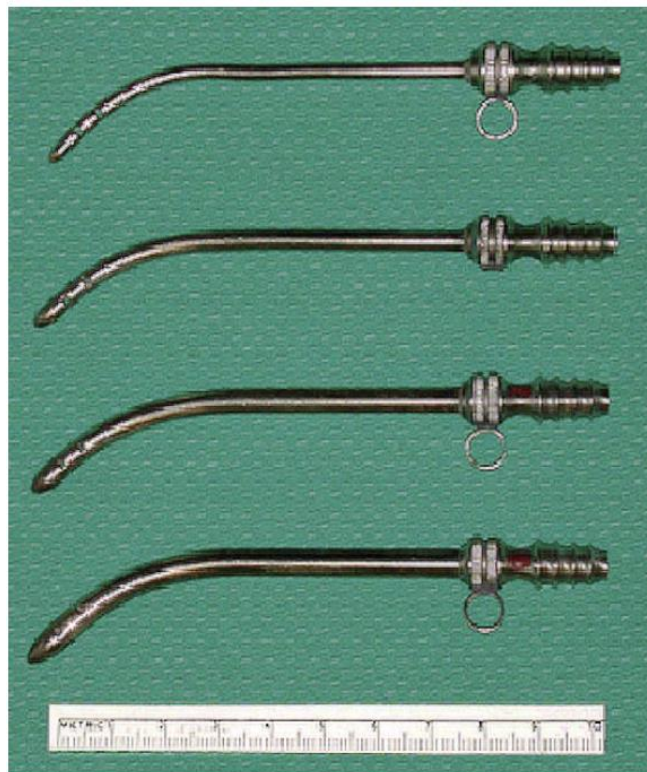
- Choanal space < 6 mm across in the coronal plane is consistent with choanal stenosis.
- Mean width of posterior choanal airspace, measured from lateral wall of nasal cavity to vomer:
 - 0.67 cm in newborns.
 - 0.86 cm by 6 years of age.
 - 1.13 cm by 16 years of age.
- Mean vomer width measures:
 - 0.23 cm in width and should not exceed 0.34 cm in children below age of 8 years
 - 0.28 cm in width and should not exceed 0.55 cm in children above age of 8 years
- **Management:**
- Bilateral choanal atresia presents as a medical emergency at birth.
 - **Emergency management:**
 - Consider an intraoral nipple with large opening (McGovern nipple) or oral airway.
 - Intubation for severe respiratory distress.
 - Tracheostomy may be needed if definitive surgery needs to be delayed due to other health concerns (e.g. cardiologic issues).



- **Surgical Repair:**
 - Timing:
 - Bilateral atresia must be addressed during first weeks of life to establish nasal airway and feeding.
 - Unilateral atresia may be repaired electively late until reaching age of one year.
 - Approaches:
 - Transnasal Puncture.
 - Transpalatal Repair.
 - Endoscopic Repair (Trans-nasal or Retrograde).
 - Balloon dilation for stenosis.

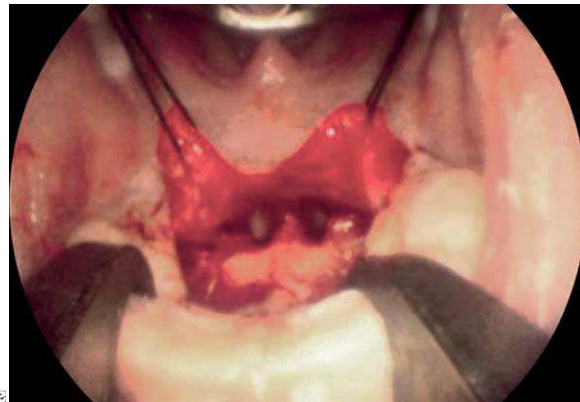
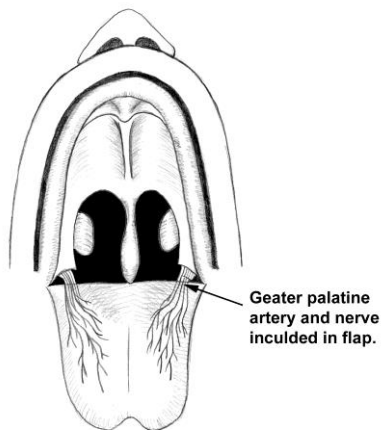
- **Trans-Nasal Puncture:**

- Long-used technique.
- Utilises Fearon dilators or urethral sounds to establish an airway.
- Puncture aimed at junction of septum and floor of nose, pointing down.
- Must direct the dissection inferomedially.
- Should be done under direct vision with mirror examining the posterior aspect of the atretic plate.
- Advantages:
 - Simple to perform quickly and effectively, even in very small infants.
 - Very useful technique for the treatment of membranous and thin bony stenosis.
- Disadvantages:
 - High recurrence rate of re-stenosis.
 - Difficult to pass the dilators if atretic plate is thick, and other techniques may be required, including drilling.



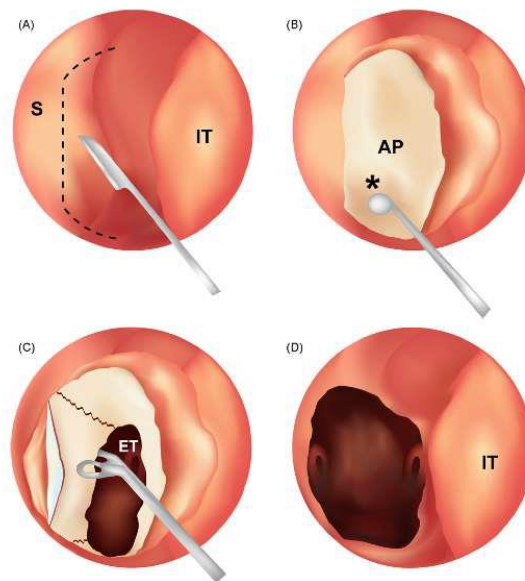
- **Trans-Palatal Repair:**

- Main technique alongside simple dilation until the advent of endoscopic techniques.
- A U-shaped flap in palate is made, with care to preserve the greater palatine artery and nerve.
- Anterior palatine bone, vomer and atretic plate are removed under direct vision with drill/rongeur.
- Stent placement
- Mucosa over palate used to resurface new choana.
- Advantages:
 - Provides good exposure.
 - Allows correction of all of anatomical abnormalities, including medialization of lateral wall at anterior pterygoid plates.
 - Success rate of more than 80%.
 - Decreased duration of stenting needed
- Disadvantages:
 - More operative time.
 - High risk of complications:
 - Blood loss.
 - Palatal flap breakdown and Fistula.
 - Stunt/alter palate growth
 - Crossbite abnormalities (50%).
 - High arched palate
 - Recommend only in kids > 5 years of age after most palatal growth finished.



- **Endoscopic Repair (Trans-Nasal or Retrograde):**

- Preferred surgical approach.
- 2.8- or 4-mm endoscope is used to visualize the atretic plate.
- Plate is perforated either under direct vision through nose (Trans-nasal) or observed via the postnasal space with 120 endoscope (Retrograde) especially in narrow nasal cavity.
- Create anterior mucosal flaps with knife over atretic plate.
- Dill out plate (diamond burr) focusing inferomedially on junction of atretic plate, hard palate, and vomer
- Remaining inferior to the posterior end of middle turbinate reduces the risk of skull base violation.
- After a passage is made into the postnasal space, opening is enlarged, typically by removing the posterior part of the septum with back-biting through-cutters, microdebrider or guarded drill.
- Most authors agree that it is essential to make a generous opening through the posterior nose, as there is a tendency for it to close over time.
- Success rates are generally high, with a low revision rate.



- Successful surgery defined as less than 50% re-stenosis.
- Risk factors for Re-stenosis in Choanal Atresia Repair:
 - Gastroesophageal reflux disease (GERD).
 - Age younger than 10 days at the time of surgery.
 - Insufficient postoperative endoscopic microdebridement of crustations.

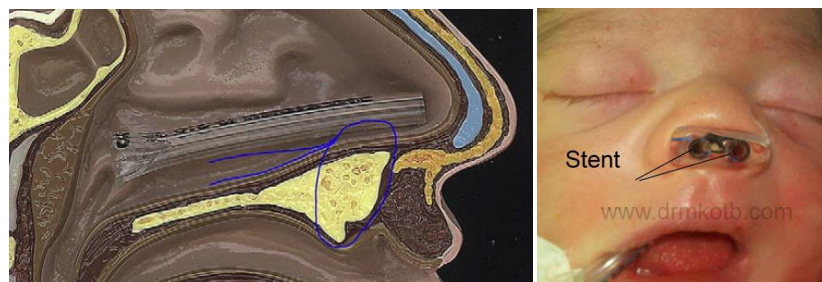
- **Aids to Surgery:**

○ **Mitomycin C:**

- Anti-proliferative/Anti-neoplastic/Alkylating agent.
- Inhibits fibroblast growth and proliferation.
- Used to prevent scar tissue and granulation formation.
- Intra-op topical application of Mitomycin C at concentration of 4 mg/ml for 4 minutes reduces scar formation and re-stenosis post airway surgeries.

○ **Stents:**

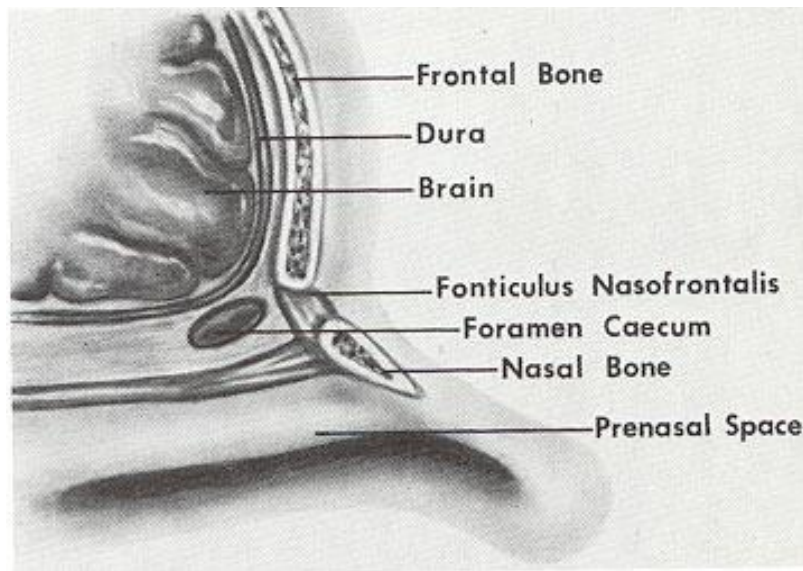
- Controversy of Stenting.
 - To Stent:
 - Favorable prognosis with less revision operations if stenting >12 weeks.
 - Not to Stent:
 - No clear-cut evidence that stents prevent stenosis after removal.
 - Stent complications:
 - Foreign body reaction.
 - Increase mucosal trauma and granulation. formation.
 - Alar erosions and pressure necrosis from columella stitch.
- In neonates, ETT size 3.0 or 3.5 is folded and inserted in both nasal cavity and a posterior fenestration is created.
- The fenestrated, folded end straddles the vomer and a mirror exam of the nasopharynx assures precise positioning of the stent.
- The stent is fixed in position with two independent 4 - 0 prolene sutures: one for each side of the nose.
- Each separate suture is placed around the alveolar ridge for extra tissue support, thus maintaining an inferior force on the stent minimizing any alar or septal pressure ulceration and/or necrosis.
- Cleaning consists of saline irrigation and suctioning as required.



- **Computer-Assisted Surgery:**
 - Most cases of choanal atresia can be safely managed simply with good quality pre-operative CT images.
 - CT-navigation is indicated in difficult cases with other skull base abnormalities.
- **Laser-Assisted Surgery:**
 - Lasers allow accurate dissection of tissue and may minimize bleeding with decreased scar formation.
 - Utilizing CO2, potassium titanyl phosphate (KTP) and, more recently, the contact diode laser (CDL).
 - Several deaths have occurred after nasal use of Nd-YAG lasers from air embolism, and there is probably little advantage over conventional instruments.
- **Post-op Care:**
 - Good post-op care will decrease the risk of infection, formation of granulation tissue and restenosis.
 - Frequent irrigation with saline drops.
 - Frequent gentle suction of the nasal cavities.
 - Topical corticosteroids starting on 2nd week post-op.
 - **Successful surgery defined as less than 50% re-stenosis.**
 - **Risk factors for Re-stenosis in Choanal Atresia Repair:**
 - Gastroesophageal reflux disease (GERD).
 - Age younger than 10 days at the time of surgery.
 - Insufficient postoperative endoscopic microdebridement of crustations.

Congenital Midline Nasal Masses:

- Rare lesions, occurs in 1 out of 40,000 births.
- Share aberrant skull base development, errors in formation of the anterior neuropore (primitive frontonasal region).
- Maintain a central nervous system (CNS) connection.
- Present at birth but some are not found until later in childhood or even adulthood when they become symptomatic
- Any unilateral nasal mass in a child should be evaluated for a congenital midline mass.
- **Embryology:**
 - During formation of skull base and nose, mesenchymal structures are formed from several centers which fuse and ossify.
 - Before their fusion, there are recognized spaces which are important in the development of congenital midline nasal masses:
 - Fonticulus frontalis
 - Prenasal space
 - Foramen cecum



- Fonticulus frontalis is space between the frontal and nasal bones which fuses with foramen cecum to create a separation between intracranial and extracranial structures.
- Prenasal space is between the nasal bones and the nasal capsule (precursor of the septum and nasal cartilages).

- **Congenital Midline Nasal Masses include:**

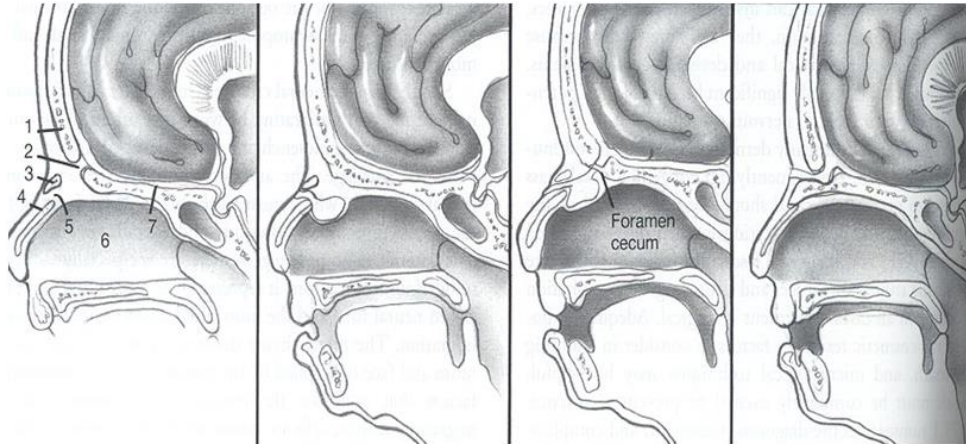
1. Nasal Dermoid
2. Glioma
3. Encephalocele

Nasal Dermoid:

- **Most common congenital midline nasal masses.**
 - o Nasal dermoid represent 8-10% of H&N dermoids.
 - o 1-3% of all dermoids
- Dermoid contain **Ectodermal** and **Mesodermal** components including adnexal tissue (Hair follicles, sweat glands, sebaceous glands).
 - o Presence of adnexal structures distinguishes dermoid from Epidermoid cysts, which do not have the same potential for transcranial extension.
 - o Unlike teratomas, dermoids do not show any components of endodermal origin.

Teratoma	Dermoid	Epidermoid
Ectoderm	Ectoderm	Ectoderm
Mesoderm	Mesoderm	-
Endoderm	-	-

- Intracranial connection in **25%** of nasal dermoids.
- Associated congenital anomalies in **5-45%** of nasal dermoids.
- **Development:**
 - o During normal development, a projection of dura protrudes through fonticulus frontalis or inferiorly into the prenasal space.
 - o This projection normally regresses but if it does not, dura can remain attached to epidermis and result in trapped ectodermal elements.
 - o Thus, dermoid occur when skin elements are pulled into prenasal space along with the regressing dural diverticulum.
 - o A sinus tract or mass may form anywhere along the course of the diverticulum from columella to anterior cranial fossa.
 - o Usually, nasal dermoids terminate in a single subcutaneous tract which can sometimes have hair protruding from the site (**pathognomonic**) .
 - o Intracranial extension of the tract most often passes through foramen cecum or cribriform plate to the base of anterior cranial fossa and adheres to the leaves of the falx cerebri extradurally.



Normal embryonic anatomy of nose and anterior skull base.

- 1, frontal cartilage;
- 2, fonticulus nasofrontalis;
- 3, nasal bone;
- 4, nasal cartilage;
- 5, prenasal space;
- 6, nasal capsule;
- 7, dura.

Normal closure of fonticulus, foramen cecum, with a sinus tract extending to the beginning of the prenasal space.

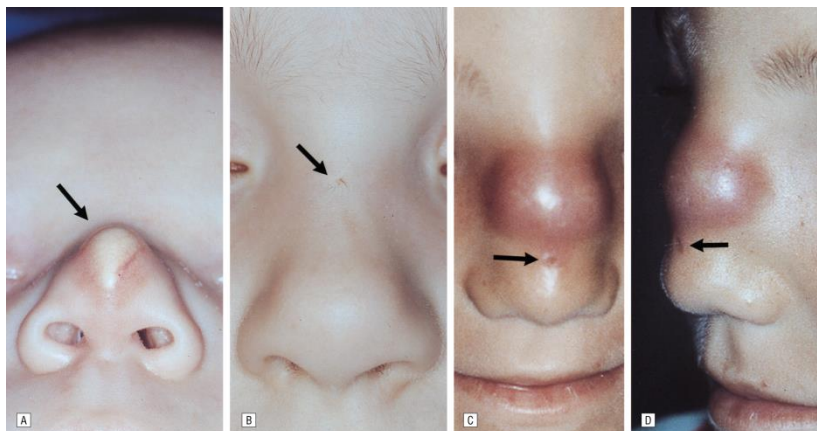
Patent foramen cecum with fistula from nasal dorsum through prenasal space.

Patent fonticulus and sinus tract to glabellar skin.

- **Clinical picture:**

o Presentation:

- Pit or sinus tract opening on the nasal dorsum:
 - Tuft of hair may protrude from pit
 - May secrete sebaceous material or pus if infected.
- Slow growing isolated cyst on nasal dorsum:
 - Firm
 - Non-compressible
 - Non-pulsatile
 - Does not trans-illuminate
 - Does not change in size with crying
 - Does not change in size with compression of bilateral internal jugular veins (**Negative Furstenberg test**).



- Location:

- Most commonly found as extranasal mass in nasal dorsum at the osteocartilaginous junction.
- May occur anywhere from glabella to columella.



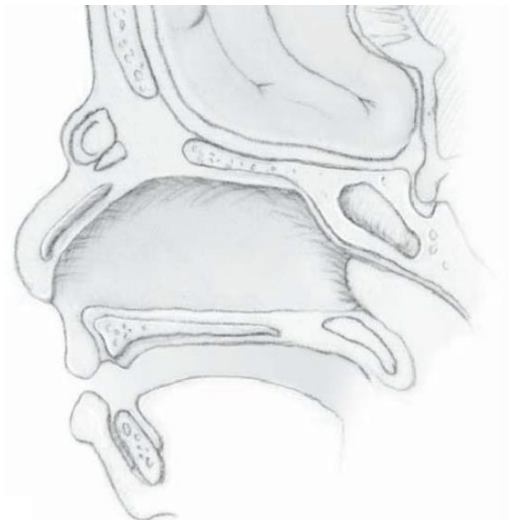
- Complications:

- Nasal obstruction
- Repeated infection
- Abscess formation
- Broaden nasal root
- Hypertelorism
- Destruction of nasal bones
- CSF leak
- Meningitis
- Brain abscess

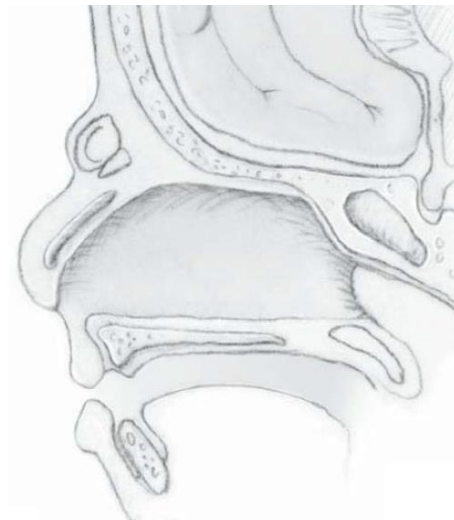


Glioma:

- Glial cells in a connective tissue matrix with or without a fibrous connection to the dura.
- Does not maintain a CSF filled subarachnoid connection.
- Intracranial connection in **15%** of gliomas.
- **Development:**
 - Abnormal closure of the fonticulus frontalis can lead to an ectopic rest of glial tissue being left extracranially.
 - Not always has an intracranial connection.



Intranasal glioma with connection to CNS.



Closure of bones to form glioma.

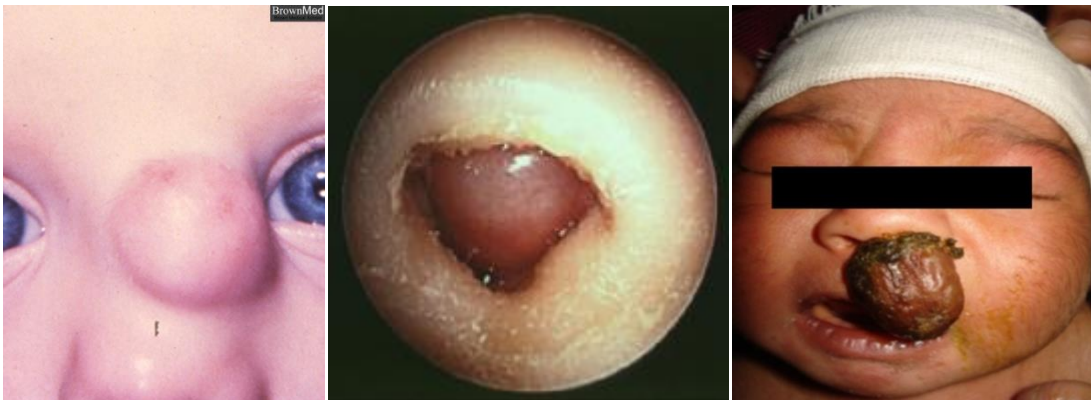
- **Clinical picture:**

○ Presentation:

- Slow growing isolated mass on nasal dorsum or intranasally:
 - Firm
 - Non-compressible
 - Non-pulsatile
 - Does not trans-illuminate
 - Does not change in size with crying
 - Does not change in size with compression of bilateral internal jugular veins (**Negative Furstenberg test**).
 - Does not associate with sinus tract.
 - Overlying skin may have telangiectasias.

○ Location:

- Most commonly found as extranasal mass along the nasomaxillary suture (**Not midline**) in 60%.
- Intranasal mass in 30%.
 - Most often arise from lateral nasal wall
 - Can pass a probe medial to it.
 - Less often arise from nasal septum.
 - Has more risk of intracranial connection.
- Combined (extranasal +intranasal) in 10%.



○ Complications:

- Nasal obstruction
- Broaden nasal root
- Hypertelorism
- Destruction of nasal bones
- CSF leak
- Meningitis
- Brain abscess

Encephalocele:

- Extracranial herniations of the meninges and/or brain.
- Maintains a CSF filled subarachnoid connection.
- Intracranial connection in **ALL** of Encephaloceles.
- Associated congenital anomalies in **30-40%** of encephalocele.

Development:

- Originate from faulty closure of fonticulus frontalis.
- Unlike gliomas, with encephaloceles the bone never closes, leaving a persistent intracranial-extracranial defect.

Types of Encephalocele by contents:

1. Meningocele:

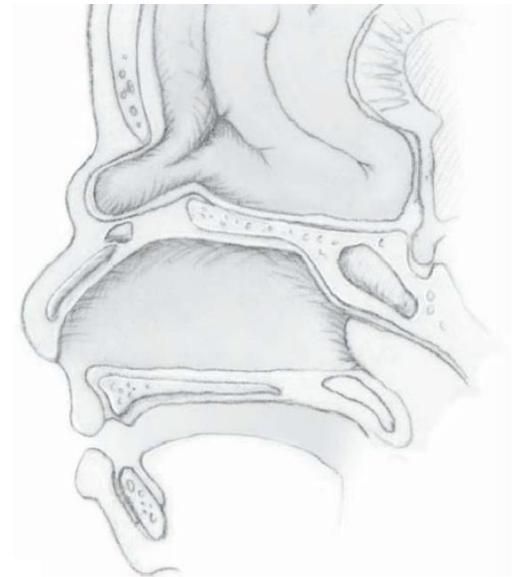
- Contains meninges only.

2. Meningoencephalocele:

- Contains meninges and brain elements.

3. Meningoencephalocystocele:

- Contains meninges, brain, and a part of the ventricular system.



Herniation of dura and glial tissue (encephalocele) through fonticulus.

Clinical picture:

○ Presentation:

- Isolated mass on nasal dorsum:
 - Soft
 - Compressible
 - Pulsatile
 - Trans-illuminate
 - Increase in size with crying or straining.
 - Increase in size with compression of bilateral internal jugular veins (**Positive Furstenberg test**) due to increased CSF pressure.
 - Does not associate with sinus tract.
- Intranasal mass that most often arise medially from nasal septum and can NOT pass a probe medial to it.



- **Types of Encephalocele by location of skull base defect:**

1. Occipital Encephaloceles: (75%)

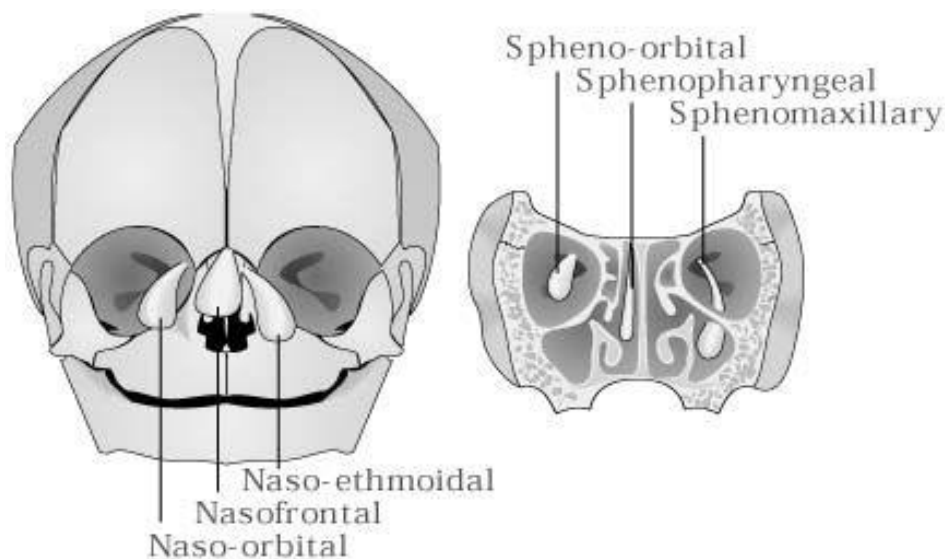
- Most common.
- Not related to the nose.

2. Sincipital Encephaloceles: (15%)

- Frontoethmoidal Encephaloceles.
- Present as Extranasal masses.
- Defect occurs between frontal and ethmoid bones at the foramen cecum immediately anterior to cribriform plate.
- Subtypes:
 - Nasofrontal (Glabellar lesion)
 - Nasoethmoidal (Lateral nose lesion)
 - Nasoorbital (Medial orbital wall lesion)

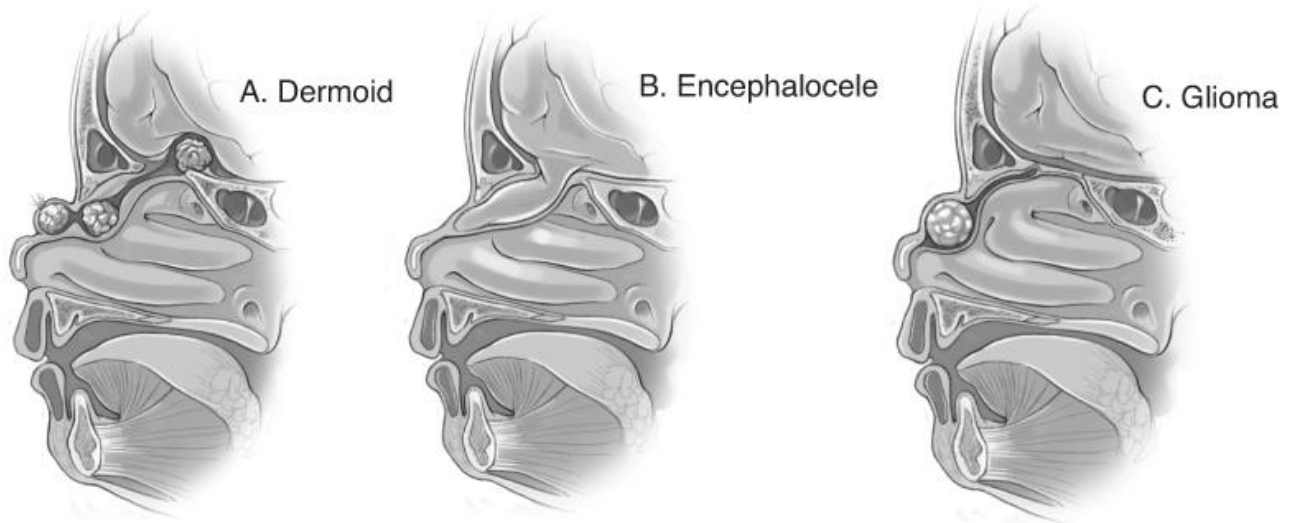
3. Basal Encephaloceles: (10%)

- Present as intranasal masses.
- Potential for airway obstruction in neonate.
- Subtypes:
 - Transethmoidal:
 - Most common subtype.
 - Extends through cribriform plate into nasal cavity medial to superior turbinate.
 - Sphenothmoidal:
 - Extends through cranial defect between posterior ethmoids and anterior sphenoid wall into nasopharynx.
 - Transsphenoidal:
 - Extends through an open craniopharyngeal canal into nasopharynx.
 - Sphenomaxillary:
 - Extends through superior and inferior orbital fissures to sphenomaxillary fossa



○ Complications of Encephaloceles:

- Nasal obstruction
- Broaden nasal root
- Hypertelorism
- Destruction of nasal bones
- CSF leak
- Meningitis
- Brain abscess



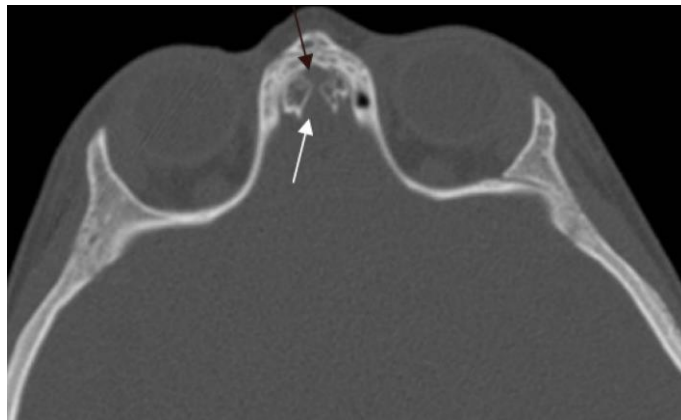
**TABLE
96.1**

CLINICAL FEATURES OF CONGENITAL NASAL MASSES

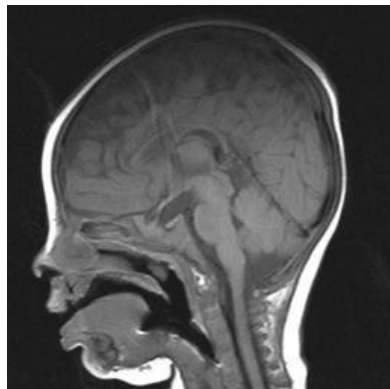
	Associated Anomalies	Midline	Cosmetic Defect	Infection Risk (Soft Tissue)	Meningitis Risk	Sinus Tract
Dermoid	S	U	Y	Y	Y	U
Glioma	N	S	S	N	Y	N
Encephalocele	S	S	S	N	Y	N
	Intracranial Communications	Intranasal Presentation	Compressible/Expandable	Skull Base Dehiscence	Low Ethmoid Roof	
Dermoid	S	N	N	S	N	
Glioma	R	S	N	N	N	
Encephalocele	Y	S	Y	Y	S	

Y, yes; N, no; S, sometimes; U, usually; R, rarely.

- **Diagnosis of Congenital Midline Nasal Masses:**
 - High index of suspicion is required for any unilateral intranasal mass in children.
 - **NEVER** do a biopsy for extra or intranasal mass in a child before imaging due to Risk of meningitis or CSF leak if there is an intracranial connection.
 - **CT Sinuses with contrast:**
 - Best to evaluate skull base defect.
 - Risk of radiation exposure.
 - CT findings:
 - Fluid filled cyst
 - Soft tissue mass
 - Intracranial mass
 - Enlargement of foramen cecum
 - Distortion of crista galli
 - CT findings of intracranial involvement:
 - Bifid crista galli
 - Enlarged foramen cecum
 - False-positive and false-negative results regarding intracranial involvement are common.



- **MRI:**
 - Best to evaluate soft tissue.
 - More sensitive and specific and valuable to identify an intracranial connection.
 - No risk of false negative
 - No risk of radiation exposure.
 - Recommended to start with CT followed by MRI to confirm intracranial connection.
 - Due to the increase cost of two tests and delay in diagnosis, MRI is preferred to be used as the initial imaging study.
 - **Dermoids:**
 - Hyper-intense on T1
 - Isointense on T2
 - **Gliomas:**
 - Variable intensity on T1
 - Hyper-intense on T2
 - **Encephalocels:**
 - Continuous with brain tissue.
 - Variable intensity on T1
 - Hyper-intense on T2



- **Management of Congenital Midline Nasal Masses:**
 - Complete surgical excision.
 - Generally, early surgical intervention is recommended to avoid distortion of nose or bony atrophy caused by growth of the mass, recurrent inflammation or meningitis.
 - Timing of surgery:
 - **If connected intracranially:**
 - Should operate immediately.
 - Joint management by ENT and neurosurgery.
 - ENT initiates with midface approach; neurosurgery then performs craniotomy (bifrontal).
 - **If no connection or infection:**
 - May delay up to 2-5 years
 - Postponing surgery allows:
 - Time for growth
 - Facilitating the surgical dissection
 - Diminishing the risks of altered nasal growth from tissue disruption.
 - **Dermoids:**
 - Entire lesion with any tract must be excised to prevent recurrence.
 - Recurrence rate 5.5%.
 - Intra-op frozen section can be done for dermoids stalk where it cannot be followed to base.
 - If only fibrous tissue is found, stalk is ligated.
 - If dermoid tissue is found, an intracranial approach is performed.
 - Open rhinoplasty:
 - Excellent exposure and good cosmesis
 - Allows simple reconstruction of the nasal dorsum as needed.
 - Vertical midline osteotomy:
 - Combined with open rhinoplasty for skull base access.
 - Transglabellar approach:
 - If tract over glabella.
 - **Gliomas:**
 - Usually excised intranasally; get the stalk too.
 - Have neurosurgery on standby.
 - **Encephaloceles:**
 - Require combined approach with neurosurgery
 - Frontal craniotomy is performed
 - Intracranial mass excised and bone-dura defect is repaired
 - Extracranial mass is then removed

Aerodigestive Foreign Bodies:

- High level of clinical suspicion can prevent delays in diagnosis and subsequent complications.
- Prevention is the most important intervention for potential aerodigestive FB.
- Epidemiology:
 - **Most commonly in children between 2-4 years old:**
 - Increased mobility.
 - High tendency for placing objects in their noses, mouths or ears.
 - Playing while eating.
 - Lack of cognitive recognition of objects.
 - Lack posterior dentition.
 - Immature abilities to swallow.
 - Adults with intellectual or behavioral disabilities.
 - Normal adults during:
 - Coma
 - Deep sleep
 - Alcoholic intoxication.
- Stages of aerodigestive FB:
 - **FB impaction:**
 - Initial period of choking, coughing or gagging.
 - Lasts for a short time.
 - **Symptomless stage:**
 - Symptoms wane as FB settles into a stationary location and tracheoesophageal reflexes tire out.
 - Last hours to weeks which may delay the diagnosis.
 - **Complication stage:**
 - Caused by obstruction, inflammation or trauma.



- **Nasal FB:**

- More common than other aerodigestive FB.

- **Type of FB:**

○ **Inorganic:**

- Most common.
- Pieces of paper, chalk, button, button batteries.

○ **Organic:**

- Nuts, seeds and beans.

- **Location:**

- **Right side nasal cavity** is the most common side due to predominance of right-handed children:
 - On the floor of the nasal passage under the inferior turbinate.
 - Superiorly in front of the middle turbinate.

- **Clinical picture:**

○ **Witnessed (80%):**

- Asymptomatic.

○ **Unwitnessed (20%):**

- Unilateral mucopurulent nasal discharge.
- Foul odor.
- Epistaxis.
- Nasal obstruction.
- Mouth breathing.

- **Diagnosis:**

○ **Direct nasal examination:**

- Establishes the diagnosis.
- Accomplished with headlight or endoscope.

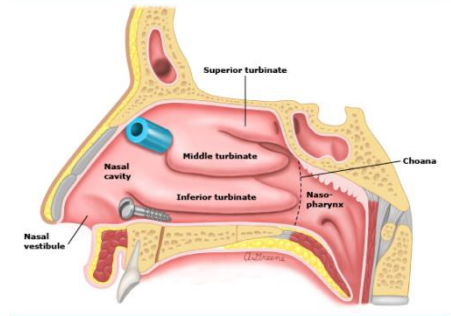
○ **X-Ray:**

- Most nasal FBs are radiolucent (Black).
- Not routinely needed when FB is visualized on examination.

▪ **Indication:**

- Suspected presence of button battery or magnet which is not obvious on physical examination:
 - Significant epistaxis, black nasal discharge, pain, or facial swelling.

Common locations of nasal foreign bodies

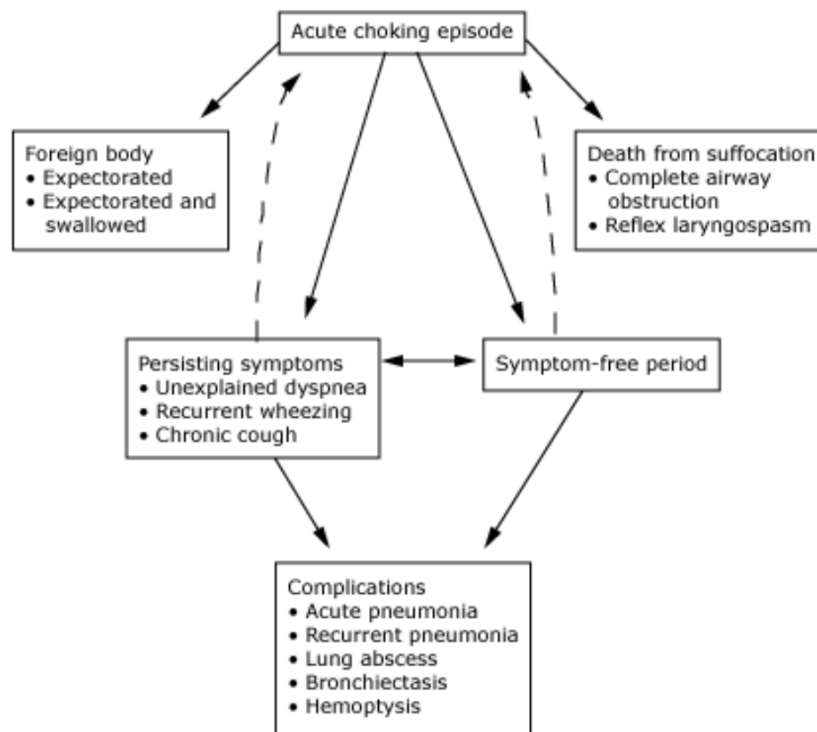


- Removal of FB:
 - **Timing:**
 - Nasal FB removal is an elective procedure **EXCEPT (Urgent):**
 - Button batteries.
 - Paired magnets attached across the nasal septum.
 - Aspiration of FB from nasal cavity into trachea:
 - Spontaneous aspiration is NOT warranted in a normal healthy patient with intact airway reflexes.
 - Risk of < 6 in 10,000 cases.
 - No reports of bronchial FBs spontaneously arising from nasal FBs in the literature.
 - Risk of aspiration is increased during FB removal if:
 - Previous attempts done by inexperienced clinicians.
 - Lacking of appropriate instruments.
 - Lacking of adequate restraint.
 - **ENT referral:**
 - Most nasal FBs can be removed by pediatric emergency clinicians without referral to ENT.
 - Indications for ENT referral:
 - Posterior FBs (not visualized by anterior rhinoscopy).
 - Chronic or impacted FBs.
 - Penetrating or hooked FBs.
 - Any FB that cannot be removed at initial attempt.
 - **Instrumentation:**
 - Alligator forceps:
 - Indicated for non-occlusive compressible FB (foam rubber or FB with rough surfaces).
 - Avoid it for removal of smooth FB that cannot be easily grasped due to risk of pushing it deeper into nasal cavity.
 - Blunt right-angle hook:
 - Indicated for FB with smooth surface (bead, stones or other small objects).
- Complications of FB:
 - **Tissue destruction:**
 - Occurs more commonly with button batteries:
 - Cause alkaline tissue necrosis due to strong electrical currents (rather than leakage of battery contents).
 - Result in:
 - **Septal perforation (within < 4 hours).**
 - Saddle nose deformity, nasal meatal stenosis and inferior turbinate necrosis.
 - Paired disc magnets across the nasal septum can cause septal perforation due to chronic compression if lasted for weeks.
 - **Rhinolith:** (Stone formation from Ca and Mg deposition around FB).
 - **Unilateral Sinusitis**
 - **FB aspiration into tracheobronchial tree during FB removal.**

- **Airway FB Aspiration:**
- Most airway FB are expelled immediately by protective cough reflex.
- Considered as a diagnostic challenge because their presentation can vary from life-threatening airway compromise to subtle respiratory symptoms that are often misdiagnosed.
- Multiple airway FB is found in **5%** of cases.
- Type of FB:
 - o **Organic:**
 - Most common aspirated objects are food products.
 - Peanut is the most common vegetable FB.
 - Nuts, seeds and beans.
 - Worse than inorganic FB due to:
 - Absorbs water over time with subsequent swelling leading to complete bronchial obstruction.
 - Illicit inflammatory reaction in surrounding tissue leading to bronchitis.
 - o **Inorganic:**
 - Pearls, latex balloon, pins and needles.
 - Loose teeth or denture in adults.
 - Less inflammatory and remain in one position for extended periods of time without causing increasing degrees of obstruction.
- Locations:
 - o **Laryngotracheal:**
 - Less common site of FB aspiration.
 - Mainly as a result of large FB aspiration.
 - Patients commonly present with acute respiratory distress with stridor, coughing and wheezing.
 - May totally obstruct the airway in the larynx leading to sudden death.
 - Occurs in larynx in **(5%)** and trachea/carina in **(15%)** of FB aspirations.
 - o **Large bronchi:**
 - Patients commonly present with coughing and wheezing.
 - Right main bronchus is the most common location **(60%)** for airway FB aspiration:
 - Wider and more vertical in line with the tracheal lumen.
 - Left main bronchus accounts for **(25%)** of FB aspirations.
 - o **Distal airways:**
 - Less commonly.
 - Patients commonly present with less acute respiratory distress.

- Clinical picture:
 - **Witnessed (50%):**
 - Choking episode:
 - Sudden onset of cough and/or dyspnea and/or cyanosis in a previously healthy child.
 - Lasts for few seconds to several minutes.
 - Sensitivity of 90% diagnosis of FB Aspiration.
 - Followed by either:
 - Complete airway obstruction.
 - Acute respiratory distress.
 - Symptom-free period.
 - Chronic wheezing, and/or persistent coughing caused by inflammation and edema formation.
 - **Unwitnessed (50%):**
 - Present either:
 - **Immediately after the aspiration:**
 - Acute respiratory distress.
 - **Days or weeks after the aspiration:**
 - Intermittent SOB and/or wheeze and/or cough.
 - Recurrent asthma exacerbation not responding to routine management.
 - Recurrent pneumonia.

Natural course of foreign body aspiration



- Diagnosis:

○ **Detailed history and physical examination:**

- Triad suggestive of FB aspiration:
 1. Cough.
 2. Localized wheeze.
 3. Localized diminished breath sounds.

○ **Neck and CXR:**

- Most important non-invasive study.
- Protocol of CXR for suspected FB aspiration:
 1. **PA and lateral views:**
 - Differentiate tracheal from esophageal FB.
 2. **Inspiratory and Expiratory films:**
 - Increase sensitivity of detecting Radio-lucent FB.
 - Lung hyperinflation during the expiratory film at the same side of FB aspiration.
 3. **Left/Right Lateral decubitus films:**
 - Dependent lung simulate expiratory films in an uncooperative young children.
 - Hyperinflation of dependent lung suggest FB.

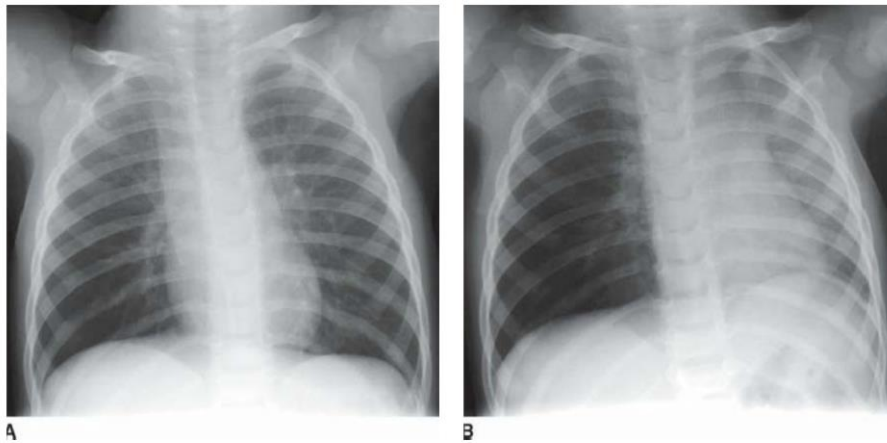


Figure 93.2 Inspiratory and expiratory radiographs of a child who aspirated a peanut into his right mainstem bronchus. **A:** Inspiratory view. **B:** Expiratory view. Note how the right lung remains inflated on expiration. Image courtesy of Dr. C. Branstetter.

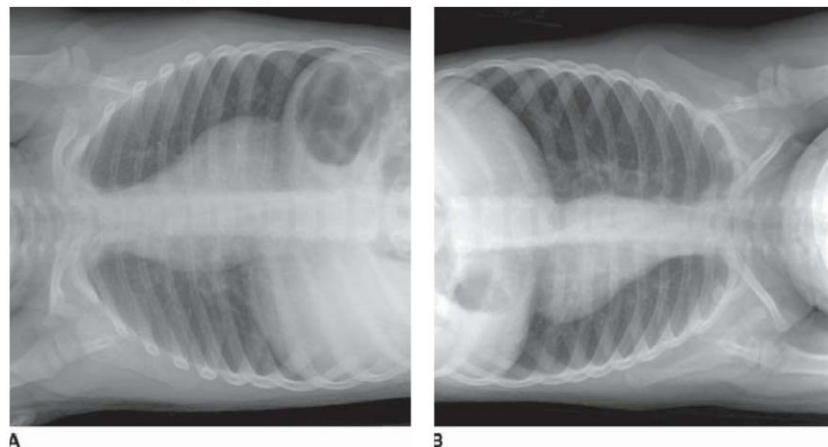
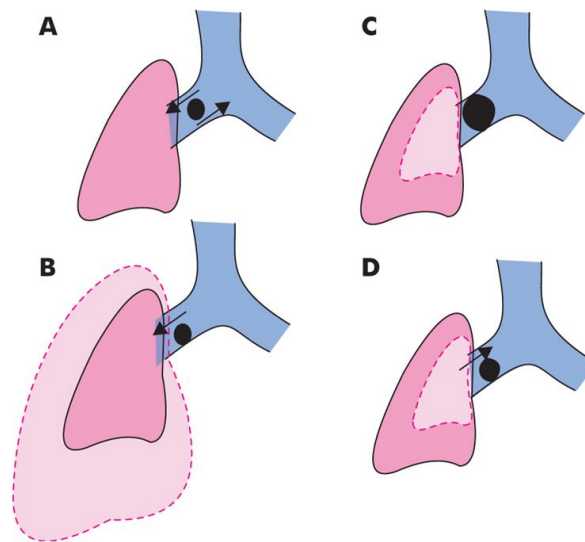


Figure 93.3 Right and left lateral decubitus radiographs of child who aspirated a pumpkin seed into her left mainstem bronchus. **A:** Right lateral decubitus position. **B:** Left lateral decubitus position. Note how the left lung does not change in volume despite changes in patient position. Image courtesy of Dr. J. Crowe.

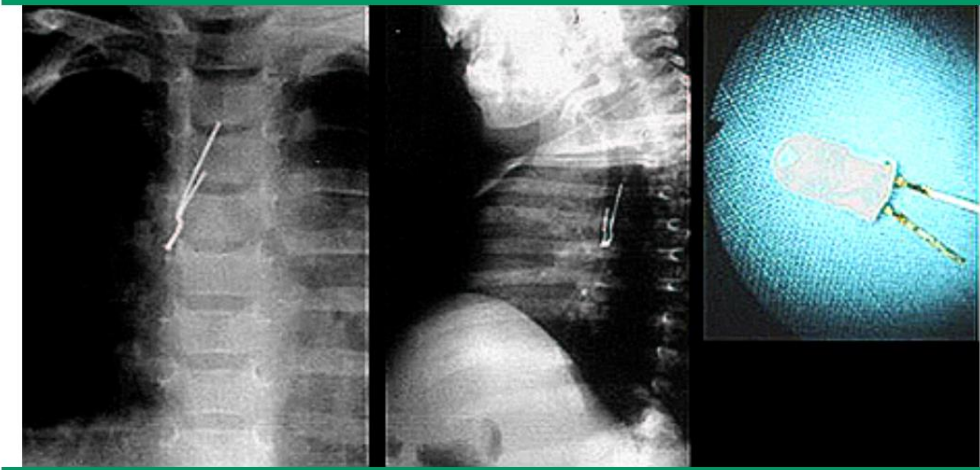
- Findings of CXR in patients with FB aspiration:
 - **Normal CXR:**
 - Found in **25%** of patients with proven FB aspiration.
 - Most common aspirated FB are Radio-lucent (Black) which is NOT detected with plain x-ray.
 - **Normal CXR does not rule out FB aspiration.**
 - **Abnormal CXR:**
 - Found in **75%** of patients with proven FB aspiration.
 - Signs:
 - 1. Radio-opaque (White) FB:**
 - Presents in **10%** of all airway FBs.
 - 2. Unilateral Hyperinflation:**
 - Partial airway obstruction with air trapping.
 - Better evaluated during expiratory or lateral decubitus films.
 - 3. Localized Atelectasis:**
 - Due to complete airway obstruction proximal to the side of collapse.
 - 4. Mediastinal Shift:**
 - Away from FB side.
 - 5. Localized Infiltrates:**
 - Distal to obstructed airway.



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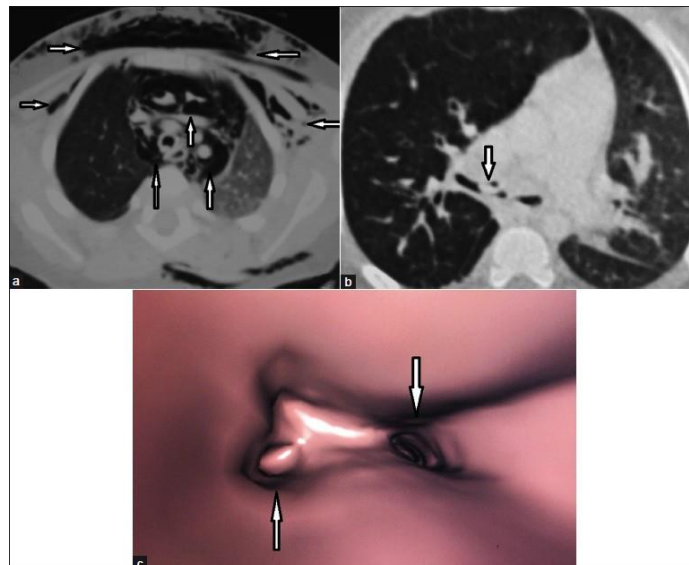
Figure 64.2 Types of bronchial obstruction by a foreign body.
 (A) Partial obstruction. Air can pass in and out, causing only wheeze.
 (B) One way obstruction. Air can go in (during inspiration) but not out, causing emphysema of lungs. (C) Total obstruction. Air can neither go in nor out, causing obstructive atelectasis. (D) One way obstruction (reverse of B). Air can only go out, causing atelectasis. Dark pink shows normal size of lung while lighter pink indicates effect of obstruction.

Radiopaque foreign body in the airway



○ CT Chest:

- Indication:
 - Persistent symptomatic but stable patients with low clinical suspicion of FB aspiration and normal or inconclusive CXR.
- Advantages:
 - **Avoid bronchoscopy in low suspicion of FB aspiration.**
 - Detect **ALL** Radio-lucent FB.
 - Generate 3D images of the large airways (Virtual bronchoscopy).
- Disadvantages:
 - Radiation exposure.
 - Delay therapeutic bronchoscopy in case of proven positive FB aspiration.



- **Criteria of High clinical suspicion of FB aspiration:**
 - Presence of ANY of the following:
 1. Witnessed FB aspiration regardless of symptoms.
 2. History of choking with either:
 - Subsequent respiratory symptoms
 - Suspicious characteristics on imaging.
 3. Triad suggestive of FB aspiration (wheeze, cough, and/or diminished breath sounds) in a young child without other explanation.
 - Next step:
 - **Rigid Bronchoscopy.**
- **Criteria of Low clinical suspicion of FB aspiration:**
 - NONE of the High clinical suspicion features are present.
 - Normal CXR is sufficient to exclude FB aspiration.
 - CT scan can be done if persistent symptoms.
- Treatment:
 - **FB with complete Airway obstruction:**
 - Occurs in Large laryngotracheal FB.
 - Patients is unable to speak or cough.
 - Life-threatening condition due to asphyxia.
 - Treatment:
 - **First aid maneuvers for dislodgement of FB:**
 - Methods:
 - Back blows and chest compressions in infants.
 - Heimlich maneuver in older children and adults.
 - Should not be done for patients with partial obstruction (can speak and cough) due to the risk of convert it to complete obstruction.
 - Blind sweeping of the mouth should be avoided for the same reason.
 - **Intubation:**
 - Done after failed FB dislodgment with first aid maneuvers and FB is located below VC.
 - **Cricothyrotomy or emergency tracheostomy:**
 - Done after failed FB dislodgment with first aid maneuvers and FB is located above VC.

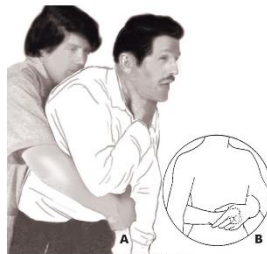
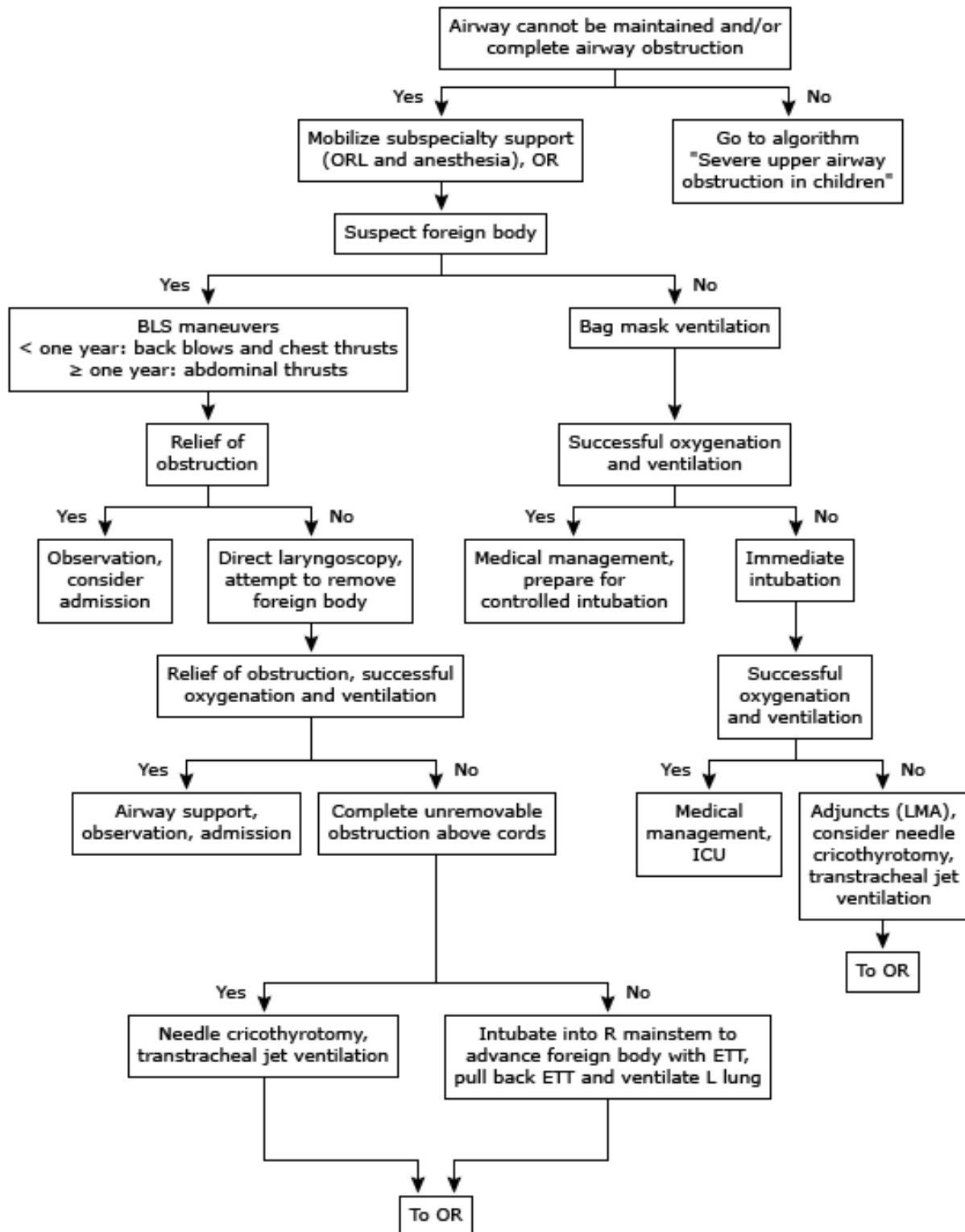


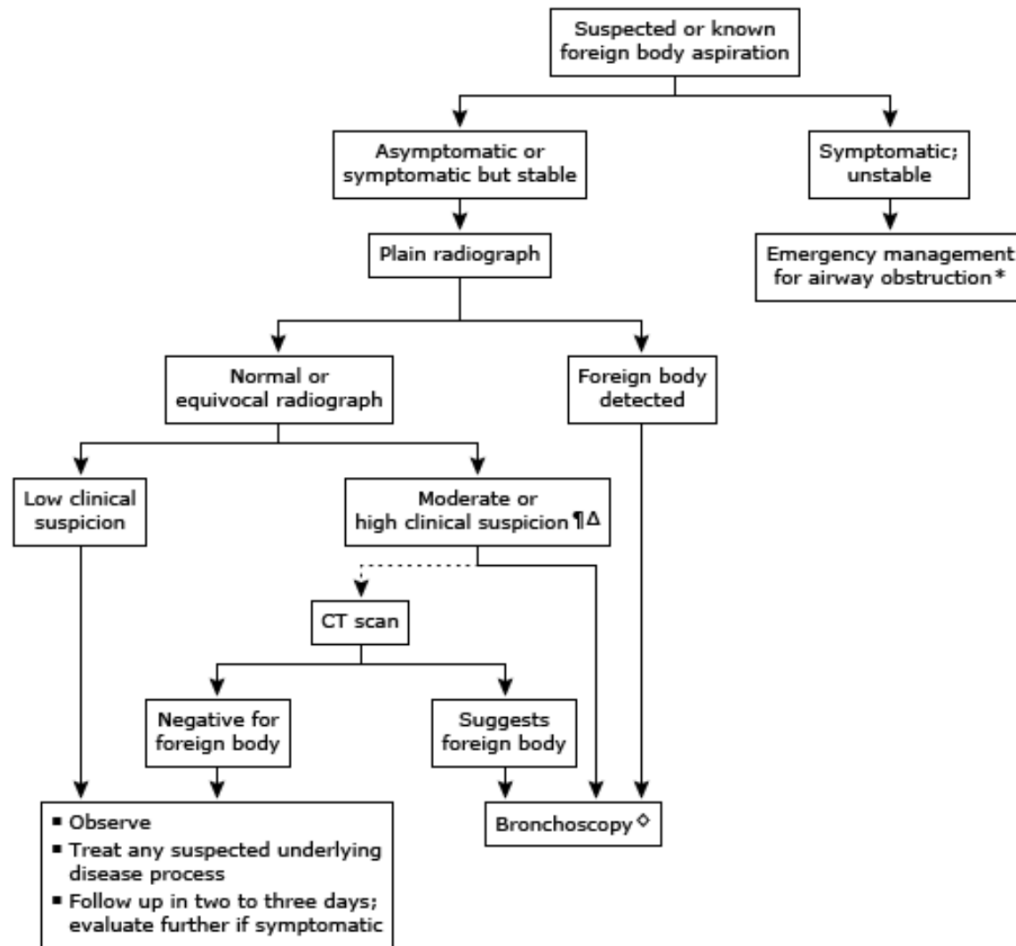
Figure 64.6 Heimlich's manoeuvre. Sudden thrust directed upwards and backwards, below the epigastrium, squeezes the air from the lungs, sufficient to dislodge a foreign body.



- **Bronchoscopy:**
 - Indicated for ALL patients with proven FB aspiration or with High clinical suspicion of FB aspiration.
 - **Rigid Bronchoscopy:**
 - Successful in 95% of cases.
 - Done in spontaneous ventilation anesthesia.
 - Should be repeated after removal of one FB to rule out presence of second FB (5%).
 - **Advantages:**
 - Permits ventilation.
 - Good visualization.
 - Manipulation of FB with a wide variety of forceps.
 - Management of mucosal hemorrhage
 - **Complications (1%):**
 - Pneumothorax
 - Hemorrhage
 - Respiratory arrest
 - **Disadvantages:**
 - Requires GA.
 - Does not reach sub-segmental airway FB.
 - **Flexible Bronchoscopy:**
 - Alternative method in some centers.
 - Successful in 90% of cases.
 - **Recommend to be done after FB removal with rigid bronchoscopy to evaluate distal tracheobronchial tree.**
 - **Advantages:**
 - Can avoids GA.
 - Reach sub-segmental airway FB.
 - **Disadvantages:**
 - Limited to older adolescents and adults.
 - Does not permits ventilation.
 - Limited variety of forceps.
- **Thoracotomy and Bronchotomy:**
 - Indicated for distal FB not reachable with rigid or flexible bronchoscopy.
- **Post-op care:**
 - IV dexamethasone
 - Nebulized epinephrine.
 - IV antibiotics

If clinical symptoms, signs, or abnormal radiographs persist after the FB removal and treatment of infection, repeat bronchoscopic examination is warranted to look for a second, previously unseen FB.

Algorithm for suspected foreign body aspiration in children



CT: computerized tomography; FBA: foreign body aspiration.

* Refer to algorithm for complete airway obstruction in children.

¶ A moderate or high suspicion of foreign body aspiration includes all children with a witnessed FBA (regardless of symptoms), and those with suggestive respiratory symptoms or suspicious characteristics on imaging, especially if there is a history of choking.

Δ For stable patients with a high clinical suspicion of aspiration, it is reasonable to proceed directly to bronchoscopy, even if the plain radiographs are normal or inconclusive. Alternatively, computed tomography (CT) can be performed first to help clarify the diagnosis (dotted line), if the provider judges that negative imaging would be sufficient to preclude bronchoscopy.

◇ Rigid bronchoscopy is the procedure of choice to remove a foreign body. In cases where the diagnosis or location of the foreign body is unclear, it is usually preferable to perform flexible bronchoscopy first, then proceed to rigid bronchoscopy for foreign body removal.

- **Esophageal FB Ingestion:**
- Twice as common as airway FB aspiration.
- Most GIT FB pass spontaneously **80%**.
- Only **20%** will require endoscopic removal.
- Less than **1%** require surgical intervention.
- Type of FB:
 - **Inorganic:**
 - Most common ingested FB in pediatrics are coins.
 - Magnets, batteries, safety pins, screws, marbles.
 - Loose teeth or denture in adults.
 - **Organic:**
 - Most common ingested FB resulting in fatality in pediatrics is a segment of hotdog.
 - Most common ingested FB in adults are fish bones and meat.
 - Food impaction should raise the suspicion of Esophageal Narrowing or Eosinophilic Esophagitis.
- Locations:
 - **Cricopharyngeus Muscle (70%):**
 - 1st normal constrictions (upper esophagus).
 - 15 cm from the upper incisors.
 - At level of (C6).
 - **At Level of Arch of Aorta and Left main bronchus (20%):**
 - 2nd normal constrictions (mid esophagus).
 - 25 cm from the upper incisors.
 - At level of (T4).
 - **Lower esophageal sphincter (20%):**
 - 3rd normal constrictions (lower esophagus).
 - 40 cm from the upper incisors.
 - At level of (T10).

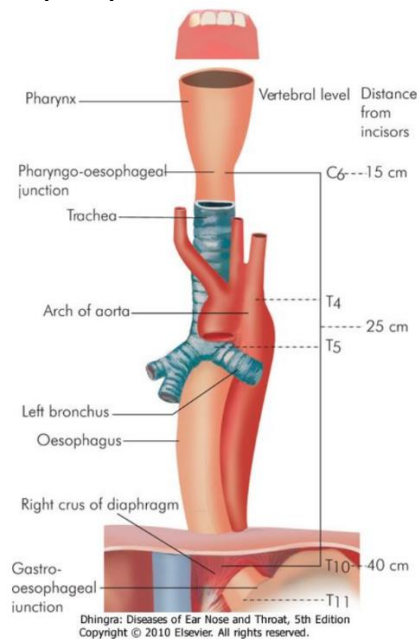


Figure 65.1 Anatomy of oesophagus and levels of normal constrictions from the upper incisors.

- Clinical picture:
 - **Witnessed (90%):**
 - Asymptomatic in 50% of patients.
 - **Unwitnessed (10%):**
 - Present either:
 - **Complete esophageal obstruction:**
 - Present immediately after ingestion.
 - Symptoms:
 - Stridor (posterior tracheal compression)
 - Absolute dysphagia (Drooling of saliva)
 - Vomiting
 - **Partial esophageal obstruction:**
 - Present days to weeks after ingestion.
 - Symptoms:
 - Decreased oral intake.
 - Weight loss.
 - Aspiration pneumonia:
 - Poor handling of oral secretions, or
 - Tracheoesophageal fistula formation.
 - **Esophageal perforation:**
 - Present with sharp or chemical FB.
 - Symptoms:
 - Neck swelling
 - Crepitus
 - Fever
 - Pneumomediastinum
- Diagnosis:
 - **Neck/Chest/Abdomen X-Ray:**
 - AP and Lateral views.
 - Findings:
 - Radio-opaque FB **(60%)**.
 - Widened prevertebral shadow in lateral view **(40%)**.
 - Loss of lordosis in lateral view **(40%)**.
 - Differentiate between esophageal and tracheal FB:
 - 1. Location of FB in AP and lateral views:**
 - Anterior in Trachea.
 - Posterior in Esophagus.
 - 2. Orientation of Flat FB (coins or button batteries):**
 - Coronal plane in Esophagus (Round in AP view).
 - Sagittal plane in Trachea (Round in Lateral view).
 - Differentiate between Button battery and Coin:
 - 1.** Button battery has double ring on AP view.
 - 2.** Button battery has step-off in its lateral edge on lateral view.

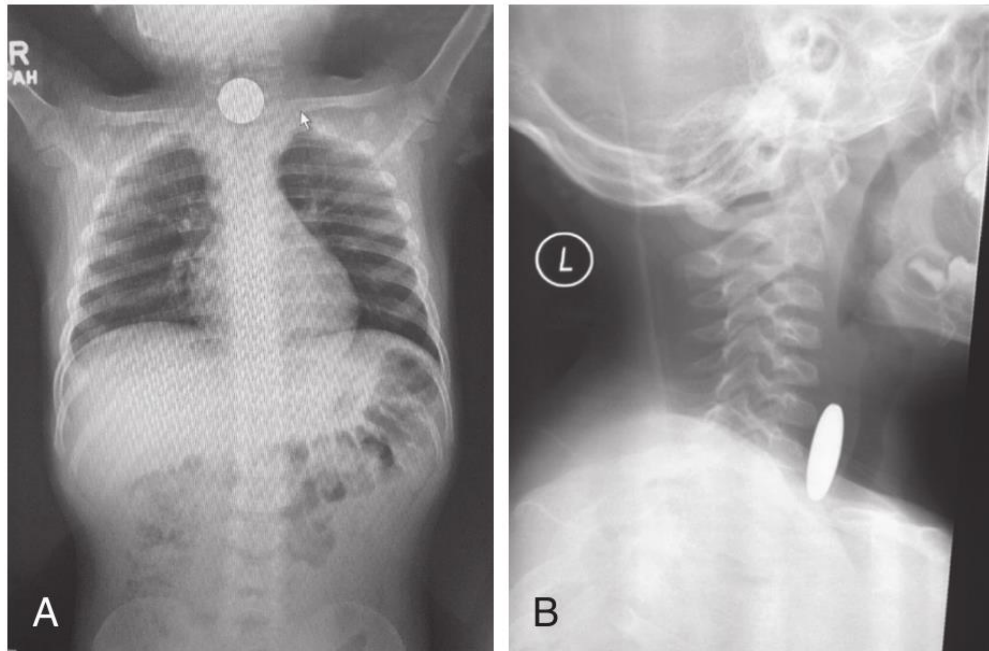


FIGURE 207-1. A posteroanterior radiograph demonstrates a coin lodged in the aerodigestive tract and oriented in the coronal plane, indicating it is likely in the esophagus. **B,** A lateral radiograph confirms the location of the coin to be posterior to the trachea in the esophagus. (Courtesy Andrew Murr, MD.)

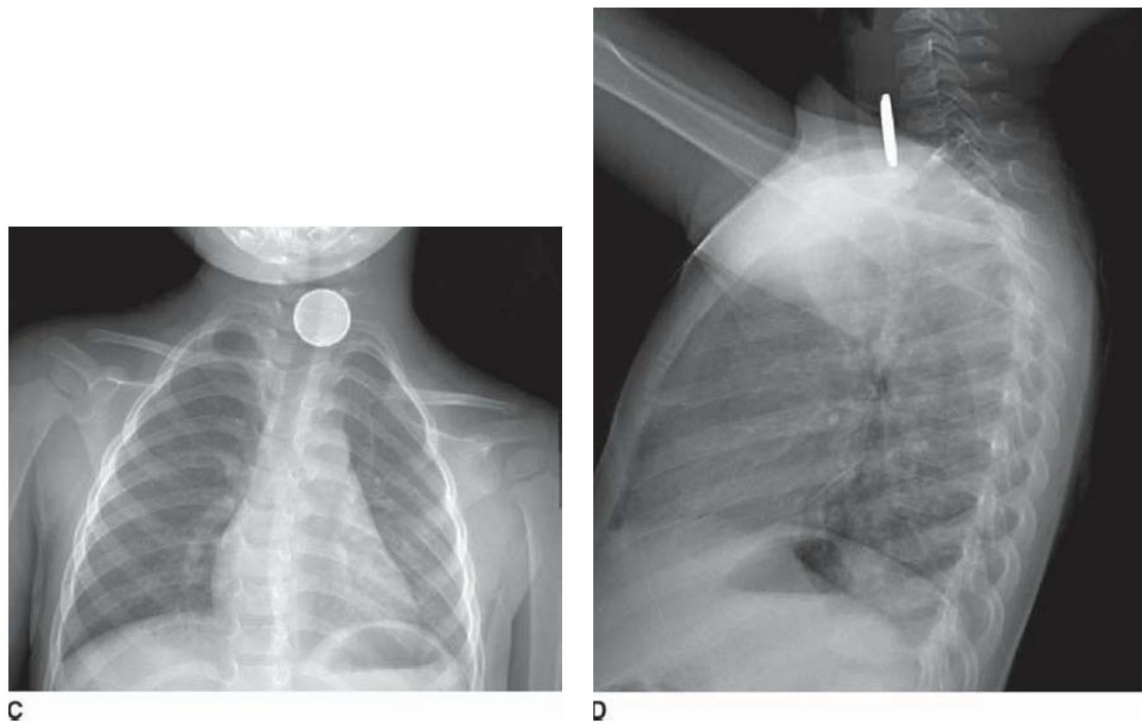


Figure 93.5 (Continued) (C) and lateral (D) views of a disc battery lodged at the UES. Note the "double-ring" that is characteristic of a disc battery in C. Also note the step that can be seen along the edge of the disc battery in D as compared to the profile of the coin in B.

- Treatment:
 - **Conservative:**
 - Observation for 12-24 hours to allow for spontaneous passage.
 - Indication:
 - Food impaction with no respiratory distress or absolute dysphagia.
 - Asymptomatic non high-risk FBs.
 - Glucagon:
 - **1mg IV.**
 - Induce relaxation of distal esophagus.
 - Mainly used in impacted esophageal food bolus in adults.
 - **Esophagoscopy:**
 - Repeat radiograph should be done immediately prior to the procedure to verify that FB didn't pass into the stomach.
 - Timing:
 - **Emergent (within few hours):**
 - Indications:
 1. Respiratory distress.
 2. Absolute dysphagia (drooling of saliva).
 3. High-risk FB (battery, sharp, long, wide).
 - **Urgent (within 24 hours):**
 - Allows the procedure to be done in more controlled environment (Morning time).
 - Indications:
 1. Failure of 24-hour conservative management for food impaction.
 2. Symptomatic non high-risk FB.
 - Techniques:
 - **Flexible Endoscopy:**
 - Done under conscious sedation.
 - Preferred in distal FB.
 - **Rigid Endoscopy:**
 - Done under GA.
 - Preferred if FB located at the level of cricopharyngeus muscle (C6).
 - Sedation and anesthesia can relax the esophageal sphincters causing spontaneous migration of FB into the stomach.
 - Higher risk of complications:
 - Mucosal abrasion
 - Esophageal perforation and mediastinitis.
 - Post-op care:
 - Avoid post-op antibiotics/strong analgesia/steroid for the first 24 hours (masking signs of perforation).
 - Monitor signs of mediastinitis (Fever, Tachycardia and Tachypnea).
 - CXR if suspecting complications.

- Specific management according to FB Type:
 - **Coins:**
 - Most common FB in children.
 - 70% will pass spontaneously.
 - Coins in distal esophagus have better chance for spontaneous passage.
 - If it reached the stomach, most will pass out uneventfully within 1 week.
 - **Food impaction:**
 - Most common FB in adults.
 - IV glucagon facilitate spontaneous passage.
 - Occurs mainly in:
 - Patients with underlying esophageal pathology (strictures or stenosis).
 - Patients with Eosinophilic esophagitis.
 - Avoid forceful pushing of food particles into the stomach due to risk of perforation in patients with pre-existing esophageal narrowing.
 - Mucosal biopsies can be taken at the time of endoscopy if suspecting Eosinophilic esophagitis.
 - **Batteries:**
 - Medical emergency.
 - Associated with significant morbidity.
 - High risk of complication from:
 - Leaking of alkaline contents.
 - Low-voltage electrical discharge.
 - Direct pressure necrosis
 - Complications:
 - Mucosal necrosis (1 hour).
 - Mucosal ulceration (2 hours).
 - Perforation (8 hours).
 - Batteries passed into the stomach do not require intervention unless it remain in the stomach for longer than 48 hours.
 - **Sharps:**
 - Medical emergency.
 - Associated with significant morbidity.
 - Straight pins, needles and fish bones.
 - Radiography is unlikely to detect fish bones and wood toothpicks.
 - High risk of perforation (35%).
 - If passed into the stomach, it should be removed promptly with flexible endoscope.
 - **Long or wide FB:**
 - Long (>6cm) or wide (>2cm) FBs should be removed from the esophagus or stomach due to the high risk of impaction at ileocecal valve.

- Complications of Esophageal FB:
 - **Perforation:**
 - Risk factors:
 - FB duration > 24 hours.
 - High-risk FB (Battery or Sharp).
 - **Transmural Erosion**
 - **Stricture Formation**
 - **Tracheoesophageal Fistula**

TABLE 207-1. Overview of the Management of Aerodigestive Foreign Bodies

	Airway Foreign Body	Esophageal Foreign Body
History	Witnessed aspiration Cough, dyspnea, wheezing, stridor Refractory asthma	Witnessed ingestion Vomiting, drooling, dysphagia, odynophagia, emesis, food refusal, chest pain
Physical examination	Decreased lung sounds, wheezing, crackles Tachypnea, hypoxemia	Drooling, poor feeding, choking
Imaging	PA and lateral radiographs (radiopaque foreign body, unilateral emphysema or hyperinflation, localized atelectasis or infiltrate)	PA and lateral radiographs (radiopaque foreign body, widened prevertebral shadow, loss of lordosis)
Treatment	If adequate suspicion, proceed immediately to rigid bronchoscopy for removal	<i>Young symptomatic children:</i> FB present >24 hr or sharp metallic or caustic objects should undergo endoscopic removal <i>Asymptomatic children:</i> recent ingestion (<24 hr), no esophageal disorders can be observed for 8 to 16 hours

FB, foreign body; PA, posteroanterior.

TABLE 2. Timing of endoscopy for ingested foreign bodies

Emergent endoscopy

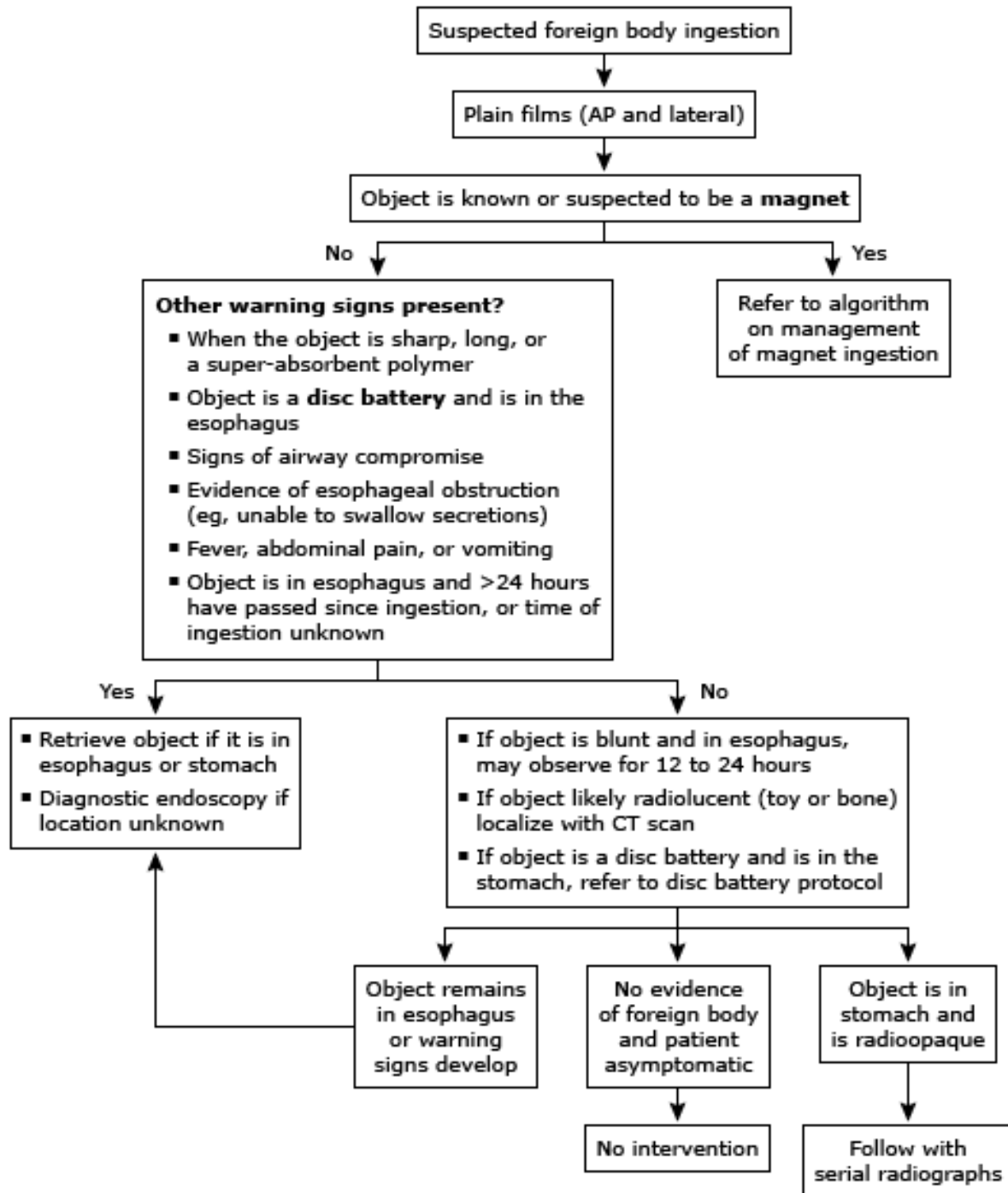
Patients with esophageal obstruction (ie, unable to manage secretions)
Disk batteries in the esophagus
Sharp-pointed objects in the esophagus

Urgent endoscopy

Esophageal foreign objects that are not sharp-pointed
Esophageal food impaction in patients without complete obstruction
Sharp-pointed objects in the stomach or duodenum
Objects >6 cm in length at or above the proximal duodenum
Magnets within endoscopic reach

Nonurgent endoscopy

Coins in the esophagus may be observed for 12-24 hours before endoscopic removal in an asymptomatic patient
Objects in the stomach with diameter >2.5 cm
Disk batteries and cylindrical batteries that are in the stomach of patients without signs of GI injury may be observed for as long as 48 hours. Batteries remaining in the stomach longer than 48 hours should be removed.



Caustic and Corrosive Ingestions:

- A caustic or corrosive substance is a dry or liquid chemical agent that induce immediate injury on contact with living tissue.
- A low concentration of a corrosive is usually an irritant.
- May be swallowed accidentally in children or taken with the purpose of suicide in adults.

- Types of responsible chemical agents:

- **Alkali (Caustic):**

- **pH >7.**
 - Odorless and tasteless agents.
 - Allow for the consumption of large volumes.
 - Destruction by **Liquefaction Necrosis:**
 - Rapid dis-integration of mucosa with deep penetration and formation of viscous liquid (**saponification**).
 - Blood vessel thrombosis compromises the tissue blood supply, which contributes to perforation.
 - Results in more oral and upper esophageal injury and surrounding tissues.
 - Examples:
 - Disk/button batteries
 - Laundry detergent
 - Lime
 - Hair straightener

- **Acidic (Corrosive):**

- **pH < 7.**
 - Bitter taste.
 - Lower the volumes of accidental ingestion.
 - Destruction by **Coagulation Necrosis:**
 - More superficial penetration with less damage to esophagus due to coagulum formation on the mucosa limiting deeper absorption until the agent reaches the stomach.
 - Results in skip areas in the esophagus and more severe damage to the stomach.
 - Examples:
 - Toilet cleaners
 - Sulfuric acid

- **Bleaches:**

- **pH = 7 (Neutral).**
 - Considered mild irritants.
 - Has no significant morbidity or mortality.
 - Doesn't require extensive workup patients following ingestion.
 - Examples:
 - Sodium hypochlorite.

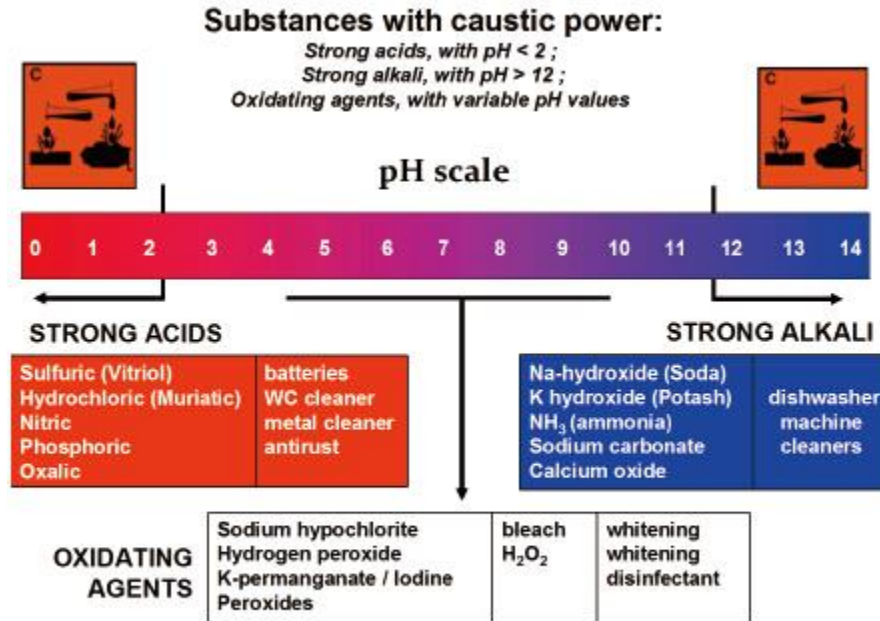


Figure 1 legend Synopsis of caustic agents: categories and most common utilization.

- Pathophysiology:
 - **Phases of injury:**
 - Acute phase:
 - Initial contact will produces immediate changes that progress over the following 2-3 days.
 - Superficial mucosal burns tend to heal without sequelae.
 - Deeper burns disrupt the submucosa and muscular layer leading to intense inflammatory response develops with accompanying esophageal dysmotility.
 - Latent phase:
 - In response to injury, fibroblasts produce a matrix of newly formed collagen fibers, which begin to contract 3-4 weeks after initial insult.
 - Contraction enables adhesive bands to form, which lead to development of:
 - Pseudo-divertirula
 - Endoluminal strictures.
 - **Factors affecting the severity of injury:**
 1. Type of chemical agent
 2. pH concentration.
 3. Amount
 4. Contact time
 5. Presence of food in the stomach
 6. Presence of GERD

- Clinical picture:
 - Presence or absence of symptoms does not reflect the degree of GIT injury.
 - Presence or absence of oral lesions does not predict esophageal injury.
 - Most common symptoms:
 - Vomiting
 - Dysphagia
 - Drooling
 - Odynophagia
 - Dysphonia
 - Stridor
 - Symptoms suggesting perforation:
 - Fever
 - Chest or Abdominal pain
 - Hypotension

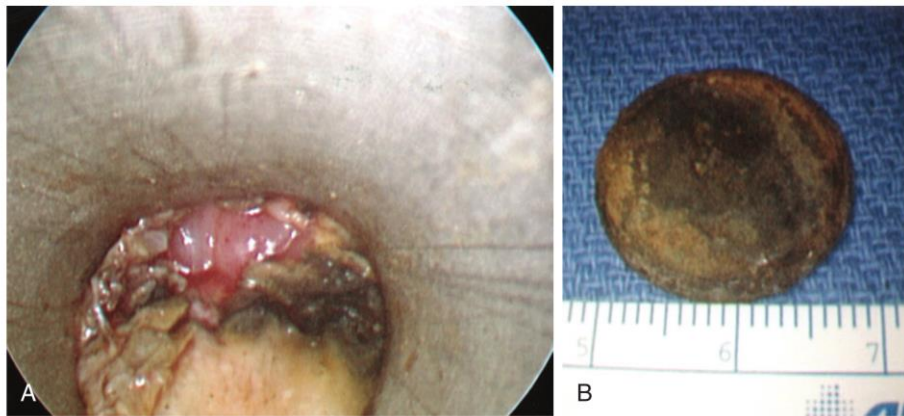


FIGURE 207-9. **A**, Esophageal injury from lithium battery. Endoscopic photo of second-degree injury from hydrolysis reaction. **B**, Lithium battery retrieved from the esophagus: note the degree of mucosal reaction on the battery's surface.

- Management:

○ **General Principles:**

- ABC
- If any respiratory distress, tracheotomy should be done.
- NPO
- IV fluid hydration
- Avoid:
 - **ET Intubation:**
 - In patients with severe laryngopharyngeal injuries.
 - **Charcoal:**
 - Does not adsorb to caustic agents.
 - **Steroid:**
 - Doesn't decrease risk of strictures formation.
 - May worsen the clinical course of severe injury leading to perforation
 - **Vomiting:**
 - Risk of re-exposure to the agent.
 - **Oral Dilution therapy:**
 - Done with water or milk.
 - May induce vomiting especially if > 15 mL/kg of weight.
 - **Blind NGT insertion:**
 - Risk of perforation.
 - **Contrast Esophagogram within <48h:**
 - Not useful as it is not sensitive to determine the initial degree of injury.
 - **Esophagoscopy after >48h:**
 - High risk of perforation.
 - Necrotic tissue sloughs off and esophageal wall becomes weak and instrumentation may easily pass through the weakened esophageal wall.

- **Hemodynamically stable and presented <48 h:**
 - Identify the chemical agent and determine pH if possible.
 - Investigations:
 - **Chest and Abdominal X-ray:**
 - Rule out free air in mediastinum or peritoneum which indicates perforation.
 - Absence of these findings does not exclude a perforation or serious injury on a visceral level.
 - **Flexible Fiberoptic Laryngoscopy:**
 - Indicated in patients with respiratory symptoms to evaluate the laryngeal mucosa.
 - **Upper GI Endoscopy:**
 - Most reliable and accurate method of determining extent of esophageal injury.
 - Should be done in ALL patients.
 - Performed between **24-48 hours** post ingestion:
 - If < 12 hours may miss evolving lesion.
 - If > 48 hours may risk perforation.
 - Flexible endoscopy is preferred over rigid:
 - Safest.
 - Less trauma.
 - Decreases risk of perforation.
 - Allows inspection of stomach and duodenum.
 - Avoid advancing endoscopy beyond areas of transmural or circumferential injury due to risk of perforation.

- Staging and Management of Esophageal Injuries:
 - **Grade I:**
 - Mucosal edema and hyperemia.
 - **Treatment:**
 - NPO for 24-48 hours.
 - Followed by regular diet.
 - **Medications:**
 - Anti-reflux medications (PPI).
 - **Follow-up:**
 - After 3 weeks.
 - If still symptomatic, consider contrast esophagram to evaluate for strictures.
 - **Grade II:**
 - Superficial mucosal sloughing and ulceration.
 - **Treatment:**
 - NPO for 7-10 days.
 - Insertion of NGT under direct vision in OR.
 - Followed by contrast esophagram before removing NGT and starting regular diet.
 - **Medications:**
 - Anti-reflux medications (PPI).
 - Antibiotics.
 - **Follow-up:**
 - After 3 weeks.
 - Consider contrast esophagram to evaluate for strictures.
 - **Grade III:**
 - Deep ulceration and necrosis.
 - **Treatment:**
 - NPO for 7-10 days.
 - Insertion of NGT under direct vision in OR.
 - Followed by contrast esophagram before removing NGT and starting regular diet.
 - **Medications:**
 - Anti-reflux medications (PPI).
 - Antibiotics.
 - **Follow-up:**
 - After 3 weeks.
 - Consider contrast esophagram to evaluate for strictures.

• **Grade IV:**

- Perforation
- **Treatment:**
 - NPO.
 - Emergency surgical exploration:
 - Surgical repair.
 - Esophagectomy and esophageal replacement
 - **Medications:**
 - Anti-reflux medications (PPI).
 - Antibiotics.

score.	mucosal layer.	lesions.	motility.	sphincters.	picture.
0.	normal.	~.	normal.	normal.	
1.	redness, oedema.	~.	normal.	normal.	
2.	redness, oedema, superficial necrosis (whitish mucosa).	superficial erosions.	normal.	normal, decreased.	
3.	diffuse necrosis, stripping, hemorrhage.	ulcers even confluent.	decreased.	decreased.	
4.	black mucosa, diffuse necrosis, deep ulcers, severe hemorrhage, areas of impending perforation.		absent.	atonic.	

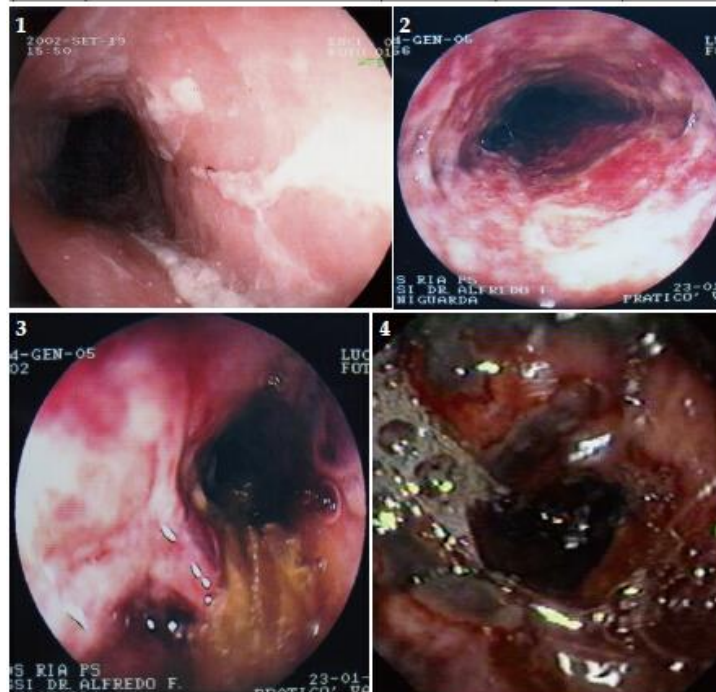


Figure 3 The "Niguarda 90" endoscopic classification of caustic burns: pictures of corresponding injuries.

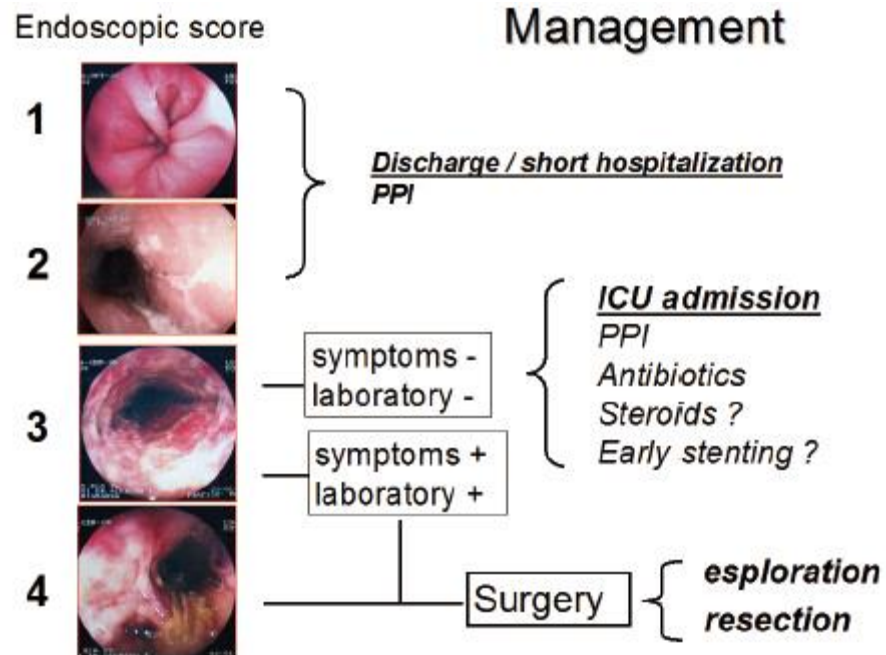


Figure 4 Management of caustic injuries according to the endoscopic score.



FIGURE 207-7. Feeding tube placement in management of esophageal injury. Endoscopic photo of a nasogastric feeding tube placed beyond the injury site to avoid stricture development.

- **Hemodynamically stable and presented >48 h:**
 - Investigations:
 - **Contrast Esophagogram:**
 - First diagnostic tool.
 - Evaluate presence of strictures.
 - Water-soluble contrast (Gastrografin) should be used in fluoroscopy whenever a perforation is suspected.
 - Strictures are dilated once found.
 - **Technetium 99m-labeled sucralfate:**
 - High sensitivity and specificity for presence of esophageal injury.
 - Does not determine the extent or severity of the injury.

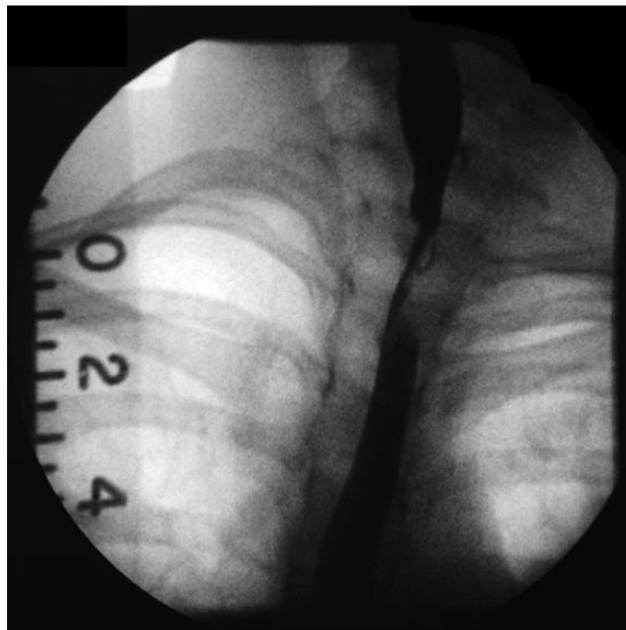


FIGURE 207-5. Esophageal stricture. Barium esophagogram demonstrates an esophageal stricture after caustic ingestion.

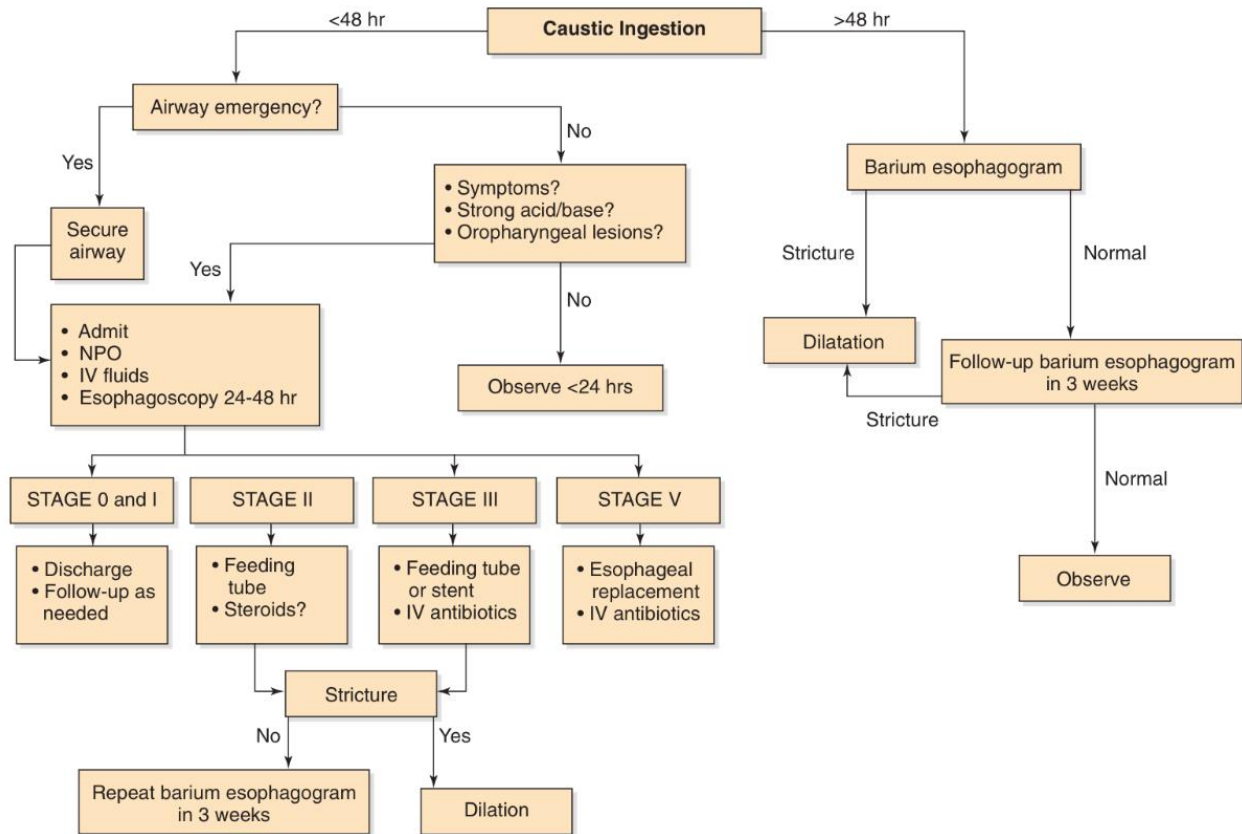


FIGURE 207-6. Caustic ingestion algorithm. IV, intravenous; NPO, nothing by mouth.

- Serious complications:
 - Airway obstruction
 - Perforation
 - Mediastinitis
 - Peritonitis
 - DIC
 - Renal failure
 - Death
- Late complications:
 - Stricture formation
 - Pseudo-diverticula
 - Tracheoesophageal fistula
 - Esophageal carcinoma (increases risk 1000 fold)

Congenital Pediatric Neck Lesions:

- Failure of involution embryologic structures and duplication of structures can lead to fistulas, sinuses, and cysts.
 - **Fistula:** Has an opening from pharynx to skin.
 - **Sinus:** Incomplete fistula, has an opening to either pharynx or skin.
 - **Cyst:** Enclosed structure with no opening to pharynx or skin.
- Failure of migration or aberrant migration of embryologic structures results in ectopic structures such as Lingual and Ectopic Thyroid.
- May remain asymptomatic and never require any treatment.
- Enlargement of the lesion may lead to concern for possible malignancy or cosmetic deformity.

- **Definitive surgical excision delayed until 3-4 years of age unless the severity warrants earlier excision.**
 - Complete surgical excisions with its deep tract to prevent recurrence.
 - Lacrimal probe or injection of Methylene blue into the tract to aid dissection.
 - If uninfected lesion is breached, Antibiotics should be given.
 - No need for Antibiotics for clean cases with no breach of the lesion.

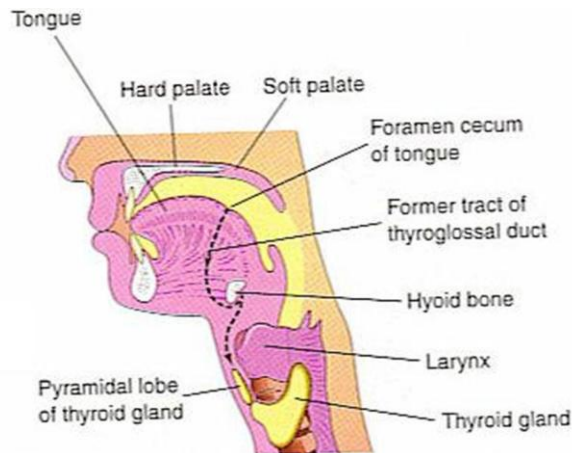
- **Infected congenital lesions:**
 - Treated with Antibiotics and aspiration if needed.
 - **I&D should be avoided** as much as possible to avoid violation of lesion walls which may make complete excision more difficult.

- **Midline Congenital Neck Masses:**
 1. Thyroglossal duct cyst (TGDC)
 2. Dermoid Cyst
 3. Teratoma
 4. Plunging Ranula
 5. Thymic cyst
 6. Ectopic Thyroid

- **Lateral Congenital Neck Masses:**
 1. Branchial Anomalies
 2. Preauricular pit/sinus/cyst
 3. Laryngocele
 4. Pseudotumor of infancy

Midline Congenital Neck Masses:

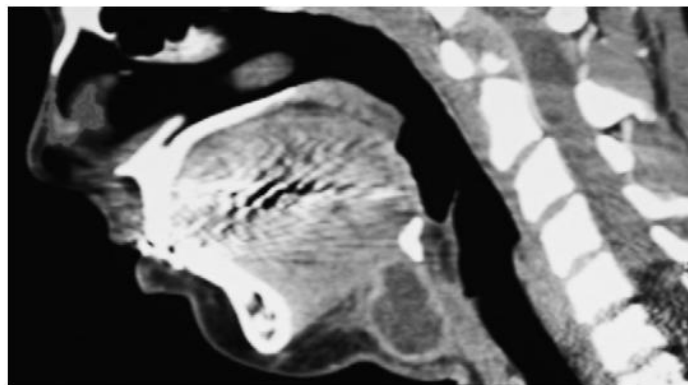
- **Thyroglossal Duct Cyst (TGDC):**
- Most common congenital cervical anomaly (**70%**).
- 2nd most common benign neck mass after Lymphadenopathy.
- 90% occurs before the age of 10.
- Failure of obliteration of Thyroglossal duct.
 - o Normally obliterates entirely during gestational weeks 7-10.



- **Clinical Picture:**
 - o Painless Midline Cystic Mass located anywhere from Thyroid cartilage up the Base of Tongue.
 - Location:
 - Suprahyoid: 20-25%
 - At level of hyoid bone: 15-50%
 - **Infrahyoid: 25-65% (Most common)**
 - If ruptures, it may form sinus or fistula that exits through overlying skin.
 - If infected, red warm painful mass.
 - Enlarge slowly with time.
 - Elevates with swallowing (attached to hyoid bone).
 - Elevates with tongue protrusion (attached to foramen cecum).
 - o Dysphagia and globus sensation.
 - o Normal thyroid function.
 - o **Median Ectopic Thyroid:**
 - Found in 1-2% of TGDC.
 - All functional thyroid tissue are located within the cyst.
 - Hypothyroidism
 - Diagnosed with Thyroid scan.
 - Removal of TGDC would make patient permanently dependent on thyroid replacement.



- Histopathology:
 - Lined with respiratory and squamous epithelium.
- Complications:
 - **Recurrent infections:**
 - Infected by way of the mouth.
 - Most common organisms are:
 - H. influenza.
 - Staph. Aureus.
 - Staph. Epidermidis.
 - **Malignancy:**
 - **Rare (1%).**
 - All types of thyroid ca except Medullary been reported.
 - Most common is Papillary Thyroid Ca (PTC).
 - Mainly in adults.
 - If TGDC is positive for PTC.
 - Do Thyroid Scan:
 - If Negative → No need for further intervention.
 - If Positive → Total Thyroidectomy with post-op Radiation.
- Investigations:
 1. **TSH:**
 - Most are Euthyroid.
 - If High TSH, Thyroid scan should be done to rule out median ectopic thyroid.
 2. **U/S Neck and Thyroid:**
 - Evaluate the TGDC.
 - Confirm the presence of normal thyroid gland.
 - Rule out presence of any ectopic thyroid tissue in the cyst.
 3. **FNA:**
 - Evaluate the TGDC.
 4. **CT Neck with contrast:**
 - Evaluate the TGDC.
 - Evaluate the anatomy
 5. **Thyroid Scan:**
 - Most sensitive to detect ectopic thyroid tissue.



- **Management:**

○ **Infected TGDC:**

- Oral Antibiotic.
- Aspiration if needed.
- Avoid I&D.

○ **Indications of Surgical Excision:**

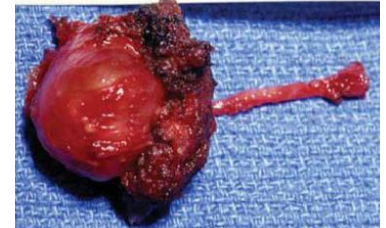
1. Confirm the diagnosis.
2. Prevent Recurrent infection.
3. Rule out presence of Malignancy.
4. Cosmetic.

○ **Sistrunk Procedure:**

- Resection of:
 1. Cystic mass
 2. Central portion of Hyoid.
 3. Deep tract to Foramen cecum.
- Low risk of recurrence (**5%**).

○ **Risk factors for TGDC Recurrence after surgery:**

- Incomplete (Simple) Excision (**50% Risk**).
- Intra-op cyst rupture.
- Infection.



- **Dermoid Cyst:**

- Benign lesions result from entrapment of epithelial elements along embryonic lines of fusion.
- Contains Ectodermal and Mesodermal elements only.
- Lined by epithelium but contain epithelial appendages, such as hair, hair follicles, or sebaceous glands.
- Diagnosed before age of 3 years old.
- Dermoid cysts of head and neck occur in T distribution.
 - Anterior neck and Midline of face and Orbits and Periauricular area.
 - Cervical dermoids are 20% of all head and neck dermoids.

- **Clinical Picture:**

- Painless superficial subcutaneous mass in anterior neck.
- Moves with the skin.
- Moves with swallowing or tongue protrusion if it was close to hyoid bone.
- Gradually increase in size over time due to accumulation of sebum.
- Infection is rare.
- Can rupture and present with granulomatous inflammation.

- **Investigations:**

1. U/S Neck:

- Delineate depth of lesion and its relationship to hyoid.

2. FNA:

- Helpful to distinguish between ruptured dermoid cyst and infected TGDC.

- **Management:**

- Complete simple excision.
 - If symptomatic, enlarging, or has ruptured.
- If it is attached to hyoid bone a Sistrunk procedure should be performed.
- Rate of recurrence is increased by incomplete resection or intraop rupture.

- **Teratoma:**

- Rare benign germ cell tumor (GCT).
- Composed of tissue foreign to site of origin.
- Contains all three germ layer (Ectoderm, Endoderm and Mesoderm).
- Most common germ cell tumor (GCT) in the pediatric age group.
- Head and neck teratoma represents less than 5% of all teratomas.

- **Typically displace rather than invade adjacent structures**

- **Clinical Picture:**

- Midline neck mass with lateral extension.
- Present at birth.
- Large and bulky
- Firm, mobile, multilobular, cystic mass with well-defined margins.
- Obstructive symptoms with rapid growth.
- Diagnosed at birth or even prenatally.
- Prenatal Maternal–Fetal Symptoms:
 - Polyhydramnios (30%).
 - Dystocia (difficult labour).
 - Preterm delivery or early fetal demise.

- **Investigations:**

1. Antenatal U/S:

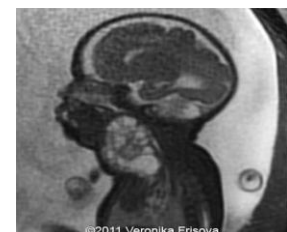
- Mixed echogenicity with multiloculated cystic and solid masses.
- Calcification in 50% of cases.

2. Fetal MRI:

- Better define the airway anatomy and the extent of the mass.
- Signal characteristics are heterogeneous on both T1 and T2 weighted sequences and highly variable dependent on tumor components.

3. CT Neck with Contrast:

- Defines location and extension of tumor.
- Reveals different densities and multiple calcified foci.
- Differentiate from lymphatic malformation.



- Histological Classifications:

1. Mature Teratomas:

- Most common.
- Completely differentiated.
- No risk for malignant transformation.

2. Immature Teratomas:

- Incompletely differentiated.
- **Risk of malignant transformation (5%).**
- Monitored by (AFP) and (b-hCG).

- Management:

- Airway management at birth is critical
- Delivery done in center prepared for high-risk deliveries.
- **EXIT Procedure:**
 - Ex Utero Intrapartum Treatment.
 - Elective modified C-section delivery under GA.
 - Infant is partially delivered but remains attached by its umbilical cord to placenta to ensure supplying oxygenated blood to the baby while the airway is being secured with ET tube or surgical tracheostomy.
 - Uterus and umbilical cord must stay relaxed.
 - Once the EXIT is complete, umbilical cord is clamped then cut and infant is fully delivered.
 - **Provides up to 30-45 minutes to secure a safe access to the airway.**



- **If No obvious airway obstruction at birth:**
 - Endotracheal intubation is advised to prevent risk of spontaneous intratumoral hemorrhage with subsequent airway obstruction.
- **Early definitive surgical resection:**
 - Performed after initial stabilization within 24–48 h.
 - Minimizes associated events such as pneumonia, atelectasis and aspiration.

- **Plunging Ranula:**
- Ranula describes a blue translucent painless swelling in floor of mouth reminiscent of underbelly of a frog.
- Plunging ranulas arise in neck due to penetration of oral ranula through dehiscence in anterior two thirds of Mylohyoid muscle.
- **Causes:**
 - **Congenital ranulas:**
 - Rare.
 - Imperforate salivary duct or ostial adhesion.
 - Resolve spontaneously.
 - **Post-traumatic ranulas:**
 - Trauma to sublingual gland leading to duct obstruction.
 - Secretory backpressure builds and acini rupture leading to mucus extravasation and formation of a **pseudocyst**.
 - Does not have an epithelial lining.



- **Clinical Picture:**
 - Midline submental neck mass.
 - Painless.
 - Gradually increases in size.
 - In conjunction with, or independent of oral ranula.
 - History of oral trauma or surgery.
- **Investigations:**
 1. **U/S:**
 - Confirms cystic nature of the lesion.
 - Evaluates integrity of mylohyoid muscle.
 2. **CT Neck:**
 - Homogeneous cyst in submandibular or parapharyngeal space that abuts sublingual space.
 3. **Aspiration:**
 - Mucus with prominent histiocytes.
 - High amylase and protein content.

- **Management:**

○ **Ranula:**

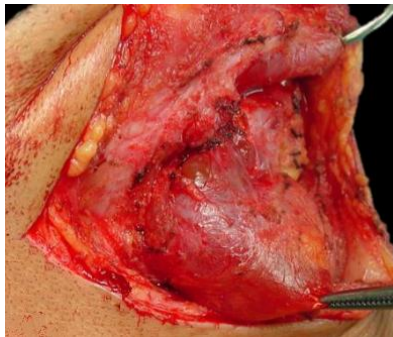
- Observation.
- Marsupialization:
 - Most widely treatment for ranulas.
 - Unroofing the cyst and tacking edges of the cyst to adjacent tissue.
 - Good for small cysts <1.5 cm are usually more superficial in nature and may respond more readily to marsupialization.
 - Failure rates range from 60%.
 - Packing the cyst cavity with gauze for 7-10 days improves the success rate.
- Marsupialization with placement of suture:
 - With micro-marsupialization, silk suture can be placed through surface of ranula.
 - Left in place a minimum of 7 days while an epithelial tract forms to allow for mucus drainage between the surface and the underlying salivary glandular tissue. Morbidity is minimal to nonexistent, and recurrence or treatment failure is the primary complication. This can also be performed in the office.
- Excision of ranula:
 - Recurrence rates 60%
- Sclerosing agents:
 - Bleomycin and OK-432 have been used with success in treatment of ranulas.
- Sublingual gland excision:
 - Standard for treatment of ranulas is excision of the sublingual gland.
 - Removes source of mucus and significantly decreases risk for recurrence.
 - Recurrence rates of 2%.

○ **Plunging Ranula:**

- Transoral approach:
 - Provides better access for complete removal of the sublingual gland.
 - Some surgeons advocate simply draining the cervical portion of the ranula and excising the gland transorally.
 - Complete excision of the cyst is not necessary if the gland itself is excised.
 - A biopsy of the cyst wall is recommended for tissue confirmation.

▪ Transcervical approach:

- Complete removal of sublingual gland is difficult with this approach.
- Requires division of mylohyoid muscle and dissection up to floor of mouth.
- Some surgeons recommend a transoral excision of sublingual gland with drainage of the cyst first.
 - If that is unsuccessful, complete excision of cyst via a transcervical approach is indicated.
 - A transcervical approach is also indicated for ranulas located exclusively in the neck.



- **Thymic Cyst:**

- Thymus developed from left and right ventral part of 3rd Pharyngeal pouch then descends to below clavicles.
- Thymic cyst is remnant of thymus along migration path.
- Present is midline neck mass in lower neck.
- Very rare.

- **Investigations:**

1. U/S

2. CT Neck

3. MRI:

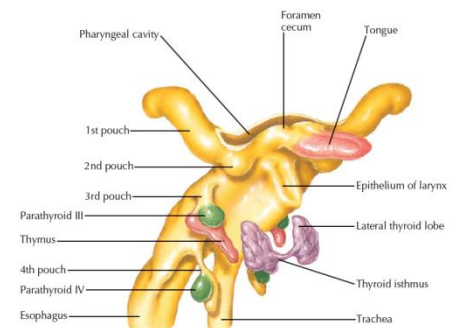
- Unicystic → Thymic cyst.
- Multicystic → Lymphatic malformation.

4. Serum Calcium:

- Associated parathyroid disorders.
- DiGeorge's syndrome.

- **Management:**

- **Surgical Excision (may require thoracic surgery).**



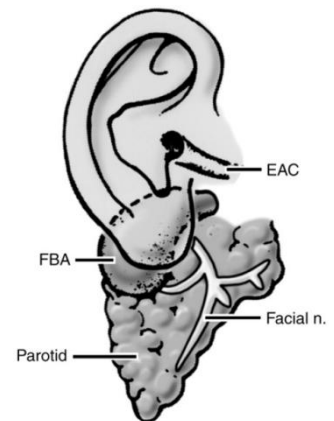
- **Ectopic Thyroid:**
- Occurs anywhere along the path of initial descent of the thyroid from Foramen cecum to Sternal notch when migration is arrested.
- **Sites:**
 - Base of tongue (Lingual Thyroid) in 90%.
 - Within the tongue.
 - Anterior neck.
- **Clinical Picture:**
 - Lingual thyroid:
 - Dysphagia
 - Dysphonia
 - Airway obstruction
 - Bleeding.
 - Midline neck mass
 - Hypothyroidism
- 70-100% of patients with Lingual thyroid have no other thyroid tissue.



- **Management of Lingual thyroid:**
 - **Thyroid suppression:**
 - For mild cases.
 - **Surgical Excision:**
 - For severe cases with airway obstruction or bleeding.
 - Approaches:
 - Transoral approach.
 - Transhyoid pharyngotomy.
 - Lateral pharyngotomy.
 - Place sutures around Lingual thyroid prior to excision that are left long and then tied after mass is excised to decrease bleeding.
 - Autotransplantation of excised ectopic thyroid is not advised due to small chance of presence of Thyroid Ca.

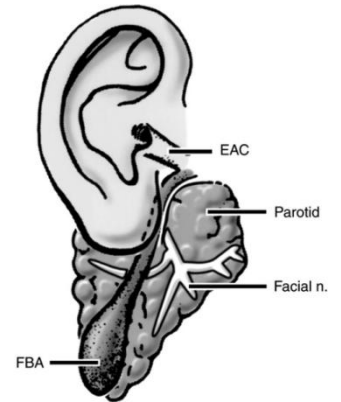
Lateral Congenital Neck Masses:

- **Branchial Cleft Anomalies:**
- 30% of congenital neck masses.
- Equally common in males and females.
- Present in childhood or early adulthood.
- **Pathophysiology:**
 - o Incomplete obliteration of branchial clefts and pouches.
 - o Present as cysts, sinuses, or fistulae in lateral neck.
 - Cysts are lined by squamous epithelium.
 - Sinuses and fistulae are lined by ciliated columnar epithelium.
- Squamous Cell Ca can be found within branchial lesions in adults.
 - o Rare.
 - o Difficult to distinguish between a primary lesion from an anomaly and a metastatic lesion from an occult primary.
- **Clinical Picture:**
 - o Painless Lateral Cystic Mass.
 - o Located anterior to SCM, deep to platysma.
 - o Fistulas and sinuses may express mucoid discharge.
- **Investigations:**
 1. **U/S**
 2. **FNA:**
 - Confirm the diagnosis.
 - Rule out carcinoma
 3. **CT Neck with contrast:**
 - Study of choice.
 - Demonstrate the fistula in 65%.
 4. **Upper Airway Endoscopy:**
 - Locate the internal opening.
 5. **Modified Barium Swallow:**
 - 50-80% sensitivity for third and fourth branchial fistulae.
- **1st Branchial Cleft Cyst:**
- Rare.
- 1% of branchial cleft malformations.
- Related to auricle.
 1. **Work Type I:**
 - Less common.
 - Duplicated EAC.
 - Contains ectodermal elements only.
 - Begin periauricularly, pass lateral (superior) to Facial nerve, parallel to EAC, End as a blind sac near mesotympanum.



2. Work Type I:

- Most common.
- Contains ectodermal and mesodermal elements.
- Presents near angle of mandible.
- Passes through Parotid, lateral or medial to Facial nerve, end near or into EAC.

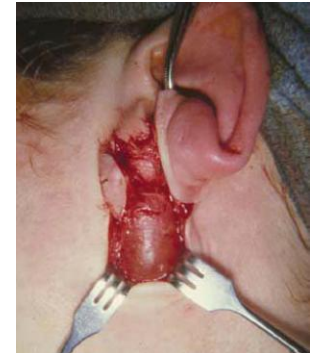


- Management:

- Complete surgical excision to reduce recurrence.
- Risk of Facial nerve injury.
- May need superficial parotidectomy.

- 2nd Branchial Cleft Cyst:

- Most common type.
- 95% of all brachial cleft malformations.
- Runs deep to 2nd arch structures and superficial to 3rd arch structures.



- Pathway:

- Cyst along Anterior border of SCM in lateral mid neck region.
- Runs between Internal and External Carotid arteries.
- Superior to Glossopharyngeal and Hypoglossal Nerve.
- Ends at Tonsillar fossa.

- Management:

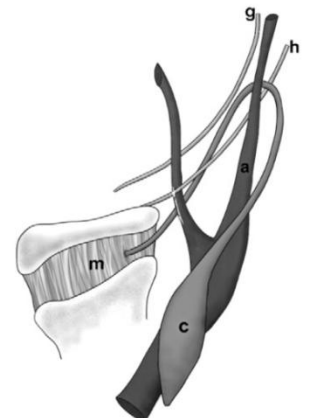
- Complete surgical excision to reduce recurrence.
- Some advocate excision of Tonsil on side of lesion.
- Most surgeons do not excise tonsil and tie off the tract close to tonsillar fossa.

- 3rd Branchial Cleft Cyst:

- Rare.
- Misdiagnosed as neck abscess or recurrent thyroiditis.
- Impossible to differentiate between 3rd and 4th BCA without surgical dissection.
- Runs deep to 3rd arch structures and superficial to 4th arch structures.
- Over 90% occur on Left side.
- Can cause hypoglossal nerve palsy if infected.

- Pathway:

- Cyst along Anterior border of SCM in lateral lower neck.
- Tract ascends deep to Internal carotid artery.
- Passes medially between Hypoglossal and Glossopharyngeal nerves.
- Pierces thyroid membrane above Internal branch of Superior Laryngeal Nerve.
- Enter the Upper Pyriform sinus.



- **Management:**

- Complete surgical excision to reduce recurrence.
- Inspection of Pyriform sinus should precede surgical exploration.
- Hemi-thyroidectomy may be required for lesions associated with thyroid gland.



- **4th Branchial Cleft Cyst:**

- Very rare.
- Misdiagnosed as neck abscess or recurrent thyroiditis.
- Impossible to differentiate between 3rd and 4th BCA without surgical dissection.
- Over 90% occur on Left side.

- **Pathway:**

- Cyst along Anterior border of SCM in lateral lower neck.
- Loops around Aortic arch medial to ligamentum Arteriosus in Left side.
- Loops around Subclavian artery in the Right side
- Ascends to the level of Hypoglossal nerve.
- Enter the pharynx at Pyriform apex or cervical esophagus.



- **Management:**

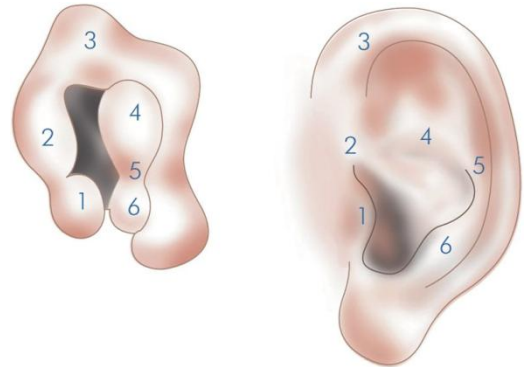
- Complete surgical excision to reduce recurrence.
- Inspection of Pyriform sinus should precede surgical exploration.
- Hemi-thyroidectomy may be required for lesions associated with thyroid gland.

TABLE 106.1	ANATOMIC PATHWAYS FOR DEEP TRACT OF FIRST, SECOND, THIRD, AND FOURTH BCAS
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- First BCA
 - The tract may run medial or lateral to the main trunk of the facial nerve or between branches.
- Second BCA
 - The deep tract runs between the internal and external carotid arteries and superior to the glossopharyngeal and hypoglossal nerves and finally ends at the tonsillar fossa.
- Third BCA
 - The deep tract passes posterior to the internal and external carotid arteries, between the 9th and 12th cranial nerves, and ends in the apex of the pyriform sinus.
- Fourth BCA
 - The deep tract begins with a sinus at the apex of the pyriform sinus and travels inferiorly in the tracheoesophageal groove, posterior to the thyroid gland, and into the thorax. Next, it loops below the aorta on the left or below the subclavian artery on the right and then ascends posterior to the common carotid artery to loop over the hypoglossal nerve and ends at the anterior border of the sternocleidomastoid muscle.

- **Preauricular Cyst and Sinus:**

- Auricle is formed from fusion of 6 Hillocks of His which derived from 1st and 2nd Branchial arches.
 - Tragus develops from tubercle of 1st Branchial Arch.
 - Rest of Auricle develops from remaining 5 tubercles of 2nd Branchial Arch.
- Faulty fusion between 1st and 2nd Branchial arches tubercles causes Preauricular sinus or cyst which is commonly seen between tragus and crus of helix.



- **Pathway:**

- Punctum anterior to Root of Helix.
- Short tract and sinus which ends with an attachment to perichondrium of Root of Helix.



- **Clinical Picture:**

- Asymptomatic.
- Discharging a small amount of mucoid discharge.
- Repeated infection.

- **Management:**

- Complete surgical excision to reduce recurrence.
 - Punctum is excised by Elliptical incision.
 - Tract and sinus are traced to Perichondrium of Root of Helix.
 - Perichondrium is scored sharply and peeled off the cartilage.
 - Full thickness of cartilage is excised to assure complete removal in case of recurrence.
 - **No risk for Facial nerve injury since it is deeper and inferior to area of dissection.**
- Infected preauricular sinus should be managed with antibiotics then a period of 6 weeks is allowed before surgical excision.



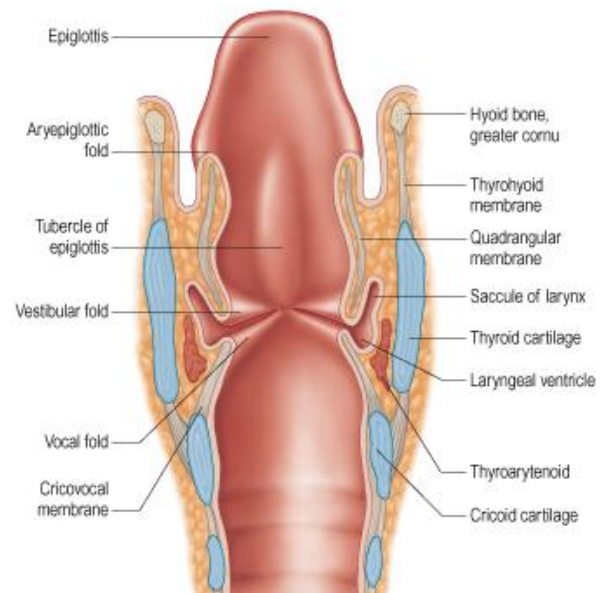
- **Branchio-Oto-Renal Syndrome (BOR):**
- Melnick Fraser Syndrome.
- Abnormal development of branchial arches and kidneys.
- Autosomal dominant.
- Can be detected prenatally with genetic testing.
- Family members should be examined and tested.
- Clinical Picture:
 - o Preauricular sinus.
 - o Branchial Cleft anomalies.
 - o Pinna deformities (maybe Normal).
 - o Hearing loss (CHL or SNHL).
 - o Renal anomalies (maybe Normal).

**TABLE
106.2**

**CLINICAL FINDINGS IN BOR
(MELNICK FRASER) SYNDROME (10)**

- Hearing loss
- Preauricular pits
- Branchial fistulas or cysts
- Anomalous pinna
- Renal dysplasia

- **Laryngocele:**
- Laryngocele is **Expanding air-filled cyst** formed in Laryngeal Saccule with communication to laryngeal lumen.
- Ventricle of larynx opens into **Sacculle**.
 - o blind-ended pouch ascends forwards from ventricle between vestibular fold and thyroid cartilage.
 - o 60-70 mucous glands open onto its luminal surface help to lubricate vocal cords which lack mucous glands.



- **Laryngopyoce** is **infected pus-filled** laryngocele.
- **Saccular Cyst** is **Fluid-filled dilation** of Saccule without communication to Laryngeal lumen.

- **Pathophysiology:**

- Congenital or acquired Laryngocele is due to expansion from prolonged increased intra-glottic pressure of laryngeal saccule (glass blowers, trumpet players).
- Congenital or acquired Saccular cyst is due to obstruction of ventricular aditus by inflammation, scarring or by tumor.

- **Types:**

- **External Laryngocele:**
 - Laryngocele sac protrudes through Thyrohyoid membrane at point of entry superior laryngeal vessels and Internal branch of superior laryngeal nerve.
 - Presents as Lateral Neck mass.
 - More common.
- **Internal Laryngocele:**
 - Laryngocele sac remains within thyroid cartilage.
 - Expands into Paraglottic space and extend superiorly to expand Aryepiglottic fold and reach Vallecula.
 - Less common.
- **Combined.**

- **Clinical Picture:**

- Lateral Neck mass.
 - Compressible.
 - Increases in size with increasing intra-laryngeal pressure (Valsava).
- Dysphonia.
- Dysphagia.
- Stridor.
- Chronic cough.



- **Investigations:**

1. Flexible Nasopharyngoscopy.

2. U/S.

3. CT Neck:

- Study of choice.
- Presence of air within the lesion differentiates Laryngocele from Saccular cysts.

- **Management:**
 - **Endoscopic Excision:**
 - Mainstay of treatment for Internal Laryngocele.
 - Low recurrence rate.
 - **Open surgical approach:**
 - For External Laryngocele.
- **Pseudotumor of infancy (Fibromatosis Colli):**
- Congenital muscular torticollis.
- Firm lateral neck mass within SCM in neonatal period.
- Affected babies aged 2-4 weeks.
- **Pathophysiology:**
 - Intrauterine or birth trauma (Breech delivery or Forceps)
 - Causing SCM muscle injury, hematoma, and resultant fibrosis.
 - Fibrosis will cause shortening of SCM results in Torticollis.
- **Clinical Picture:**
 - Lateral Neck mass.
 - Firm and often rock-hard.
 - Unilateral (Right side in 70%).
 - Noticed 2-4 weeks after birth.
 - Prominent with turning of infant's face in opposite direction and tilting of head toward the mass.
 - Tilting of head to diseased side.
- **Investigations:**
 1. **U/S:**
 - Confirm the diagnosis.
 2. **CT Neck:**
 - Rule out malignancy or a congenital lesion.
 - Presence of air within the lesion differentiates Laryngocele from Saccular cysts.
- **Management:**
 - **Conservative:**
 - Usually resolves within 4-8 months.
 - When torticollis is present, Physical therapy is recommended.
 - 10-20% of patients may progress to persistent muscular torticollis with risk of craniofacial deformity.
 - **Surgical Release of SCM muscle:**
 - Indications:
 - Persistent beyond 12 months of age despite adequate physical therapy.
 - Presence of early signs of Craniofacial asymmetry.



Pediatric Syndromes:

Terminology:

1. Isolated Anomaly:

- Defect in a single system.
- Cleft Palate.

2. Syndrome:

- Recognizable pattern of multiple anomalies due to a single known etiology.
- Down Syndrome:
 - Trisomy 21.

3. Sequence:

- Group of anomalies rising secondary to an initial malformation.
- Pierre-Robin sequence:
 - Underdevelopment of the lower jaw causes displacement of the tongue and subsequent cleft palate.

4. Association:

- Multiple anomalies occurring at a high frequency together without a known etiology.
- VACTERL Association:
 - Vertebral, Anal, Cardiac, TEF, Renal, Limb defects.

Summary of Syndromes:

1. Down syndrome (Trisomy 21):

○ **Manifestations:**

▪ **Airway (Multiple level obstruction):**

- Macroglossia and glossoptosis
- Abnormally narrow upper airway
- Adenotonsillar and lingual tonsils hypertrophy
- Hypopharyngeal collapse
- Laryngomalacia
- Subglottic stenosis
- Laryngeal cleft



▪ **Otology:**

- CHL (EAC stenosis, cerumen impaction, OME, CSOM, ossicular abnormalities).
- Inner ear dysplasia

▪ **Others:**

- Obesity
- Generalized hypotonia
- Atlantoaxial instability (25%)

○ **Treatment:**

▪ **Airway:**

• Adenotonsillectomy (+/- CPAP):

- Pre-op polysomnography.
- Pre-op flexion and extension lateral cervical x-ray.
- Intubation with ETT 2 sizes smaller.
- Head should to be remain in a neutral position.
- No improvement post-op (50%).
- Higher risk of adenoid recurrence.
- Need post-op overnight observation

- GERD treatment.

▪ **Otology:**

- Regular follow up with hearing assessment (3-6 months).
- Regular dewaxing.
- Hearing Aids.
- VTs (can be repeated multiple times).
- Tympanomastoidectomy.

2. CHARGE syndrome (AD / CHD7 gene mutation in chromosome 8):

- **Airway:**
 - Choanal atresia (60% vs 100%)
 - Tracheoesophageal Fistula
 - Laryngeal cleft
- **Otology:**
 - External ear anomalies.
 - CHL (Ossicular abnormalities)
 - SNHL (Mondini malformation)
- **Others:**
 - **C**oloboma
 - **H**ear disease
 - **A**tresia of choanae
 - **R**etardation (CNS)
 - **G**enital hypoplasia
 - **E**ar

Major	Minor	Diagnosis
1. Ocular coloboma	5. Cardiovascular malformations	Typical CHARGE: four majors or three majors and three minor
2. Choanal atresia	6. Genital hypoplasia	
3. Characteristic ear abnormalities	7. Cleft lip/palate	
4. Cranial nerve abnormalities including SNHL	8. Tracheoesophageal fistula	
	9. Hypothalamo-hypophyseal dysfunction	
	10. Distinctive CHARGE facies	
	11. Developmental delay	

3. VACTERL association:

- **Airway:**
 - Tracheoesophageal Fistula
 - Laryngeal cleft
 - Choanal atresia
- **Otology:**
 - Ear anomalies
- **Others:**
 - **V**ertebral anomalies
 - **A**nal Atresia
 - **C**ardiac anomalies
 - **T**racheo-esophageal fistula
 - **E**ar anomalies
 - **R**enal anomalies
 - **L**imb anomalies

4. **Opitz G/BBB Syndrome** (MID1 gene mutation of chromosome 22):

- **Airway:**
 - Laryngeal cleft
- **Others (Midline defects):**
 - Cleft lip/palate
 - Hypospadias
 - Hypertelorism

5. **Pallister-Hall syndrome** (GLI3 gene mutation):

- **Airway:**
 - Laryngeal cleft
 - Bifid epiglottis
- **Others:**
 - Hypothalamus abnormalities (Pan-hypopituitarism).
 - Distal extremity anomalies (syndactyly and postaxial polydactyly).
 - Imperforate anus.

6. **DiGeorge/Velo-cardio-facial syndrome** (Microdeletion of 22q11):

- **Diagnosis:**
 - Fluorescent In Situ Hybridization (FISH)
- **Airway:**
 - Anterior glottis web
 - Velopharyngeal insufficiency
 - Medialized carotid arteries:
 - Contraindication for pharyngeal flap surgery for VPI.
- **Others (CATCH-22):**
 - **C**ardiac (Congenital heart disease)
 - **A**bnormal facies
 - **T**hymic aplasia (immunodeficiency)
 - **C**left lip/palate
 - **H**ypo-parathyroidism (Hypocalcemia, agenesis of parathyroid glands)
 - **22q11**



7. Pierre-Robin Sequence:

- **Diagnosis (Triad):**
 - Micrognathia
 - Glossoptosis
 - Cleft palate
- **Associations:**
 - Stickler
 - DiGeorge/Velo-cardio-facial syndrome
 - Treacher Collins
 - Goldenhar
- **Treatment:**
 - Conservative:
 - Prone or decubitus positioning
 - Nasopharyngeal airway
 - CPAP
 - Surgery:
 - Tracheostomy
 - Mandibular distraction osteogenesis
 - Tongue–lip adhesion
 - Floor of mouth muscular release

8. Usher Syndrome (AR):

- Most common cause of autosomal recessive syndromic HL.
- Most common cause of deaf-blindness (dual sensory impairment).
- **Manifestations:**
 - **Otology:**
 - Congenital SNHL.
 - **Others:**
 - Progressive retinitis pigmentosa.

9. Pendred Syndrome (AR):

- 2nd most common cause of autosomal recessive syndromic HL.
- **Manifestations:**
 - **Otology:**
 - Congenital SNHL (Mondini and enlarged VAD).
 - **Others:**
 - Multinodular goiter.

10. Goldenhar Syndrome/Hemifacial Microsomia (AR):

- Oculo-auriculo-vertebral Syndrome.
- Affecting 1st and 2nd branchial arch structures.
- Typically unilateral on RIGHT side.
- **Manifestations:**
 - **Otology:**
 - Microtia/aural atresia
 - Ossicular malformation
 - Abnormal FN
 - SNHL
 - **Others:**
 - Hemifacial Microsomia.
 - Ocular abnormalities.
 - Vertebral abnormalities (fusion or absence of cervical vertebrae).



11. Waardenburg Syndrome (AD):

- Most common cause of autosomal dominant syndromic HL.
- **Manifestations:**
 - **Otology:**
 - SNHL
 - **Others:**
 - Heterochromia iridis
 - White forelock
 - Vitiligo



12. Stickler Syndrome (AD):

- Progressive Arthro-Ophthalmopathy.
- **Manifestations:**
 - **Otology:**
 - Progressive SNHL
 - **Others:**
 - Ocular (myopia, retinal detachment, cataracts)
 - Marfanoid habitus (tall and thin)
 - Arthritic abnormalities (joint hypermobility, arthritis)
 - Pierre-Robin sequence

13. Branchio-Oto-Renal/Melnick-Fraser Syndrome (AD):

- Progressive Arthro-Ophthalmopathy.
- **Manifestations:**
 - **Otology:**
 - Pinna deformities
 - Preauricular ear pits, fistulas, or tags
 - Ossicular malformations
 - Cochlear malformations (Mondini or enlarged VAD)
 - **Others:**
 - Renal abnormalities (agenesis, dysplasia)
 - Branchial anomalies
 - Lacrimal duct stenosis

14. Treacher Collins Syndrome (AD):

- Mandibulofacial Dysostosis.
- Malformation of 1st and 2nd branchial arches.
- Normal IQ.
- **Manifestations:**
 - **Airway:**
 - Choanal atresia
 - **Otology:**
 - Microtia/aural atresia.
 - Ossicular abnormalities.
 - **Others:**
 - Retro-gnathia
 - Macrostomia.
 - Lower eyelid coloboma



15. Crouzon Syndrome (AD):

- Craniofacial Dysostosis.
- Normal IQ.
- **Manifestations:**
 - **Airway:**
 - Choanal atresia
 - **Otology:**
 - CHL (EAC stenosis, ossicular abnormalities).
 - **Others:**
 - Craniosynostosis (premature closure of cranial sutures)
 - Proptosis
 - Hypertelorism
 - Malocclusion
 - Cleft palate
 - Short upper lip



16. Alport Syndrome (X-linked):

- Mutation in type IV collagen gene.
- **Manifestations:**
 - **Otology:**
 - Progressive SNHL
 - **Others:**
 - Renal dysplasia/agenesis
 - Progressive nephritis

Table 19-1. Syndromes associated with hearing loss

Syndrome	Features	Type of hearing loss	Onset (Congenital/Delayed)	Type of inheritance
1. Waardenburg's syndrome (Fig. 19.1)	- White forelock - Heterochromia iridis - Vitiligo - Dystopia canthorum	Unilateral or bilateral SNHL	Congenital	AD
2. Usher syndrome	- Retinitis pigmentosa - Night-blindness	SNHL	Delayed	AR
3. Jervell and Lange-Nielson's syndrome	- Repeated syncopal attacks - Prolonged QT interval in ECG	SNHL	Congenital	AR
4. Pendred syndrome	- Goitre (non-toxic) usually evident before puberty - Perchlorate discharge test shows defect in organic binding of iodine	SNHL	Congenital	AR
5. Alport syndrome	- Hereditary progressive glomerulonephritis - Corneal dystrophy	Progressive SNHL	Delayed	AD or X-linked
6. Treacher-Collins syndrome (mandibulofacial dysostosis)	- Antimongoloid palpebral fissures - Coloboma of lower lid - Hypoplasia of mandible and malar bones - Malformed pinna and meatal atresia - Malformed malleus and incus (stapes normal)	Conductive	Congenital	AD
7. Crouzon's syndrome (craniofacial dysostosis)	- Frog eyes (exophthalmos with divergent squint) - Hypertelorism - Parrot-beak nose - Mandibular prognathism - Premature closure of cranial sutures with mental retardation	Conductive or mixed	Congenital	AD
8. Apert's syndrome	- Syndactyly - All other features of Crouzon's syndrome	Conductive (Stapes fixation)		AD
9. Klippel-Feil syndrome	- Short neck - Fused cervical vertebrae - Spina bifida - Atresia of ear canal	SNHL or mixed	Congenital	AR
10. Wildervanck syndrome	- Klippel-Feil syndrome - SNHL - CN VI Paralysis	SNHL	Congenital	X-linked
11. Branchio-oto-renal syndrome	- Branchial fistulas/cysts - Malformed pinnae with preauricular pits or sinuses - Renal abnormalities	Conductive or mixed	Congenital	AD
12. Stickler's syndrome	- Small jaw - Cleft palate (Pierre-Robin sequence) - Myopia ≥ retinal detachment - Cataract - Juvenile onset arthritis	Conductive or SNHL	Delayed	AD
13. Van der Hoeve's syndrome	- Osteogenesis imperfecta with history of fractures - Blue sclera - Hearing loss (delayed onset)	Conductive, SNHL or mixed (like otosclerosis)	Delayed	AD
14. Pierre-Robin sequence*	- Micrognathia - Glossoptosis - Cleft palate - Often part of Stickler's syndrome	SNHL Conductive loss		AD
15. Goldenhar's syndrome (Facio-auriculo-vertebral dysplasia) or (oculo-auriculo-vertebral [OAV] syndrome)	- Facial asymmetry - Low set ears, atresia of ear canal - Cardiac abnormalities - Preauricular tags/pits - Hemivertebrae in cervical region - Epibulbar dermoid - Coloboma of upper lid	Mixed or Conductive	Congenital	AD or sporadic Extra chromosome Multifactorial (genetic and environmental)
16. Down's syndrome (Trisomy 21)	- Microcephaly - Mental retardation/delayed development - Short stature - Epicentral folds - Stenosis of ear canal - High incidence of serous otitis media - Atlanto-axial instability	Conductive	Congenital	Extra chromosome

Syndromes associated with Airway anomalies:

- **SGS:**
 1. Down syndrome
- **Laryngeal Web:**
 1. DiGeorge/Velo-cardio-facial syndrome
- **Laryngeal Cleft:**
 1. VACTERL association
 2. CHARGE syndrome
 3. Opitz G/BBB Syndrome
 4. Pallister-Hall syndrome
 5. Down syndrome
- **Tracheoesophageal Fistula:**
 1. VACTERL association
 2. CHARGE syndrome
- **Choanal Atresia:**
 1. CHARGE syndrome
 2. VACTERL association
 3. Crouzon Syndrome (Craniofacial Dysostosis)
 4. Treacher Collins Syndrome (Mandibulofacial Dysostosis)
- **Cleft Palate:**
 1. Pierre-Robin Sequence
 2. CHARGE syndrome
 3. DiGeorge/Velo-cardio-facial syndrome
 4. Opitz G/BBB Syndrome
 5. Crouzon Syndrome

Syndromes associated with Ear anomalies:

- **Microtia/Aural atresia:**
 1. Treacher Collins Syndrome (Mandibulofacial Dysostosis)
 2. Goldenhar's/Hemifacial macrosomia
 3. CHARGE syndrome
 4. Crouzon Syndrome (Craniofacial Dysostosis)
 5. Branchio-Oto-Renal/Melnick-Fraser Syndrome
- **HL and Renal disease:**
 1. Branchio-Oto-Renal Syndrome (Melnick-Fraser Syndrome)
 2. Alport Syndrome
- **HL and Retinitis pigmentosa:**
 1. Usher Syndrome
- **HL and Thyroid disease:**
 1. Pendred Syndrome
- **HL and Facial nerve abnormalities:**
 1. Goldenhar Syndrome/Hemifacial Microsomia
- **HL and Pigmentary abnormalities:**
 1. Waardenburg Syndrome
- **HL and Joint abnormalities:**
 1. Stickler Syndrome
- **HL and Cleft palate:**
 - CHARGE syndrome/Stickler Syndrome / Crouzon Syndrome

Head and Neck:

- 1- Principles of Head and Neck Oncology**
- 2- Thyroid - Embryology, Anatomy and Physiology**
- 3- Approach for Evaluation of Thyroid Nodules**
- 4- Benign Thyroid Diseases**
- 5- Malignant Thyroid Tumors**
- 6- Parathyroid - Embryology, Anatomy and Physiology**
- 7- Diseases of Parathyroid Glands**
- 8- Salivary Glands - Embryology, Anatomy and Physiology**
- 9- Approach to Salivary Gland Pathology**
- 10- Benign Salivary Gland Diseases**
- 11- Salivary Glands Tumors**
- 12- Parapharyngeal Space Tumors**
- 13- Paraganglioma**
- 14- Anatomy of Oral Cavity**
- 15- Tongue - Embryology, Anatomy and Physiology**
- 16- Common Disorders of Oral Cavity**
- 17- Premalignant Oral Ulcers & Lesions**
- 18- Oral Cavity Cancer**
- 19- Nasopharyngeal Tumors**
- 20- Oropharyngeal Tumors**
- 21- Hypopharyngeal Cancer**
- 22- Early Laryngeal Cancer**
- 23- Advanced Laryngeal Cancer**
- 24- Lymphoma**
- 25- Orbital Anatomy and Tumors**
- 26- Odontogenic Cysts and Tumors**
- 27- Cutaneous Malignancy**
- 28- Melanoma**
- 29- Head and Neck Reconstruction Pearls**

Principles of Head and Neck Oncology:

- H&N cancers makes up about 10% of all malignancies excluding skin.
- SCC arises from genetic alterations to genes regulates cellular growth and death.
 - Mainly acquired from exposure to environmental agents.
 - Less commonly inherited.

- Risk Factors for H&N Cancers:

1. Alcohol and Tobacco Abuse:

- Common etiologic factors in cancers of oral cavity, oropharynx, hypopharynx, and larynx.
- Tobacco abuse includes smoking cigarettes, cigars, pipes or chewing and snuffing tobacco.
- High risk for developing second primary neoplasms of H&N, lung, esophagus due to exposure of entire aerodigestive tract to these carcinogens.

2. Human Papilloma Virus (HPV):

- Risk factor for oropharyngeal SCC (Lingual and palatine tonsils, and base of tongue) in younger men who are nonusers of tobacco and alcohol.
- Mainly HPV type 16.

3. Epstein-Barr Virus (EBV):

- Primary risk factor for Nasopharyngeal carcinoma and oral hairy leukoplakia.

4. Human Immunodeficiency Virus (HIV):

- 2-3 fold increase in incidence of HNSCC.
- Increases incidence of non-AIDS-defining malignancies:
 - Anogenital Cancer, Hodgkin Lymphoma, Testicular Germ Cell Tumors, Non-small-cell Lung Cancer, Hepatocellular Cancer.

5. Herpes Simplex Virus (HSV):

- Less strongly correlated with development of Oral Cancer than EBV or HPV.
- HSV can transform cells in vitro to a malignant phenotype.

6. Radiation:

- Prior irradiation is risk factor for thyroid, salivary gland tumors, HNSCC and sarcomas.

7. Betel Nut Chewing:

- Independent risk factor for HNSCC, esophageal and hepatocellular Cancer.
- Synergistic effect with tobacco and alcohol.

8. Occupational Rxposure:

- Dry cleaning agent perchloroethylene, asbestos, pesticides, wood workers, plastic and rubber products, naphthalene refiners, ethanol, formaldehyde, sulfuric acid mist, leather and paint workers, automobile mechanics, cement and metal workers.

9. Diet:

- Protective effect of H&N cancers is associated with increased consumption of fruits and vegetables.
- Risk of Nasopharyngeal carcinoma is increased in frequent consumers of preserved meats contains high levels of added nitrites.

10. Genetic Factors:

- H&N cancers in these patients tend to arise at an earlier age and in the absence of other risk factors.

11. Other Risk Factors:

- Poor oral hygiene and Periodontal disease are linked with carcinoma of oral cavity.
 - Dental prostheses or poorly fitting dentures do not appear to be an independent risk factor for development of oral carcinoma.
- HNSCC starts on the surface and spreads superficially, deeply, and submucosally.
- Lymphatic metastasis are common in advanced tumors.
 - Distant metastasis are rare
 - Status of the cervical lymph nodes is the most important prognostic factor in SCCA of the upper aerodigestive tract.
 - **Cure rates drop in half (50%) when there is regional lymph node involvement.**
- **General Classification of H&N Tumors:**
- **Mucosal Tumors:**
 1. Sinonasal
 2. Oral.
 3. Pharyngeal.
 4. Laryngeal
 - **Non-Mucosal Tumors:**
 1. Thyroid and Parathyroid.
 2. Salivary Glands.
 3. Paragangliomas.
 4. Skin.

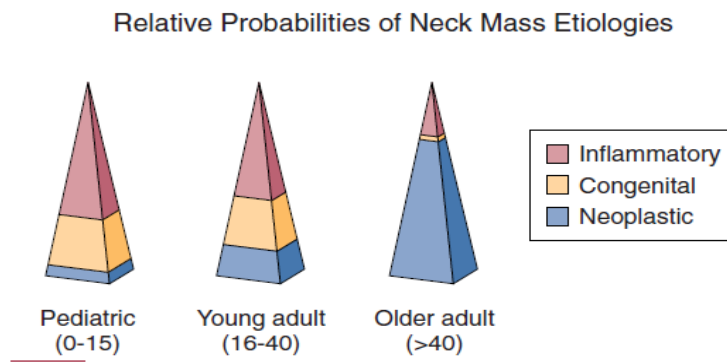
Evaluation of H&N Patient in Clinic:

History (8 Points):

1. Age and Gender
2. Chief complain
3. History of Present Illness:
 - Site
 - Onset
 - Duration
 - Progression
 - Associated symptoms.
4. Past Medical and Surgical History.
5. Medications:
 - Blood Thinners.
6. Allergies
7. Social History:
 - Alcohol and smoking
8. Family History of Tumors.

- Important Keypoints in History:

- **Any neck mass in an adult over the age of 40 years is malignant until proven otherwise.**
- **Any unilateral ENT Sign and Symptom is malignant until proven otherwise.**



- **Otological Symptoms:**
 - Persistent otalgia (referred pain with normal otologic exam).
 - Hearing loss.
 - Aural fullness.
 - Pulsatile tinnitus.
- **Nasal/Paranasal/Nasopharyngeal Symptoms:**
 - Recurrent Epistaxis.
 - Unilateral Nasal Obstruction.
 - Persistent Rhinorrhea or Sinusitis.
- **Oral/Pharyngeal Symptoms:**
 - Persistent sore throat (>3 weeks).
 - Odynophagia.
 - Dysphagia.
 - Trismus.
 - Presence of nonhealing ulcers.
 - Halitosis.
 - Numbness in the lower teeth.
- **Laryngeal Symptoms:**
 - Persistent hoarseness and throat pain (>3 weeks).
 - Difficulty breathing.
- **Neck Symptoms:**
 - Character and duration of neck masses.
- **Neurological Symptoms:**
 - Diplopia.
 - Cranial nerve palsies.
 - Mental status changes.
- **Risk Factors:**
 - Pack-year history of smoking.
 - Alcohol use.
 - Tobacco abuse.
 - Sun exposure.
 - Previous cancers.
 - Family history of cancer.
 - Radiation exposure.
 - Exposure to wood dust or heavy metals.
- **Constitutional Symptoms:**
 - Extent of weight loss.
 - Bone pain.
 - Hemoptysis
 - Malaise.
 - Anorexia.

Physical Examination (8 Areas):

- Complete H&N examination including All cranial nerves.

1. Head:

1. Skin and Scalp.
2. Parotid.

2. Neck:

1. Five Lymph Node Levels.
2. Thyroid.

3. Oral Cavity (7 sub-sites):

1. Floor of mouth.
2. Tongue.
3. Alveolar Ridge.
4. Lips
5. Hard Palate.
6. Buccal area.
7. Retro-molar triangle.

4. Nasal cavity:

1. Vestibule.
2. Septum.
3. Floor.
4. Lateral wall.

5. Nasopharynx:

1. Postero-superior wall (Skull base to soft palate).
2. Lateral wall (Fossa of Rosenmuller).
3. Inferior wall (superior surface of soft palate).

6. Oropharynx:

1. Base of Tongue.
2. Soft palate.
3. Tonsils.
4. Posterior pharyngeal wall.

7. Hypopharynx:

1. Postcricoid area.
2. Piriform sinus.
3. Posterior pharyngeal wall.

8. Larynx:

1. Supraglottis:

1. Epiglottis.
2. Aryepiglottic fold.
3. Arytenoids.
4. Ventricular bands.

2. Glottis:

1. Vocal cords.

3. Subglottis:

1. From lower border of glottis to inferior border of cricoid.

- **Important Keypoints in Physical Examination:**

- Ipsilateral otalgia with normal otoscopy:
 - Direct attention to:
 1. Tonsil.
 2. Tongue base.
 3. Supraglottis.
 4. Hypopharynx.
- Unilateral OME:
 - Direct examination of Nasopharynx.

Levels of Investigations:

1. Diagnostic:

- CT with Contrast from Skull base to thoracic inlet for all neck masses **EXCEPT** Thyroid (US Thyroid).
- FNA.

2. Staging (TNM):

- CXR or CT Chest.
- LFT or CT Abdomen.

3. Treatment (Pre-op Assessment):

- CBC.
- U&E.
- Coagulation profile.

- **Ultrasonography (US):**

- **Indications:**
 1. Thyroid masses.
 2. Pediatric Neck masses
- **Advantages:**
 1. Differentiates between Solid vs. cystic masses.
 2. Noninvasive
 3. Avoids ionizing radiation
 4. Not required sedation.
- **Characteristics of Benign Lymph node in US:**
 1. Small
 2. Well-defined
 3. Oval shape
 4. Presence of fatty hilum.
- **Characteristics of Pathologic Lymph node in US:**
 1. Large
 2. Ill defined
 3. Round shape
 4. Loss of fatty hilum
 5. Microcalcifications
 6. Focal necrosis

Table I. Classic US criteria used in differentiating benign vs. malignant lymph nodes.

Criterion	Benign	Malignant
B scan criteria		
Size	small	large
shape	oval	rounded
hilum	present	absent
echogenicity	moderate or low	marked hypoechoic
margins	sharp	irregular, blurred, angular, invasive
Structural changes	absent	present
– focal cortical nodules		
– intranodal necrosis		
– reticulation		
– calcification		
– matting		
Soft tissue edema	may be present	absent
Doppler criteria		
Flow	absent	present
Vessel location	central	peripheral
Vascular pedicles	single	multiple
Vascular pattern	regular	chaotic
Impedance values	low	high

- **Computed Tomography (CT) with Contrast:**

○ **Indications:**

1. 1st imaging modality in diagnosis of H&N masses.
2. Helps in staging.
3. Detects Recurrence and Metastasis.

○ **Advantages:**

1. Evaluates characteristics and extent of primary tumor.
2. Evaluates involvement of adjacent structures.
3. Best for evaluation of bone invasion.
4. Evaluates lymph nodes status (Requires contrast).
5. Evaluates vascularity.

○ **Disadvantages:**

1. Ionizing radiation.
2. Can miss small metastases and early recurrence.
3. Poor soft tissue and Peri-neural evaluation.
4. Better to avoid contrast in Thyroid lesions.

- **Pathologic LN in CT:**
 1. Size > 1cm, EXCEPT:
 - > 1.5cm in Level 1-2.
 - > 8mm in Retropharynx.
 2. Ill defined
 3. Round shape.
 4. Central Necrosis
 5. Enhancement with contrast
 6. Rim enhancement
 7. Extra Capsular Invasion
- **Indications of CT in Thyroid :**
 1. Huge Thyroid Mass.
 2. Retrosternal Extension.
 3. For completion thyroidectomy.
 4. Pathological Lymph Node.
 5. Involvement of Adjacent Structures.

- **Magnetic Resonance Imaging (MRI) with Contrast:**

- **Advantages:**
 - Evaluation of Soft tissue.
 - Evaluation of Cartilage and Pre-vertebral fascia (Larynx).
 - Evaluation of Bone marrow invasion.
 - Evaluation of Peri-neural invasion:
 1. Abnormal nerve thickening.
 2. Enhancement of nerve sheath.
 3. Expansion of skull base foramina.
 4. Sclerotic changes in nerve canal.
- **Disadvantages:**
 1. Expensive.
 2. Can miss small metastases and early recurrence.
 3. Poor evaluation of Bone invasion.
 4. Artifacts with breathing or swallowing.
- **Indications of MRI in H&N Tumors:**
 1. Sinonasal tumors with intra-cranial or intra-orbital involvement.
 2. Nasopharyngeal tumors.
 3. Deep parotid lobe tumors.
 4. Parapharyngeal tumors.
 5. Vascular tumors.
 6. Laryngeal tumors with cartilage or pre-vertebral fascia involvement.
 7. Parathyroid adenoma.

- **Positron Emission Tomography (PET):**
 - Functional imaging with 18F-fluorodeoxyglucose (FDG).
 - Defines tissues with increased metabolic rate.
 - Improve characterization of both primary tumors and metastatic disease.
 - Poor spatial resolution of PET and the lack of anatomic landmarks can make exact localization of disease difficult.
- **Combined PET/CT:**
 - **Advantages:**
 - Allows for both anatomic and functional characterization of disease at the same time.
 - More accurate than either modality alone for detection of malignancy in H&N.
 - Improved localization of abnormalities.
 - Better differentiation of therapeutic changes from residual disease.
 - Improved assessment of tumor extent.
 - **Disadvantages:**
 - More expensive.
 - Some H&N cancers do not uniformly accumulate FDG.
 - Numerous false positive results.
 - Inflammatory and infectious lesions.
 - Warthin's and pleomorphic adenomas.
 - **Indications of PET/CT in H&N:**
 1. Modality of choice for evaluation of **Recurrence**.
 - Initial surveillance should be performed at least 3 months after conclusion of therapy.
 2. Modality of choice for identifying **Second primary**.
 3. Useful in detection of **Unknown primary** tumors.
 - PET/CT is a supplement, not a substitute, for endoscopy and biopsy in unknown primary tumors.
 - Finds 20-30% of unknown primary tumors above and beyond the traditional search.
 4. Treated Differentiated Thyroid Carcinoma with elevated thyroglobulin levels and negative RAI scan.
 5. Treated Medullary Thyroid Carcinoma with persistent measurable calcitonin levels and equivocal or negative sonography.

- **Keypoints:**
 - PET/CT is superior to standard anatomic imaging modalities in staging advance (T3 and T4) primary tumor.
 - PET/CT is not sufficiently accurate to avoid neck dissection in patients with advanced primaries and N0.
 - PET/CT is superior to conventional imaging modalities for radiation treatment planning, allowing for improved tumor coverage and sparing of normal tissues.
 - PET/CT is useful for monitoring treatment response in HNSCC.
 - PET/CT is useful for restaging, including nodal disease and distant metastases.
 - PET/CT has the potential to avoid unnecessary neck dissections in patients with nodal disease that responds completely to therapy.
- **Miscellaneous investigations:**
 - **Modified Barium Swallow with Esophagram:**
 - Evaluates aspiration and swallow.
 - Indicated for suspicion of esophageal carcinoma or lesions.
 - **Videostroboscopy:**
 - Provides documentation of laryngeal tumors and allows for patient education.
 - **Nuclear Medicine Studies:**
 - Considered in evaluating thyroid, parathyroid, mandible invasion, and metastatic bone disease.
 - Usually not indicated as a screening tool for head and neck cancer.

- **Biopsy:**

- Histologic confirmation of the diagnosis is mandatory before pursuing definitive therapy.
- **Fine Needle Aspiration (FNA):**
 - Standard of diagnosis.
 - Utilizes small (21-25) gauge needle.
 - Allows cytology evaluation of cells aspirated from the lesion.
 - **Indications:**
 1. Any neck mass that is not an obvious abscess.
 2. Persistence of Neck mass after a 2 weeks course of antibiotics.
 - **Advantages:**
 - Easy, quick, inexpensive.
 - Less pain.
 - Low risk of hematoma
 - Low risk of seeding of tumor along the biopsy tract.
 - Can be performed in a palpable node without US.
 - **Disadvantages:**
 - Provides information on cytology rather than nodal architecture.
 - Does not allow for histological subtyping.
 - Accuracy varies depending on disease entity.
 - Contraindicated in vascular lesions.
 - **Steps required to improve accuracy of FNA results:**
 1. Proper collection and skilled cytopathologist.
 2. Minimum of 4 separate passes.
 3. On-site review for adequacy.
 - **Sensitivity and specificity rates are 85-90%.**
 - FNA results may contribute to establishing the diagnosis but should not be accepted as absolute when clinical or other information contradicts the FNA findings.
 - Measurement of Thyroglobulin (Tg) in needle washout fluid enhances the sensitivity of FNA of cervical nodes in patients with thyroid nodule and suspicious LN on Neck US.

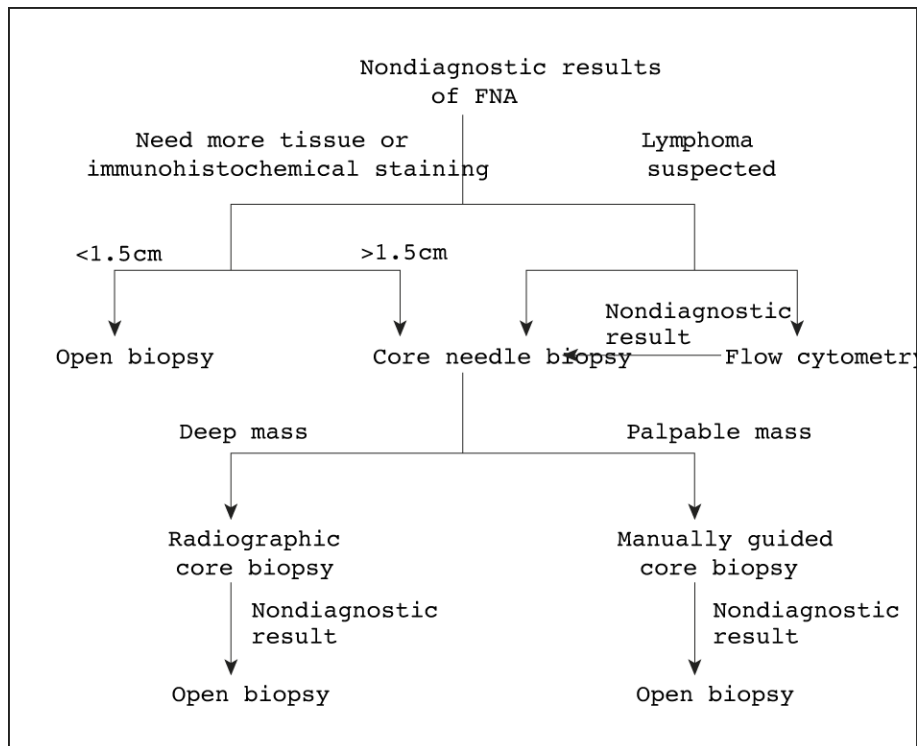
- **Selective results with FNA:**
 - **Follicular Thyroid Neoplasms:**
 - Difficult to differentiate between Follicular Thyroid Adenoma and Carcinoma.
 - Does not evaluate for capsular or vascular invasion.
 - **Salivary Gland Neoplasms:**
 - Interpretation of salivary gland pathology is difficult and requires greater experience.
 - On average, FNA has high specificity (98%) but lower sensitivity (80%).
 - **Lymphomas:**
 - Accurate diagnosis of lymphoma depends on changes in architecture, which requires morphologic examination of entire node.
 - Combination FNA with flow cytometry and immunohistochemistry may increase the accuracy of lymphoma diagnosis without the need for excisional biopsies.
- **Core Needle Aspiration (CNA):**
 - Utilizes larger (18-20) gauge needle.
 - **Advantages:**
 - Improves the accuracy of diagnosis.
 - Provides a sufficient amount of material for conventional histopathology and immunotyping as well as for molecular examinations.
 - **Higher diagnostic accuracy for diagnosis of the following:**
 1. Lymphoma.
 2. Salivary gland tumors.
 3. Tuberculosis
 - **Disadvantages:**
 - Expensive.
 - Requires experience.
 - Painful.
 - High risk of hematoma.
 - Requires U/S to avoid complications and to ensure suitable targeting.
 - Risk for seeding malignant cells along needle tract.
 - Lower incidence in Lymphoma compared to SCC.
 - One treatment strategy is to consider excision of the needle tract site at the time of definitive surgery and/or inclusion of the biopsy site in the radiation field.

○ **Excisional Biopsy:**

- Remove the tumor with its capsule.
- No risk for tumor seeding.
- Indications:
 1. Three negative FNA with highly suspicion of malignancy.
 2. Lymphoma.

○ **Incisional Biopsy:**

- Violates tumor capsule.
- High Potential risk for tumor seeding.
 - **Increase the mortality 50%.**
- Indicated only when all diagnostic modalities have failed to establish a diagnosis and excisional biopsy of the mass is not technically feasible.



- **Immunohistochemistry:**

- Utilizes antigen-antibodies reactions that bind to specific cellular components that aid in the histological diagnosis.
- Most markers are not tumor-specific and not utilized on a routine basis (due to expense).
- Useful for paranasal malignancy and poorly differentiated cancers (small- and large-cell tumors).
- SCC typically is not difficult to distinguish, possible False Positives include:
 - Necrotizing sialometaplasia
 - Mucoepidermoid carcinoma

○ **Common Immunohistochemical Markers:**

- **Lymphoma:**
 - Common leukocyte antigens
 - T-cell and B-cell markers
- **Carcinoma:**
 - Cytokeratin.
- **Melanoma:**
 - S-100 (High sensitivity, Low specificity, also found in neural and cartilaginous tumors).
 - HMB-45 (Sensitive and specific, does not stain spindle cell type).
 - Mitf and Tyrosinase (Sensitive and specific).
 - MART-1 and melan-A (Newer, sensitive and specific for melanocytes).
- **Neuroendocrine:**
 - Chromogranin.
 - Neuronspecific endolase (NSE).
- **Sarcomas:**
 - Vimentin.
 - Desmin (smooth and skeletal muscle).
 - Myoglobin (skeletal muscle, rhabdomyosarcoma).
 - Actin (Skeletal muscle).
 - S-100.

- **Diagnostic and Ancillary Procedures:**

- **Triple Endoscopy:**
 - Direct Laryngoscopy, Esophagoscopy, and Bronchoscopy.
 - Considered as routine screening for:
 1. Unknown primary
 2. Second primary.
 - Controversial for negative CXR and no signs or symptoms of esophageal or tracheobronchial involvement.
- **Feeding Tube:**
 - Allows enteral feedings with lower risk of aspiration, may place with anticipation of radiation effects, postoperative effect, and tumor growth.
 - Temporary (Nasogastric tube) or Permanent (Gastrostomy tube).
- **Tracheostomy:**
 - Low threshold for surgical airway management in anticipation of radiation effects, postoperative effect, and tumor growth.

- **Determine Classification of Neoplasms:**
- Based on histological specimen and evaluation of size and spread of primary tumor (tumor mapping).
 - **Histological Grading:**
 - Categorizes the histological type of cancer according to the degree of differentiation.
 - **Not significant to prognosis.**
 - Classification:
 - **Gx:** Grade cannot be assessed.
 - **G1:** Well-differentiated.
 - **G2:** Moderately differentiated.
 - **G3:** Poorly differentiated.
 - **G4:** Undifferentiated.
 - **TNM:**
 - Categorizes size and spread of cancer.
 - **Extent of Primary Tumor (T0-4):**
 - Criteria varies for each type of cancer.
 - **Tx:** Primary tumor cannot be assessed.
 - **T0:** No evidence of primary tumor.
 - **Tis:** Carcinoma in situ.
 - **Lip and Oral Cavity (T):**
 - **T1:** Tumor $\leq 2\text{cm}$.
 - **T2:** Tumor $> 2 \leq 4\text{cm}$.
 - **T3:** Tumor $> 4\text{cm}$.
 - **T4 (Lip):** Tumor invades through cortical bone, inferior alveolar nerve, floor of mouth, or skin of face.
 - **T4a (Oral Cavity):** Tumor invades adjacent structures , cortical bone, into deep [extrinsic] muscle of tongue, maxillary sinus, skin of face.
 - **T4b (Oral Cavity):** Tumor invades masticator space, pterygoid plates, or skull base and/or encases the internal carotid artery.

- **Nasopharynx (T):**
 - **T1:** Tumor is confined to nasopharynx or tumor extends to oropharynx and/or nasal cavity, without parapharyngeal extension.
 - **T2:** Tumor extends into Parapharyngeal space.
 - **T3:** Tumor involves bony structures and/or paranasal sinuses.
 - **T4:** Tumor has intracranial extension and/or involves cranial nerves, infratemporal fossa, hypopharynx, orbit, or masticator space.
- **Oropharynx (T):**
 - **T1:** Tumor $\leq 2\text{cm}$.
 - **T2:** Tumor $> 2 \leq 4\text{cm}$.
 - **T3:** Tumor $> 4\text{cm}$.
 - **T4a:** Tumor invades the larynx, deep/extrinsic muscle of the tongue, medial pterygoid, hard palate, or mandible.
 - **T4b:** Tumor invades the lateral pterygoid muscle, pterygoid plates, lateral nasopharynx, or skull base or encases the carotid artery.
- **Hypopharynx (T):**
 - **T1:** Tumor limited to one subsite and/or $\leq 2\text{cm}$.
 - **T2:** Tumor invades more than one subsite or an adjacent site, or measures $> 2 \leq 4\text{cm}$ without fixation of hemilarynx.
 - **T3:** Tumor $> 4\text{cm}$ or with fixation of hemilarynx or extension to esophagus.
 - **T4a:** Tumor invades thyroid/cricoid cartilage, hyoid bone, thyroid gland, or central compartment soft tissue.
 - **T4b:** Tumor invades prevertebral fascia, encases the carotid artery, or involves mediastinal structures.

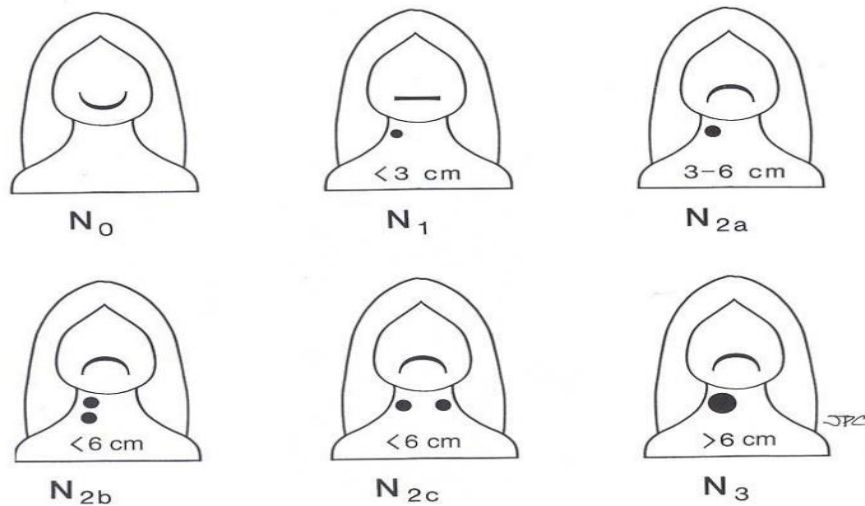
- **Supraglottis (T):**
 - **T1:** Tumor limited to one subsite with normal vocal cord mobility.
 - **T2:** Tumor invades mucosa of more than one adjacent subsite of the supraglottis or glottis or region outside the supraglottis without fixation of the larynx.
 - **T3:** Tumor limited to the larynx with vocal cord fixation and/or invades any of the following: postcricoid area, pre-epiglottic tissues, paraglottic space, and/or inner cortex of thyroid cartilage.
 - **T4a:** Tumor invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck, including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus).
 - **T4b:** Tumor invades prevertebral space, encases the carotid artery, or invades mediastinal structures.

- **Glottis (T):**
 - **T1a:** Tumor limited to one vocal cord with normal mobility.
 - **T1b:** Tumor involves both vocal cords with normal mobility.
 - **T2:** Tumor extends to the supraglottis and/or subglottis, and/or with **impaired vocal cord mobility**.
 - **T3:** Tumor limited to the larynx with vocal cord fixation and/or invades paraglottic space and/or inner cortex of thyroid cartilage.
 - **T4a:** Tumor invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck, including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus).
 - **T4b:** Tumor invades prevertebral space, encases the carotid artery, or invades mediastinal structures.

- **Subglottis (T):**
 - **T1:** Tumor limited subglottis.
 - **T2:** Tumor extends to the vocal cords with normal or impaired mobility.
 - **T3:** Tumor limited to the larynx with vocal cord fixation.
 - **T4a:** Tumor invades cricoid or thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck, including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus).
 - **T4b:** Tumor invades prevertebral space, encases the carotid artery, or invades mediastinal structures.
- **Maxillary Sinus (T):**
 - **T1:** Tumor limited to the maxillary sinus mucosa, with no erosion or destruction of bone.
 - **T2:** Tumor causing bone erosion or destruction, including extension into the hard palate and/or middle nasal meatus, except extension to the posterior wall of the maxillary sinus and pterygoid plates.
 - **T3:** Tumor invades any of the following: bone of the posterior wall of the maxillary sinus, subcutaneous tissues, floor, or medial wall of the orbit, pterygoid fossa, or ethmoid sinuses.
 - **T4a:** Tumor invades anterior orbital contents, skin of cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses.
 - **T4b:** Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve (V2), nasopharynx, or clivus.

- **Nasal Cavity and Ethmoid Sinus (T):**
 - **T1:** Tumor limited to any one subsite with or without bone erosion.
 - **T2:** Tumor invades two subsites in a single region or extends to involve an adjacent region within the nasoethmoidal complex, with or without bony invasion.
 - **T3:** Tumor extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribriform plate.
 - **T4a:** Tumor invades anterior orbital contents, skin of cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses.
 - **T4b:** Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve (V2), nasopharynx, or clivus.
- **Salivary Glands (T):**
 - **T1:** Tumor **≤2cm** without extraparenchymal extension.
 - **T2:** Tumor **> 2 ≤4cm** without extraparenchymal extension.
 - **T3:** Tumor **> 4cm** and/or with extraparenchymal extension.
 - **T4a:** Tumor invades the skin, mandible, ear canal, and/or facial nerve.
 - **T4b:** Tumor invades the skull base and/or pterygoid plates and/or encases the carotid artery.
- **Thyroid Gland (T):**
 - **T1:** Tumor **≤2cm** Limited to Thyroid only.
 - **T2:** Tumor **> 2 ≤4cm** Limited to Thyroid only.
 - **T3:** Tumor **> 4cm** Limited to Thyroid only or Tumor of Any size with Minimal Extra-thyroid Extension (e.g., extension to SCM or Perithyroid soft tissues).
 - **T4a:** Tumor of Any size extends beyond Thyroid capsule to invade Subcutaneous soft tissues, Larynx, Trachea, Esophagus, or RLN.
 - **T4b:** Tumor invades Prevertebral Fascia or encases Carotid Artery or Mediastinal vessels.
 - **All Anaplastic carcinomas are considered T4.**

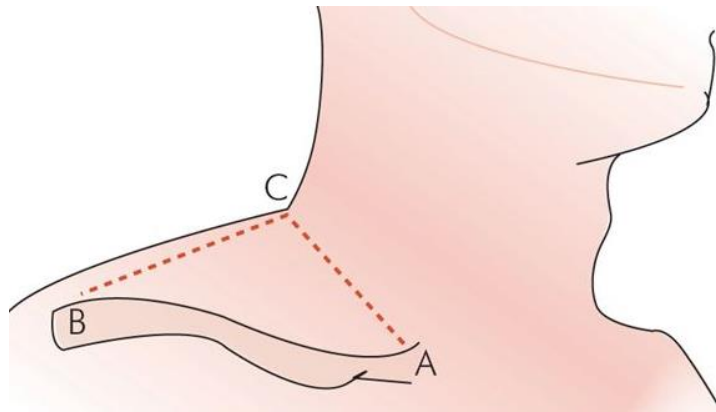
- **Mucosal Melanoma (T):**
 - **T3:** Mucosal disease.
 - **T4a:** Tumor involving deep soft tissue, cartilage, bone, or overlying skin.
 - **T4b:** Tumor involving brain, dura, skull base, lower cranial nerves (IX, X, XI, XII), masticator space, carotid artery, prevertebral space, or mediastinal structures.
- **Regional Lymph Node Metastasis (N0-3):**
 - Most important prognostic indicator for HNSCC.
 - Consistent for all Head and Neck Mucosal tumors **EXCEPT** Thyroid, Nasopharynx and Mucosal Melanoma.
 - **Nx:** Regional LN cannot be assessed.
 - **N0:** No regional LN metastasis.
 - **N1:** Single ipsilateral LN ≤ 3 cm.
 - **N2a:** Single ipsilateral LN > 3 -6cm.
 - **N2b:** Multiple ipsilateral LN ≤ 6 cm.
 - **N2c:** Bilateral or contralateral LN ≤ 6 cm.
 - **N3:** Any LN > 6 cm.



- **Thyroid Regional Lymph Node Metastasis (N):**
 - **N0:** No regional LN metastasis.
 - **N1a:** Metastasis to **Level VI** (pretracheal, paratracheal, prelaryngeal/Delphian LN).
 - **N1b:** Metastasis to cervical (**Level I-V**) or superior Mediastinal lymph nodes (**Level VII**).

- **Nasopharynx Regional Lymph Node Metastasis (N):**

- **N0:** No regional LN metastasis.
- **N1:** Unilateral LN $\leq 6\text{cm}$
- **N2:** Bilateral LN $\leq 6\text{cm}$
- **N3a:** Any LN $> 6\text{cm}$
- **N3b:** Extension to **Supraclavicular fossa:**
 - Supraclavicular fossa (Ho's Triangle) is bounded by medial (A) and lateral (B) ends of clavicle and the point (C) where neck meets the shoulder.
 - It includes caudal portions of levels IV and V



- **Mucosal Melanoma Regional Lymph Node Metastasis (N0-1):**

- **N0:** No regional LN metastasis.
- **N1:** Regional LN metastasis present.

- **Distant Metastasis (M0-1):**

- Any SCC involves Level VII lymph node is considered Distant Metastasis for palliative therapy.
- Lung Metastasis is NOT a contraindication for surgical excision in Thyroid and Salivary glands tumors.
 - **Mx:** Distant metastasis cannot be assessed.
 - **M0:** No distant metastasis.
 - **M1:** Distant metastasis present.

- **Staging:**
 - Grouped TNM classification.
 - Used to guide treatment plan and apply statistical data such as prognosis and treatment effectiveness.
 - **Early Stage (I-II):**
 - Single modality treatment plan:
 - Surgery **or** Radiation.
 - **Advanced Stage (III-IV):**
 - Double modality treatment plan:
 - Surgery **and** Radiation
 - or
 - Chemotherapy **and** Radiation.

	N0	N1	N2-3,M+	
T1	I			
T2	II			
T3	III			
T4	IV			

- TNM Staging Table is applied for All Head and Neck Cancers **Except** Thyroid and Nasopharynx.
 - Any T4 is considered Stage IV (Advance Stage).
 - Any N1 is considered Stage III (Advance Stage).
 - Any N2-3 is considered Stage IV (Advance Stage).
 - Any M1 is considered Stage IV (Advance Stage).
- **Pathological staging** > Radiological > Clinical staging.
- **Up-Staging Theory:**
 - If in doubt about the staging → Upstage.

Management Plan of H&N Cancers:

- **Discuss the case in H&N Tumor Board.**
- **Consultations:**
 - **Dental:**
 - Evaluate dentition and possibility of teeth extraction before radiation therapy.
 - **Plastic/Reconstructive Surgery:**
 - Evaluate complex reconstructive issues including free-flap transfer if needed.
 - **Neurosurgery:**
 - Assist in certain skull base and cranial procedures.
 - **Ophthalmology:**
 - Assist in surgical management of the eye.
 - **Prosthodontics:**
 - Preoperative consultation if oral-maxillary, orbital, and other head and neck prostheses needed.
 - **Dermatology:**
 - Mohs micrographic surgery for dermal lesions.
 - **Vascular Surgery:**
 - Consultation for possible carotid resection or bypass.
 - **Medicine:**
 - Preoperative evaluation (preoperative clearance), management of coexisting medical conditions.
 - **Dietitian:**
 - Considered preoperatively for malnourished patients to assess nutrition status and recommend methods to obtain a positive nitrogen balance.
 - **Speech and Swallow Therapy:**
 - Considered postoperatively to assess swallowing and speech and provide therapy.
- **Treatment Concepts:**
 - Goal of treatment is maximizing survival with preservation of form and function.
 - Treat the Neck mets with same modality used for treatment of the primary tumor.
 - **Single-modality Therapy (Stage I-II):**
 - Primary surgery **or** radiation therapy alone.
 - **Double-modality Therapy (Stage III-IV):**
 - Surgery **and** Radiation
 - Chemotherapy **and** Radiation.
 - Surgical treatment is better for:
 - Sinonasal + Oral.
 - Non-Surgical treatment is better for:
 - Pharyngeal + Laryngeal.

Principles of Oncologic Surgical Therapy:

- The goal of oncologic surgery is complete tumor resection with histologic verification of tumor-free margins.
- If surgery is the chosen modality of treatment, think of 4 points:
 - 1. Primary Tumor:**
 - Resection with 1cm safe margin.
 - Approach.
 - 2. Neck Dissection:**
 - Type.
 - Unilateral or Bilateral.
 - 3. Reconstruction.**
 - 4. Airway i.e. Tracheotomy.**
- **Operability vs Resectability:**
- Operability is with regard to the patient:
 - **Inoperable:** Operation cannot be done because of either:
 - Poor general condition of the patient (unfit), or
 - Surgery can not cure the malignant disease.
 - Ex: Distant metastasis.
- Resectability is with regard to the tumor:
 - **Unresectable:** Tumor infiltrates vital structures (T4b).
 - Ex: Encasement of common or internal carotid artery.
- Inoperable disease may still be resectable, but that is not going to offer a cure for the malignant disease.
 - SCC with distant metastasis is considered inoperable, but the lesion may be locally resectable.
- **Assessment of Resectability:**
- Unresectable tumors are associated with poor prognosis.
- Based on ability to obtain clear margins due to involvement of (**T4b**):
 - 1. Skull base:**
 - Erosion of pterygoid plates or sphenoid, widening of foramen ovale.
 - 2. Nasopharynx:**
 - Deep extension into eustachian tube and lateral nasopharyngeal walls.
 - 3. Pterygoid muscles with severe trismus or pterygopalatine fossa involvement with cranial neuropathy.**
 - 4. Direct extension to mediastinum , prevertebral fascia, or cervical vertebrae.**
 - 5. Encasement of common or internal carotid artery.**
 - Encasement is assessed radiologically and defined as a tumor surrounding the carotid artery by 270 degrees or greater.
 - 6. Direct extension of neck disease to involve external skin.**
 - 7. Presence of subdermal metastases.**

- **Primary Tumor Resection:**
- **En-bloc resection** is recommended.
- Peri-neural invasion is suspected when tumors are adjacent to nerves.
 - Nerve should be dissected both proximally and distally and should be resected to obtain clearance of disease.
 - Frozen section determination of proximal and distal nerve margins may prove helpful to facilitate tumor clearance.
- Primary tumor should be assessed histologically for:
 - Depth of invasion.
 - Distance from invasive tumor to margin of resection, including peripheral and deep margins.
- **Tumor Margins:**
- Tumor-free margins are essential to decrease risk of local recurrence.
 - Positive margins are an indication for post-op adjuvant therapy.
- Achievement of adequate wide margins may require resection of an adjacent structure.
- **Clear Margin:** $\geq 5\text{mm}$ from invasive tumor to resection margin.
- **Close Margin:** $< 5\text{mm}$ from invasive tumor to resection margin.
- **Positive Margin:** Carcinoma in situ or invasive carcinoma at resection margin.
- Margin assessment can be done by:
 - 1. Frozen section:**
 - Done intra-op (Real time).
 - Indicated if there is uncertain clear margins due to:
 - Indistinct tumor margins
 - Suspected residual disease (soft tissue, cartilage, carotid artery, or mucosal irregularity).
 - 2. Formalin-fixed tissues.**
- **Surgical Management of Cranial Nerves (VII, X, XI, XII):**
- Influenced by pre-op clinical function.
 - **Pre-op Functioning Nerve:**
 - Preserve structure and function of the nerve even if otherwise adequate tumor margins are not achieved.
 - Should not leave any gross residual disease.
 - Adjuvant post-op Radiation or Chemoradiation is prescribed when a microscopic residual or gross residual tumor is suspected.
 - **Pre-op Paralyzed Nerve (Direct Nerve Invasion):**
 - Segmental resection (and nerve grafting) if tumor-free margins are assured throughout the remainder of the procedure.

- **Neck Dissection:**
- Removal of regional lymphatics and surrounding fibrofatty tissue.
- Indicated in positive neck or tumors with high risk to develop occult metastasis.

- Indications of Neck Dissection:

1. Tumors with N+.
2. Tumors with N0 and high risk of occult disease.
3. Salvage surgery

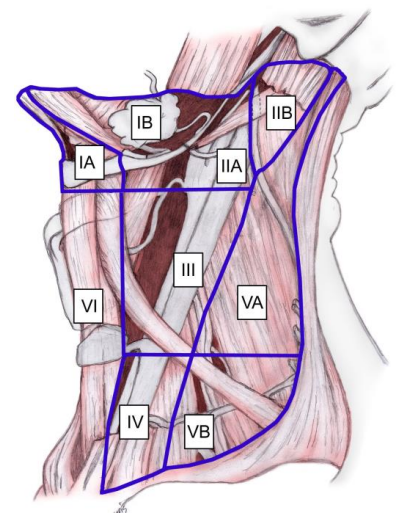
- Indications of Bilateral Neck Dissection:

1. Midline tumors
2. Tumors crossing the midline

- Regional Lymph Nodes of the Neck:

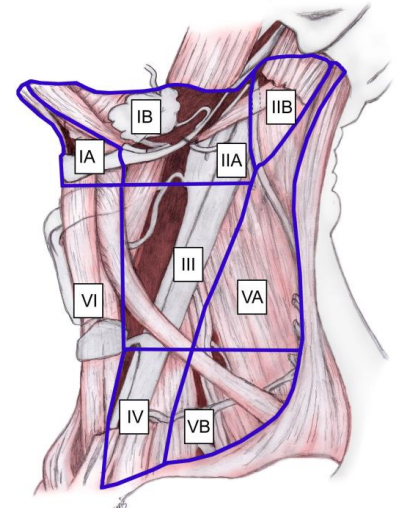
- **Level Ia (Submental LN):**

- **Anatomical Landmark:**
 - Bounded by bilateral anterior bellies of digastric muscles and hyoid.
- **Radiological Landmark:**
 - Lymph nodes located antero-medially to horizontal line passing from posterior border of submandibular gland.
- **Drainage Sites:**
 - Floor of mouth, Anterior tongue, Anterior mandibular alveolar ridge, and Lower Lip.



- **Level Ib (Submandibular LN):**

- **Anatomical Landmark:**
 - Bounded by anterior and posterior bellies of digastric and body of Mandible.
- **Radiological Landmark:**
 - Lymph nodes located antero-laterally to horizontal line passing from posterior border of submandibular gland.
- **Drainage Sites:**
 - Oral cavity, Anterior Nasal Cavity, soft tissue structures of the mid face, and Submandibular gland.

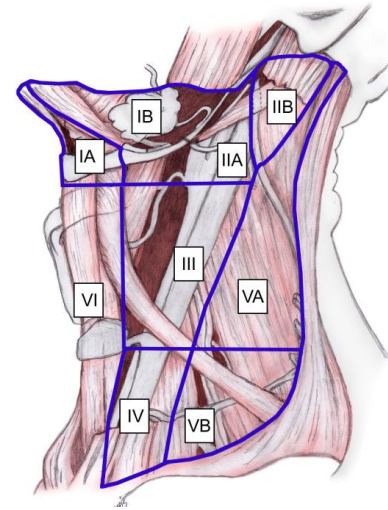


- **Level II (Upper Jugular / Jugulodigastric LN):**

- Related to upper third of internal jugular vein.
- Spinal Accessory Nerve (CN-XI) travels obliquely across this area.

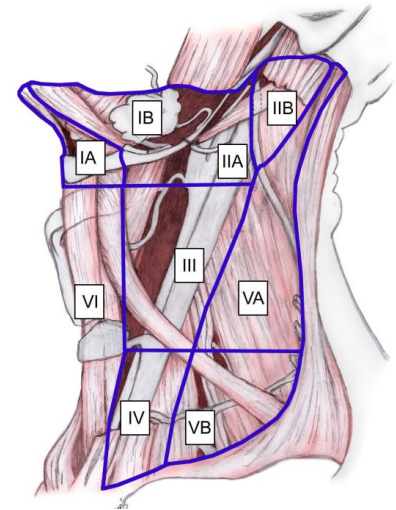
○ **Level IIa:**

- **Clinical Landmark:**
 - From Skull base to Hyoid bone.
- **Surgical Landmark:**
 - From posterior belly of digastric to Carotid bifurcation.
 - **Anterior** to Spinal Accessory Nerve (CN-XI).
- **Radiological Landmark:**
 - Lymph nodes located between 2 horizontal lines passing from posterior border of submandibular gland and posterior border of SCM from Skull base to hyoid.
- **Drainage Sites:**
 - Oral cavity, nasal cavity, nasopharynx, oropharynx, hypopharynx, larynx, and parotid gland.



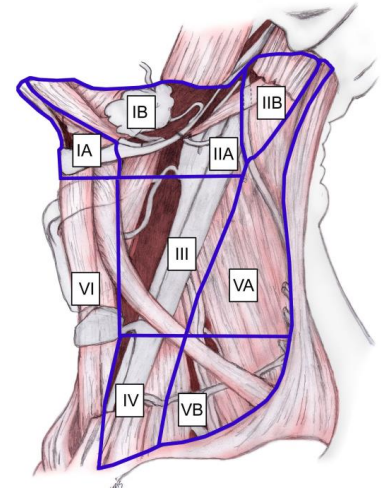
○ **Level IIb:**

- **Clinical Landmark:**
 - From Skull base to Hyoid bone.
- **Surgical Landmark:**
 - From posterior belly of digastric to Carotid bifurcation.
 - **Posterior** to Spinal Accessory Nerve (CN-XI).
- **Radiological Landmark:**
 - Lymph nodes located between 2 horizontal lines passing from posterior border of submandibular gland and posterior border of SCM from Skull base to hyoid.
- **Drainage Sites:**
 - Oral cavity, nasal cavity, nasopharynx, oropharynx, hypopharynx, larynx, and parotid gland.



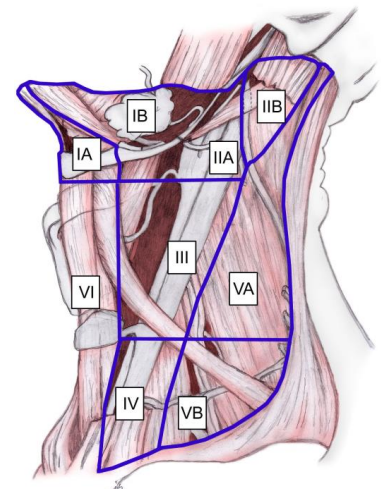
- **Level III (Middle Jugular LN):**

- Related to Middle third of internal jugular vein.
- **Clinical Landmark:**
 - From Hyoid bone to Cricoid.
- **Surgical Landmark:**
 - From Carotid bifurcation to Omohyoid.
- **Radiological Landmark:**
 - Lymph nodes located between 2 horizontal lines passing from posterior border of submandibular gland and posterior border of SCM from hyoid to cricoid.
- **Drainage Sites:**
 - Oral cavity, nasopharynx, oropharynx, hypopharynx, and larynx.



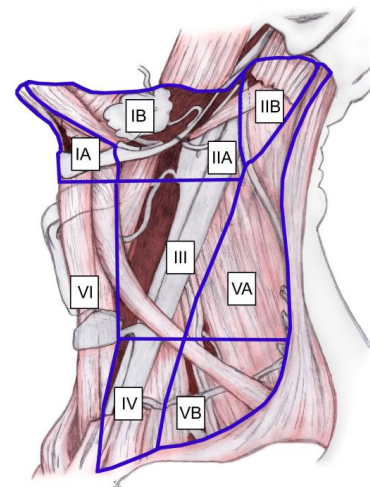
- **Level IV (Lower Jugular LN):**

- Related to Lower third of internal jugular vein.
- **Clinical Landmark:**
 - From Cricoid to Clavicle.
- **Surgical Landmark:**
 - From Omohyoid muscle to Clavicle.
- **Radiological Landmark:**
 - Lymph nodes located anterior to horizontal line passing from posterior border of SCM from hyoid to cricoid lateral the carotids.
- **Drainage Sites:**
 - Larynx, hypopharynx, thyroid, and cervical esophagus.



- **Level V (Posterior Triangle LN):**

- Located along lower half of the spinal accessory nerve and transverse cervical artery.
- **Level Va:**
 - **Anatomical Landmark:**
 - From Apex of Convergence of SCM and trapezius muscle to horizontal plane crossing the inferior border of cricoid.
 - **LN Associated with Spinal accessory nerve (CN-XI)**
 - **Radiological Landmark:**
 - Lymph nodes located posterior to horizontal line passing from posterior border of SCM.
 - **Drainage Sites:**
 - Drains nasopharynx, oropharynx, and skin of the posterior scalp and neck.



○ **Level Vb:**

▪ **Anatomical Landmark:**

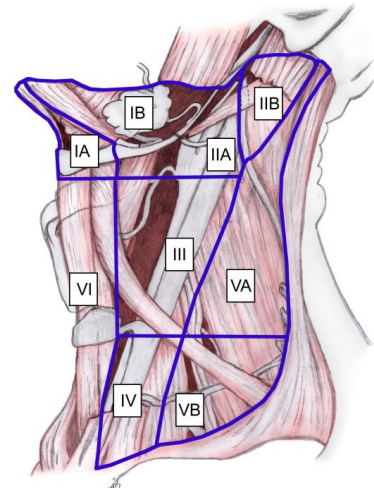
- From horizontal plane crossing the inferior border of cricoid to Clavicle.
- **LN Associated with Transverse Cervical and supraclavicular nodes.**

▪ **Radiological Landmark:**

- Lymph nodes located posterior to horizontal line passing from posterior border of SCM.

▪ **Drainage Sites:**

- Drains nasopharynx, oropharynx, and skin of the posterior scalp and neck.
- **Virchow's lymph node** is located in left supraclavicular fossa and drains internal abdominal organs (Stomach mainly).



- **Level VI (Central Compartment):**

○ **Anatomical Landmark:**

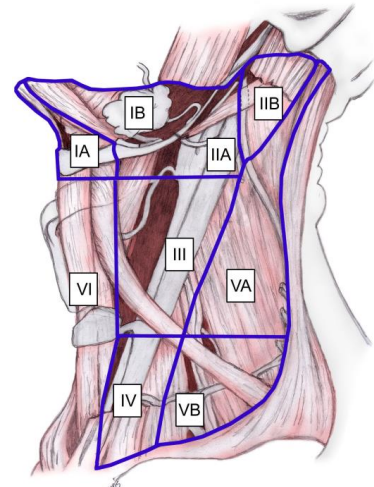
- Extend in midline from hyoid bone to Suprasternal notch between the carotids.
 - **Paratracheal LN:**
 - Located in Tracheoesophageal groove.
 - **Pretracheal LN:**
 - Located in front of the trachea
 - **Parathyroidal LN:**
 - Located around the thyroid gland
 - **Precricoid or Delphian LN:**
 - Located on Cricothyroid membrane.

○ **Radiological Landmark:**

- Lymph nodes located anterior to horizontal line passing from posterior border of SCM medial to carotids.

○ **Drainage Sites:**

- Drains Thyroid gland, subglottic larynx, cervical trachea, hypopharynx, and cervical esophagus.



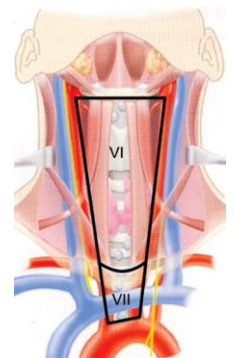
- **Level VII (Anterior Superior Mediastinal):**

▪ **Landmark:**

- Extends from suprasternal notch to Innominate artery, behind manubrium and between Common carotid arteries.

▪ **Clinical Importance:**

- Any SCC involves Level VII is considered Distant Metastasis for palliative therapy.



- **Classification of Neck Dissections:**

1. Radical Neck Dissection (RND):

- Removes all the followings:
 1. Level (I-V) Lymph Nodes.
 2. Spinal **A**ccessory Nerve (SAN)
 3. **I**nternal Jugular Vein (IJV).
 4. Sternocleidomastoid **M**uscle (SCM).
- Indications:
 1. Recurrence or Residual disease.
 2. Involvement of SAN, IJV and SCM.
 3. Advance nodal disease (**N3**).
- Advantages:
 1. Technically easier.
 2. Lower risk of residual disease
- Disadvantages:
 1. Neck deformity (Removal of SCM).
 2. Shoulder drop (Removal of SAN).
 3. Risk of facial edema (Removal of IJV).
 4. Hyposthesia of Neck and periauricular region.

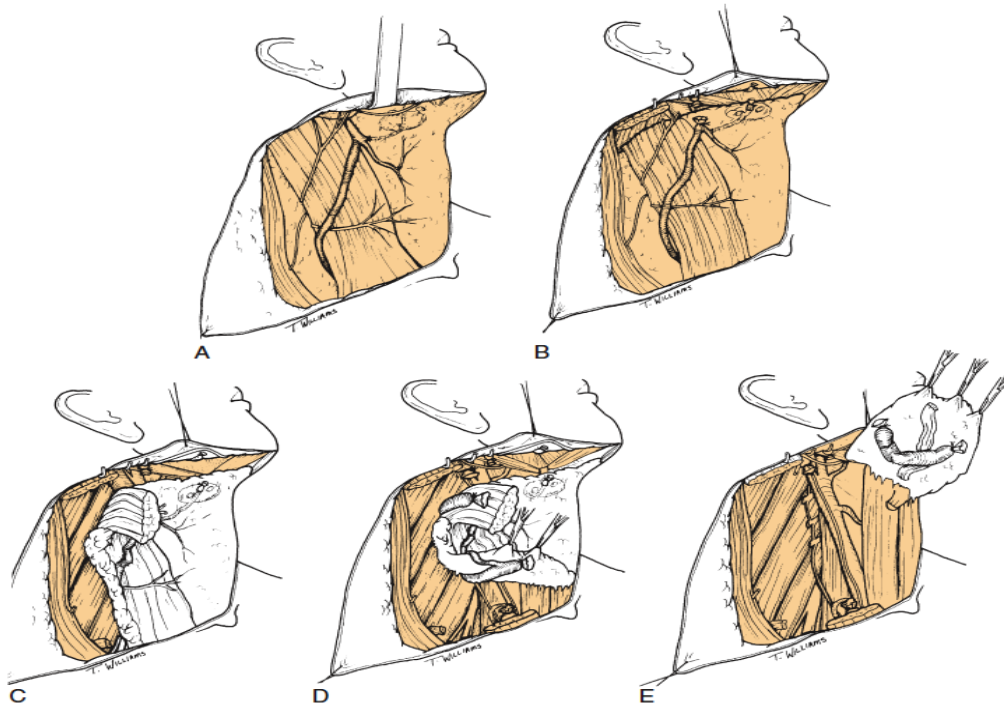


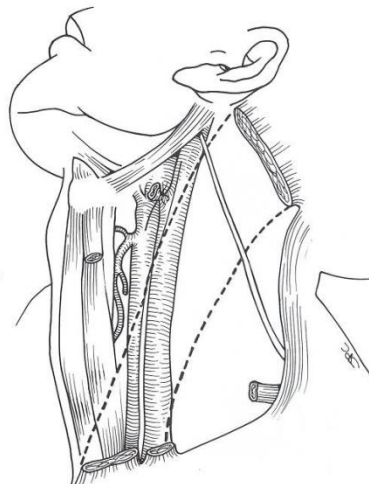
Figure 121-5. Steps of the radical neck dissection. (See text for discussion.)

2. Modified Radical Neck Dissection (MRND):

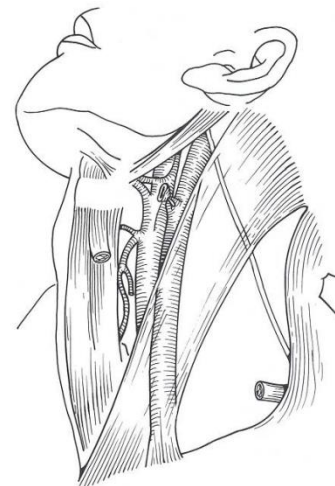
- Indicated in patients with N1-N2.
- Types:
 1. **Type I:**
 - Removes the following:
 1. Level (I-V).
 2. **IJV.**
 3. **SCM.**
 - Spares **SAN.**
 - Indicated in **N1 and N2** with involvement of these structures (**Intra-op Decision**).
 2. **Type II:**
 - Removes the following:
 1. Level (I-V).
 2. **SCM.**
 - Spares **SAN** and **IJV.**
 - Indicated in **N1 and N2** with involvement of these structures (**Intra-op Decision**).
 3. **Type III (Functional/Bocca):**
 - Removes level (I-V) only.
 - Spares **SAN, IJV** and **SCM.**
 - Indicated in **N1 and N2** without involvement of these structures.
- Advantages:
 1. Lower morbidity.
- Disadvantages:
 1. Technically more difficult.
 2. Higher risk of residual disease.



Type I MRND



Type II MRND



Type III MRND

3. Selective Neck Dissection:

- Neck dissection with preservation of one or more LN groups.
- Indicated only in **N0** with high risk of occult metastasis ($\geq 20\%$).
- Levels removed depend on the location of the primary lesion and its known pattern of spread.
- Subtypes:

1. Supraomohyoid (Anterolateral) Neck Dissection:

- Removes levels **I–III**.
- Extended supraomohyoid removes level IV.
- Indications:
 1. N0 Oral cavity.
 2. N0 Salivary glands.

2. Lateral Neck Dissection:

- Removes levels **II–IV**.
- Typically bilateral.
- Indications:
 1. N0 Oropharynx.
 2. N0 Hypopharynx.
 3. N0 Larynx.

3. Posterlateral Neck Dissection:

- Removes levels **II–V**.
- Indications:
 1. Scalp.
 2. Nasopharynx.
 3. **N1b** Thyroid.

4. Anterior (Central) Neck Dissection:

- Removes Level **VI**.
- Indications:
 1. **N1a** Thyroid.
 2. Parathyroid.
 3. Subglottic.
 4. Cervical esophagus.

4. Extended Neck Dissection:

- Any Neck dissection with additional removal of any structure other than Lymph node levels I-V, SAN, IJV and SCM.

- **Clinically Negative Neck (N0):**
 - Tumors with Low risk (<20%) of occult metastasis:
 - Management is observation.
 - **No Neck Dissection in N0 Sinonasal tumors.**
 - Lymphatic drainage is mainly to Retropharyngeal.
 - Tumors with High risk (≥20%) of occult metastasis:
 - All tumors with **N0 and ≥T2**, Except:
 - **N0 and ≥T1:**
 - Oral tongue.
 - Piriform sinus.
 - **N0 and ≥T3:**
 - Glottic.
 - **N0 and ≥T4:**
 - Hard palate.
 - Subglottic.
 - Management:
 - Selective Neck Dissection (SND).
- **Indications of ND in N0 Oral cavity Ca (Supraomohyoid ND):**
 - Oral Tongue **≥T1**
 - Others **≥T2**
 - Hard Palate **≥T4**
- **Indications of ND in N0 Pharyngeal Ca (Bilateral Lateral ND):**
 - Piriform sinus **≥T1**
 - Hypopharynx and Oropharynx **≥T2**
- **Indications of ND in N0 Laryngeal Ca (Bilateral Lateral ND):**
 - Supraglottic **≥T2**
 - Glottic **≥T3**
 - Subglottic **≥T4**
- **Indications of ND in Nasopharyngeal Ca:**
 - Recurrence.
 - If primary tumor was treated surgically.
- **Indications of ND in Thyroid Ca:**
 - **DTC N1a** → Central (VI) ND.
 - **DTC N1b:**
 - **If Ipsilateral LN:**
 - Ipsilateral Posterolateral + Central (II-VI) ND.
 - **If Contralateral LN:**
 - Ipsilateral Posterolateral + Central (II-VI) ND.
 - Contralateral Lateral (II-VI) ND.
 - **Medullary Thyroid Ca:**
 - Bilateral Posterolateral + Central (II-VI) ND.

- **Indications of ND in Salivary Glands Ca:**

- High Grade Tumors:
 - **N0 and $\geq T1$** → Supraomohyoid ND.
 - **N+ve** → MND.
- Low Grade Tumors:
 - **N0 and $\geq T3$** → Supraomohyoid ND.
 - **N+ve** → MND.

- **Complications of Neck Dissection:**

- **Wound infection:**

- Higher incidence in:
 - Irradiated neck
 - Soilage of wound by saliva, tracheal or gastric secretions.
 - Tight wound closure
 - Immunocompromised
 - Malnourished
 - Presence of foreign body
 - Hematoma
 - Seroma
- Treatment:
 - Aggressive antibiotic regimen
 - Monitor for fistula
 - Control diabetes
 - Optimize nutrition
 - Meticulous wound care (debridement, wet to dry dressings)
 - Evaluate potential for carotid blowout.

- **Shoulder dysfunction:**

- Most commonly in patients underwent RND due to removal of spinal accessory nerve supplying the trapezius muscle.
- However, any type of neck dissection may result in impairment of function of the shoulder.
- Results in:
 - Shoulder pain
 - Shoulder drop
 - Winged scapula
 - Inability to abduct the shoulder above 90 degrees.
- If any deficit is detected, the patient should be properly counseled and coached to ensure proper rehabilitation of the shoulder.

- **Hematoma:**

- Prevented by:
 - Meticulous hemostasis
 - Placement of suction drains.
- If detected early:
 - Milking the drains may result in evacuation of accumulated blood.
- If massive hematoma or blood re-accumulates quickly:
 - Best to be managed in OR by exploration of the wound under sterile conditions and evacuating the hematoma after controlling the bleeding.
- Failure to recognize or manage postoperative hematoma properly may predispose to development of wound infection.

- **Facial/Cerebral Edema:**

- Resulted from synchronous bilateral RNDs due to IJVs ligation.
- Facial or cerebral edema results from a mechanical problem of venous drainage which may resolves with time as collateral circulation is established.
- More common and more severe in previously irradiated patients.
- Cerebral edema presents with syndrome of inappropriate secretion of antidiuretic hormone (SIADH), impaired neurologic function or coma that occur after bilateral RND.
- Prevented by:
 - Preserving at least one EJV whenever bilateral RND is anticipated.
 - Reconstructing one IJV using saphenous vein graft.

- **Chyle Leak:**

- Occurs in 1-2% of neck dissections.
- Typically left-sided in 95–97%.
- Chyle consists of fat, protein, electrolytes and lymphocytes.
- Results from injury to the thoracic duct:
 - Thoracic duct is the conduit for lymph and dietary fat to reach the venous bloodstream.
- Clinical picture:
 - Odorless milky appearance fluid in the drain.
 - Apparent after few days of starting enteral feeding.
- Fluid analysis:
 - Total fat composition of 0.4-4 g/L.
 - Total protein greater than 30 g/L.
 - Lymphocyte predominance.
- Complications:
 - Electrolyte disturbance
 - Hypovolemia
 - Hypoalbuminemia
 - Wound infection
 - Chylothorax

- Prevented intraoperatively by:
 - Bloodless operative field while dissecting around the thoracic duct.
 - Observing the area of the thoracic duct while asking the anesthesiologist to do Valsalva maneuver.
 - Intraoperative control of any apparent leak with ligation or clipping of any visualized or potential lymphatic tributaries.
- Management:
 - **Conservative management:**
 - Head elevation.
 - Closed-wound drainage
 - Pressure dressings
 - Enteral diet modification:
 - Low-fat nutritional support.
 - Medium-chain triglycerides:
 - Absorbed directly into the portal circulation bypassing the lymphatic system.
 - Replace fluid and electrolytes.
 - Octreotide administration.
 - Parenteral alimentation through central line:
 - Indicated for high-output or intractable fistulae.
 - **Indications of early surgical exploration:**
 - Daily output of chyle exceeds 600 ml.
 - Failure of conservative measures (>1 week).
 - Cachexia.
 - Chylothorax.

- **Surgical Defect Reconstruction:**
- **Primary closure** is recommended.
 - Should not be done at expense of obtaining wide, tumor-free margins.
- **Follow the Reconstructive ladder:**
 1. Primary closure
 2. Delayed closure
 3. Split thickness skin graft (STSG)
 4. Full thickness skin graft (FTSG)
 5. Random pattern flap
 6. Pedicled flap
 7. Free flap
- **Indications of Post-op Adjuvant Radiation alone in H&N Cancers:**
 1. Advance Stage (III -IV).
 2. Peri-neural invasion.
 3. Vascular invasion
 4. Positive margin.
 5. Depth > 4mm (**Oral cavity**)
 6. High grade (**Salivary glands**)
- **Indications of Post-op Adjuvant ChemoRadiation in H&N Cancers:**
 1. Nodal extra-capsular invasion.
 2. Positive margin.

- **Post-treatment Follow-up Plan:**
- **Regular Follow-up every:**
 - 1 month for 1st year post-therapy.
 - 2 months for 2nd year post-therapy.
 - 3 months for 3rd year post-therapy.
 - 6 months for 4th year post-therapy.
 - 1 year for 5th year and afterward post-therapy.
- **PET/CT:**
 - Initial post-treatment surveillance done **>3 months** after conclusion of therapy then **Annually**.
- **Annual investigations:**
 - Chest Radiographs (Consider CT of primary site and chest)
 - Thyroid function tests (Post radiation therapy to the neck)
 - Liver enzymes.
 - EBV monitoring for Nasopharynx.

Prognostic Evaluation of H&N Cancers:

- Overall 5-year survival of H&N cancers is **<40%**.
 - 50–60% of mortality is from **failed loco-regional control**.
 - 20–30% of mortality is from metastatic disease.
 - 10–20% of mortality is from second primaries.
- Premorbid conditions account for mortality of:
 - 30–35% of early-staged H&N cancers patients.
 - 10–15% of late-staged H&N cancers patients.

- **Risk Factors for Poor Prognosis in H&N cancer:**

1. Presence of regional nodal disease

- The strongest predictor of prognosis for H&N cancers.
- Decrease survival as much as 50%.

2. Invasiveness (T) and metastasis (M) within each site:

- T1 H&N cancers survival rates is 80%.
- T2 H&N cancers survival rates is 60%.
- T3 H&N cancers survival rates is 40%.
- T4 H&N cancers survival rates is 20%.

3. Increased tumor thickness and volume determined by CT/MRI:

- Especially if >4mm in depth.

4. Presence of abundant lymphatic drainage at primary site:

- Increases risk of regional disease.

5. Character of Nodal Disease:

1. Presence of nodal extracapsular spread.
2. Increased number of positive nodes.
3. Presence of metastasis to lower nodal levels (level IV-V).
4. Skipped nodes (Nodal involvement of level II and IV without involvement of level III).
5. Lymphocyte-depleted pattern immunomorphology of the lymph node.

6. Vascular and Perineural Invasion:

- Histological predictors of cancer spreading beyond the margins of resection, indicate aggressive cancer behavior.

7. Cytomorphometric Parameters: (controversial)

- Number of chromosome set (ploidy) may predict prognosis.
- Aneuploid is associated with worse prognosis than diploidy.

- **Grade of Tumor differentiation is not an important determinant in prognosis for most head and neck cancers.**

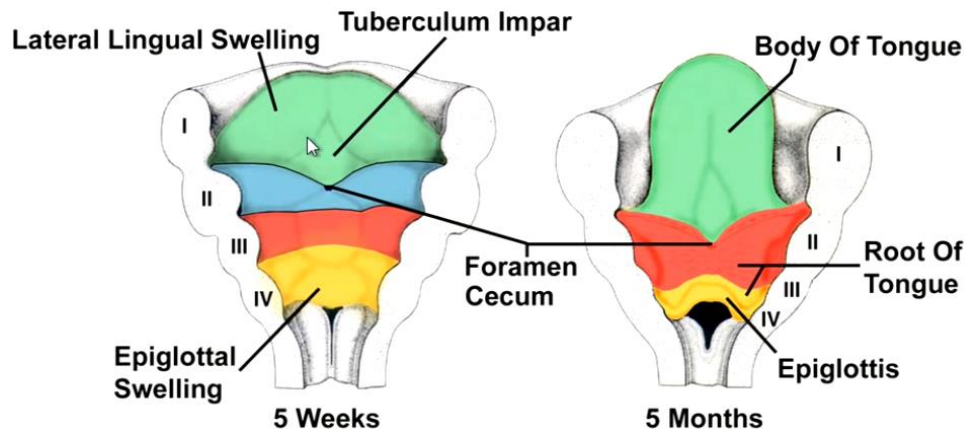
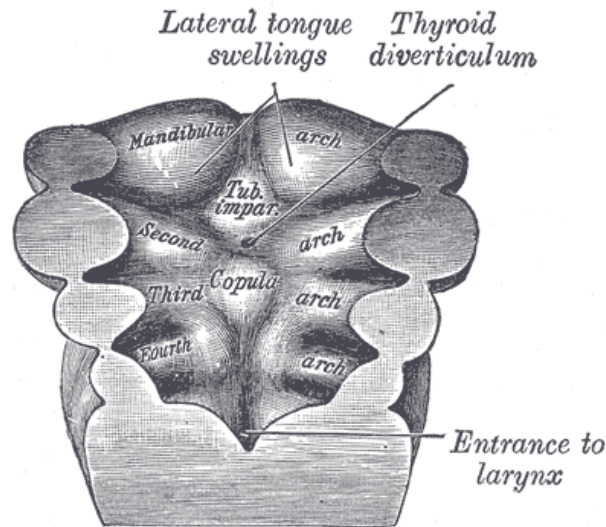
- **Unknown Primary Tumors:**
- Malignant neoplasm metastatic to cervical lymph nodes without an identifiable primary tumor following a comprehensive evaluation.
 - In most cases, primary tumor will be found by physical exam, imaging, or endoscopic evaluation.
- **5-10 % of patients presenting with nodal metastases will have an occult primary tumor.**
 - Tumor regression due to patient immunity.
 - Too small tumor (5 mm) to be seen on endoscopy and imaging.
 - Tumor located in a location that may escape notice:
 - Tonsillar crypts.
 - Cutaneous primary tumors.
 - Tumors outside H&N.
- **Squamous cell carcinoma (SCC)** is the most common histotype, followed by adenocarcinoma, undifferentiated carcinoma and others.
- Pattern of Nodal involvement:
 - Most commonly **Level II** then **level III**.
 - Unilateral lymph node involvement is more common.
 - Bilateral adenopathy is present in about 10% of patients.
 - Location of neck nodes may provide information regarding location of primary tumor:
 - **Level I:** Not Oropharynx.
 - **Levels II-III:** Oropharynx.
 - **Level IV:** Thyroid, Infraclavicular primary.
 - **Level V:** Nasopharynx.
- By using a thorough physical examination, Panendoscopy, and imaging findings, Occult primary site is discovered in 20–40% of cases.
- Most common site of occult primary tumor:
 - **Tonsil:**
 - 75% from ipsilateral tonsillar fossa.
 - 5% from contralateral tonsillar fossa.
 - 10% from bilateral tonsillar fossa.
 - **Base of tongue.**
 - **Pyramidal sinus.**
- Evaluation of patients with unknown primary:
 - **Full History and Complete H&N physical examination:**
 1. All cranial nerves.
 2. Head and scalp.
 3. Neck.
 4. Oral cavity.
 5. Nasal cavity.
 6. Nasopharynx.
 7. Oropharynx.
 8. Hypopharynx.
 9. Larynx.

- **Imaging studies:**
 - CT or/and MRI with contrast.
 - Head and Neck.
 - Chest, Abdomen and Pelvis.
 - PET/CT:
 - Detects tumor $\geq 5\text{mm}$ in size.
 - Helps to direct site of the biopsy.
 - Supplement but not a substitute, for endoscopy and biopsy in the setting of an unknown primary due to risk of false negative results.
 - False positive results:
 - Lymphoid tissue.
 - Salivary glands
 - Prior biopsy.
- **FNA:**
 - U/S guidance helps to target solid component.
 - Immunohistochemical stain to exclude Lymphoma.
 - EBV detection for Nasopharyngeal primary.
 - HPV detection for Oropharyngeal primary.
- **EUA + Panendoscopy:**
 1. Direct Laryngoscopy.
 2. Esophagoscopy.
 3. Bronchoscopy.
- **Multiple Biopsies:**
 - Ideally, biopsies should be performed after PET scan.
 - Allows biopsy of suspected area in PET.
 - Avoids false positive PET-scans at biopsy site.
 - Biopsy sites:
 1. All suspicious sites clinically and radiologically.
 2. Sites of possible origin of the primary:
 1. Nasopharynx.
 2. Base of the tongue.
 3. Pyriform sinus.
 4. Supraglottic area
 5. Bilateral Tonsillectomy
- **Other diagnostic procedures:**
 - Upper and Lower GI endoscopy mainly in patients with metastases to the left lower cervical (supraclavicular) lymph nodes.

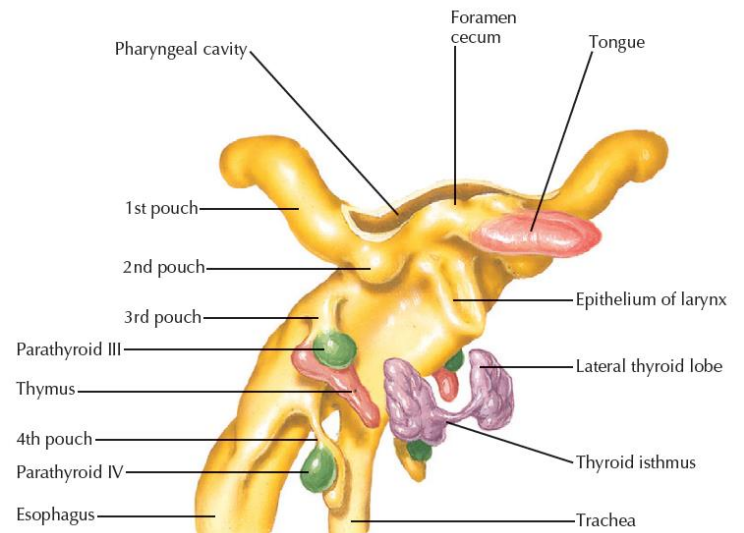
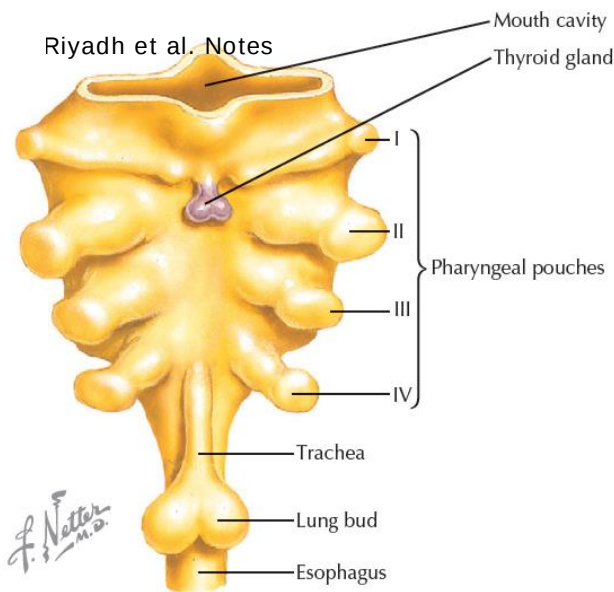
- Management of Unknown Primary Tumors:
- If no primary tumor site was identified (**Tx**), the standard therapy consists of:
 - 1. Radiation therapy to all mucosal surfaces of H&N.**
 - 2. Neck:**
 - **If N1:**
 - Ipsilateral Modified Radical Neck Dissection (MRND).
 - OR**
 - Ipsilateral radiation of the neck.
 - **If N2-3:**
 - Ipsilateral Modified Radical Neck Dissection (MRND).
 - AND**
 - Ipsilateral radiation of the neck.
- If occult primary tumor can be identified, treatment will be directed according the primary site.
- **Second Primary Tumors:**
- Patients with primary HNSCC have a high rate (3-5% per year) of developing second primary tumors than any other group of cancer patients.
 - Due to distribution of toxic effects from tobacco and alcohol.
- Major sites of involvement:
 - Other H&N sites.
 - Lung.
 - Esophagus.
- Types of Second primary:
 - 1. Synchronous:**
 - Second cancer occurs around the same time as the primary tumor.
 - 2. Metachronous:**
 - Second cancer occurs ≥ 6 months after the primary tumor.
- Post-treatment surveillance can diagnose second primary lesions earlier and more accurately allowing for potentially higher cure rates.
 - **PET/CT is modality of choice for identifying second primary tumors.**
 - Routine post-treatment Panendoscopy has not been shown to be effective in detecting second primary tumors.

Thyroid Embryology:

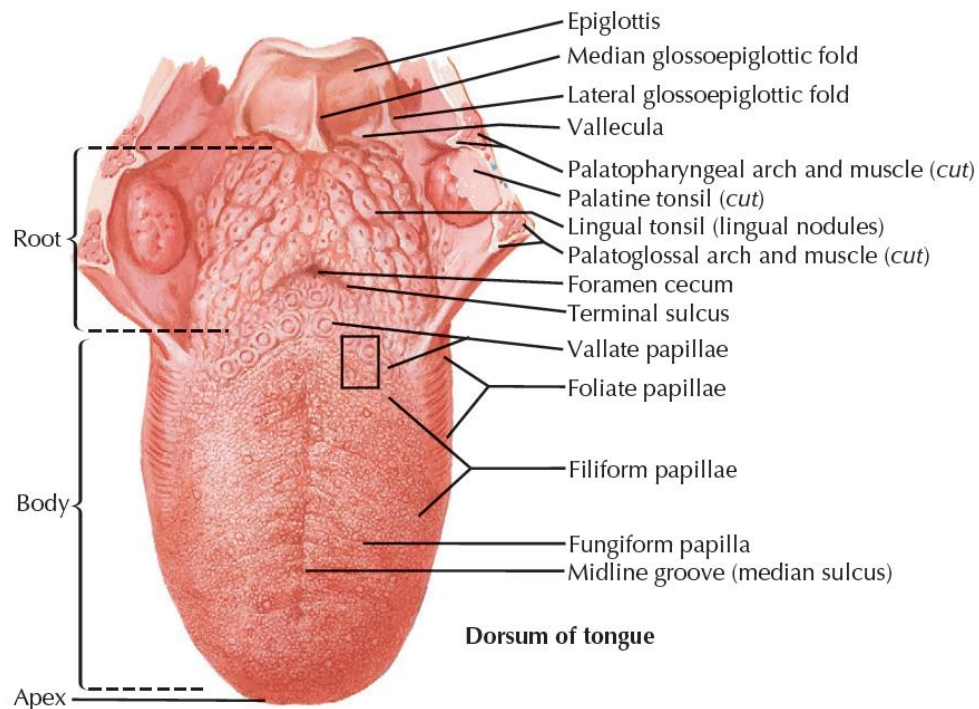
- 1st Endocrine glands to develop, on 24th day of gestation.
- Follicular element of Thyroid originates in **Foramen Cecum** as a proliferation of Endodermal epithelial cells on the median surface of the developing pharyngeal floor.
 - Between 1st and 2nd Pharyngeal Pouches.
 - Caudal to: **Tuberculum Impar** (Median Tongue Bud).
 - Rostral to: **Copula** (Hypobranchial eminence).

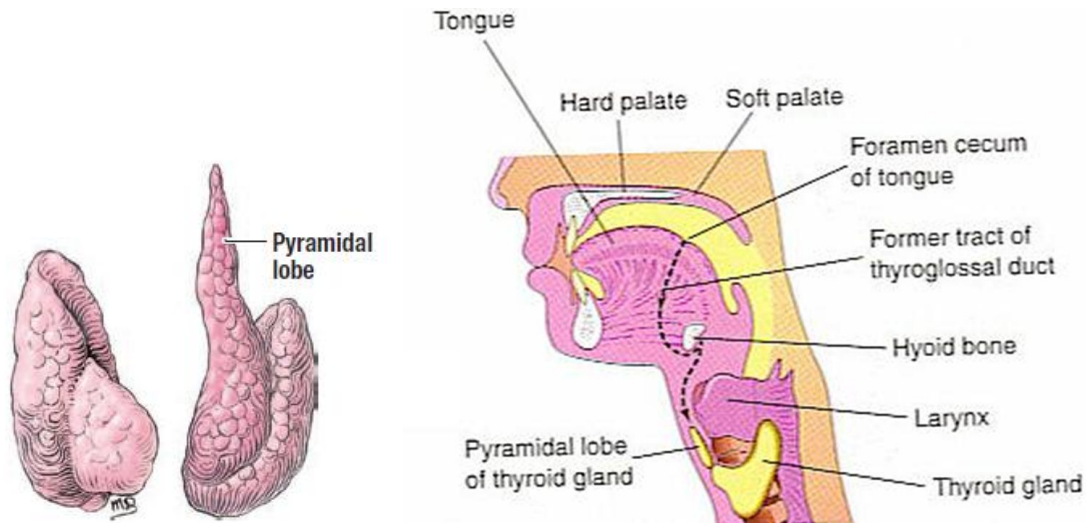


- Thyroid primordium starts as a simple midline thickening and develops to form Thyroid Diverticulum.
 - Initially hollow, later solidifies and becomes bilobed.
 - 2 lobes are located on either side of the midline and are connected via an Isthmus.

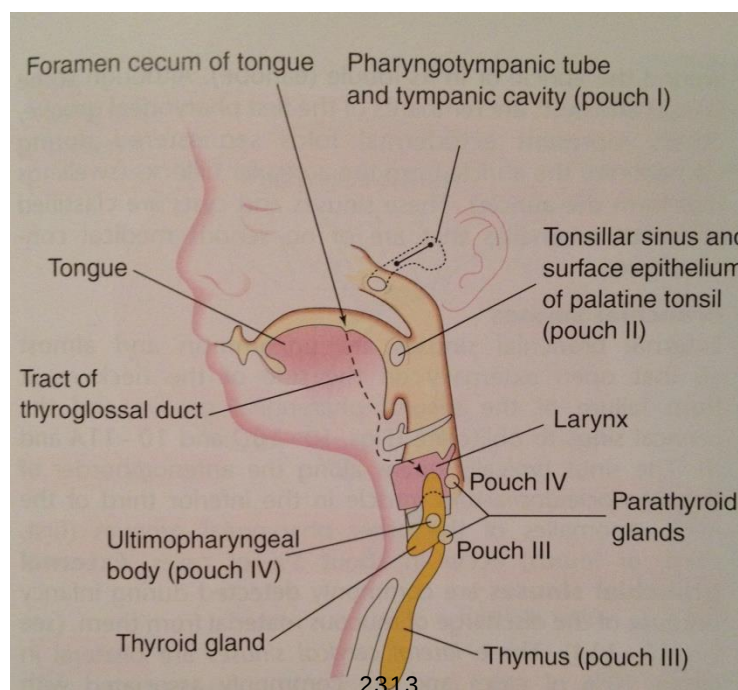


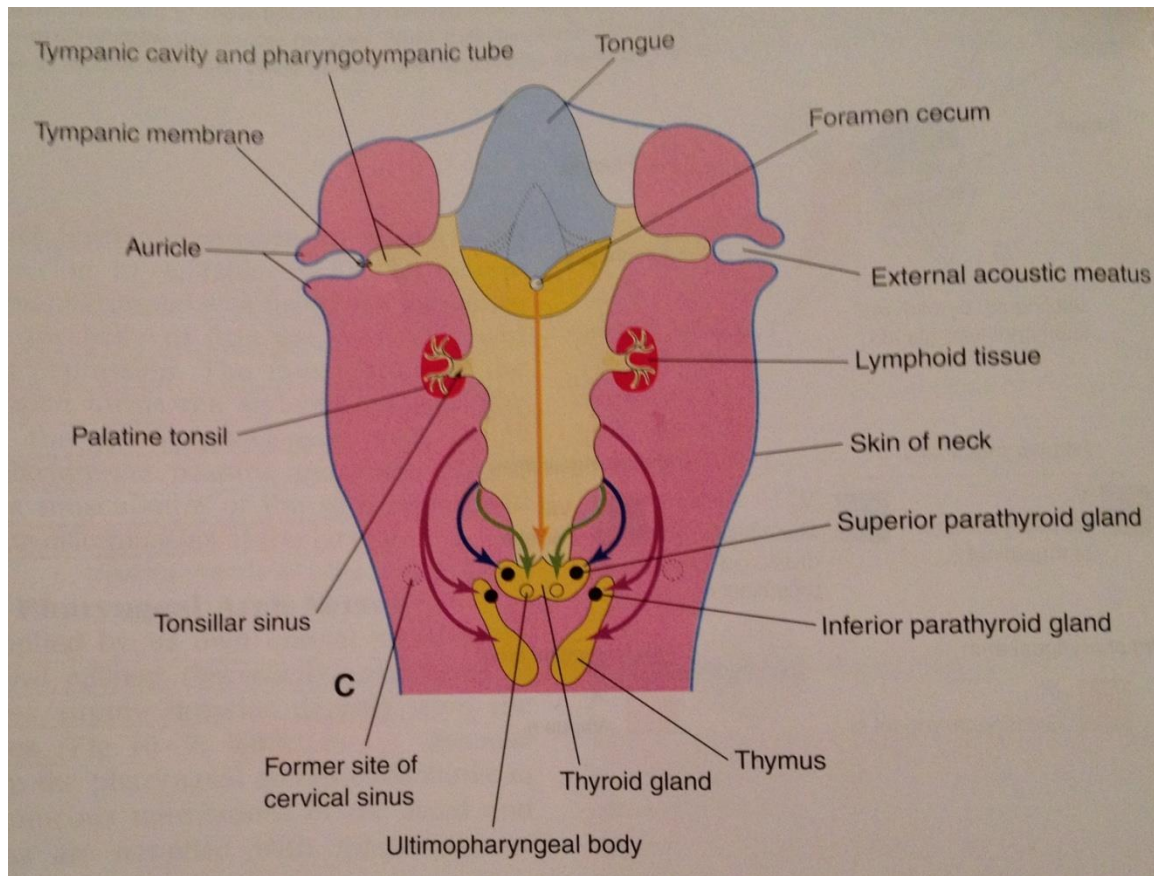
- Initial descent of Thyroid gland occurs Anterior to Pharyngeal gut.
- At this point, Thyroid is still connected to the Tongue via Thyroglossal duct.
 - Thyroglossal duct later solidifies and subsequently obliterates entirely during gestational weeks 7-10.
 - Remnants of this duct may still persist.
 - Foramen Cecum represents opening of Thyroglossal duct into the tongue and may be observed as a small blind pit in Midline between Anterior 2/3 and Posterior 1/3 of the tongue.
 - A pyramidal lobe may be observed in 50% of patients which represents a persistence of Inferior end of Thyroglossal duct that has failed to obliterate.





- Further descent of Thyroid gland carries it Anterior to Hyoid bone and, subsequently, Anterior to Laryngeal cartilages.
- As Thyroid gland descends, it forms its mature shape, with a median isthmus connecting 2 lateral lobes.
- Thyroid completes its descent in 7th gestational week, coming to rest in its final location immediately Anterior to Trachea.
- **Parafollicular cells (C cells)** are special subset of cells within Thyroid gland.
 - o Arise from Ventral part of 4th Pharyngeal Pouch as **Ultimopharyngeal body**.
 - o Consists of Neural crest cells which infiltrate Ultimopharyngeal body then incorporated into Thyroid gland.
 - o Secrete Calcitonin hormone which is necessary for regulation of Calcium.





- **Thyroid Embryology Clinical Correlations:**

- Athyreosis:

- Absent Thyroid gland.
- Rare.

- Thyroglossal Duct Cyst (TGDC):

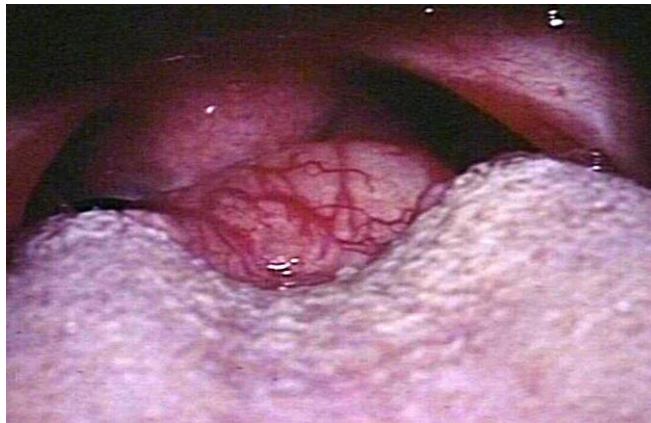
- Persistent of Thyroglossal duct that has failed to obliterate.
- Most common congenital cervical anomaly.
- Midline cystic masses located anywhere from Thyroid cartilage up the Base of Tongue.
 - 50% are located At or just below Hyoid bone.
- If TGDC ruptures, it may go on to form Thyroglossal duct sinus or a Thyroglossal duct fistula that exits through the overlying skin.
- 1% incidence of carcinoma within the cyst.
- May be only functioning thyroid tissue.
- Because Hyoid bone develops in an anterior direction and may surround Thyroglossal duct, Surgeon should resect Central portion of Hyoid bone along with the cyst (Sistrunk procedure).

- Pyramidal Lobe:

- Observed in 50% of patients.
- Represents a persistence of Inferior end of Thyroglossal duct that has failed to obliterate.

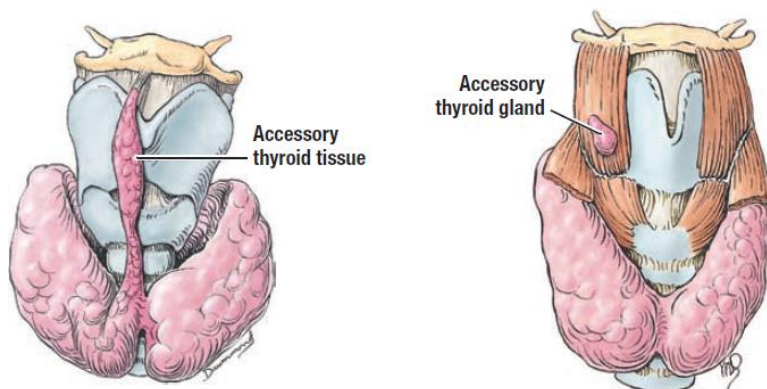
- Aberrant or Ectopic Thyroid:

- Occurs anywhere along the path of initial descent of the thyroid from Foramen cecum to Sternal notch.
 - Most commonly at Base of Tongue, just posterior to Foramen cecum (**Lingual Thyroid**).
 - Represents Failure of the thyroid to descend.
 - Most of patients are having hypothyroidism.
- Incomplete descent of Thyroid results in final resting point of Thyroid gland to be high in Neck or just below Hyoid bone.



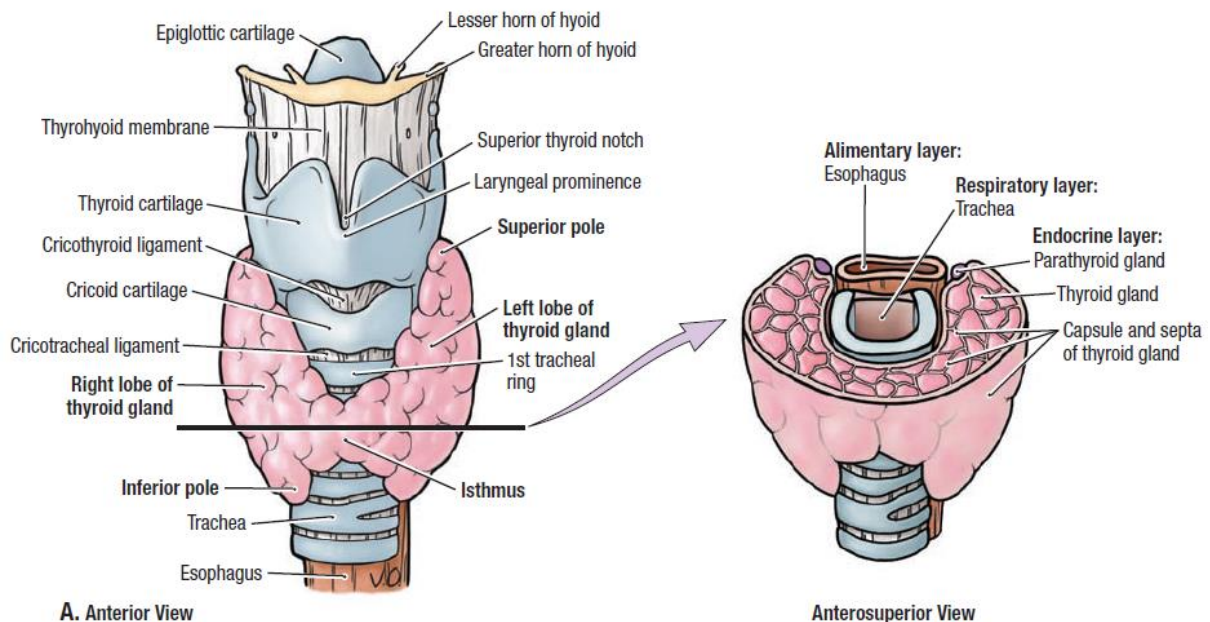
- Accessory Thyroid tissue:

- Arises from remnants of Thyroglossal duct anywhere along the path of Thyroglossal duct tract.
- May be functional but generally insufficient for normal function if the main Thyroid gland is entirely removed.

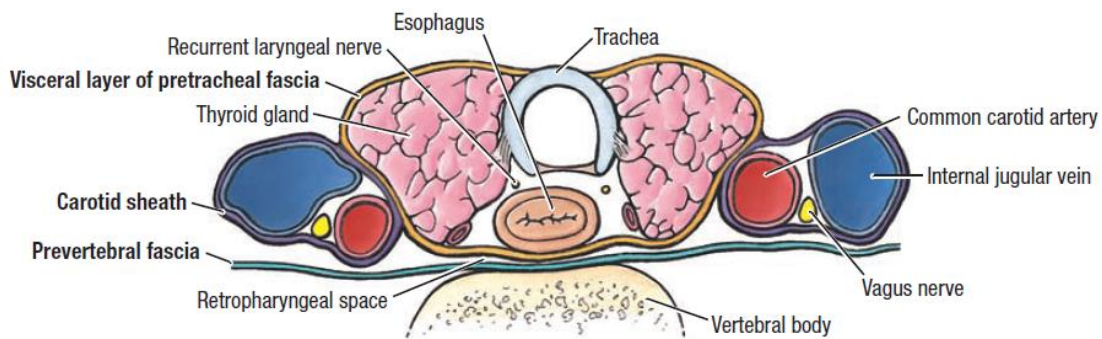


Thyroid Anatomy:

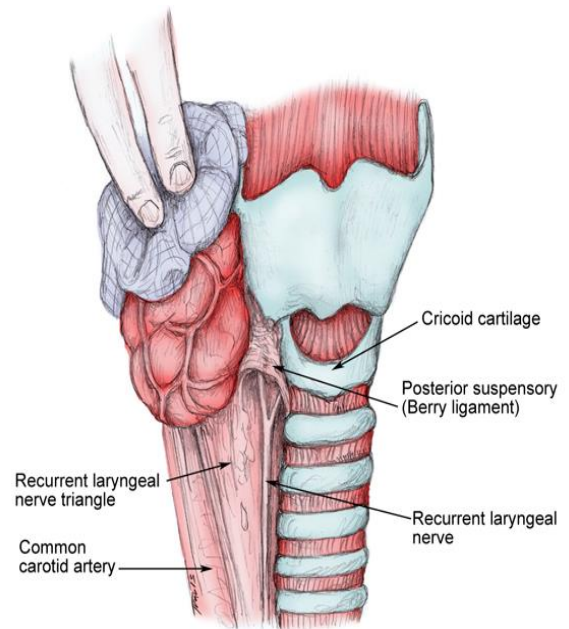
- Highly vascular, brownish-red gland, located Anteriorly in lower neck.
- Extends from level of C5 to T1.
- Varies from an H to a U shape.
- Formed by 2 elongated lateral lobes with superior and inferior poles.
- Both lobes are connected by Isthmus overlying 2nd to 3rd tracheal rings.
 - o Isthmus is encountered during Tracheotomy and must be retracted (superiorly or inferiorly) or divided.
 - o Occasionally, isthmus is absent, and the gland exists as 2 distinct lobes.



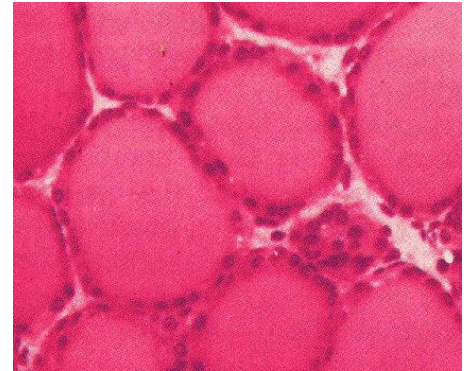
- Thyroid is ensheathed by Visceral layer of Pretracheal Fascia (Middle layer of Deep Cervical Fascia), which attaches it firmly to the laryngoskeleton.
 - o Responsible for movement of Thyroid gland and related structures during swallowing.



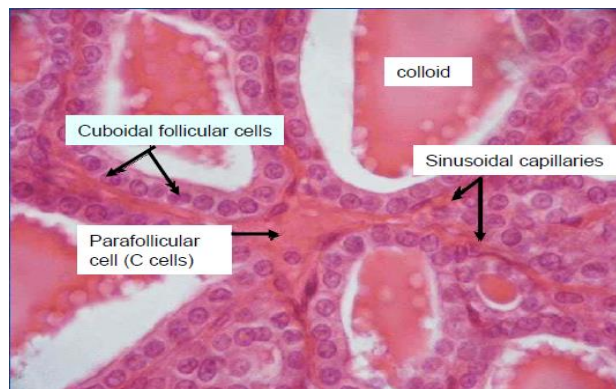
- Anterior Suspensory Ligament:
 - Connects Superior-medial aspect of each thyroid lobe to Cricoid and Thyroid cartilage.
- Posterior Suspensory Ligament (Berry's Ligament)
 - Connects Posteromedial aspect of thyroid gland to the side of Cricoid Cartilage, 1st and 2nd Tracheal rings.
 - Recurrent Laryngeal Nerve (RLN) usually passes deep to Berry ligament.



- Thyroid has an inner true capsule, which is thin and adheres closely to the gland.
- Extensions of this capsule within the substance of the gland form numerous septae, which divide it into lobes and lobules.
- Lobules are composed of Follicles which consist of a layer of simple epithelium enclosing a colloid-filled cavity.



- Types of Thyroid Epithelial cells:
 - 1. Follicular cells:**
 - Responsible for formation of Colloid.
 - Colloid contains Iodothyroglobulin, a precursor of thyroid hormones.
 - 2. Parafollicular cells (C-cells):**
 - Lie adjacent to the follicles within the basal lamina.
 - Produce the hormone Calcitonin, a protein central to calcium homeostasis.



- **Blood Supply:**

1. Superior Thyroid Artery:

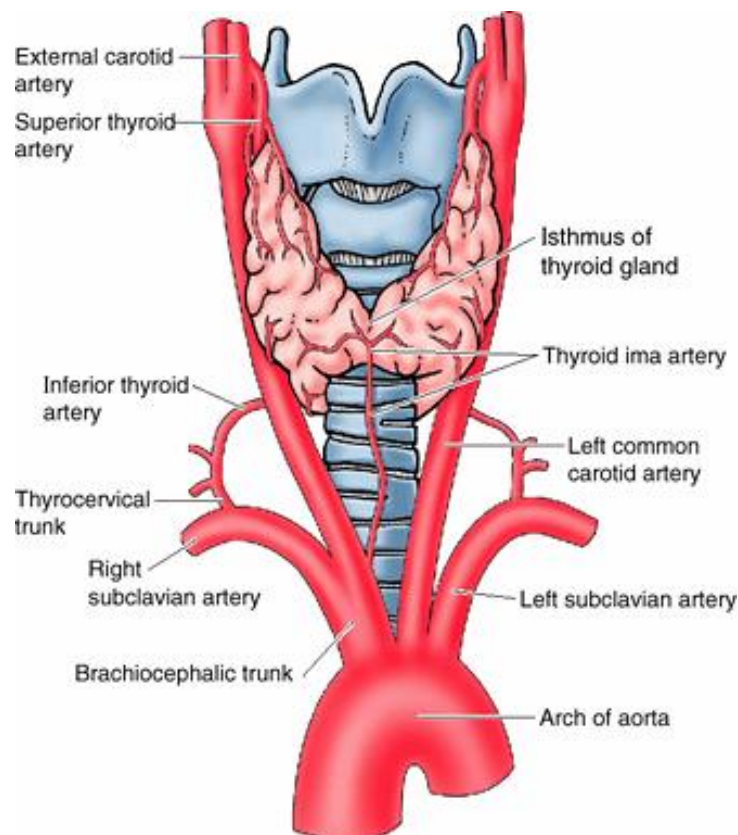
- 1st branch of External Carotid Artery.
- Arises just below Greater cornu of Hyoid bone.
- Descends along Inferior constrictor muscle to reach upper pole of Thyroid gland.
- Cephalic to Upper pole, External branch of Superior Laryngeal Nerve runs with Superior Thyroid Artery before turning medially to supply Cricothyroid muscle.
 - High ligation of Superior Thyroid Artery during Thyroidectomy places this nerve at risk of inadvertent injury, which would produce dysphonia by altering pitch regulation.
- It is the primary blood supply to approximately 15% of superior parathyroid glands.
- Gives rise to:

1. Superior Laryngeal Artery:

- Prices Thyrohyoid membrane to supply the larynx.
- Accompanied with Internal branch of Superior laryngeal nerve.

2. Cricothyroid Artery:

- Runs cephalic to upper pole and runs toward the midline on the cricothyroid ligament.
- Lacerated during emergent cricothyroidotomy.

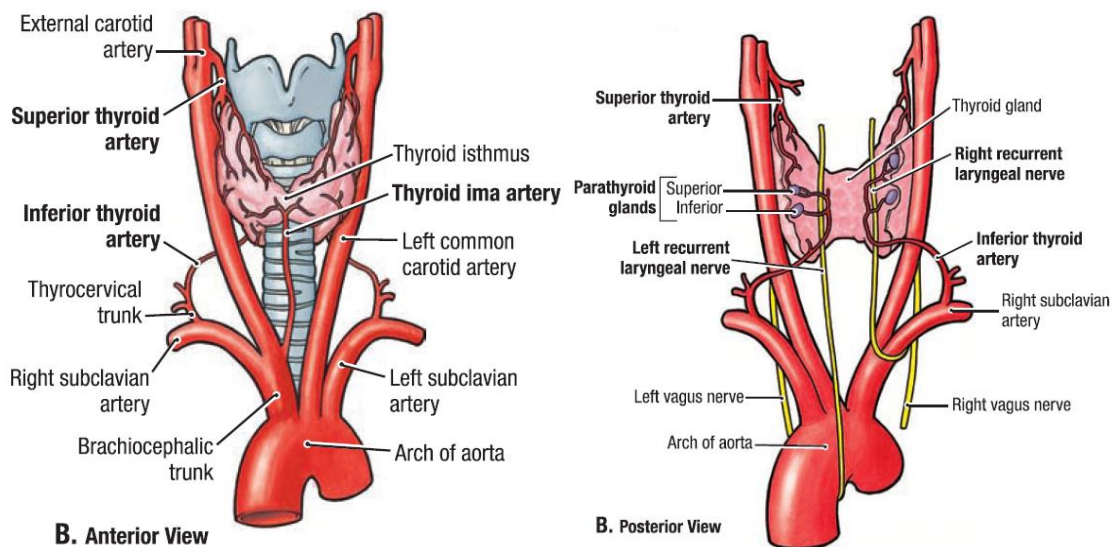


2. Inferior Thyroid Artery:

- Branch of Thyrocervical trunk of Subclavian Artery.
- Ascends vertically along Medial border of Anterior Scalene Muscle to enter Tracheoesophageal groove.
- Penetrates posterior aspect of Lower lobes.
- Runs Posterior to Carotid sheath and Anterior to Vertebral Artery.
- Has a variable branching pattern.
- Supplies the inferior parathyroid glands and 85% of superior parathyroid glands.
- Accompanied by Recurrent laryngeal Nerve (RLN).

3. Thyroidea ima Artery:

- Presents in 3% of population.
- Arise from Aortic Arch or Brachiocephalic (Innominate) Artery.
- Enter Thyroid gland at inferior border of isthmus.



- Venous Drainage:

1. Superior Thyroid Vein:

- Ascends along Superior Thyroid Artery.
- Drains into Internal Jugular Vein.

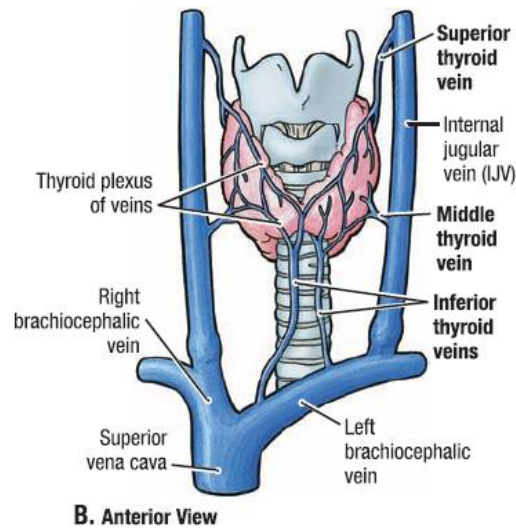
2. Middle Thyroid Vein:

- Follows a direct course laterally to drain into Internal Jugular Vein.

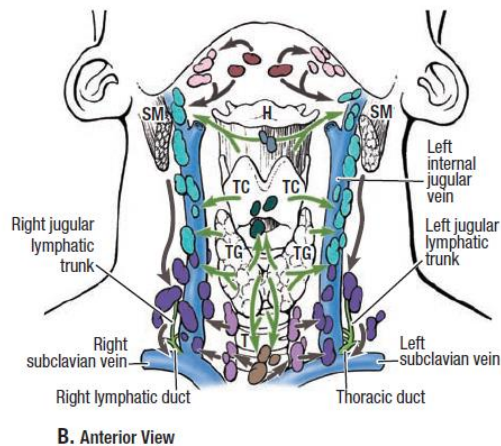
3. Inferior Thyroid Vein:

- Drains into Left Brachiocephalic Vein.

- Occasionally, both inferior veins form a common trunk called Thyroid ima vein, which empties into the left brachiocephalic vein.



- **Lymphatic Drainage:**
- Extensive and flows multidirectionally.
- Immediate lymphatic drainage to Central LN compartment (Level VI).
 - Precricoid (Delphian) LN.
 - Pretracheal LN.
 - Paratracheal LN.

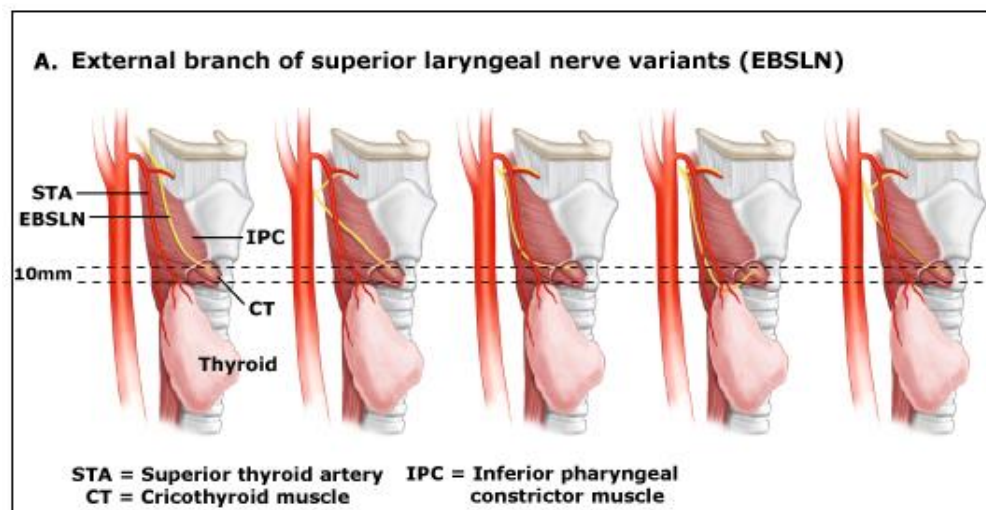


Lymph nodes:			
Buccinator	Paratracheal	Superficial cervical	SM Sternocleidomastoid
Inferior deep cervical	Parotid	Superior deep cervical	T Trachea
Infrathyoid	Prelaryngeal	Structures:	TC Thyroid cartilage
Jugulodigastric	Pretracheal		TG Thyroid gland
Jugulo-omohyoid	Retropharyngeal	→ Initial drainage	P Palatine tonsil
Mastoid (retro-auricular)	Submandibular	→ Secondary (subsequent) drainage	PG Parotid gland
Occipital	Submental	H Hyoid	Ph Pharyngeal tonsil

- **Important Anatomical Relations:**

- **Superior Laryngeal Nerves (SLN):**

- Originate from vagus nerve as it exits base of the skull.
- Courses with Superior thyroid artery until approximately 1 cm before the artery enters the capsule of Superior pole of the thyroid.
- Injury to SLN during surgery can be minimized by dissecting the superior thyroid vessels at level of thyroid capsule.
 - Stay as close as possible to thyroid capsule.
- Maintaining hemostasis is critical because control of bleeding from superior thyroid vessels with hemostats may put SLN at risk.
- Two primary branches:
 - **External branch:**
 - Primarily motor:
 - Inferior constrictor muscle.
 - Cricothyroid muscle.
 - Travels with Superior thyroid artery until approximately 1 cm before the artery enters superior thyroid pole.
 - Divides into branches that enter Lateral inferior pharyngeal constrictor muscle and Cricothyroid muscle.
 - Rates of injury are as high as 30%.
 - **Internal branch:**
 - Sensory to the larynx.
 - Enters the larynx through Thyrohyoid membrane superior to the external branch.



- **Recurrent Laryngeal Nerve (RLN):**

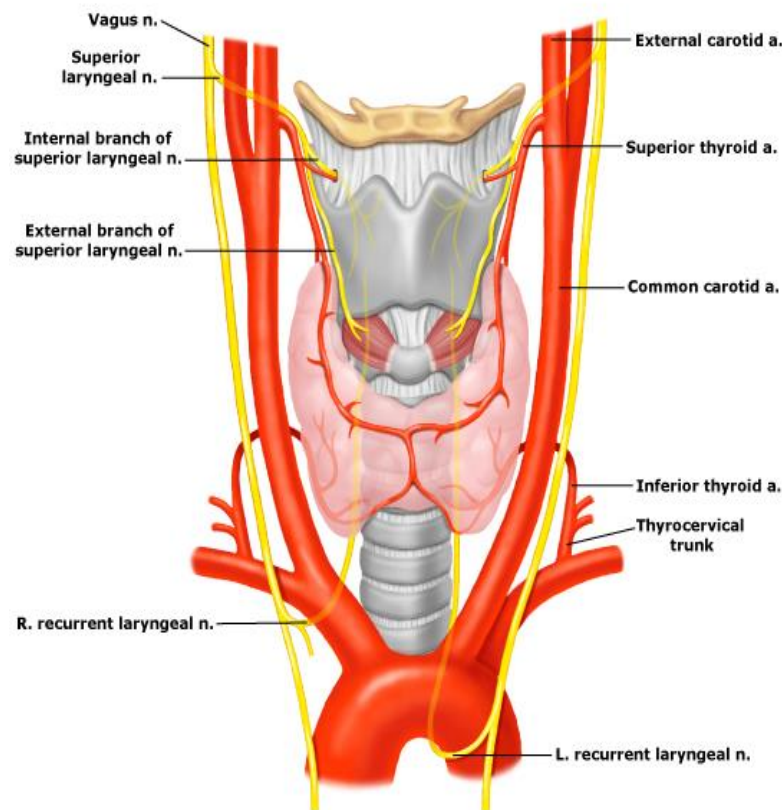
- Provides both sensory and motor function to Larynx.
- Sensory to Subglottic region.
- Innervates all muscles to the larynx Except Cricothyroid muscle.
- Associated with Inferior thyroid artery at junction of lower and middle thirds of thyroid gland.

○ **Left RLN:**

- Originates from Left Vagus nerve below Arch of Aorta at the point where Ligamentum Arteriosum (**6th arch**) is attached.
- Loops posterior to Arch of Aorta
- Ascends to the side of Trachea at Tracheoesophageal groove.
- Crosses deep to Inferior thyroid artery.

○ **Right RLN:**

- Originates from Right Vagus nerve at level of Subclavian Artery (**4th arch**).
- Loops posterior to Subclavian Artery.
- Ascends to the side of Trachea at Tracheoesophageal groove.
- Courses through Ligament of Berry and enters larynx through first tracheal ring, inferior to the cricothyroid muscle.



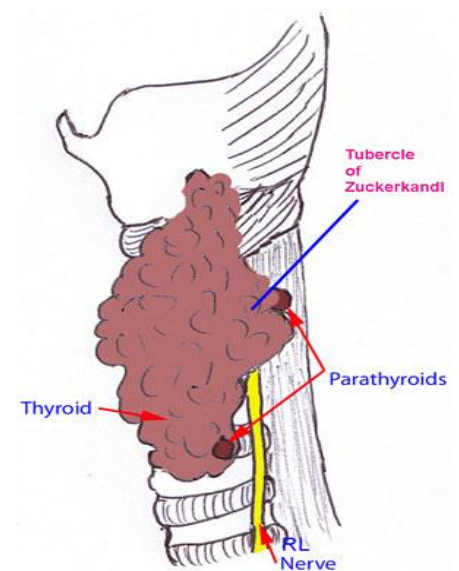
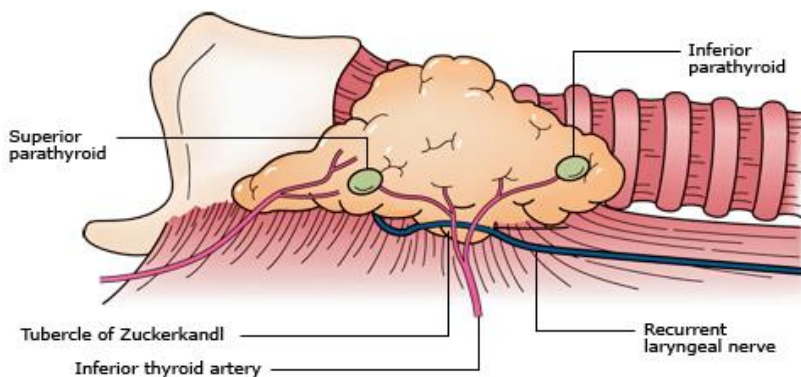
- **Reed classification (Relation between RLN and Inferior Thyroid Artery):**

1. RLN Posterior to Inferior Thyroid Artery (**40%**).
2. Between branches of the artery (**35%**)
3. RLN Anterior to Inferior Thyroid Artery (**20%**)

- Relationship between RLN and Inferior Thyroid Artery on one side of the neck is similar to that found on the other side in only 17% of the population.

B. Recurrent laryngeal nerve variants (RLN)		Right (percent)	Left (percent)
Anterior to inferior thyroid artery		24.1	19.7
Posterior to inferior thyroid artery		64.1	71.5
Between inferior thyroid artery branches		7.6	5.4

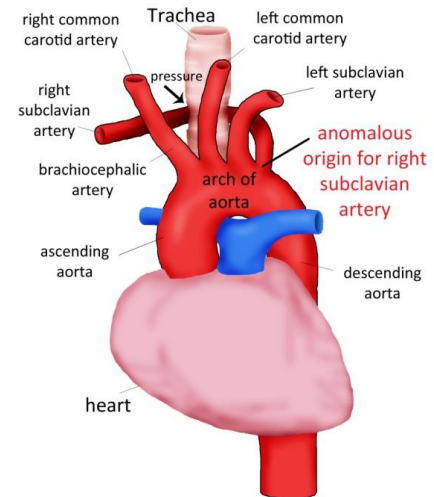
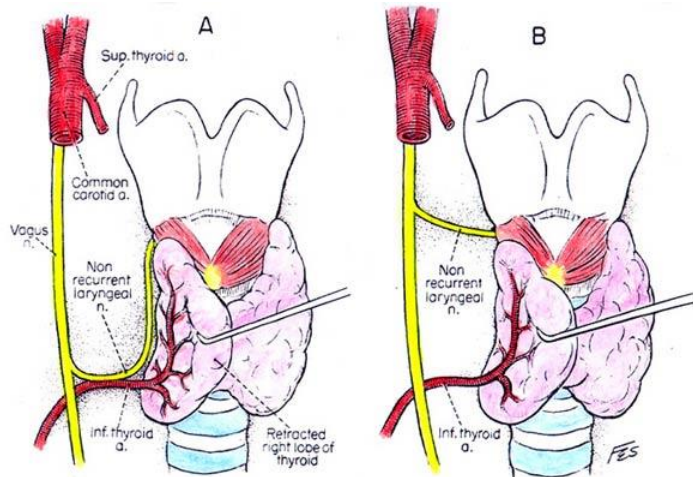
- Another hint to RLN location is **Zuckerkindl tubercle**.
- Pyramidal extension of thyroid on its posterior aspect.
 - RLN usually pass into a cleft medial to it.



- **Non-Recurrent Laryngeal Nerve:**

○ Right Non-Recurrent:

- 0.5–0.6% of Patients.
- More common than left.
- Occurs with **Arteria Lusoria**.
 - Right Retroesophageal Subclavian Artery and Absent Innominate Artery.



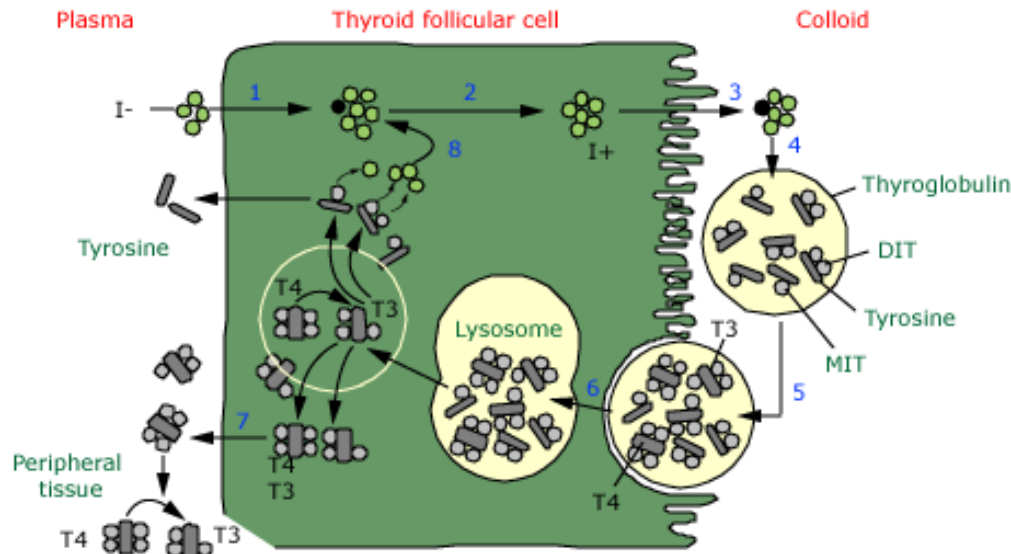
○ Left Non-Recurrent:

- Very rare.
- 0.004% of Patients.
- Patient must have the following anomalies to occur:
 1. Right-sided Aortic Arch (Situs Inversus)
 2. Left Subclavian Artery must also be Lsoria (Absent Aortic segment between Left Carotid & Subclavian).
 3. Arterial ligament must be on Right side

Thyroid Physiology:

- Thyroid hormones are critical for brain and somatic development in infants and for metabolic activity in adults.
- Two biologically active thyroid hormones:
 - 1. Tetra-iodotyronine (T4):**
 - 2 iodine atoms on tyrosine (inner) ring.
 - 2 iodine atoms on phenyl (outer) ring.
 - Thyroxine
 - **Most Numerous form (90% of thyroid output).**
 - Produced only by Thyroid.
 - Half life is one week.
 - 99% is found bound in the serum.
 - 1. Thyroxine-Binding Globulin (TBG):**
 - Binds 75% of T4
 - Very high affinity for T4.
 - Made in Liver.
 - Increased with increased estrogen and pregnancy.
 - 2. Thyroxine Binding Pre-Albumin (TBPA):**
 - Binds 15% of T4
 - 3. Albumin:**
 - Binds 5% of T4.
 - 2. Tri-iodotyronine (T3):**
 - 2 iodine atoms on tyrosine (inner) ring.
 - 1 iodine atom on phenyl (outer) ring.
 - **Most Active form (4-times more active than T4).**
 - Half life is one day.
 - Produced by:
 - Thyroid (20%).
 - Conversion of T4 in Extrathyroidal tissues (**80%**)
 - Liver, Kidneys, Muscles, and Pituitary.
 - 99% is found bound in the serum.
- Iodine is essential for normal thyroid function.
- Obtained only by consumption of foods that contain it.
 - Iodized salt, Dairy products and some vegetables.
 - Dietary iodine is absorbed as Iodide.

- **Steps of Thyroid hormones synthesis:**



1. Iodide Uptake by Thyroid:

- Stimulated by TSH.

2. Oxidation of Iodide to Iodine.

- Catalyzed by Peroxidase.
 - **Inhibited by Thioamides (Methimazol+ Propylthiouracil).**

3. Iodination of Thyroglobulin:

- Mono-iodotyrosine (**MIT**).
- Di-iodotyrosine (**DIT**).
- Catalyzed by Peroxidase.
 - **Inhibited by Thioamides (Methimazol+ Propylthiouracil).**

4. Coupling of Iodinated tyrosine:

- 1 DIT + 1 MIT couple → Tri-iodotyronine (**T3**)
- DITs couple → Tetra-iodotyronine (**T4**)
- Catalyzed by Peroxidase.
 - **Inhibited by Thioamides (Methimazol+ Propylthiouracil).**

5. Storage of Thyroid hormone:

- Stored as Iodinated Thyroglobulin in Follicular colloid.
- Last for 2-3 months.
 - **Inhibited by High dose of Iodide.**

6. Proteolytic release of T3,T4,DIT, and MIT from Thyroglobulin.

7. Deiodination of DIT and MIT:

- Yield tyrosine and iodide.
- Iodide is recycled for thyroid hormone synthesis.

8. Secretion of T3 and T4 into the blood.

9. Conversion of T4 to T3 in Peripheral tissues:

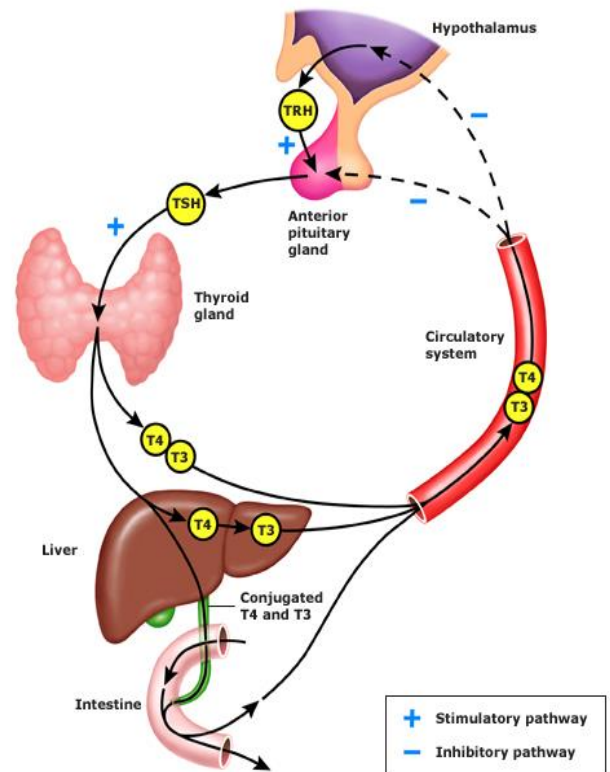
- Liver, Kidneys, Muscles, and Pituitary.
- Catalyzed by 5'deiodinase.
 - **Inhibited by Propranolol and Propylthiouracil.**

Type	Percent
bound to thyroxine-binding globulin (TBG)	70%
bound to transthyretin or "thyroxine-binding prealbumin" (TTR or TBPA)	10-15%
albumin	15-20%
unbound T ₄ (fT ₄)	0.03%
unbound T ₃ (fT ₃)	0.3%

- Most of T₄ and T₃ in serum is bound.
- Changes in serum concentrations of binding proteins (TBG) have a large effect on serum TOTAL T₄ and T₃ concentrations.
 - o They do not alter free hormone concentrations.
 - Estrogen-induced TBG excess cause high total T₄ concentrations due to increased TBG-bound hormone, but the physiologically important free T₄ concentrations are normal.
 - It is necessary to estimate free hormone concentrations.
- Serum free T₄ and T₃ concentrations determine the hormones' biological activity.
- Free Thyroid hormones are available for uptake into cells and interaction with nuclear receptors.
- Bound Thyroid hormones represent a circulating storage pool that are not immediately available for uptake into cells.
- Measuring concentrations of free thyroid hormones is of great diagnostic value.

- **Regulation of thyroid hormones production:**
- Regulation of thyroidal biosynthesis and secretion of T4 and T3:

- Thyrotropin-Releasing Hormone (**TRH**):
 - Secreted by Hypothalamus.
 - Stimulates TSH Secretion from Pituitary.
 - **Inhibited by:**
 - High serum T4 and T3.
 - **Stimulated by:**
 - Low serum T4 and T3.
- Thyroid-stimulating Hormone (**TSH**).
 - Secreted by Anterior Pituitary.
 - Stimulates Thyroid hormones synthesis and secretion.
 - Activates growth of thyroid gland.
 - **Inhibited by:**
 - High serum T4 and T3.
 - **Stimulated by:**
 - Low serum T4 and T3.
 - Thyrotropin-Releasing Hormone (TRH).



- Regulation of extrathyroidal conversion of T4 to T3:
 - by nutritional, hormonal, and illness-related factors.
- **Thyroid hormone effects:**
 - Elevates metabolic rate.
 - Thermogenesis.
 - Increases oxygen consumption.
 - Essential for normal neural and skeletal development.
 - Stimulates chondrocytes, Bone reabsorption, growth of neuronal tissue.
 - Increases sympathetic activity.
 - Increases heart rate and contractility.
 - Releases steroid hormones.
 - Stimulates erythropoiesis.

- **Thyroid Pharmacology:**

- Thinamides:

- MOA:
 - Inhibits T4 conversion.
 - Inhibits Oxidation and Organification of Iodine.
- S/E:
 - Hepatitis.
 - Agranulocytosis.
 - Parotiditis
- Ex:
 - Propylthiouracil (PTU).
 - Methimazole (Tapazole):
 - Contraindicated in Pregnancy.

- Iodine (Lugol's solution):

- MOA:
 - Excess iodine inhibits thyroid hormone
- Contraindicated in Rheumatoid arthritis.Pregnancy.

- Glucocorticoids:

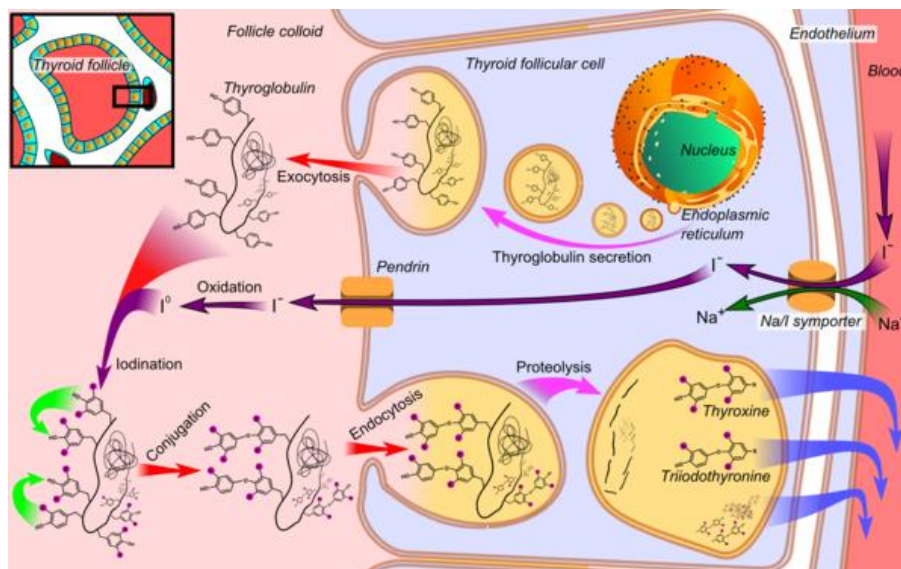
- Suppress Hypothalamic–pituitary–thyroid axis.

- Lithium:

- Inhibits thyroid hormone release.
- Contraindicated in renal failure and cardiovascular disease.

- B-blockers:

- Used to control peripheral manifestation of sympathetic overactivity (inhibits thyrotoxicosis)
- Ex:
 - Propanolol.
 - Metoprolol.



- **Thyroid Function Test:**

1. TSH concentration:

- Most sensitive for Hypo/Hyperthyroidism.

2. Total T₄ concentration.

3. Total T₃ concentration.

4. Free T₄:

- More specific for Hypo/Hyperthyroidism.

5. Resin T₃ Uptake (RT₃U):

- Measures the binding capacity of existing TBG.
- Indirect measurement of TBG.
- High RT₃U:
 - Low total TBG.
 - Low total T₄.
 - Normal free T₄ (euthyroid)
- Low RT₃U:
 - High total TBG (pregnancy or estrogen from OCP).
 - High total T₄.
 - Normal free T₄ (euthyroid)

TABLE 4-1. Thyroid Function Test Results

Condition	TBG	Total T ₄	RT ₃ U	Free T ₄
Normal	Normal	Normal	Normal	Normal
Pregnant	↑	↑	↓	Normal
Liver/Renal disease	↓	↓	↑	Normal
Hyperthyroid	Normal	↑	↑	↑
Hypothyroid	Normal	↓	↓	↓

Patterns of thyroid function tests during assessment of thyroid function

Serum TSH	Serum Free T ₄	Serum T ₃	Assessment
Normal hypothalamic-pituitary function			
Normal	Normal	Normal	Euthyroid
Normal	Normal or high	Normal or high	Euthyroid hyperthyroxinemia
Normal	Normal or low	Normal or low	Euthyroid hypothyroxinemia
Normal	Low	Normal or high	Euthyroid: triiodothyronine therapy
Normal	Low normal or low	Normal or high	Euthyroid: thyroid extract therapy
High	Low	Normal or low	Primary hypothyroidism
High	Normal	Normal	Subclinical hypothyroidism
Low	High or normal	High	Hyperthyroidism
Low	Normal	Normal	Subclinical hyperthyroidism
Abnormal hypothalamic-pituitary function			
Normal or high	High	High	TSH-mediated hyperthyroidism
Normal or low*	Low or low-normal	Low or normal	Central hypothyroidism

* In central hypothyroidism, serum TSH may be low, normal or slightly high.

- [Screening for thyroid dysfunction:](#)
- **Normal TSH:**
 - No further test.
- **High TSH:**
 - Free T4 is requested to determine degree of Hypothyroidism.
- **Low TSH:**
 - Free T4 and T3 requested to determine degree of Hyperthyroidism.

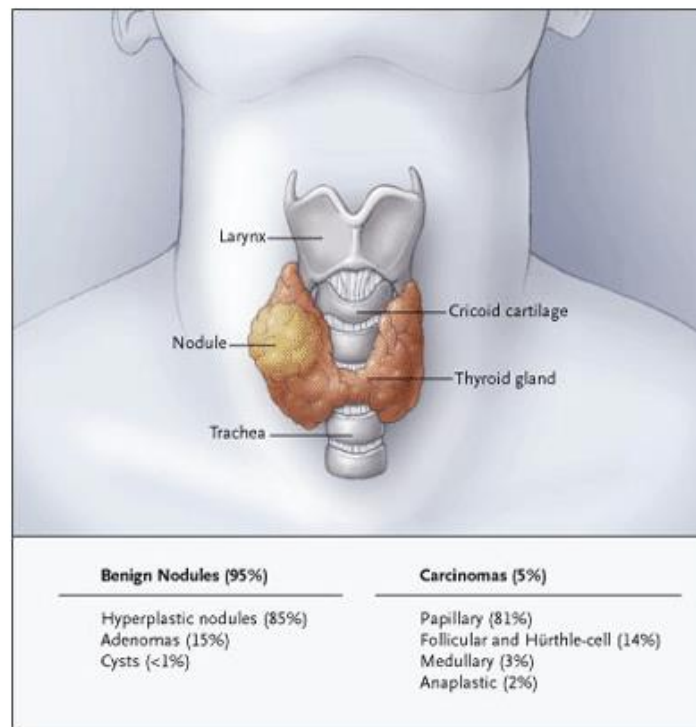
- [Monitoring levothyroxine therapy:](#)
- **Normal TSH:**
 - Continue same levothyroxine dose.
- **High TSH:**
 - Increase levothyroxine dose.
- **Low TSH:**
 - Decrease levothyroxine dose.

- [Monitoring suppressive therapy:](#)
- **Normal TSH:**
 - Increase levothyroxine dose.
- **TSH (0.06 - 0.5 mU/L):**
 - Appropriate for suppressive therapy.
- **TSH (<0.05 mU/L):**
 - Request serum free T4 to assess degree of potentially excessive therapy.

- **Anti-thyroid Antibodies:**
- 1. Thyroglobulin Antibodies (Tg):
 - Synthesized by follicular cells.
 - Stored as colloid.
 - Found in All patients with Hashimoto's thyroiditis.
- 2. Thyroid peroxidase Antibodies (TPO):
 - Catalyzes Iodination of tyrosine residues of Thyroglobulin to form MIT and DIT.
 - Found in All patients with Hashimoto's thyroiditis.
- 3. TSH receptor Antibodies:
 - Found in patients with Graves' disease.

Approach for Evaluation of Thyroid Nodules:

- Thyroid nodule is a discrete lesion within thyroid gland that is radiologically distinct from surrounding thyroid parenchyma.
- **(5%)** of population have Palpable Thyroid Nodules.
 - o **(5%)** of these palpable thyroid nodules are Malignant.
- Non-palpable nodules detected on US or other imaging studies are termed incidentally discovered nodules or "incidentalomas".
 - o Have same risk of malignancy as palpable nodules with the same size **(5%)**.



- Indications to Investigated Thyroid Nodules:

1. Thyroid Nodules >1cm.

- Have a greater potential to be clinically significant cancers.

2. Thyroid Nodules <1cm with one of the following:

1. Suspicious US findings.
2. Positive LN.
3. History of H&N irradiation.
4. Family History of Thyroid Ca.

- Approach to clinically and incidentally discovered thyroid nodules:

1. Complete History and Physical assessment.
2. Laboratory investigations.
3. Imaging studies.
4. FNA biopsy.

History:

- Neck Mass:
 - Midline.
 - Rate of growth.
 - Pattern of growth.
 - Pain or Painless
 - Moving with swallowing.
 - LN enlargement.
- Associated Symptoms:
 - **Hyperthyroidism:**
 - Elevated Metabolic Rate:
 - Weight loss, fatigue, sweating, and heat intolerance.
 - Increased Sympathetic Activity:
 - Palpitations, tachycardia, and tremor.
 - Increased Protein Degradation:
 - Weakness, fine hair.
 - Neurologic Effects:
 - Increased deep tendon reflex, nervousness.
 - Reproductive Effects:
 - Abnormal menstrual cycle, decreased libido
 - **Hypothyroidism:**
 - Reduced Metabolic Rate:
 - Weight gain, cold intolerance, lethargy.
 - Decreased Sympathetic Activity:
 - Bradycardia, constipation.
 - Decreased Protein Degradation:
 - Weakness, fine hair, hoarseness.
 - Decreased Neurologic Response:
 - Slowed deep tendon reflex, depression.
 - Myxedema:
 - Non-pitting edema due to mucopolysaccharides deposition.
 - Cretinism:
 - Hypothyroidism in children; mental retardation, impaired physical growth, macroglossia, protruberant abdomen.
 - **Compressive or invasive symptoms:**
 - Dysphagia.
 - Airway obstruction.
 - Hoarseness.
 - Hemoptysis.
- History of Risk Factors:
 - Radiation exposure.
 - Family history of thyroid disorders.

Physical Exam:

- Complete Head and Neck exam:
 - Palpation of Thyroid Nodule:
 - Number.
 - Size.
 - Location.
 - Consistency.
 - Mobility
 - Tenderness.
 - Skin changes.
 - Movement with swallowing.
 - Complete Lymph Nodes and Parotid palpation.
 - Vocal folds Assessment.
- Signs suspicious for malignancy in physical examination:
 1. Fixed nodule.
 2. Lateral cervical lymphadenopathy.
 3. Vocal fold paralysis

Laboratory Investigations:

- Thyroid Function Test:
 - TSH and Free T4.
 - Evaluate the functional status of thyroid gland.
 - Only laboratory test indicated for initial evaluation of thyroid nodule.
- Serum Thyroglobulin (Tg) level:
 - Routine measurement is **NOT Recommended**.
 - Elevated in most thyroid diseases.
 - Non-sensitive and non-specific for thyroid cancer.
- Serum Calcitonin level:
 - Routine measurement is **NOT Recommended**.
 - Indicated only if suspecting MTC.

Imaging Studies:

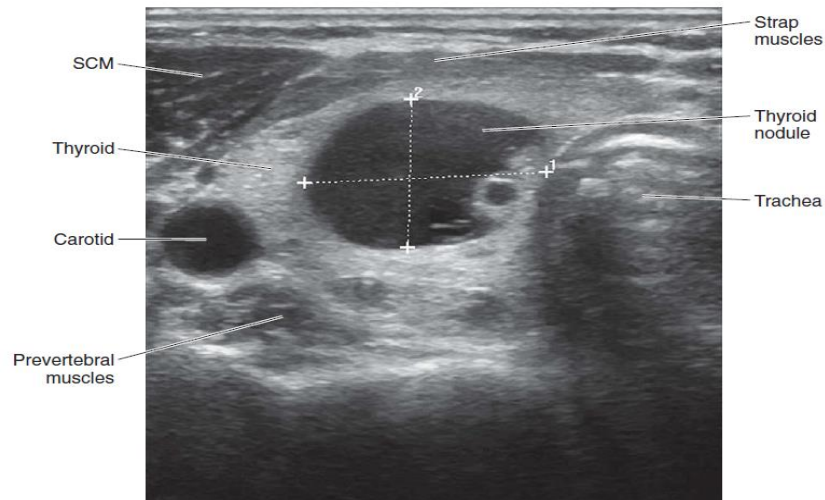
- Thyroid US:
 - **Imaging study of choice for ALL known or suspected thyroid nodules**
 - Advantages:
 1. Identify Thyroid size and anatomy
 2. Identify Thyroid nodules features.
 3. Gives accurate measurements for interval monitoring.
 4. Helps to select nodules for FNA biopsy.
 5. Evaluate the status of Neck LN.

- US features for suspicious Thyroid Nodule:
 1. Hypoechoic.
 2. Micro-calcifications.
 3. Irregular margins.
 4. Increased intranodular flow.
 5. Central vascularity.
 6. Absence halo.
 7. Nodule is taller than wide.
 8. Documented enlargement of a nodule.
- NO single sonographic feature or combinations of features is adequately sensitive or specific to identify all malignant nodules.
 - Microcalcifications are highly specific for PTC.

Table 1 Reported sensitivities and specificities of sonographic characteristics for detection of thyroid cancer		
	Median Sensitivity (%)	Median Specificity (%)
Microcalcifications	52	83
Absence of halo	66	54
Irregular margins	55	79
Hypoechoic	81	53
Increased intranodular flow	67	81

Data from Fish SA, Langer JE, Mandel SJ. Sonographic imaging of thyroid nodules and cervical lymph nodes. Endocrinol Metab Clin North Am 2008;37(2):401–17.

- US features for suspicious metastatic LN:
 1. Loss of fatty hilum
 2. Rounded shape.
 3. Hypoechogenicity
 4. Cystic change
 5. Calcifications,
 6. Peripheral vascularity.
- NO single sonographic feature is adequately sensitive for detection of lymph nodes with metastatic thyroid cancer.
 - Loss of fatty hilum is highly sensitivite for PTC.



○ **CT Neck (without contrast):**

- Not routinely done.
- CT with IV contrast (contains iodine) should be avoided.
 - Reduces subsequent uptake of iodine molecules and interfere with Nuclear scintigraphy (^{123}I) or postoperative Radioiodine Ablation Therapy (^{131}I) for malignant nodules for a period of 1 to 2 months.

- **Indications of CT in Thyroid:**

1. Huge Thyroid Mass.
2. Retrosternal Extension.
3. For completion thyroidectomy.
4. Enlarged Lymph Node.
5. Involvement of Adjacent Structures.

○ **Thyroid Scan:**

- **General Indications:**

1. Determine function of Thyroid Gland or Nodule.
 - Toxic nodule vs. Grave's in hyperthyroid patients.
2. Select Cold nodules which require FNA among multiple thyroid nodules.
3. Identify Ectopic thyroid tissue.
 - Retrosternal goiter.
 - Lingual thyroid.
 - Metastasis.

- Only indication for Thyroid scan in patient with thyroid nodule is when they have Hyper-thyroidism (Low TSH), aiming to evaluate the functional status of the nodule:

1. Hyper-functioning (Hot):

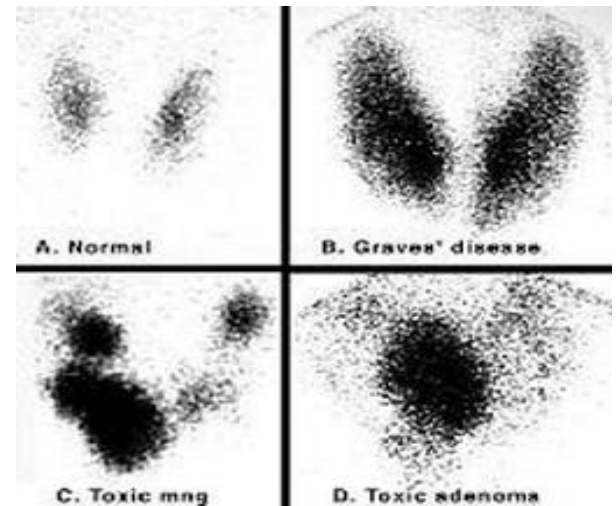
- Uptake is greater than surrounding thyroid.
- Risk of malignancy is 5%.

2. Iso-functioning (Warm):

- Uptake is equal to surrounding thyroid.
- Risk of malignancy is 5-10%.

3. Non-functioning (Cold):

- Uptake is less than surrounding thyroid.
- Risk of malignancy is up to 20%.



- Types of Thyroid scan:

▪ **131 I:**

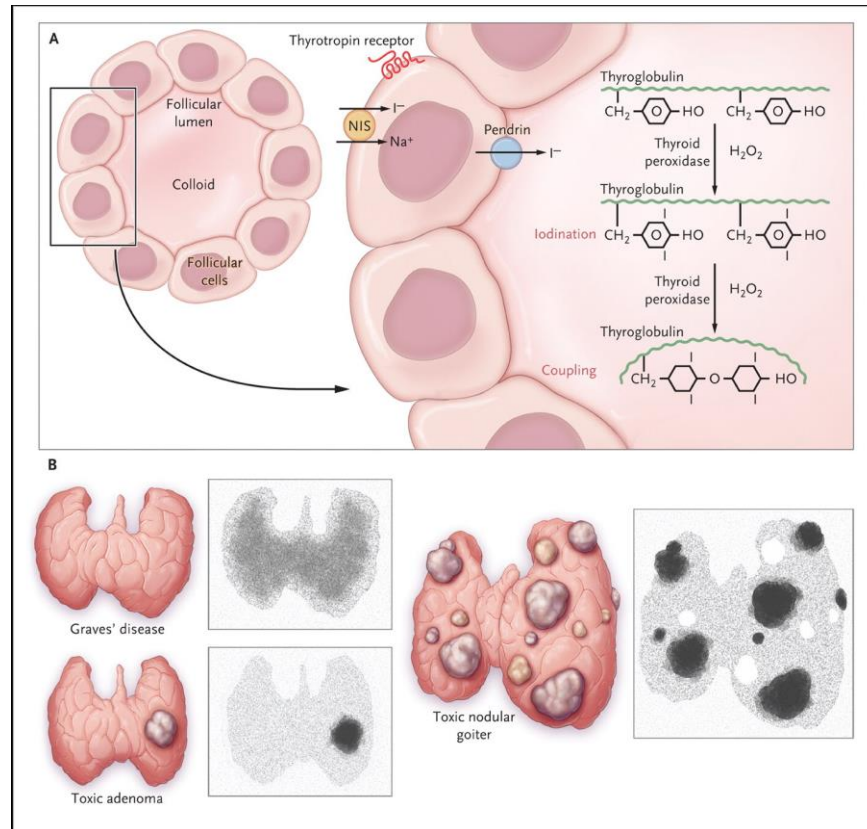
- High Radiation burden.
- Results available in 48–72 hours.
- Tracer of choice for:
 1. Evaluate Metastasis.
 2. Evaluate Residual disease.
 3. Ablation for hyperthyroidism.

▪ **123 I:**

- Expensive.
- Must be delivered daily.
- Testing requires 2 visits.
 - At 4 and 24 hours.
 - Shorter half-life.

▪ **99m Tc:**

- Trapped by follicular cells.
- Does not measure uptake.
- Low dose radiation.
- Less expensive.
- Image obtained in single visit within 30 minutes.



Fine Needle Aspiration (FNA) Biopsy:

- Single most important diagnostic tool.
- Most accurate and cost-effective method for evaluating thyroid nodules.
- Most reliable technique to differentiate benign from malignant.
- Requires adequate specimen and a well-trained pathologist
- 1–10% False-Negative results, depending on:
 - Size of needle.
 - Experience of pathologist and technician.
- Fine needle biopsies using 23 to 27 gauge (commonly 25 gauge) needles are preferred over large-bore needles.
 - Reduce risk of malignant seeding.
 - Allow for multiple biopsies.
- Larger needle biopsies may be considered for failed fine needle aspiration.

Statistical features of thyroid fine-needle aspiration

Statistical feature	Mean (%)	Range (%)
Sensitivity	83	65–98
Specificity	92	72–100
PPV	75	50–96
False-negative rate	5	1–11
False-positive rate	5	0–7

Abbreviation: PPV, positive predictive value.

- Indications for FNA in Patients with Thyroid nodule:

1. Thyroid Nodules >1cm:

- Great potential to be clinically significant cancers.

2. Thyroid Nodules <1cm with one of the following:

1. Suspicious US findings.
2. Positive LN.
3. History of H&N irradiation.
4. Family History of Thyroid Ca.

3. Suspicious LN:

- Measurement of Tg in the needle washout fluid enhances sensitivity of FNA of cervical nodes.
 - This FNA measurement of Tg is valid even in patients with circulating Tg Auto-antibodies.

- Indications for US-guided FNA biopsy:

1. Non-palpable nodules.
2. Predominantly cystic nodules (Directed biopsy of solid component)
3. Posteriorly located nodules.
4. Taking multiple biopsy from Large nodules (>4 cm) to reduce risk of a false negative results.

TABLE 3. SONOGRAPHIC AND CLINICAL FEATURES OF THYROID NODULES AND RECOMMENDATIONS FOR FNA

<i>Nodule sonographic or clinical features</i>	<i>Recommended nodule threshold size for FNA</i>	
High-risk history ^a		
Nodule WITH suspicious sonographic features ^b	>5 mm	Recommendation A
Nodule WITHOUT suspicious sonographic features ^b	>5 mm	Recommendation I
Abnormal cervical lymph nodes	All ^c	Recommendation A
Microcalcifications present in nodule	≥1 cm	Recommendation B
Solid nodule		
AND hypoechoic	>1 cm	Recommendation B
AND iso- or hyperechoic	≥1–1.5 cm	Recommendation C
Mixed cystic–solid nodule		
WITH any suspicious ultrasound features ^b	≥1.5–2.0 cm	Recommendation B
WITHOUT suspicious ultrasound features	≥2.0 cm	Recommendation C
Spongiform nodule	≥2.0 cm ^d	Recommendation C
Purely cystic nodule	FNA not indicated ^e	Recommendation E

^aHigh-risk history: History of thyroid cancer in one or more first degree relatives; history of external beam radiation as a child; exposure to ionizing radiation in childhood or adolescence; prior hemithyroidectomy with discovery of thyroid cancer, ¹⁸F-DG avidity on PET scanning; MEN2/FMTC-associated RET protooncogene mutation, calcitonin >100 pg/mL. MEN, multiple endocrine neoplasia; FMTC, familial medullary thyroid cancer.

^bSuspicious features: microcalcifications; hypoechoic; increased nodular vascularity; infiltrative margins; taller than wide on transverse view.

^cFNA cytology may be obtained from the abnormal lymph node in lieu of the thyroid nodule.

^dSonographic monitoring without biopsy may be an acceptable alternative (see text) (48).

^eUnless indicated as therapeutic modality (see text).

- Patients with MNG have the same risk of malignancy as those with solitary nodules.
 - FNA in patients with MNG should be done for:
 1. Nodules with suspicious US features.
 2. Largest nodules in size if no suspicious US features.
 3. Warm (iso-functioning) and Cold (non-functioning) nodules found in Thyroid scan done for patients with MNG and Hyperthyroidism (Low TSH).
- Indications to repeat FNA in Thyroid nodule:
 1. Initial FNA non-diagnostic.
 2. Enlarging FNA-benign nodule.
 - >50% increase in volume, or
 - 20% increase in at least two nodule dimensions with a minimal increase of 2mm in solid nodules or in the solid portion of mixed cystic–solid nodules.
 3. Recurrent cysts (vs. Hemi-thyroidectomy).
 4. Large (>4 cm) nodule (vs. Hemi-thyroidectomy).

Interpretation of FNA Results (Bethesda System):

1. Bethesda I (Non-Diagnostic or Unsatisfactory):

- Risk of Malignancy is 5%
- Cytology:
 - Cyst fluid only.
 - Virtually acellular specimen.
 - Other (obscuring blood, clotting artifact, etc).
- Management:
 - Repeat FNA under US guidance.
 - If repeated US-guided FNA is non-diagnostic:
 - US-guided core needle biopsy, or
 - Clinical follow up 6-12 months with US for small partially cystic nodules.
 - Hemi-thyroidectomy for large solid nodules with sonographically suspicious features.

2. Bethesda II (**Benign**):

- **Risk of Malignancy is 3%**
- Cytology:
 - Consistent with a benign follicular nodule (includes adenomatoid nodule, colloid nodule, etc).
 - Consistent with lymphocytic (Hashimoto) thyroiditis in the proper clinical context.
 - Consistent with granulomatous (subacute) thyroiditis.
 - Others.
- Management:
 - Clinical follow up 6-12 months with US.
 - Small changes in nodule size on serial US do not require a repeated FNA.
 - Routine suppression therapy of benign thyroid nodules in iodine sufficient populations is NOT recommended.

3. Bethesda III (**AUS or FLUS**):

- **Risk of Malignancy is 15%.**
- Cytology:
 - Atypia of Undetermined Significance (AUS).
 - Follicular Lesion of Undetermined Significance (FLUS).
- Management:
 - Repeat FNA under US guidance.
 - If repeated results came AUS or FLUS, do Diagnostic hemithyroidectomy.

4. Bethesda IV (**Follicular Neoplasm or Suspicious for Follicular Neoplasm**):

- **Risk of Malignancy is 30%.**
- Cytology:
 - Follicular Neoplasm/Suspicious for a Follicular Neoplasm
 - Hürthle cell (oncocytic) type.
- Management:
 - Diagnostic Hemi-thyroidectomy:
 - For Low Risk patients:
 - Age <45 years.
 - Isolated thyroid nodule.
 - No LN involvement.
 - No family History of Thyroid Ca.
 - No history of radiation.
 - Total Thyroidectomy for high risk patients.

5. Bethesda V (**Suspicious for Malignancy**):

- Risk of Malignancy is 75%.
- Cytology:
 - Suspicious for papillary carcinoma.
 - Suspicious for medullary carcinoma.
 - Suspicious for metastatic carcinoma.
 - Suspicious for lymphoma.
 - Others.
- Management:
 - Total Thyroidectomy.

6. Bethesda VI (**Malignant**):

- Risk of Malignancy is 99%.
- Cytology:
 - Papillary thyroid carcinoma.
 - Poorly differentiated carcinoma.
 - Medullary thyroid carcinoma.
 - Undifferentiated (anaplastic) carcinoma.
 - Squamous cell carcinoma.
 - Carcinoma with mixed features.
 - Metastatic carcinoma.
 - Non-Hodgkin lymphoma.
 - Other.
- Management:
 - Total Thyroidectomy.

TABLE 133.1 THE BETHESDA SYSTEM FOR REPORTING THYROID CYTOPATHOLOGY

Diagnostic Category	Risk of Malignancy (%)
Nondiagnostic or unsatisfactory	1–4
Benign	0–3
Atypia of undetermined significance or FLUS	5–15
Follicular neoplasm or suspicious for a follicular neoplasm	15–30
Suspicious for malignancy	60–75
Malignancy	97–99

Modified from Cibas ES, Ali SZ. The Bethesda system for reporting thyroid cytopathology. *Am J Clin Pathol* 2009;132:658–665.

- **Indications of Surgery in Thyroid nodules:**

- Benign Pathology:
 1. Suspicious US features.
 2. Hyperthyroidism.
 3. Compressive symptoms.
 4. Cosmetic reasons.
 5. Large Thyroid nodule >4cm
 6. Recurrent cyst.
- Suspicious or malignant thyroid Ca.

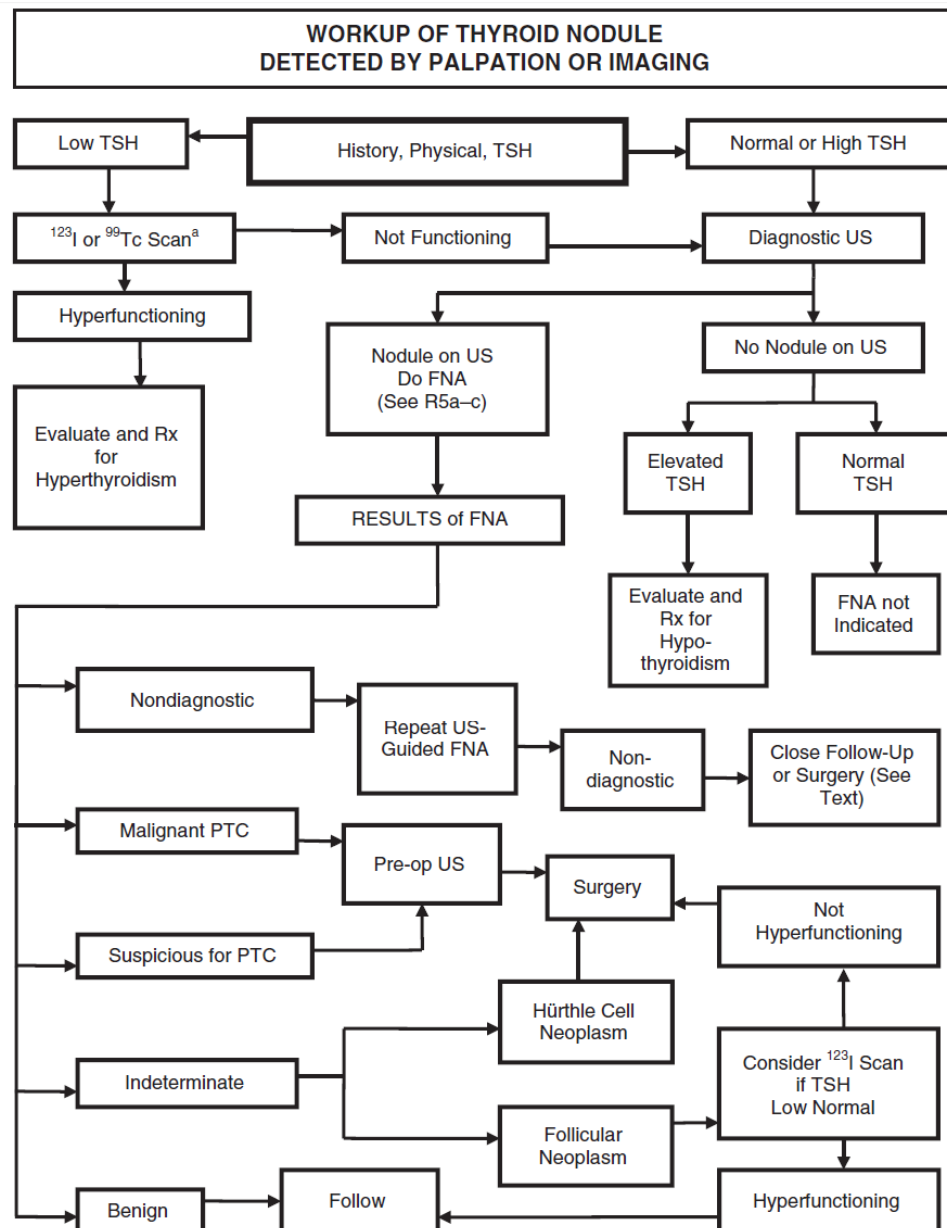
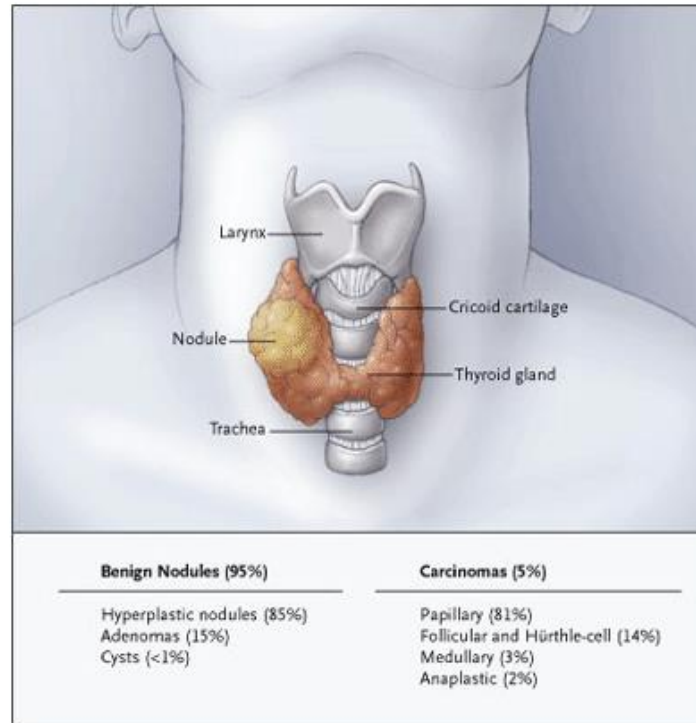


FIG. 1. Algorithm for the evaluation of patients with one or more thyroid nodules.

^aIf the scan does not show uniform distribution of tracer activity, ultrasound may be considered to assess for the presence of a cystic component.

Benign Thyroid Diseases:

- Overall, Women are affected more than men (5:1).
- 5% of population have Palpable Thyroid Nodules.
- Only 5% of these palpable thyroid nodules are Malignant.



- **Thyroid Cysts:**
- 20-50% of all Thyroid Nodules are Cystic.
- Majority of cystic thyroid nodules are Benign degenerating thyroid adenomas.
- Purely cystic lesions rarely contain cancer.
- Risk of cancer in complex (cystic and solid) nodules approaches that of solid nodules (5-10%).
 - o Risk of CA decreases as the cystic component increases.
 - o Papillary thyroid cancer is most common Cancer associated with Thyroid cyst.
- Clinical Picture:
 - o Asymptomatic:
 - Incidental finding.
 - Euthyroidism.
 - Most cystic thyroid nodules are Nonfunctioning.
 - o Midline neck mass:
 - Smooth and Round Thyroid mass.
 - Compressive symptoms if Large.
 - o Thyroid Pain:
 - **Thyroid cysts are Most common cause of thyroid pain.**
 - Maybe due to Sudden hemorrhage.
- Diagnosis:
 - 1. Serum TSH:**
 - o Typically normal.
 - o Low TSH:
 - Rarely, hemorrhage into thyroid nodule can result in transient Thyrotoxicosis.
 - Some cystic nodules are hyperfunctioning thyroid adenomas that have undergone cystic degeneration.
 - 2. Thyroid Ultrasound:**
 - Identify predominantly cystic nodules.
 - Identify suspicious features.
 - 3. Fine Needle Aspiration (FNA):**
 - US-guided FNA is preferred in Predominantly cystic nodules.
 - Indications:
 - 1.** ≥ 1.5 cm Mixed cystic-solid nodule WITH any suspicious ultrasound features.
 - 2.** ≥ 2.0 cm Mixed cystic-solid nodule WITHOUT suspicious ultrasound features.
 - FNA is NOT indicated for Purely cystic nodule.
 - Diagnostic biopsy usually requires tissue obtained from the solid component of the nodule, but occasionally, malignant cells are present in the cyst fluid.

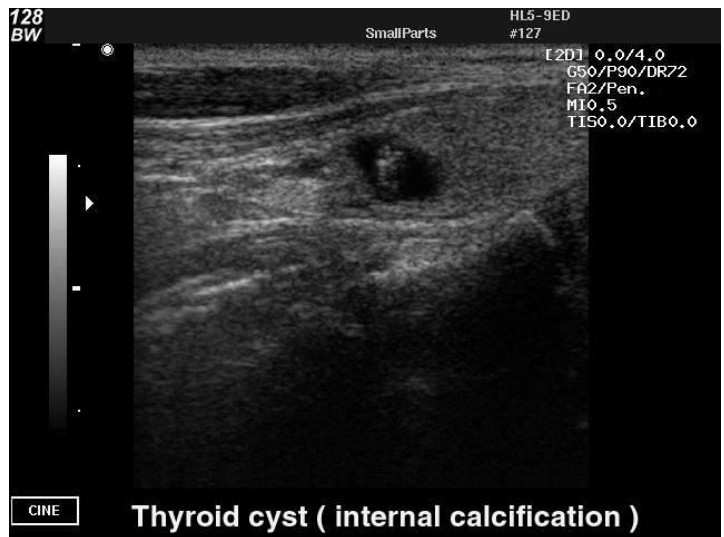
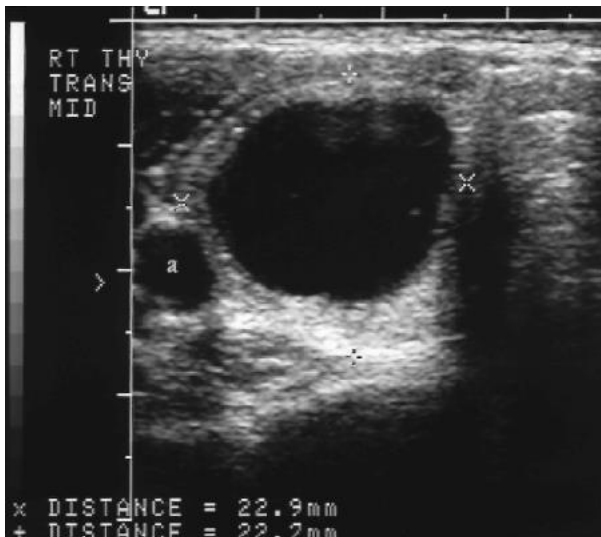
- Color of aspirate does Not help in Diagnosis, **EXCEPT**:
 - Bloody Aspirate → Cancer.
 - Clear Fluid → Parathyroid tumor.

4. CT Neck:

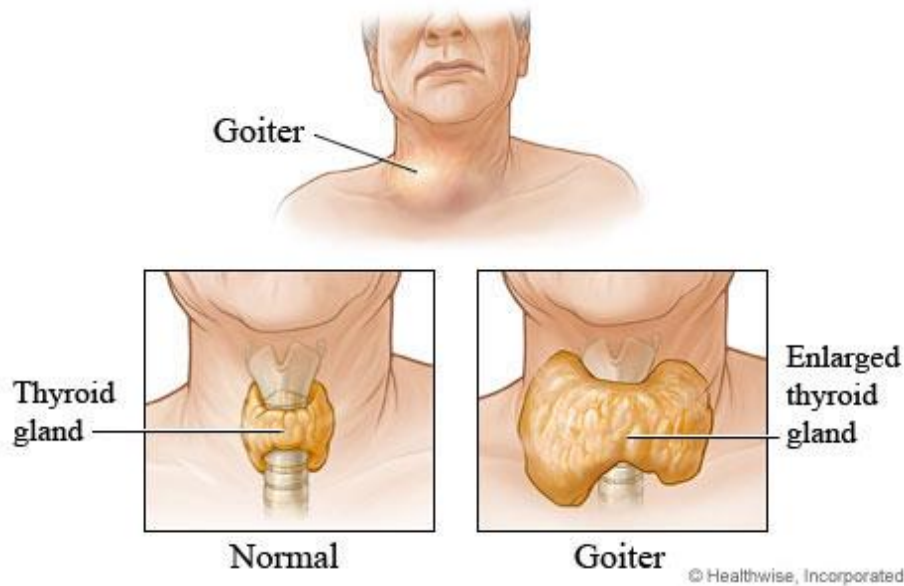
- Not routinely done.

- Management:

1. Observe for Regression.
2. FNA relieves pain and help in diagnosis.
 - If non-diagnostic FNA:
 - Repeat FNA with US-guidance.
3. Surgery:
 - Lobectomy:
 1. Cysts Recalcitrant to drainage.
 2. Cysts > 4 cm.
 3. Cysts Suspicious for CA (Papillary):
 - Bloody Aspirates.
 - Recurrent cysts.
 - Total Thyroidectomy:
 - Malignant lesion.



- **Diffuse Colloid Goiter (Multinodular Goiter)**
- Diffuse or nodular enlargement of Thyroid gland.
- Not results from an inflammatory or neoplastic process.
- Not associated with abnormal thyroid function.
- More common in women.
- Causes:
 - Iodine Deficiency:
 - Most common cause of goiter.
 - Iodine Excess:
 - Rare.
 - Goitrogens:
 - Drugs:
 - Propylthiouracil.
 - Lithium.
 - Phenylbutazone.
 - Aminogluthethimide.
 - Iodine-containing expectorants.
 - Foods:
 - Cabbage, turnips, Brussels sprouts, rutabagas
- Pathophysiology:
 - Iodine deficiency.
 - → TSH Hypersecretion.
 - → Stimulates Thyroid hyperplasia and involution.
 - → Multinodularity.
- Clinical Picture:
 - Neck Mass.
 - Euthyroidism.
 - Compressive symptoms.
 - Intrathoracic extension is the path of least resistance.
- Diagnosis:
 1. Serum TSH and Free T4 level
 2. US Thyroid
 3. FNA
- Management:
 - Observation:
 - No rapid growth.
 - No obstructive symptoms.
 - No thyrotoxicosis.
 - Surgery:
 - Cosmetic.
 - Compressive symptoms.
 - Thyrotoxicosis.



- **Toxic Multinodular Goiter (Plummer's Disease):**
- Variation of diffuse colloid goiter in which multiple nodules are hyperfunctional resulting in hyperthyroidism.
- 2nd most common cause of Hyperthyroidism after Graves.
- Management:
 1. Iodine Replacement:
 - Reverses goiter.
 2. Hormonal suppression:
 - Requires careful monitoring with serum TSH to avoid hyperthyroid, arrhythmias and osteoporosis.
 3. Radioactive iodine therapy.
 4. Surgical excision (Subtotal thyroidectomy):
 - Cosmesis.
 - Decompression.
 - Concern of Malignancy.
 - Toxicosis.

- **Most Common Causes of Hyperthyroidism:**

1. Graves' Disease.
2. Toxic Multinodular Goiter.
3. Subacute Thyroiditis.
4. Exogenous from Medications, Iodine induced.
5. Toxic adenoma.
6. Thyroid Cancer.
7. Pituitary tumors.

- **Graves' Disease (Diffuse Toxic Goiter):**

- Autoimmune condition.
- Thyroid-Stimulating Immunoglobulins (TSI) stimulate growth of Thyroid gland via TSH Receptors.
- Most common cause of hyperthyroidism.
- Graves is Triad of:
 1. Goiter
 2. Exophthalmos
 3. Pretibial myxedema.
- Risk factors:
 - o Women (adolescence or 30-40)
 - o Radiation exposure
 - o Genetic disposition
- Histopathology:
 - o Hyperplasia, Increased colloid material, papillary projections.
- Clinical Picture:
 - o Diffuse goiter.
 - o Hyperthyroidism.
 - o Dermopathy.
 - o Exophthalmos
 - Autoimmune → Extraocular muscle deposition.
 - o Blindness from optic neuropathy.
 - o Pre-tibial myxedema.
 - o Acropathy
 - Clubbing from osteoarthropathy.
 - o Compressive symptoms.



- Complications:
 - Secondary effects of hyperthyroidism.
 - Heart failure.
 - Arrhythmia
 - thyroid storm.
 - Vision loss
- Diagnosis:
 - Low TSH.
 - High free T4.
 - Thyroid-Stimulating Immunoglobulins (TSI).
 - High RAIU.
 - High Thyroglobulins.
 - Diffuse uptake in Thyroid radioactive iodine scan.
- **Treatment:**
- Medical Management:
 1. Propylthiouracil and Methimazole:
 - Considered for Small goiter and mild disease
 - Propylthiouracil:
 - Inhibits peripheral T4 to T3 conversion.
 - Methimazole, carbimazole:
 - Inhibits MIT & DIT coupling, inhibits iodine oxidation.
 2. Iodide:
 - Inhibits organification & thyroid hormone release.
 - Decreases Thyroid vascularity.
 - Most useful in thyroid storm (10-14 days pre-Op).
 - **Wolf-Chaikoff Effect:**
 - Transient Anti-thyroid effect of iodides (Autoregulatory phenomenon).
 - **Jod-Basedow Phenomenon:**
 - Development of overt hyperthyroidism in subclinical patients (Abnormal thyroid) due to exogenous iodide administration.
 - Does not occur in persons with normal thyroid glands who ingest extra iodine in any form.
 3. Propanolol (B-blocker):
 - Supplemented for severe symptoms (24-48 hrs pre op).

- Radioactive Iodine (^{131}I):

- Indicated for:
 1. Severe symptoms.
 2. Failed medical therapy.
- Side effects:
 - Hypothyroidism.
 - Thyroiditis.
 - Bladder CA.
 - Breast CA
 - AML
 - Chromosomal abnormalities.
- Contraindicated in pregnancy and Pediatrics.

- Surgical Management:

- Subtotal Thyroidectomy:
- Indicated for:
 1. Failed Medical therapy.
 2. Pregnancy (especially in 2nd Trimester).
 3. Noncompliance with medications.
 4. Suspicious non-functioning (Cold) Nodule.
 5. Compressive symptoms.
 6. Cosmetic.
 7. Desire for rapid control of toxic process.

- Management for Exophthalmos and Optic Neuropathy:

- Ophthalmology evaluation.
- Eye care:
 - Artificial tears.
 - Taping retracted lids.
 - Protective eyewear.
- If optic Neuropathy persists despite medical therapy,
 - → Trial of corticosteroids is used for 2 weeks,
 - If no improvement → Surgical decompression
- If Exophthalmos persists after 6 months despite adequate therapy,
 - Radiation therapy or Surgical correction.
 - Orbital decompression
 - Eyelid retraction release
 - Strabismus surgery.

- **Thyroid Storm:**
- Acute extreme state of thyrotoxicosis.
- Life-threatening.
- 20-50% mortality
- Diagnosed by clinical history and exam
- Etiology:
 1. Surgery (Thyroidectomy).
 2. Trauma.
 3. Childbirth.
 4. Infection.
 5. Untreated hyperthyroid.
 6. Radioactive iodine Rx.
 7. Diabetic ketoacidosis
 8. Vigorous palpation of thyroid gland.
- Clinical picture:
 - Fever.
 - Profuse sweating.
 - Tachycardia.
 - Nausea.
 - Abdominal pain.
 - Tremors.
 - Restless.
 - Psychosis.
 - Coma.
 - Stupor
- Management:
 1. ICU Admission for Cardiac monitoring.
 2. Immediate administration of:
 - Inorganic iodine.
 - Propylthiouracil.
 - Propranolol.
 - Corticosteroids
 3. Supportive measures:
 - Glucose-containing IV fluids.
 - Cooling blanket.
 - Supplemental oxygen.
 - Antipyretics.

- **Most Common Causes of Hypothyroidism:**

1. Radiation Therapy
2. Chronic Thyroiditis
3. Idiopathic Atrophy
4. Hashimoto's Thyroiditis
5. Surgical Removal.
6. Radioiodine Ablation Therapies
7. Secondary Hypothyroidism.
8. Iodine Deficiency
9. Congenital Hypothyroidism.

Risk factors for hypothyroidism

- I) Old age
- II) Female sex
- III) Grave's disease
- IV) Hashimoto's disease
- V) Other autoimmune disease
- VI) Drugs: lithium iodide, amiodarone, iodide containing drugs
- VII) Postthyroidectomy
- VIII) Euthyroid goiter
- IX) Prior head & neck radiotherapy management
- X) Laryngectomy with/without irradiation

Head & neck signs & symptoms of hypothyroidism

- I) Hearing loss: sensorineural, mixed, conductive
- II) Vertigo, tinnitus
- III) Enlarged tongue
- IV) Hoarseness (mucopolysaccharide infiltration)
- V) Blurred vision

- **Causes of Thyroiditis:**

1. Hashimoto's Thyroiditis (Chronic Autoimmune Thyroiditis)
2. Subacute Thyroiditis (De Quervain's)
3. Painless Thyroiditis.
4. Riedel's Thyroiditis.
5. Acute Suppurative Thyroiditis

TABLE 4–2. Clinical Differences Between Thyroiditis Conditions

Condition	Subacute	Hashimoto's	Riedel's	Suppurative
Incidence	common	common	rare	rare
Thyroid hormone status	hyper- then hypothyroid	hyper- then hypothyroid	hypothyroid	—
Onset	acute	gradual	gradual	rapid
Pain	common	none	rare	common
Goiter	rare	common	hard gland	rare

- **Hashimoto's Thyroiditis (Chronic Autoimmune Thyroiditis)**
- **Most common cause of Hypothyroid in the US**
- Associated with:
 - Lymphoma.
 - Neoplasms.
 - Other autoimmune disease (SLE, sjogren syndrome, scleroderma).
- Pathophysiology:
 - **Antibody to Thyroid Peroxidase.**
 - Anti-thyroglobulin Antibody.
 - Anti-microsomal Antibody.
 - Anti-TSH receptor
 - → Transient Hyperthyroid then Hypothyroidism.
- Risk factors:
 - Women
 - Genetic susceptibility (HLA-DR3).
 - Sjögren's.
 - DM,
 - Pernicious anemia
- Histopathology:
 - Fibrosis
 - Lymphocytic infiltration
- Clinical picture:
 - Slowly enlarging goiter.
 - Painless.
 - Hypothyroidism 20%.
- Diagnosis:
 - Antibody to Thyroid Peroxidase (90%).
 - Anti-thyroglobulin Antibody (20-50%).
 - Anti-microsomal Antibody.
 - ESR.
 - TFT (maybe elevated, normal, or low of T4 and TSH).
 - FNA:
 - Only for prominent nodules (>1cm) suspicious of carcinoma or lymphoma that do not resolve with medical therapy.
- Treatment:
 - Long term Thyroxine therapy with TFT monitoring.
 - Re-evaluate TSH after 4-6 wks "long half life".
 - Surgical excision for:
 - Compressive symptoms.
 - Suspicious nonfunctioning (cold) nodule.
 - Progressive disease.
 - Pregnancy.

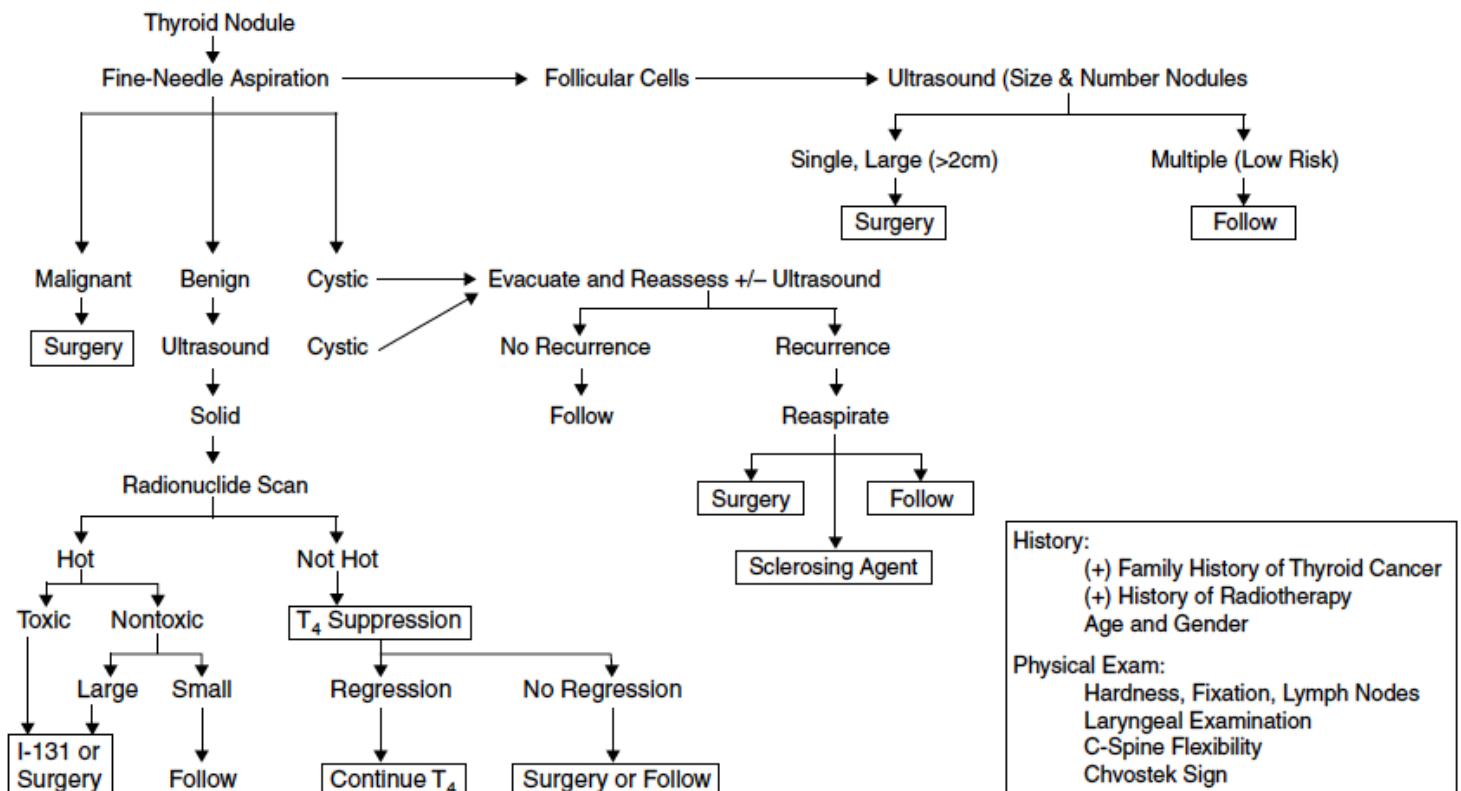
- **Subacute Thyroiditis (De Quervain's)**
- **Most common cause of Painful thyroid.**
- Pathophysiology:
 - o Viral (Mumps, Coxsackivirus) or Postviral.
 - o → Decreases iodine uptake (unlike Grave's).
- Clinical picture:
 - o Painful Enlarged thyroid.
 - o Self-limiting.
 - o Malaise.
 - o Associated URTI prior to thyroid tenderness.
 - o Transient hypothyroid 50%.
 - o Permanent hypothyroid 5%.
- Management:
 - o Symptomatic therapy (NSAIDs, steroids).
 - o Observation

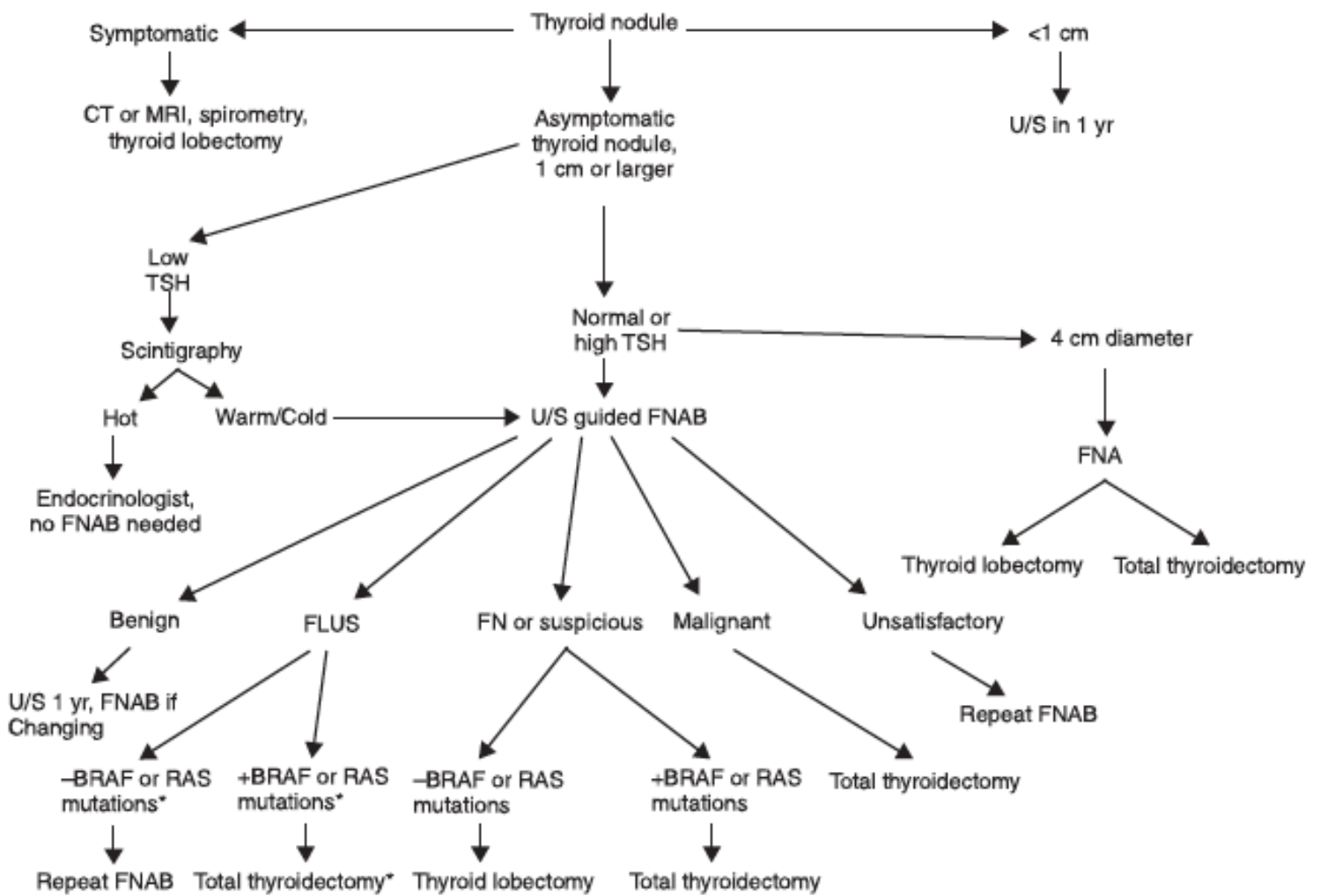
- **Painless Thyroiditis:**
- Similar to Subacute Thyroiditis but Painless.
- Common in women postpartum.

- **Riedel's Thyroiditis:**
- Very rare.
- More in Women (W:M=3.5:1).
- 30-60 years.
- Unknown etiology.
- Clinical picture:
 - o Painless.
 - o Rock hard due to Thyroid fibrosis.
 - o Euthyroid.
 - o Compressive symptoms.
 - o Associated with other syndromes of focal sclerosis (Retroorbital fibrosis, sclerosing cholangitis).
- Management:
 - o Medical Management
 - Steroids
 - Tamoxifen
 - Methotrexate
 - Raloxifene
 - o Hormone replacement.
 - o Surgical release at isthmus (Biopsy to rule-out Ca)

- **Acute Suppurative Thyroiditis:**
- Uncommon.
- Management with Systemic antibiotics, consider drainage for abscess formation.

- **Thyroid Adenomas:**
- Well-circumscribed.
- Follicle-derived.
- Different than surrounding thyroid parenchyma.
- No vascular or capsular invasion.
- Types:
 - **Follicular Adenoma:**
 - Most common Benign Thyroid Neoplasm.
 - Can be Trabecular, Follicular, Microfollicular, Solid.
 - **Hurthle cell Adenoma (Oncocytic):**
 - Homogeneous and brown when cut.
 - Large eosinophilic thyroid cells that contain a large number of **mitochondria** with a high content of oxidative enzymes.
 - **Hyalinizing trabecular:**
 - Very little follicle formation.
 - Nuclear features similar to papillary Ca.
 - **Nodular (Adenomatous):**
 - Diffuse.
 - Red/brown nodularity.
 - Coalescent nodules with dense colloid.





- **Thyroglossal Duct Cyst (TGDC):**
- Most common congenital cervical anomaly.
- Failure of complete obliteration of thyroglossal duct (created from tract of the thyroid descent from the foramen cecum "tuberculum impar" down to midline neck).
- Clinical picture:
 - Midline neck mass with cystic and solid components.
 - Elevates with tongue protrusion (attached to hyoid bone).
 - Typically inferior to hyoid bone and superior to thyroid gland.
 - Dysphagia.
 - Globus sensation
 - 20% slightly off to the left
- US is done to determine if the TGDC is pts only functional tissue
- Histopathology: lined with respiratory (columnar) and squamous epithelium.
- Complications:
 - Rare malignant potential (1%) (CT-solid tissue elements and calcifications).
 - Secondary infection
- Management:
 - Sistrunk procedure:
 - Excision of cyst and tract with cuff of tongue base and mid-portion of hyoid bone.
 - 3-4% recurrence.
 - If No hyoid bone excision → 50% recurrence.
 - Higher recurrence with young age and intra-op rupture.
- **TGDC carcinoma:**
 - Most are papillary (also mixed papillary follicular, SCC, Hurthle cell).
 - No medullary (no C cells in midline tissue).
 - 33% have concurrent intrathyroidal malignant mass → US must be done on TGDCC.
 - Sistrunk procedure is adequate if TGDCC is small, non-invasive and there is no evidence of thyroid ca or mets

Malignant Thyroid Tumors:

- 3-7% of population have Palpable Thyroid Nodules.
- Only 5-10% of these palpable thyroid nodules are Malignant.
- High Risk Thyroid Nodule for Malignancy:
 - History:
 1. Male
 2. Age <20 years or >60 years.
 3. Rapid Progress.
 4. Pressure Symptoms (Dysphagia)
 5. Hoarseness
 6. Family history of Thyroid Ca.
 7. History of Radiation Exposure.
 - Physical Exam:
 1. Fixation.
 2. Nodule > 4cm (to be investigated if 1>cm).
 3. Vocal cord paralysis.
 4. Presence of Cervical LN.
 - U/S:
 1. Hypoechogenic Nodule.
 2. Microcalcification.
 3. Irregular margins.
 4. Absent of Halo sign.
 5. Increase vascularity.
 6. Shape taller than width.
 7. Presence of LN.
- Thyroid follicular epithelial-derived Ca can be classified into:
 - **Differentiated Thyroid Ca:**
 1. Papillary (85%)
 2. Follicular (10%)
 3. Hurthle (3%).
 - **Undifferentiated Thyroid Ca:**
 1. Anaplastic (1-5%)
- In all Thyroid CA, Women>Men, **Except:**
 - Medullary thyroid Ca Men=Women.
- High Risk Criteria for Thyroid CA (AMES):
 - **A**ge:
 - Males ≥41 Years.
 - Females ≥51 Years.
 - **M**etastasis:
 - Presence of Metastasis suggests Malignancy.
 - **E**xtent:
 - Extrathyroidal.
 - Major capsular involvement
 - **S**ize:
 - Nodule ≥ 5 cm

- Others:
 - Radiation therapy.
 - Autoimmune Thyroiditis.
 - More common in **women**.
 - Children and Males have a higher risk of malignancy if presents with a thyroid nodule.
- Indication for CT scan in Thyroid:
 1. Huge mass.
 2. Retrosternal extension.
 3. Positive LN.
 4. Involvement of adjacent structures.
 5. For completion thyroidectomy
- **Staging of Thyroid CA:**
- **Primary Tumor (T):**
 - **Tx:** Primary tumor cannot be Assessed.
 - **T1:** Tumor Size $\leq 2\text{cm}$, Limited to Thyroid only.
 - **T2:** Tumor Size $> 2\text{cm}$ to 4cm , Limited to Thyroid only.
 - **T3:** Tumor Size $> 4\text{cm}$, Limited to thyroid only
OR
Tumor of Any size with Minimal Extra-thyroid Extension (e.g., extension to SCM or Perithyroid soft tissues).
 - **T4a:** Tumor of Any size extends beyond Thyroid capsule to invade Subcutaneous soft tissues, Larynx, Trachea, Esophagus, or RLN.
 - **T4b:** Tumor invades Prevertebral Fascia or encases Carotid Artery or Mediastinal vessels.
 - All Anaplastic carcinomas are considered T4 tumors
- **Lymph Nodes (N):**
 - **Nx:** Lymph nodes cannot be Assessed.
 - **N0:** No lymph node metastasis.
 - **N1a:** Metastasis to Level VI (pretracheal, paratracheal, and prelaryngeal/Delphian lymph nodes).
 - **N1b:** Metastasis to Unilateral, Bilateral, or Contralateral Cervical or Superior Mediastinal lymph nodes.
- **Distant Metastasis (M)**
 - **Mx:** Distant metastasis cannot be assessed.
 - **M0:** No distant metastasis.
 - **M1:** Distant metastasis.
- **STAGING if Age < 45 years:**
 - **Stage 1:** Any T + Any N + **M0**
 - **Stage 2:** Any T + Any N + **M1**

TABLE 4. TNM CLASSIFICATION SYSTEM FOR DIFFERENTIATED THYROID CARCINOMA

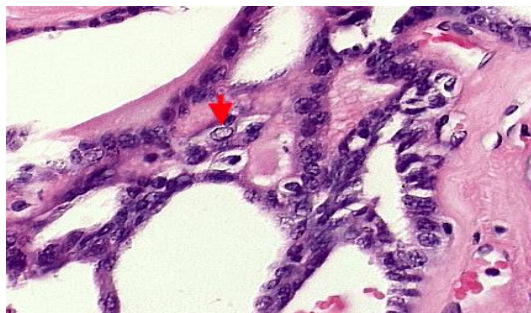
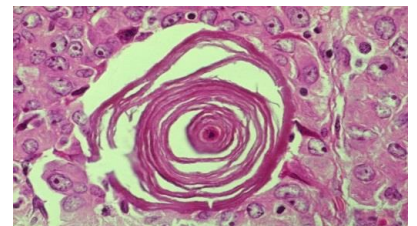
		Definition
T1	Tumor diameter 2 cm or smaller	
T2	Primary tumor diameter >2 to 4 cm	
T3	Primary tumor diameter >4 cm limited to the thyroid or with minimal extrathyroidal extension	
T4 _a	Tumor of any size extending beyond the thyroid capsule to invade subcutaneous soft tissues, larynx, trachea, esophagus, or recurrent laryngeal nerve	
T4 _b	Tumor invades prevertebral fascia or encases carotid artery or mediastinal vessels	
TX	Primary tumor size unknown, but without extrathyroidal invasion	
N0	No metastatic nodes	
N1 _a	Metastases to level VI (pretracheal, paratracheal, and prelaryngeal/Delphian lymph nodes)	
N1 _b	Metastasis to unilateral, bilateral, contralateral cervical or superior mediastinal nodes	
NX	Nodes not assessed at surgery	
M0	No distant metastases	
M1	Distant metastases	
MX	Distant metastases not assessed	
Stages		
	Patient age <45 years	Patient age 45 years or older
Stage I	Any T, any N, M0	T1, N0, M0
Stage II	Any T, any N, M1	T2, N0, M0
Stage III		T3, N0, M0
		T1, N1 _a , M0
		T2, N1 _a , M0
		T3, N1 _a , M0
Stage IVA		T4 _a , N0, M0
		T4 _a , N1 _a , M0
		T1, N1 _b , M0
		T2, N1 _b , M0
		T3, N1 _b , M0
		T4 _a , N1 _b , M0
Stage IVB		T4 _b , Any N, M0
Stage IVC		Any T, Any N, M1

- **STAGING if Age $\geq$ 45 years:**

- **Stage 1:** T1 + N0 + M0
- **Stage 2:** T2 + N0 + M0
- **Stage 3:**
 - T3 + N0 + M0
 - T1 + N1_a + M0
 - T2 + N1_a + M0
 - T3 + N1_a + M0
- **Stage 4A:**
 - T4_a + N0 + M0
 - T4_a + N1_a + M0
 - T4_a + N1_b + M0
 - T1 + N1_b + M0
 - T2 + N1_b + M0
 - T3 + N1_b + M0
- **Stage 4B:** T4_b + Any N + M0
- **Stage 4C:** Any T + Any N + M1

- **Papillary Thyroid Carcinoma (PTC):**
- Most common Thyroid Ca.
 - o Papillary = Popular.
 - o 80% of all Thyroid Ca.
- More common in Young females (F:M 3:1).
- Peak incidence in 3rd/4th decades.
- Slow-growing tumor that arises from Thyroxine and Thyroglobulin-producing Follicular cells of thyroid:
 - o TSH sensitive.
 - o Take up iodine.
 - o Produce Thyroglobulin in response to TSH stimulation.
- 70% of well differentiated thyroid cancers have mutations of ret/PTC, NTRK1, RAS, or **BRAF**.
 - o Patients with the BRAF mutation are more likely to have Extrathyroidal invasion, Lymph node metastases, Advanced stage at diagnosis, and Tumor recurrence.
- Papillary Thyroid Microcarcinoma (PTMC) is papillary carcinoma measuring ≤ 1 cm.
- **Variants of PTC:**
 - o Follicular variant:
 - Most common variant (10% of all PTC).
 - Characterized by presence of follicles, typical of follicular tumors, in addition to typical features of common-type PTC.
 - Behaves more papillary than Follicular Ca.
 - o Tall-cell variant:
 - More aggressive than common-type PTC.
 - 1% of all PTC.
 - Characterized by tumor cells with eosinophilic cytoplasm that are twice as tall as they are wide.
 - Tend to be large, invasive with local and distant metastases at time of diagnosis.
 - 5-year mortality rate is higher than common-type PTC.
- **Risk Factors to develop PTC:**
 - o **Radiation Exposure during Childhood:**
 - **Most important risk factor for differentiated thyroid cancer.**
 - 4-10% of PTC have History of prior Neck irradiation.
 - o **Family History of PTC:**
 - 10% of PTC have family History.
 - o **Gardner's Syndrome:**
 - Familial Adenomatous Polyposis.
 - Multiple polyps in Colon together with tumors outside colon include:
 - o Osteomas of Skull.
 - o Papillary Thyroid Cancer.
 - o Epidermoid cysts.
 - o Fibromas & Sebaceous cysts.

- Clinical Picture:
- Often Asymptomatic
 - o Incidental finding of non-palpable thyroid nodule.
- Painless, slow-growing solitary thyroid nodule.
- Symptoms of compression or invasion:
 - o Hoarseness.
 - o Dysphagea
 - o SOB
 - o Hemoptysis.
- Routes of PTC spread:
 - 1. Direct invasion:**
 - Through Thyroid capsule to invade surrounding structures.
 - 2. Lymphatic spread:**
 - Most common LN involvement is:
 1. Level 6 (Central compartment).
 2. Levels 2 to 5.
 - 15-30% of PTC have Palpable regional LN.
 - 70% of PTC have occult LN.
 - 3. Hematogenous spread:**
 - Rare.
 - Lung is most common site of distant mets.
 - Bone is 2nd most common.
- Diagnosis:
 - 1. US Thyroid and Neck:**
 - Evaluate the Thyroid and Presence of positive LN.
 - 2. FNA:**
 - For Suspicious Thyroid nodule and LN.
- Histopathology of PTC (None are Pathognomonic):
 - 1. Multifocal Tumor:**
 - 80% of PTC are bilateral.
 - 2. Psammoma bodies:**
 - Small laminated calcifications.
 - Found in 50% of PTC.
 - 3. Ground Glass Appearance/ Orphan Annie Eyes:**
 - Large nuclei cleared of chromatin.



- Prognosis of PTC :
- Good prognosis with 95% 5-year survival rate.
- **Poor Prognostic indicators:**
 - o Tumor > 1.5 cm.
 - o Extracapsular spread.
- Cervical mets have increased Cervical Recurrence rates **without** affecting Survival rate.

- **Follicular Thyroid Carcinoma:**
- 2nd Most common Thyroid Ca.
 - o 10-15% of all Thyroid Ca.
- More common in Elderly Female (40-60 Years).
- F:M ratio is 3:1.
- Like PTC, Follicular Ca arise from Follicular cells of Thyroid.
 - o TSH sensitive.
 - o Take up iodine.
 - o Produce Thyroglobulin in response to TSH stimulation.

- Follicular Ca is common with patients having elevated TSH in goiter endemic areas due to low iodine intake (TSH dependant).
- **RAS** mutations seen in approximately 40% of Follicular Ca.
 - o Associated with more aggressive cancers and higher mortality.
 - o Not specific to Follicular thyroid cancer.
 - o Seen in Follicular-variant PTC.

- Clinical Picture:
- More aggressive compared with PTC.
- Often Asymptomatic
 - o Incidental finding of non-palpable thyroid nodule.
- Painless, slow-growing solitary thyroid nodule.
- Symptoms of compression or invasion:
 - o Hoarseness.
 - o Dysphagea
 - o SOB
 - o Hemoptysis.

- Routes of Follicular thyroid Ca spread:
 - 1. Direct invasion:**
 - Through Thyroid capsule to invade surrounding structures.
 - 2. Hematogenous spread:**
 - More than PTC.
 - 20-50% hematogenous spread with distant mets.
 - Lung is most common site of distant mets.
 - Bone is 2nd most common.
 - 3. Lymphatic spread:**
 - Rare compared to PTC.

- Diagnosis:
 - 1. US Thyroid and Neck:**
 - Evaluate the Thyroid and Presence of positive LN.
 - 2. FNA:**
 - For Suspicious Thyroid nodule and LN.
 - Difficult to distinguish between Follicular Adenoma and Follicular Carcinoma because no definitive cytologic features.
 - 3. Open Biopsy (Lobectomy or Hemi-thyroidectomy):**
 - Indicated for Follicular Thyroid lesions.
 - Can distinguish between Follicular Carcinoma and Follicular Adenoma through identification of tumor extension by the presence of capsular and/or vascular invasion.
- Predisposing factors:
 1. Previous ionizing radiation exposure
 2. Low iodine dietary intake.
- Histopathology of Follicular thyroid Ca:
 - 1. Unifocal Tumor.**
 - 2. Absence of features suggestive of PTC.**
 - Psammoma bodies
 - Ground Glass Appearance/ Orphan Annie Eyes
 - 3. Extracapsular spread with lymphatics or vascular invasion.**
 - 4. Distant Mets.**
- Follicular tumors are divided into:
 - **Minimally invasive follicular cancer (MIFC):**
 - Microscopic penetration of tumor capsule without vascular invasion.
 - **Widely invasive follicular cancer (WIFC):**
 - Tumor extends beyond tumor capsule into blood vessels and adjacent thyroid parenchyma.
- Immunohistochemical staining for thyroglobulin and cytokeratins is nearly always positive.
- Prognosis of Follicular Ca :
- 70-85% 5-year survival rate.
- 20% with distant mets.
- Worse prognosis with vascular invasion and extracapsular spread.

- **Hurthle Cell Tumor (Oncocytic Ca):**
- Variant of Follicular Thyroid Ca.
 - Hürthle cells predominate (Large granular eosinophilic cells) in histopathology.
 - 15% of Follicular Thyroid Ca.
 - 3% of all Thyroid Ca.
 - More aggressive than conventional type of Follicular Ca.
 - Highest rate (34%) of distant metastases among other types of differentiated Thyroid Ca.
- Less sensitive to Thyroid suppression and to diagnostic and therapeutic radioactive iodine therapy.
- Hürthle cells can be found in a variety of benign thyroid conditions:
 1. Hashimoto thyroiditis.
 2. Graves disease
 3. multinodular goiter
- Prognosis of Hürthle cell Ca:
- 50% 5-year survival rate.
- high risk for recurrent and metastatic disease
- Should be monitored closely for recurrent and metastatic disease.

- **Management of Differentiated Thyroid CA (DTC):**
- Goals of initial therapy of DTC:
 - Remove primary tumor and involved cervical lymph nodes.
 - Residual metastatic LN represent the most common site of disease persistence/recurrence.
 - To facilitate post-op treatment with radioactive iodine.
 - For patients undergoing RAI remnant ablation, or RAI treatment of residual or metastatic disease, removal of all normal thyroid tissue is an important element of initial surgery.
 - Total thyroidectomy also reduce the risk for recurrence within contralateral lobe.
 - To permit accurate long-term surveillance for disease recurrence.
 - Both RAI whole-body scanning (WBS) and measurement of serum Thyroglobulin (Tg) are affected by residual normal thyroid tissue.
 - To minimize risk of disease recurrence and metastatic spread.
 - Adequate surgery is the most important treatment variable influencing prognosis.
 - Radioactive iodine treatment, TSH suppression, and external beam irradiation each play adjunctive roles.

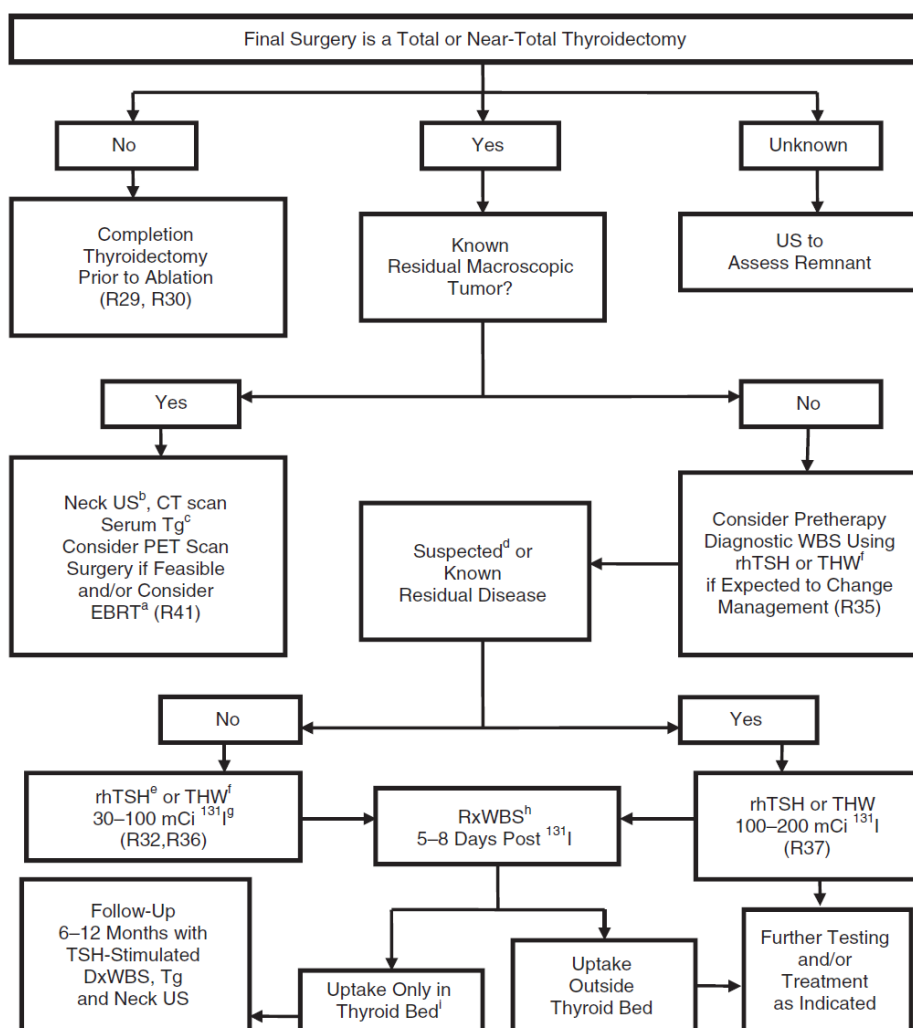
1. Primary treatment is Surgical Excision:

- Primary Thyroid Surgery:
 - **Total Thyroidectomy:**
 - Standard of care for all patients with Thyroid Ca.
 - **Hemi-Thyroidectomy:**
 - Can be offered for Low Risk patients with Follicular Ca:
 - Age <45 years.
 - Isolated thyroid nodule <1 cm.
 - No LN involvement or distant mets.
 - No family History of Thyroid Ca.
 - No history of radiation.
- Neck Dissection:
 - **Elective Central Neck Dissection (VI):**
 - Indicated only for advanced PTC (T3-T4) and negative LN involvement.
 - **Therapeutic Central Neck Dissection (VI):**
 - Indicated for positive central LN involvement.
 - **Therapeutic Modified Neck Dissection (II-V) + Central Neck Dissection (VI):**
 - Performed for positive metastatic lateral LN.

2. Post-op Radioactive iodine (RAI) Scan and Ablation:

- DTC are TSH sensitive and take up iodine.
- **131-I** or **123-I** RAI administration should be performed in HYPOTHYROID state (**High TSH >30 mU/L**) which can be achieved by the one of the following methods:
 1. Discontinuing T4 for 3 weeks and T3 for 2 weeks prior to RAI scan or ablation.
 2. Continuing on T4 with administration of rhTSH (Thyrogen) at time of scan or ablation.
 - Used mainly for patients who are unable to tolerate hypothyroidism or unable to generate an elevated TSH.
- Thyroxine therapy may be resumed on 2nd or 3rd day after RAI administration
- Pregnancy and breastfeeding are Absolute contraindications to RAI administration.
 - Avoid pregnancy for 6–12 months after RAI therapy.
 - Breastfeeding should be stopped at 6-8 weeks prior to RAI therapy.
- **RAI Whole-Body Scan:**
 - Diagnostic dose of **131-I** or **123-I** is given to detect any tissue taking up radioiodine.
 - Remnant normal thyroid tissue.
 - Neoplastic thyroid tissue.
 - Regional and Distant mets.
 - Indications of RAI WBS:
 1. If extent of thyroid remnant can't be accurately ascertained from surgical report or neck U/S.
 2. If the results would alter either the decision to treat or activity of RAI that is administered.
 - Instructions:
 - RAI scans should utilize Low-activity 131-I (1–3 mCi) or 123-I (1.5–3 mCi).
 - RAI ablation should be administered within 72 hours of the diagnostic activity.
 - Disadvantages:
 - If large thyroid remnant, RAI scan is dominated by uptake within the remnant, potentially masking presence of extrathyroidal disease, reducing sensitivity of disease detection.
 - Risk of stunning of thyroid remnants.
 - Stunning is a reduction in uptake of 131-I therapy dose induced by a pre-treatment diagnostic activity.
 - Risk factors to develop stunning:
 1. High-activity 131-I scans (5–10 mCi).
 2. Long duration between diagnostic dose and therapy (>72 hours).

- **RAI Ablation:**
 - Therapeutic dose of ¹³¹I is administered to ablate any detected normal thyroid remnant or metastatic disease.
 - Indications of RAI Ablation:
 1. Primary tumor size >4 cm (**T3**).
 2. Gross extrathyroidal extension (**T4**).
 3. Positive LN.
 4. Distant metastases.
 - Instructions:
 - Ablation with activity of (30–100 mCi) is necessary to achieve successful remnant ablation in low-risk patients.
 - Higher activities (100–200 mCi) is used if residual microscopic disease is suspected or documented, or if more aggressive tumor histology (ex, tall cell).
 - Low-iodine diet for 1–2 weeks is recommended for patients undergoing RAI ablation.
 - Avoid iodine contamination (IV contrast) to RAI therapy to increase the effective radiation dose.
 - Low Thyroglobulin (Tg) level at time of RAI ablation has excellent negative predictive value for absence of residual disease.
 - Risk of persistent disease increases with higher stimulated Tg levels.
 - Post-treatment diagnostic RAI WBS is done after 2-10 days of initial RAI ablation.
 - **Uptake in Thyroid Bed only:**
 - Indicate normal remnant tissue or residual central neck nodal metastases.
 - Follow up 6–12 Months with:
 1. RAI WBS (in High risk patients)
 2. Thyroglobulin (Tg) level.
 3. Neck US.
 - If F/U RAI WBS is **Positive**,
 - Additional RAI Ablation is given.
 - This process is repeated until the diagnostic scan is **Negative**.
 - **Uptake outside Thyroid Bed:**
 - Further Testing and/or Treatment as Indicated.
 - Successful RAI remnant ablation is defined as:
 - Absence of RAI uptake on a subsequent diagnostic RAI WBS.
 - Undetectable stimulated serum Tg.



3. 3. Algorithm for initial follow-up of patients with differentiated thyroid carcinoma.

^aEBRT, external beam radiotherapy. The usual indication for EBRT is macroscopic unresectable tumor in a patient older than 45 years; it is not usually recommended for children and adults less than age 45.

^bNeck ultrasonography of operated cervical compartments is often compromised for several months after surgery.

^cTg, thyroglobulin with anti-thyroglobulin antibody measurement; serum Tg is usually measured by immunometric assay and may be falsely elevated for several weeks by injury from surgery or by heterophile antibodies, although a very high serum Tg level after surgery usually indicates residual disease.

^dSome clinicians suspect residual disease when malignant lymph nodes, or tumors with aggressive histologies (as defined in the text) have been resected, or when there is a microscopically positive margin of resection.

^erhTSH is recombinant human TSH and is administered as follows: 0.9 mg rhTSH i.m. on two consecutive days, followed by ¹³¹I therapy on the third day.

^fTHW is levothyroxine and/or triiodothyronine withdrawal.

^gSee text for exceptions regarding remnant ablation. The smallest amount of ¹³¹I necessary to ablate normal thyroid remnant tissue should be used. DxWBS (diagnostic whole-body scintigraphy) is not usually necessary at this point, but may be performed if the outcome will change the decision to treat with radioiodine and/or the amount of administered activity.

^hRxWBS is posttreatment whole-body scan done 5 to 8 days after therapeutic ¹³¹I administration.

ⁱUptake in the thyroid bed may indicate normal remnant tissue or residual central neck nodal metastases.

- **Complications of RAI Ablation:**

- Cumulative dose-related low risk of early and late complications:

1. Sialadenitis:

- Most frequent complication of RAI therapy.
- Immediate transient bilateral parotid (mainly) swelling and pain with decreased salivary flow (xerostomia).
 - Swelling from the inflammatory infiltrate causes increased periductal pressure with salivary duct constriction and obstruction.
- Lasts for few days then subsides but may persist for long time.
- Duct lumen narrowing from inflammatory stricturing
- Treatment:
 - No permanent cure.
 - Symptomatic treatment.
 - **Aggressive external massage:**
 - Milk out retained saliva.
 - Increase salivary lavage
 - Flush out ductal debris.
 - **Antibiotics:**
 - Used if infection is present (suppurative salivary return and/or fever)
 - **Good oral Hygiene:**
 - Avoid dehydration because it leads to decreased salivary lavage and recurrent exacerbations.
 - **Constant use of sialogogic agents:**
 - Sugarless sour candy.
 - Chewing gum.
 - **Cholinergic agents:**
 - Pilocarpine.
 - **Cytoprotective adjuvant:**
 - Amifostine.

2. GI discomfort (Nausea)

3. bone marrow suppression.

4. Loss of taste sensation.

5. Dental caries.

6. Nasolacrimal duct obstruction.

7. Secondary Malignancies and Leukemia.

- Very low risk.
- Dose related.
- Significant increase in risk with cumulative ^{131}I activities above 500–600 mCi.

3. TSH Suppression Therapy:

- After Total thyroidectomy and RAI ablation, patients with Differentiated thyroid carcinoma are maintained on thyroid-suppression suppression.
- Patients take supra-physiologic doses of T4 daily sufficient to suppress TSH production by the pituitary.
 - Low TSH levels in the bloodstream reduce tumor growth rates and reduce recurrence rates of well-differentiated thyroid carcinomas.
 - **Recommended TSH levels:**
 - High risk Patients **≤ 0.1 mU/L.**
 - Low risk Patients **$0.1-0.5$ mU/L.**
- Adverse effect of TSH suppression:
 - Subclinical thyrotoxicosis.
 - Exacerbation of angina in patients with ischemic heart disease.
 - Increased risk for Atrial fibrillation in older patients.
 - Increased risk of osteoporosis in postmenopausal women.

4. External Beam Irradiation:

- Indications:
 1. Gross unresectable tumor in patients > 45 years old.
 2. None-operable symptomatic metastases:
 - Fracture.
 - Painful bone metastases.
 - Vertebral metastases.
 - CNS metastases.
 - Pelvic metastases.
 3. Adjuvant therapy with surgery for selective cases of bone or brain metastases with concomitant use of glucocorticoids.

5. Chemotherapy:

- Indications:
 1. Distal metastases, especially in the situations with the evidence of progression, RAI refractory diseases, or compromising organ function.
- Doxorubicin is the only FDA-approved agent for metastatic thyroid carcinoma.

- **Long term follow-up plan for patients with DTC:**
- **Goals of long term F/U:**
 - Accurate surveillance for possible recurrence.
 - Monitor Thyroxine suppression or replacement therapy.
- **Criteria for Absence of persistent tumor post Total Thyroidectomy and RAI Ablation:**
 1. No clinical evidence of tumor.
 2. No imaging evidence of tumor
 - No uptake outside Thyroid bed on initial post-treatment RAI WBS.
 - If uptake outside thyroid bed had been present, No imaging evidence of tumor on a recent diagnostic scan and neck US.
 3. Undetectable serum Tg levels during TSH suppression and stimulation in the absence of interfering antibodies.

1. Thyroglobulin (Tg) and Anti-Thyroglobulin Antibodies levels:

- Serum Tg level is sensitive and specific marker to detect PTC recurrence in the absence of Tg antibodies, especially post total thyroidectomy and/or RAI ablation.
- Should be done every 6-12 months with Neck US.
- Important modality to monitor patients for residual or recurrent disease.
- Anti-Thyroglobulin Antibodies are presents in 25% of thyroid cancer patients.
 - Interfere with Thyroglobulin (Tg), causing False Low Thyroglobulin (Tg) level.
 - **Always request Anti-Thyroglobulin Antibodies with Thyroglobulin (Tg) level.**
- If Negative Anti-Thyroglobulin Antibodies in patients post Total thyroidectomy and RAI ablation and during TSH suppression or stimulation:
 - **Rising Tg level over time is suspicious and has high sensitivity and specificity to detect thyroid cancer.**
 - No specific cutoff Tg levels that distinguish normal residual thyroid tissue from persistent thyroid cancer.
 1. Tg level post rhTSH stimulation is requested only if Tg level with TSH suppression is Negative.
 - Tg level >2 ng/mL following rhTSH stimulation is highly sensitive in identifying patients with persistent tumor.
 - Tg <0.5 ng/mL following rhTSH stimulation identifies patients completely free of tumor on follow-up.

- **If Tg >1 ng/mL with Negative Tg Ab:**
 - RAI WBS:
 1. If Positive:
 - → RAI Ablation.
 2. If Negative:
 - Stimulated Tg <5–10:
 - Monitor Tg and Neck US:
 - If Rising Tg with Negative US:
 - CT Neck/Chest
 - MRI Neck
 - If ALL are Negative do **PET/CT**
 - Stimulated Tg >5–10:
 - CT Neck/Chest
 - MRI Neck
 - If ALL are Negative do **PET/CT**

2. Neck US:

- Used to evaluate thyroid bed, central and lateral cervical LN.
 - Should be performed with Tg level at 6–12 months and then periodically depending on the patient's risk for recurrent disease and Tg status.
- Highly sensitive in detection of cervical metastases in patients with DTC.
 - **Can detect cervical metastases with patients with undetectable TSH-stimulated serum Tg levels.**
- If Positive results:
 - Suspicious LN should be biopsied for cytology with Tg measurement in needle washout fluid.

3. RAI WBS:

- Most useful during follow-up when there is no remaining normal thyroid tissue.
- After RAI Ablation, post-therapy RAI WBS should be done.
 - If it does not reveal uptake outside thyroid bed, subsequent RAI WBS are NOT necessary in Low-risk patients with the following:
 1. Clinically free of residual tumor.
 2. Undetectable serum Tg level on thyroid hormone.
 3. Negative Anti-Thyroglobulin Antibodies.
 4. Negative cervical US.

4. PET/CT:

- Indicated for localization of the disease in patients with:
 - 1. Positive Tg level, and**
 - Tg level is ≥ 10 ng/mL after T4 withdrawal, or
 - Tg level ≥ 5 ng/mL after rhTSH stimulation.
 - 2. Negative RAI WBS.**
- If Negative PET/CT:
 - Empiric RAI Ablation (100–200 mCi) is considered to aid localization or for therapy of surgically incurable disease.

- Management of DTC patients with metastatic disease:

- Preferred steps of treatment for metastatic disease (in order):
 - 1.** Surgical excision of locoregional disease.
 - 2.** ^{131}I RAI Ablation for RAI-avid disease.
 - 3.** External beam radiation.
 - 4.** Watchful waiting with patients with stable or slowly progressive asymptomatic disease.
- Unlike other malignancies, surgical treatment is NOT contraindication for DTC with distant metastatic disease.
- Treatment of locoregional disease confined to neck with negative distant metastasis:
 - **Therapeutic MND (L2-5) + CND (L6).**
- Treatment of Aerodigestive invasion:
 - **Surgery + RAI Ablation and/or External beam radiation.**
 - Surgery ranges from:
 - Shaving tumor off trachea or esophagus for superficial invasion.
 - Tracheal resection and anastomosis or Laryngopharyngoesophagectomy for direct intraluminal invasion.
- Treatment of Pulmonary metastases:
 - Pulmonary micrometastases should be treated with RAI Ablation and repeated every 6–12 months as long as disease continues to concentrate RAI and respond clinically.
- Treatment of Bone metastases:
 - Complete surgical resection of isolated symptomatic metastases.
 - RAI therapy of iodine-avid bone metastases.
- Treatment of Brain metastases:
 - Complete surgical resection.
 - IF not amenable to surgery, External beam irradiation and Radiosurgery.

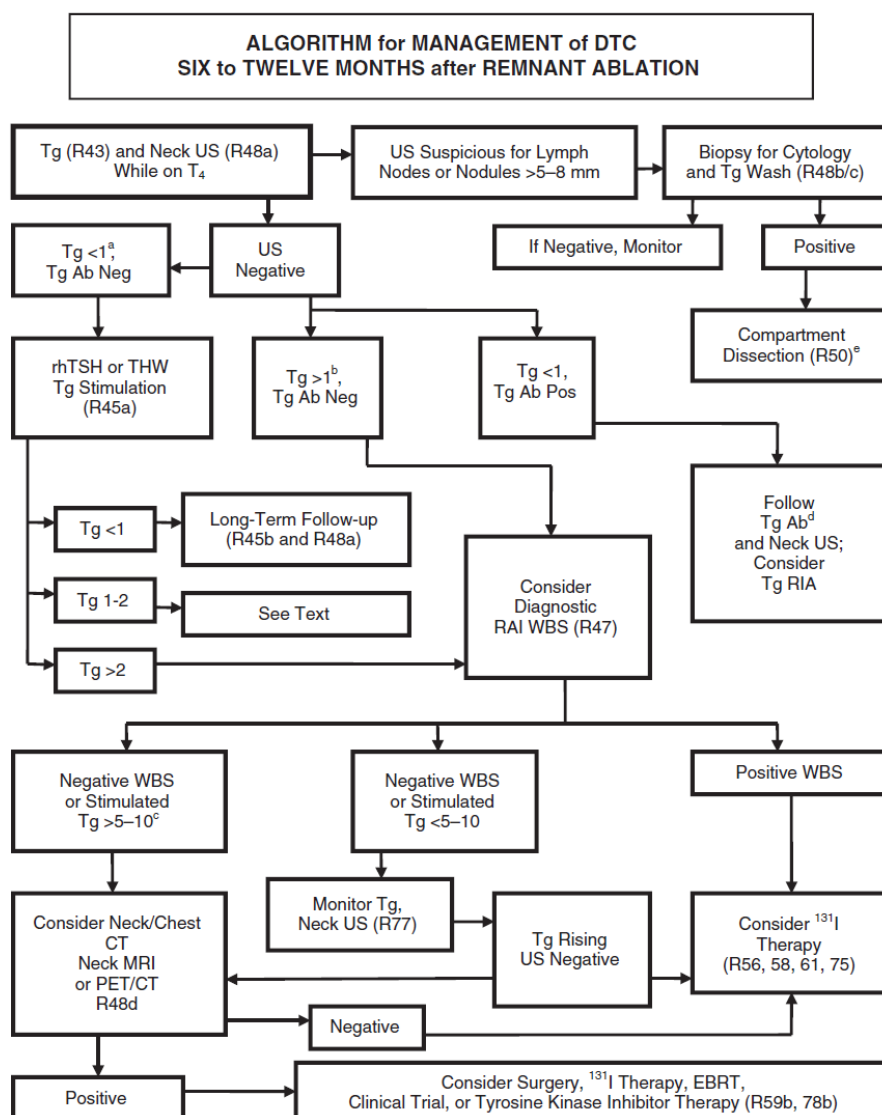


FIG. 4. Longer term follow-up of patients with differentiated thyroid carcinoma.

^aTgAb is anti-thyroglobulin antibody usually measured by immunometric assay.

^bHeterophile antibodies may be a cause of falsely elevated serum Tg levels (436,437). The use of heterophile blocking tubes or heterophile blocking reagents have reduced, but not completely eliminated this problem. Tg that rises with TSH stimulation and falls with TSH suppression is unlikely to result from heterophile antibodies.

^cSee text concerning further information regarding levels of Tg at which therapy should be considered.

^dTg radioimmunoassay (RIA) may be falsely elevated or suppressed by TgAb. Tg results following TSH stimulation with rhTSH or thyroid hormone withdrawal are invalidated by TgAb in the serum even when Tg is measured by most RIA tests. TgAb levels often decline to undetectable levels over years following surgery (306). A rising level of TgAb may be an early indication of recurrent disease (305).

^eSee text for decision regarding surgery versus medical therapy, and surgical approaches to locoregional metastases. FNA confirmation of malignancy is generally advised. Preoperative chest CT is recommended as distant metastases may change management.

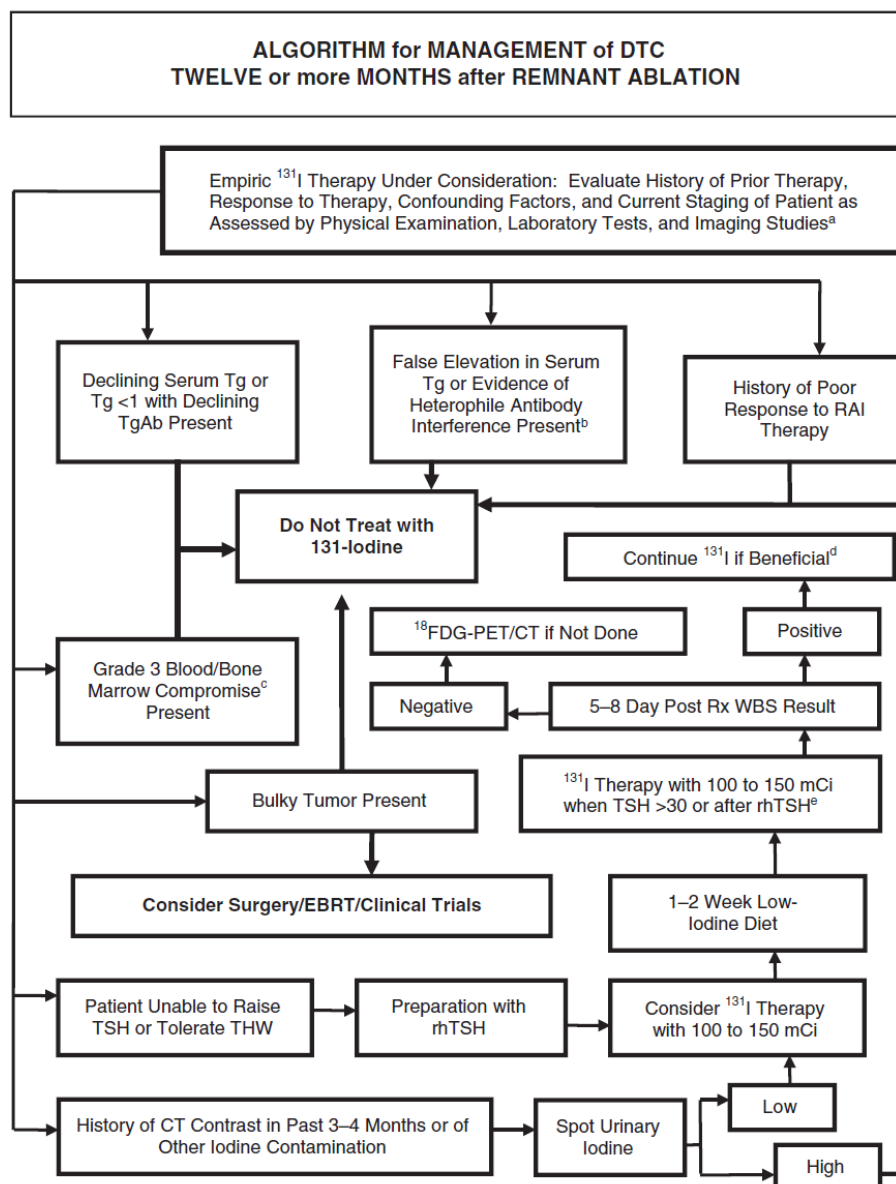


FIG. 5. Considerations for empiric treatment with radioiodine.

^aEmpiric ^{131}I therapy should be done with meticulous patient preparation, including low-iodine diet and, if iodine contamination is a possibility, urinary iodine measurements. If the RxWBS is negative or subsequent follow-up studies show no therapeutic benefit, further empiric ^{131}I should not be administered.

^bTg that rises with TSH stimulation and falls with TSH suppression is unlikely to result from heterophile antibodies.

^cNational Cancer Institute Common Terminology Criteria for Adverse Events, Version 3.0, (<http://ctep.cancer.gov>).

^dDosimetry could be considered to allow administration of maximum radioiodine activity if the tumor is life-threatening.

^eA dose of 200 mCi could exceed the maximum tolerable dose in older individuals (see Recommendation 52b).

- **Management of DTC in Special Patient Populations:**
- **Children:**
 - Diagnostic and therapeutic approaches to thyroid neoplasms in children are similar to adults.
 - Consider familial and syndromic associations.
- **Pregnancy:**
 - Thyroid nodules in pregnant women should be evaluated in the same way as nonpregnant women EXCEPT that use of RAI is contraindicated.
 - If FNA is consistent with PTC during pregnancy:
 - **Early pregnancy (< 24 weeks):**
 - Monitor with US for growth or spread.
 - If substantial growth is evident before 24 weeks gestation, surgery should be performed.
 - surgery is recommended and should be done before 24 weeks gestation.
 - **Late pregnancy (> 24 weeks):**
 - Surgery should be done after delivery.
- **Prognostic factors for Differentiated Thyroid CA:**
- ✓ **Age:**
 - Death is most likely to occur if patient is >40 years at time of diagnosis.
 - Recurrences most common in patients whose disease is diagnosed when they were < 20 years or >60 years.
- ✓ **Sex:**
 - Men are twice as likely as women to **die** from thyroid cancer.
- ✓ **Size:**
 - Primary tumors >4 cm have increased recurrence and cancer-related mortality rates.
- ✓ **Histology:**
 - Papillary carcinoma (benign cancer) **95%** 10-year survival.
 - Follicular carcinoma **70–85%** 5-year survival (20% with distant metastasis)
- ✓ **Local invasion:**
 - Extracapsular spread Invasion to surrounding tissues outside of thyroid (angioinvasion in follicular) indicates significantly worsens the patient's prognosis.
- ✓ **Lymph node metastasis:**
 - Cervical metastases have increased cervical recurrence rates without affecting survival
- ✓ **Distant metastasis:**
 - Distant metastasis at initial examination is associated with a 68.1-fold increase in the rate of disease-specific death.

- **Medullary Thyroid Ca (MTC):**
- 4% of all thyroid Ca.
- M=F
- **Pathophysiology of MTC:**
 - MTC is Neuroendocrine tumor of **Parafollicular (C-cells)**.
 - Highest concentration of C-cells are found in the Lateral upper 2/3rds of Thyroid gland.
 - Malignant C-cells release:
 1. **Calcitonin** (Useful for diagnosis and monitoring)
 2. **Carcinoembryonic Antigen (CEA)** (85%)
 3. **Adrenocorticotropin Hormone (ACTH)**
 4. Gastrin
 5. Substance P
 6. Histaminadases
 7. Prostaglandins
 8. Serotonin
 - Thyroid function tests are normal in patients with MTC.
 - Parafollicular C-cells are NOT TSH-responsive.
 - Thyroid Hormone Suppression Therapy is Not indicated in patients with MTC.
 - Parafollicular C-cells do Not take up 131-I.
 - Radioactive iodine (131-I) has NO role in follow-up or treatment of patients with MTC.

- **Two Types of MTC:**

1. Sporadic MTC (80%):

- Most common type.
- Unifocal and Unilateral.
- Age 50-60 years
- Worse Prognosis.
 - Disease has already metastasized at the time of diagnosis.
 - Early LN involvement.
 - Late vascular involvement.
 - 50% have palpable Cervical LN.
 - 15% have compression or invasion symptoms.
 - 5% have distant mets (Mediastinum , Lungs , Liver, Bone)

2. Inherited MTC (20%):

- Less common type.
- Multifocal and Bilateral (90%).
- Autosomal dominant.
- Mutations in **RET** proto-oncogene (Chromosome arm 10q)
- Part of Multiple Endocrine Neoplasia (MEN-2) which subclassified into:

1. **MEN 2A (Sipple's Syndrome):**

- Associated with: (MTC+2p)
 1. MTC
 2. Pheochromocytoma.
 3. Primary Hyperparathyroidism.
- Most common subtype of MEN2.
- MTC always develops by Third decade.
- 95% of MEN 2A have identifiable RET mutation.
- Penetrance of MTC is nearly 100%.

2. **MEN 2B (Williams-Pollock Syndrome):**

- Associated with: (MTC+1p)
 1. MTC
 2. Pheochromocytoma
 3. Mucosal neuroma
 4. Marfanoid habitus
- Least common subtype of MEN2.
- Most aggressive type of MEN due to its very early onset of the disease.
- MTC develops at Earlier age (10 year-old)
- 95% of MEN 2B have identifiable RET mutation.
- Penetrance of MTC is nearly 100%.

3. **Familial Medullary Thyroid Ca (FMTC):**

- MTC alone without other endocrine involvements.
- MTC in FMTC usually develops during Adulthood.
- 88% of FMTC have identifiable RET mutation.

TABLE 4–3. Multiple Endocrine Neoplasms

MEN I (Werner's Syndrome)	MEN II (Sipple Syndrome)	MEN IIB/III
Parathyroid hyperplasia	Medullary thyroid carcinoma	Medullary thyroid carcinoma
Pancreatic tumors (insulinomas, gastrinomas)	Pheochromocytoma	Pheochromocytoma
Pituitary adenomas	Parathyroid hyperplasia	Mucosal neuromas
		Marfanoid habitus

- Clinical Picture:
- Most common presentation is solitary thyroid nodule.
- Symptoms of compression or invasion:
 - Hoarseness.
 - Dysphagea
 - SOB
 - Hemoptysis.

- Diagnosis:

1. US Thyroid and Neck:

- Evaluate the Thyroid and Presence of positive LN.

2. FNA:

- For Suspicious Thyroid nodule and LN.
- FNA sensitivity for detection of MTC (63%) is lower than that of serum calcitonin (98%).
 - Higher sensitivity can be obtained by the addition of immunohistochemical staining for calcitonin.

3. Elevated serum Calcitonin:

- Used to diagnose MTC.
 - Serum Calcitonin concentrations correlate with tumor mass.
 - Pentagastrin-stimulated calcitonin test:
 - IV Pentagastrin increases sensitivity of the test.
 - Baseline plasma calcitonin level is measured.
 - IV administration of Pentagastrin 0.5 mg/kg.
 - Serial measurements of Calcitonin 1.5 and 5 minutes after injection.
 - Elevated basal or stimulated calcitonin levels strongly suggest MTC.
- Good screen test for family members.
- Used to determine success of surgery.
- Monitors for Recurrence.

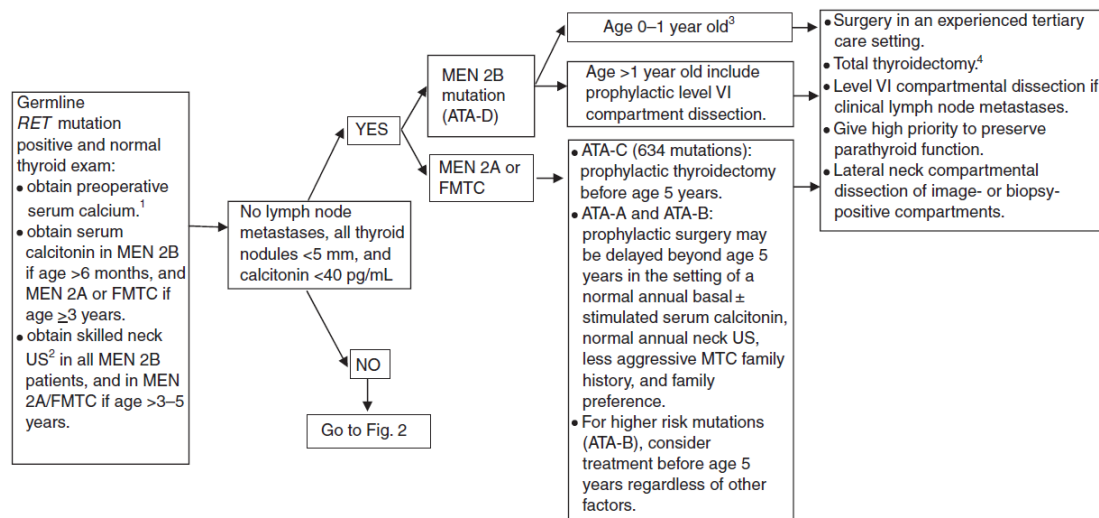
4. Elevated serum CEA.

5. Presence of RET proto-oncogene mutations:

- Indications:
 1. All patients with MTC:
 - To diagnose patients with MEN2.
 - For patients with sporadic MTC to rule out new spontaneous germline mutations.
 2. Screening of High risk patient to develop MTC:
 1. All patients with history of primary C cell hyperplasia or MEN2.
 2. All 1st degree family members of patients with MEN2 or MTC.
 - If Positive, 90% will get MTC within 1st two decades of life.

○ **Testing for coexisting tumors:**

- Parathyroid tumor (MEN 2A):
 - Elevated serum Calcium.
 - Used also to screen Family members of MEN2.
- Pheochromocytoma (MEN 2B):
 - Elevated serum and 24h urine Metanephrines and catecholamines.
 - Used also to screen Family members of MEN2.



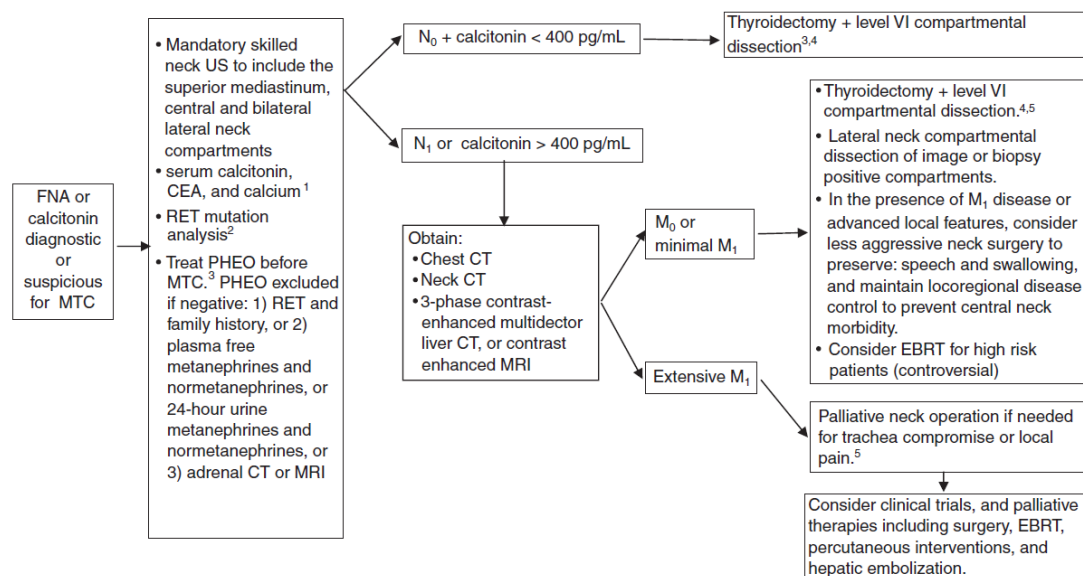
¹Treat hyperparathyroidism with 4 gland resection and autograft to heterotopic site, or subtotal parathyroidectomy. Consider cryopreservation. PHEO preoperative screening should begin by age 8 years for MEN 2B and mutated *RET* codons 634 and 630; otherwise by age 20 years for other *RET* mutations.

²Neck US to include the superior mediastinum and central and lateral neck compartments.

³Insufficient data to recommend routine prophylactic level VI compartmental dissection.

⁴Parathyroid glands resected or devascularized should be autografted in the neck in *RET*-negative, MEN 2B, and FMTC patients, while MEN 2A glands should be autografted to a heterotopic site.

FIG. 1. Initial diagnosis and therapy of pre-clinical disease.



¹Treat hyperparathyroidism with 4 gland resection and autograft to heterotopic site, or subtotal parathyroidectomy. Consider cryopreservation.

²Ideally performed with genetics counseling and completed preoperatively.

³PHEO preoperative screening should begin by age 8 years for MEN 2B and mutated *RET* codons 634 and 630; and by age 20 years for other *RET* mutations.

⁴Parathyroid glands resected or devascularized should be autografted in the neck in *RET*-negative, MEN 2B, and FMTC patients, while MEN 2A glands should be autografted to a heterotopic site.

⁵Consider external beam radiation of TNM stage T4 disease to prevent recurrent local disease.

FNA, fine-needle aspiration biopsy.

FIG. 2. Initial diagnosis and therapy of clinically apparent disease.

- Histopathology of MTC:
 1. **Amyloid stroma**
 - Typical green birefringence on Congo red staining
 2. **Small round cells**
 3. **May have calcification and fibrotic strands**
 4. **May have peripheral C-cell hyperplasia.**
- Prognosis of MTC:
 - 50–80% 10-year survival
 - 45% if cervical LN are involved.
 - MTC is the most common cause of mortality in MEN2.
 - MEN 2B have a prognosis worse than MEN 2A.
 - Worse prognosis in:
 - Unilateral
 - Sporadic type
 - Younger patient
 - MTC with Mets.
- Management of MTC:
 1. **Primary treatment is Surgical Excision:**
 - Surgery:
 - **Total Thyroidectomy.**
 - For ALL patients with MTC.
 - If the vasculature of Parathyroid gland is disrupted, Autotransplantation of Parathyroid gland into SCM or Non-dominant forearm is performed.
 - Neck Dissection:
 - **Prophylactic CND (Level VI):**
 - For ALL patients with MTC.
 - **Therapeutic MND (II-V) + CND (VI):**
 - For patients with positive lateral LN.
 - **If +ve Distant metastasis:**
 - Less aggressive neck surgery is appropriate to preserve speech, swallowing, and parathyroid function while maintaining locoregional disease control to prevent central neck morbidity.
 2. **External Beam Radiation Therapy (EBRT): (Controversial)**
 - Used as an adjuvant at some centers.
 - Can be used to treat patients with:
 1. Surgically inoperable.
 2. Recurrences and metastases.
- **MTC is relatively Insensitive to Chemotherapy**
- **In patients with MEN2, Pheochromocytoma should be surgically resected prior to surgery of MTC or primary hyperparathyroidism due to the high risk of anesthesia.**

- [Management of Patients with High Risk to develop MTC:](#)
- All 1st degree family members of MEN2 or MTC patients should go for screening tests of MTC which can be by:
 - 1. Basal and Pentagastrin-stimulated Calcitonin level:**
 - Not preferred alone due to risk of false positive test.
 - Negative test in a child at risk for MEN2 reflects only that disease is not currently detectable.
 - 2. Genetic testing of RET-mutation:**
 - Gold standard screening test.
 - Definitively establishes RET-mutation carriers.
 - Timing:
 - Before age of 5 years for relatives of patients with MEN 2A and FMTC
 - Shortly after birth for relatives of patients with MEN 2B.
 - **If Positive RET-Mutation:**
 - **Prophylactic Total Thyroidectomy with Central LN dissection (Level VI):**
 - Timing:
 - 1st year of life in MEN2B.
 - Before 5 years of life in MEN2A/FMTC.
 - 90% will get MTC within first two decades of life.
 - If children's parents refuses Surgery:
 - Annual Pentagastri-Stimulated Calcitonin level is done until at least age 35 years or until a positive test occurs.
 - Total Thyroidectomy with Central LN dissection (Level VI) should be done for patients with positive results.
 - **If Negative RET-Mutation:**
 - No further action is needed.

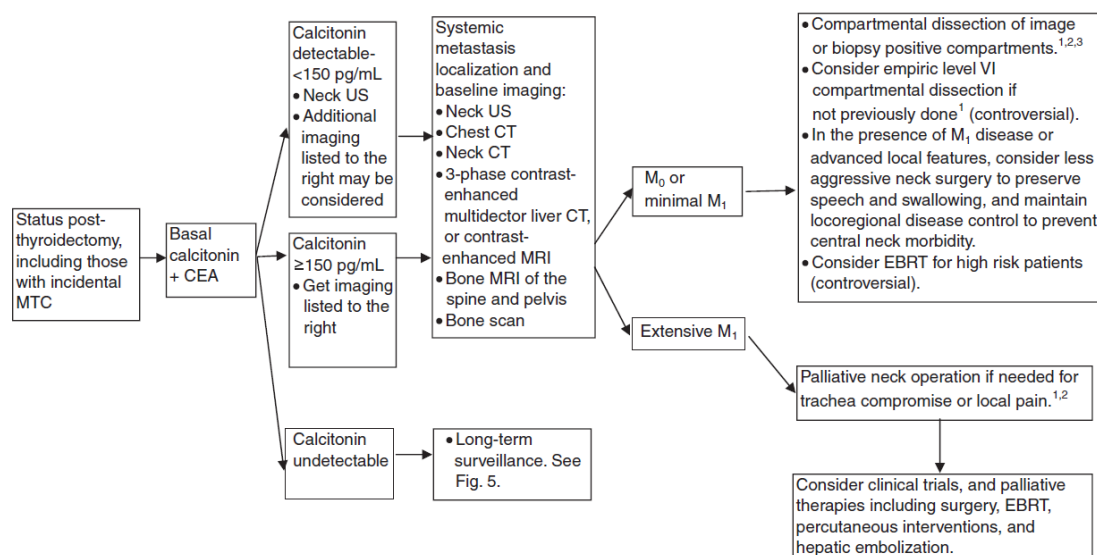
- [Post-op Follow-up Care:](#)

1. Thyroxin Replacement:

- Started immediately after surgery.
- Initial dose is 1.6 mcg/kg of body weight.
- TSH measurement after one month.
- The goal of T4 therapy is to restore and maintain Euthyroidism.
- Suppressive doses are not necessary as C cells are not TSH-responsive.

2. Serial monitoring of serum Calcitonin and CEA with Physical Exam:

- Serum calcitonin is highly sensitive method to detect persistent or recurrent disease.
- Doubling time has proven to be a useful marker of disease burden and prognosis.
- Both calcitonin and CEA play important roles in follow-up of MTC patients.
- Started 2-3 months after surgery.
- Calcitonin concentration falls slowly in some patients.
- [Normal CEA and undetectable Calcitonin values:](#)
 - Patients considered biochemically cured.
 - Have the best prognosis (5% 5-year recurrence rate).
 - Subsequent follow-up with Physical examination and Calcitonin and CEA levels every 6 months for 2 years then annually.
- [High Basal Serum Calcitonin value:](#)
 - Presumptive evidence of Residual disease.
 - Required to do Neck U/S to localize cervical disease.
 - Neck, abdominal, and pelvic CT or MRI is used to detect disease if metastasis or recurrence is suspected.
 - **If Positive Imaging for recurrence or residual with High Calcitonin level:**
 - Surgery.
 - +/- Radiotherapy.
 - **If Negative Imaging for recurrence or residual with Stable High Calcitonin level:**
 - Observation with annual Neck U/S for 2-3 years to detect any progression, then less often during long-term follow-up.
 - **If Negative Imaging for recurrence or residual with Rising High Calcitonin level:**
 - **PET/CT.**
 - +/- Chemotherapy

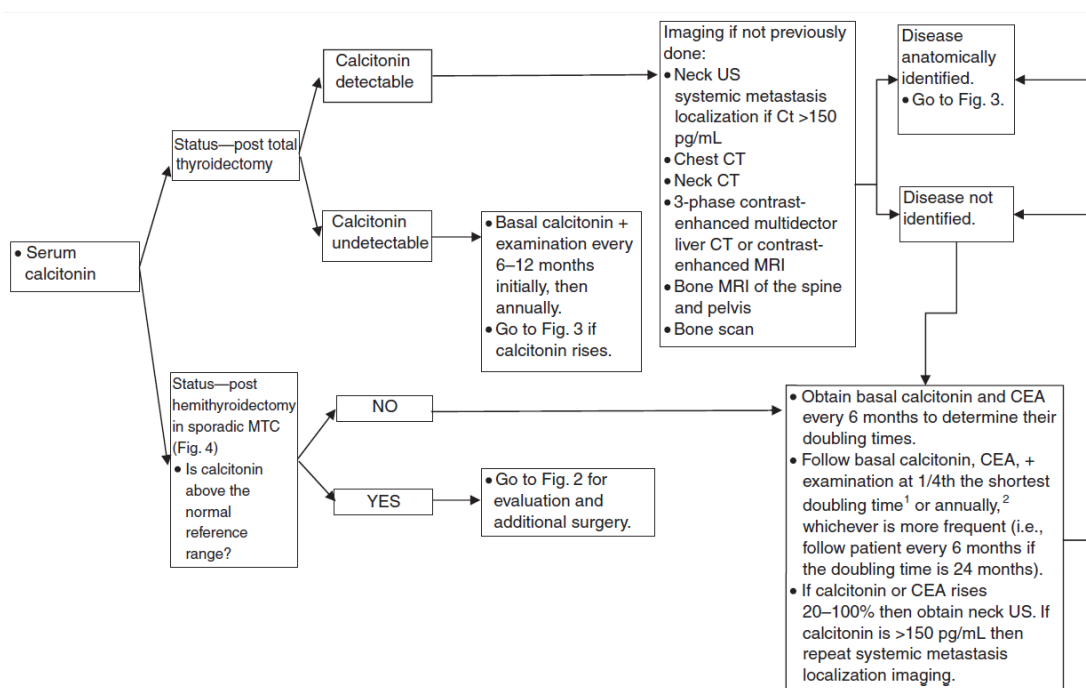


¹Parathyroid glands resected or devascularized should be autografted in the neck in *RET*-negative, MEN 2B, and FMTC patients, while MEN 2A glands should be autografted to a heterotopic site.

²Consider external beam radiation of T₄ disease to prevent recurrent local disease.

³Observation of nonthreatening locoregional disease <1 cm may be considered.

FIG. 3. Initial evaluation and treatment of postoperative patients.



¹ Doubling time may be estimated or optimally calculated by fitting data to single exponentials by nonlinear least-square analysis (calculator available at www.thyroid.org).

²Patients with *RET* mutations associated with PHEO or primary hyperparathyroidism should be screen annually beginning at age 8 years in MEN 2B and mutated *RET* codons 634 and 630, and from age 20 years in carriers of other MEN 2A *RET* mutations, while those associated only with FMTC should be screened at least periodically.

FIG. 5. Long-Term surveillance.

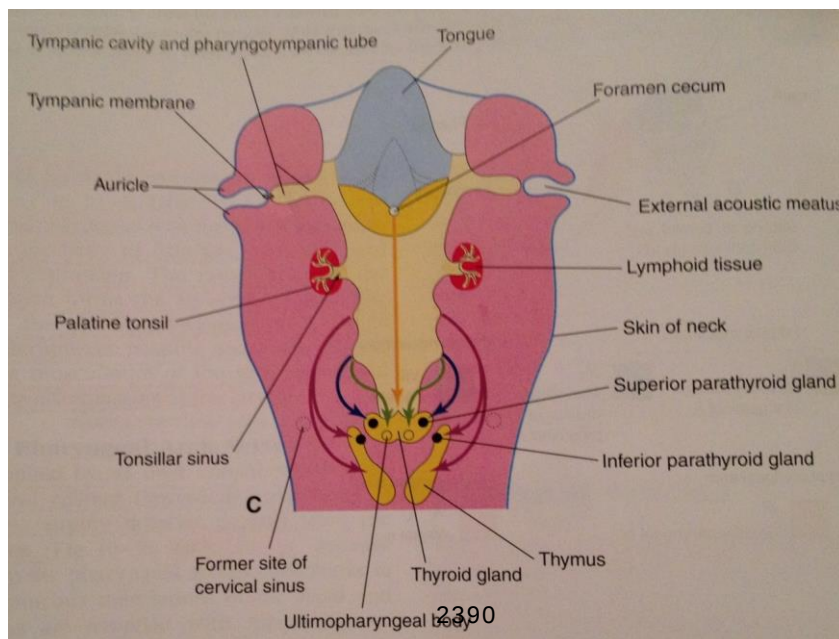
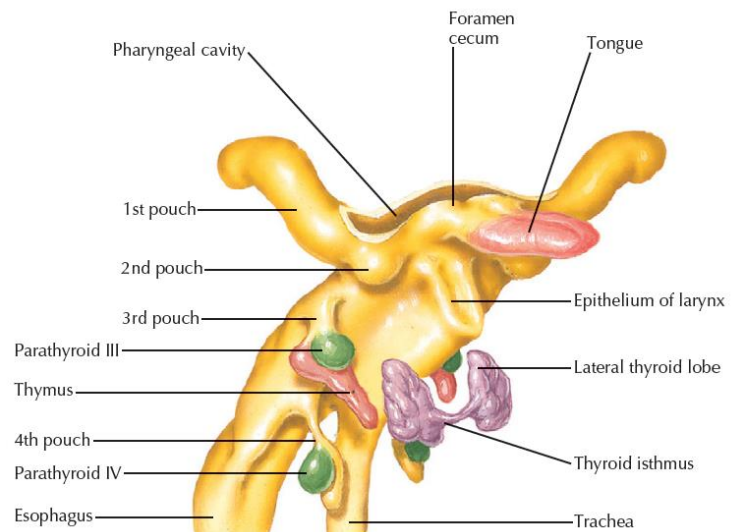
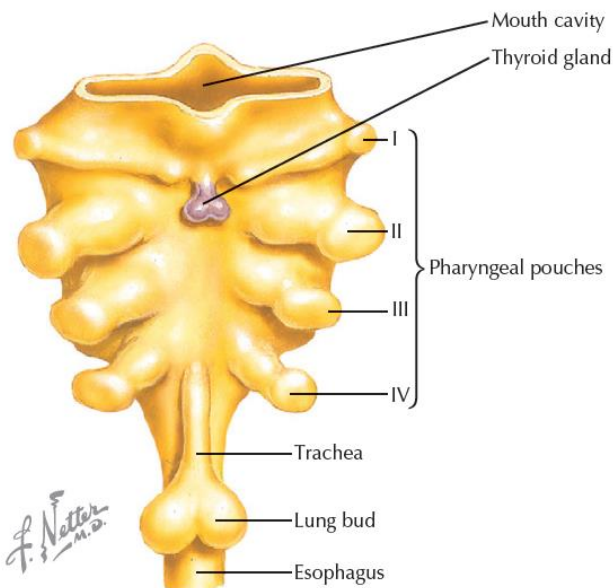
- **Anaplastic Ca:**
 - Extremely aggressive and poorly differentiated Thyroid Ca.
 - 1-5% of thyroid Ca.
 - F>M (Similar to PTC and Follicular ca).
 - Most commonly seen in Elderly (7th decade).
- Pathophysiology of Anaplastic Ca:
 - o May be from transformation of differentiated carcinoma.
 - o May find coexistent follicular or papillary carcinoma (20-30%).
 - o Rapid , Aggressive.
 - o Spread by infiltrative & direct extension.
- Clinical Picture:
 - Present later than other thyroid malignancies.
 - All patients present with rapidly enlarging Thyroid mass.
 - o Bilateral asymmetric thyroid enlargement.
 - o Hard, Nodular and fixed to surrounding structures.
 - o Most often larger than 5 cm at presentation.
 - o Causing compressive or invasive symptoms.
 - 50% of patients have enlarged cervical LN.
 - 90% of cases have regional or distant Mets at time of diagnosis.
 - o Lung is the most common site of distant Mets (90%).
 - Horner's Syndrome (Ptosis, Miosis and Anhidrosis) due to lesion or compression of one side of cervical or thoracic sympathetic chain.
- Diagnosis:
 1. **US Thyroid and Neck:**
 - Evaluate the Thyroid and Presence of positive LN.
 2. **FNA:**
 - For Suspicious Thyroid mass and LN.
- Histopathology of Anaplastic Ca:
 1. **Undifferentiated "bizarre cells".**
 2. **Squamoid ,Giant and Spindle cells variation.**
 3. **Tumor often extends through capsule of Thyroid gland itself.**
- Prognosis of Anaplastic Ca:
 - Worst survival rates of all malignancies in general.
 - Disease-specific mortality approaching 100%.
 - Median survival is 8 months after diagnosis
 - Poorly responsive to multimodality therapy.
 - Most patients die from local airway obstruction or complications of pulmonary metastases.
- Management of Anaplastic Ca:
 - No adequate therapy.
 - o Complete excision is often impossible because disease is advanced at the time of diagnosis in most patients.
 - **Tracheotomy** for airway protection.
 - Radiation and Chemotherapy (Doxorubicin).

- **Primary Thyroid Lymphoma:**
- Less than 5% of all thyroid CA.
- Most common type is **Non-Hodgkin B-cell lymphoma (NHL)**.
- 2nd Most common type is **Low-grade malignant lymphoma of mucosa-associated lymphoid tissue (MALT)**.
- Hodgkin lymphoma, Burkitt cell lymphoma, and T-cell lymphoma have also been reported.
- **Chronic Autoimmune (Hashimoto's) Thyroiditis** is the only known risk factor for primary thyroid lymphoma
 - o Found in 50% of patients with primary thyroid lymphoma.
 - o Increase risk of thyroid lymphoma at least 60 times higher than in patients without thyroiditis.
- Women are more commonly affected than men.
- Incidence peaks in the sixth decade of life.
- **Clinical Picture:**
- Similar to Anaplastic Ca.
- Most common presentation is Rapid enlarged thyroid mass.
- 10% have B-symptoms (fever, night sweats, and weight loss).
- 10% have HYPO-thyroidism,
- May have compressive or invasive symptoms.
- Regional and distant lymphadenopathy is common.
- **Diagnosis:**
 1. **US Thyroid and Neck:**
 - Evaluate the Thyroid and Presence of positive LN.
 2. **FNA:**
 - For Suspicious Thyroid nodule and LN.
 - Difficult to distinguish between Lymphoma and Chronic Lymphocytic Thyroiditis.
 3. **Open Biopsy (Lobectomy or Hemi-thyroidectomy):**
 - Indicated for all patients suspected for Thyroid lymphoma alone with lymphoma staging work-up.
- **Prognosis of Thyroid lymphoma:**
- 5-year survival rate of up to 85%.
- Spread beyond the thyroid gland reduces the 5-year survival rate to about 35%.
- **Management of Thyroid lymphoma:**
- **Chemotherapy:**
 - o Rapid response to chemotherapy with or without radiation.
 - o Most common regimen is **CHOP**.
 - Cyclophosphamide, Doxorubicin, Vincristine and Prednisone.
- **Radiation therapy:**
 - o Neck and Superior mediastinum.
- **Surgery:**
 - o Aids for diagnosis of lymphoma.
 - o Not the primary treatment modality.

Thyroid CA			
Papillary 80%	Follicular 10%	Medullary 5%	Anaplastic 1-5%
Most common Popular Young F>M Mets to Near LN Slow growing Hx of Radiation exposure Great prognosis 95% 10-year survival Rx: surgery then radioablation , ,Thyroid suppression Psammoma bodies	2nd Most common Elderly F>M Hematogenous spread with distant metastasis LN rare Slow growing Prognosis 70-85% 5-year survival Rx: surgery then radioablation , ,Thyroid suppression	Arise from Parafoallicular C-cells Familial (MEN IIA/B) RET proto-oncogene mutations F = M LN (Early) Vascular (late) Bad prognosis Rx : only surgery Produce: calcitonin , CEA , gastrin , ACTH	F>M Spread by direct extension Aggressive , rapid Bad prognosis , survival is 8.1 months after diagnosis Rx: tracheostomy

Parathyroid Embryology:

- Superior Parathyroid (Parathyroid IV):
 - Arise from **Dorsal** Endodermal cells of **4th Pharyngeal Pouch**.
 - Closely associated with lateral lobes of Thyroid.
 - Have short line of embryologic descent together with Ultimopharyngeal body.
 - More constant location in the neck.
- Inferior Parathyroid (Parathyroid III):
 - Arise from **Dorsal** Endodermal cells of **3rd Pharyngeal Pouch**.
 - Closely associated with Thymus.
 - Have longer line of embryologic descent together with Thymus.
 - Leads to more variability in their anatomic position.



- **Parathyroid Histology:**

- Composed primarily of chief cells and fat with thin fibrous capsule dividing gland into lobules.

- Chief cells:

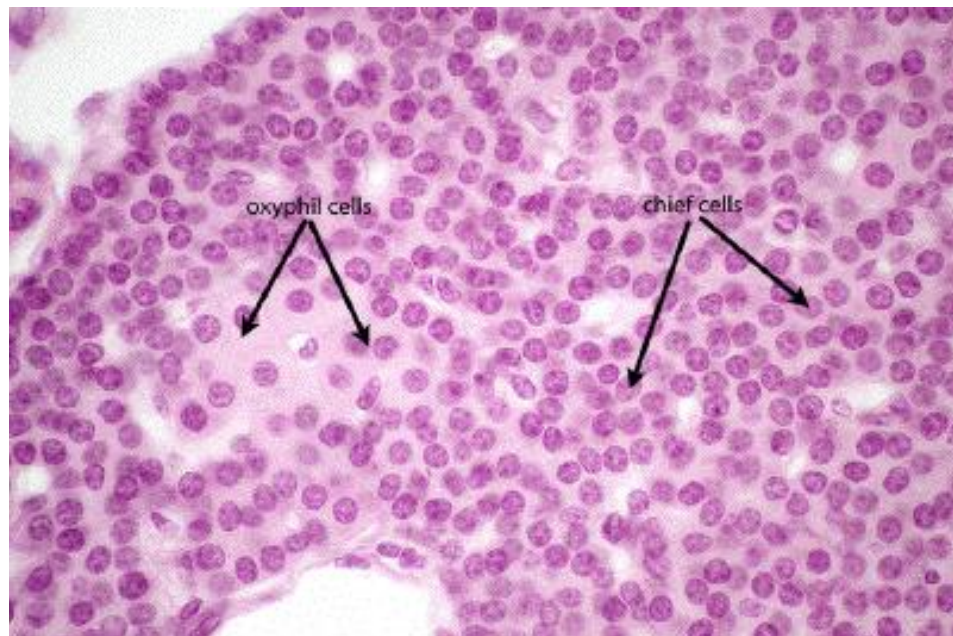
- Basic cell type of Parathyroid.
- Contain granules of parathyroid hormone (PTH)
- Chief cell is most sensitive to changes in ionized calcium

- Oxyphil cells:

- Slightly larger than chief cell.
- No secretory granules

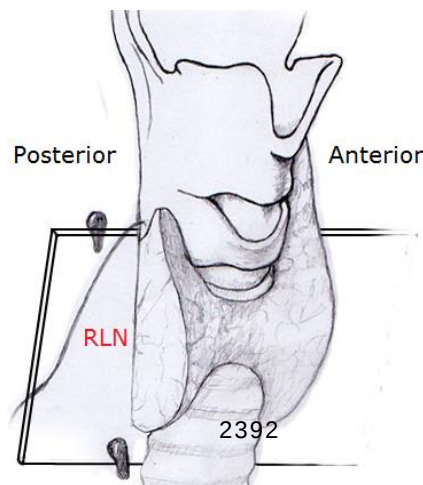
- Water clear cell:

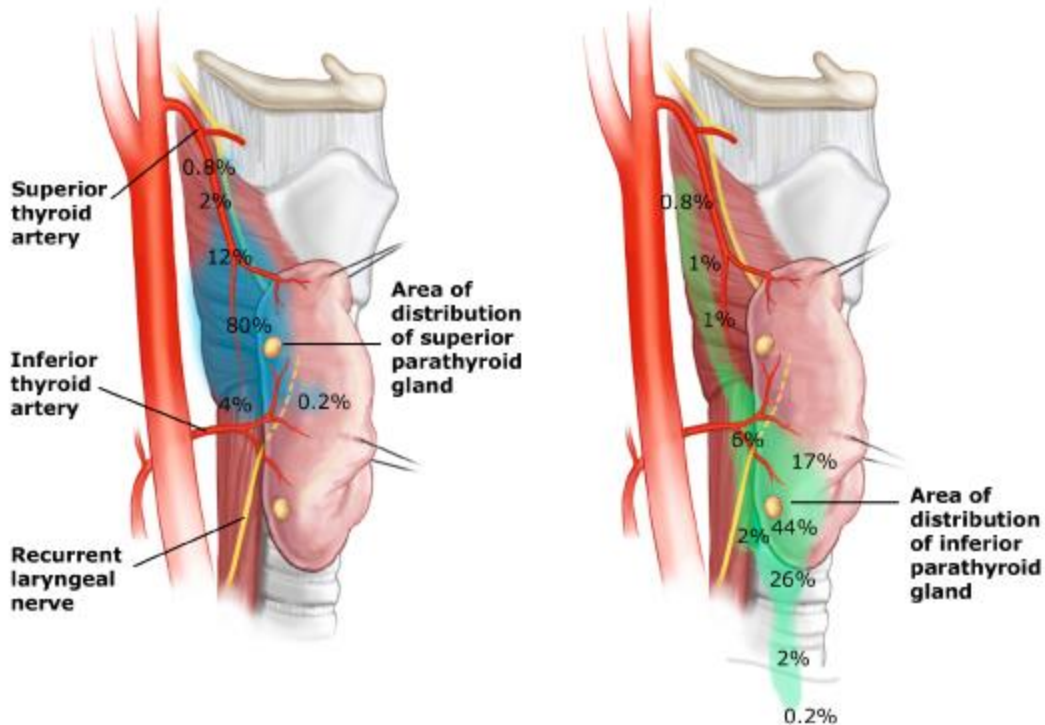
- Abundant optically clear cytoplasm and sharply defined cell membranes.
- Chief cells with excessive cytoplasmic glycogen



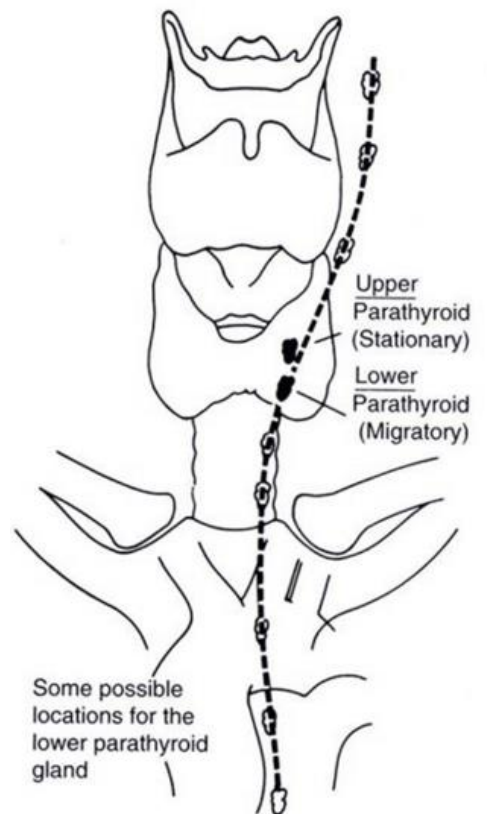
Parathyroid Anatomy:

- Most of patients (84%) have 4 Parathyroid glands (2 superior and 2 inferior glands).
 - o Additional glands are found in 13% of patients.
 - o Only three glands are found in $\leq 3\%$ of patients..
- Light yellow to reddish-brown in color.
- Varies in shape:
 - o Oval, bean shaped or spherical (83%).
 - o Elongated (11%).
 - o Bi-lobated (5%).
 - o Multilobated (1%).
- There is significant variability in the position of the glands.
- **Superior Parathyroid glands:**
 - o More constant than inferior glands.
 - o Symmetric in 80% of cases.
 - o Locations:
 - 1. Posterio-lateral to Superior thyroid lobe.**
 - Posterior to RLN (Always).
 - 1-2 cm cranial to intersection of RLN with Inferior Thyroid Artery.
 - **Most common location (85%).**
 - 2. Undescended.**
 - 3. Parapharyngeal.**
 - 4. Retropharyngeal.**
 - 5. Retrotracheal.**
- **Inferior Parathyroid glands:**
 - o More variations in anatomical position than inferior glands.
 - o Symmetric in 70% of cases.
 - o Locations:
 - 1. Posterio-lateral to Inferior thyroid lobe:**
 - Anterior to RLN (Always).
 - **Most common location (60%).**
 - 2. Thyrothymic ligament.**
 - 3. Anterior mediastinum.**
 - 4. Above Superior Parathyroid glands.**

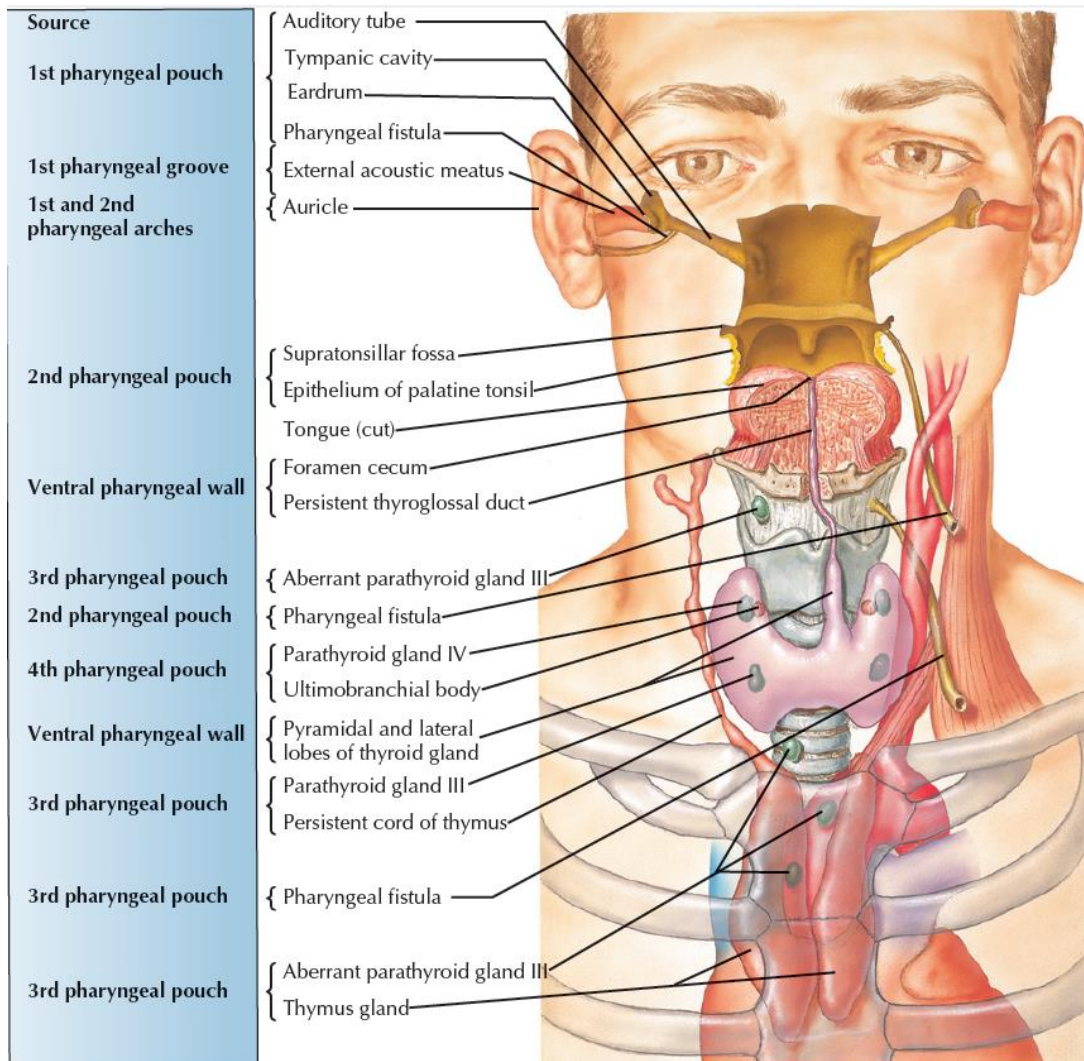




- **Ectopic parathyroid glands:**
- Undescended glands are ectopic parathyroid gland that fails to have full migration during normal development.
- May be one of the four parathyroid glands or a supernumerary gland.
- Locations:
 - **Ectopic Superior Parathyroid Glands:**
 - Paraesophageal.
 - Retroesophageal.
 - At Piriform sinus (Undescended).
 - Intrathyroidal.
 - **Ectopic Inferior Parathyroid Glands:**
 - Carotid bulb (Undescended).
 - Subcapsular.
 - Intrathyroidal.
 - Thymus.
 - Anterior Superior Mediastinum.



- **Supernumerary Parathyroid Glands:**
- More than 4 parathyroid glands.
 - o Range from 5 to 8 in number.
- 2.5 - 15% of individuals.
- Majority are small, rudimentary, or divided.
- May be responsible for persistent hyperparathyroidism after failed parathyroid exploration.
 - o Especially in patients with Secondary Hyperparathyroidism or hyperparathyroidism associated with familial syndromes.
- Locations:
 - o Anywhere from behind Thyroid down to and including within Thymus.
 - o Most common location is within Thymus or in relation to Thyrothymic ligament.
 - 2/3 of cases.
 - o Remaining are located in vicinity of mid-thyroid lobe between two other glands.



- **Blood Supply:**

- 80% of parathyroid glands have single arterial supply.
- 15% of parathyroid glands have dual arterial supply.
- 5% of parathyroid glands have multiple arterial supply.

- **Superior Parathyroid glands:**

1. **Inferior Thyroid Artery.**

- Mainly in most patients.

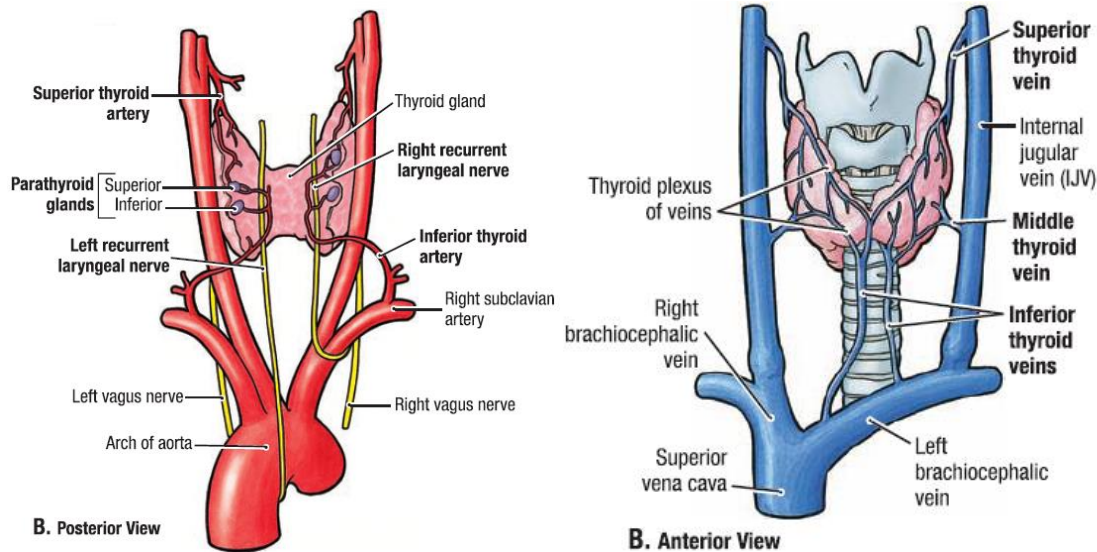
2. **Superior Thyroid Artery:**

- 15 - 20% of patients.

- **Inferior Parathyroid glands:**

1. **Inferior Thyroid Artery.**

- Mainly in most patients.
- Preserved intra-op by:
 - Gentle medial mobilization of Parathyroid rim from Thyroid capsule and preservation of lateral arteriole going to Parathyroid gland.
 - Ligation of branches of inferior thyroid artery close to thyroid and medial to RLN.



- **Venous Drainage:**

1. **Superior Thyroid Vein:**

- Ascends along Superior Thyroid Artery.
- Drains into Internal Jugular Vein.

2. **Middle Thyroid Vein:**

- Follows a direct course laterally to drain into Internal Jugular Vein.

3. **Inferior Thyroid Vein:**

- Drains into Left Brachiocephalic Vein.

Calcium Homeostasis:

- More than 99% of total body Calcium is stored in bone in form of Phosphate and hydroxide salts, predominantly as Hydroxyapatite.
- Normally, a very small portion of Calcium is available for exchange in the serum.
- Calcium Homeostasis is a complex process involving 4 components:
 1. Serum Calcium.
 2. Serum Phosphate.
 3. 1,25-dihydroxyvitamin D-3
 4. Parathyroid Hormone (PTH).

- **Parathyroid Hormone (PTH):**
- Secreted by Parathyroid glands in response to:
 - ↓ Serum Calcium.
 - ↑ Serum Phosphate.
- Feedback inhibition of PTH release in response to:
 - ↑ Serum Calcium.
 - ↓ Serum Phosphate.

- Actions of PTH:

1. Kidney:

- ↑ Calcium Reabsorption.
- ↑ Phosphate Excretion.
- Activation of Vitamin D.

• Intestine:

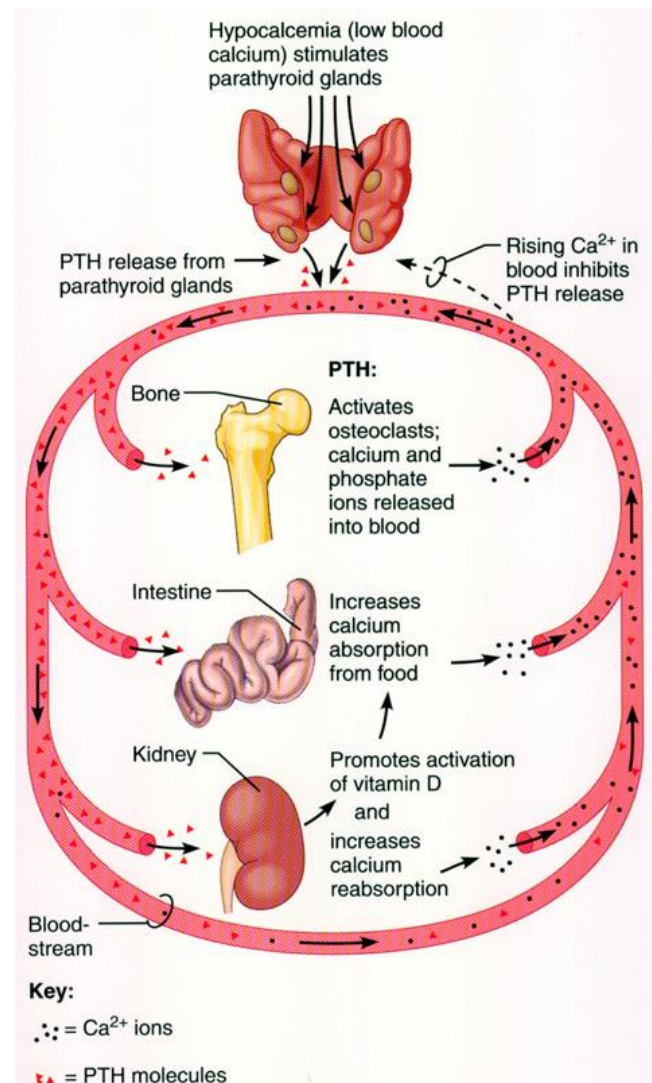
- ↑ Calcium Absorption
- ↑ Phosphate Absorption.

2. Bone:

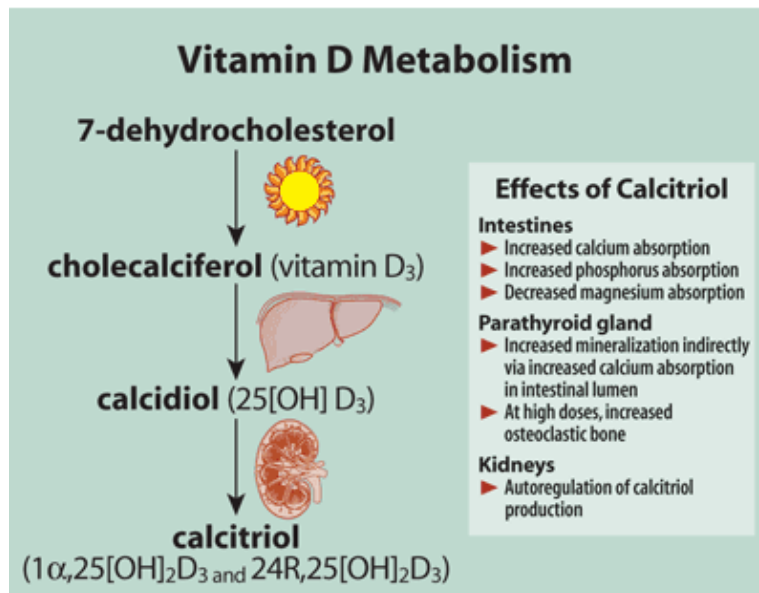
- ↑ Calcium Resorption.
- ↑ Phosphate Resorption.

- Effects of PTH:

1. ↑ Serum Calcium.
2. ↓ Serum Phosphate.
3. ↑ Urine Phosphate.



- **Vitamin D:**
- Exerts a much slower regulatory effect on calcium balance.
- Vitamin D3 (Cholecalciferol):
 - o Formed in Skin after exposure of 7-dehydrocholesterol to ultraviolet light.
- Calcidiol (25-hydroxyvitamin-D3):
 - o Formed by 25-hydroxylation of Vitamin D3 (Cholecalciferol) in Liver.
- Calcitriol (1,25-dihydroxyvitamin-D3):
 - o Formed by 1-hydroxylation in the kidney.
 - o Active form of Vitamin D.
 - o Activation is stimulated by PTH.

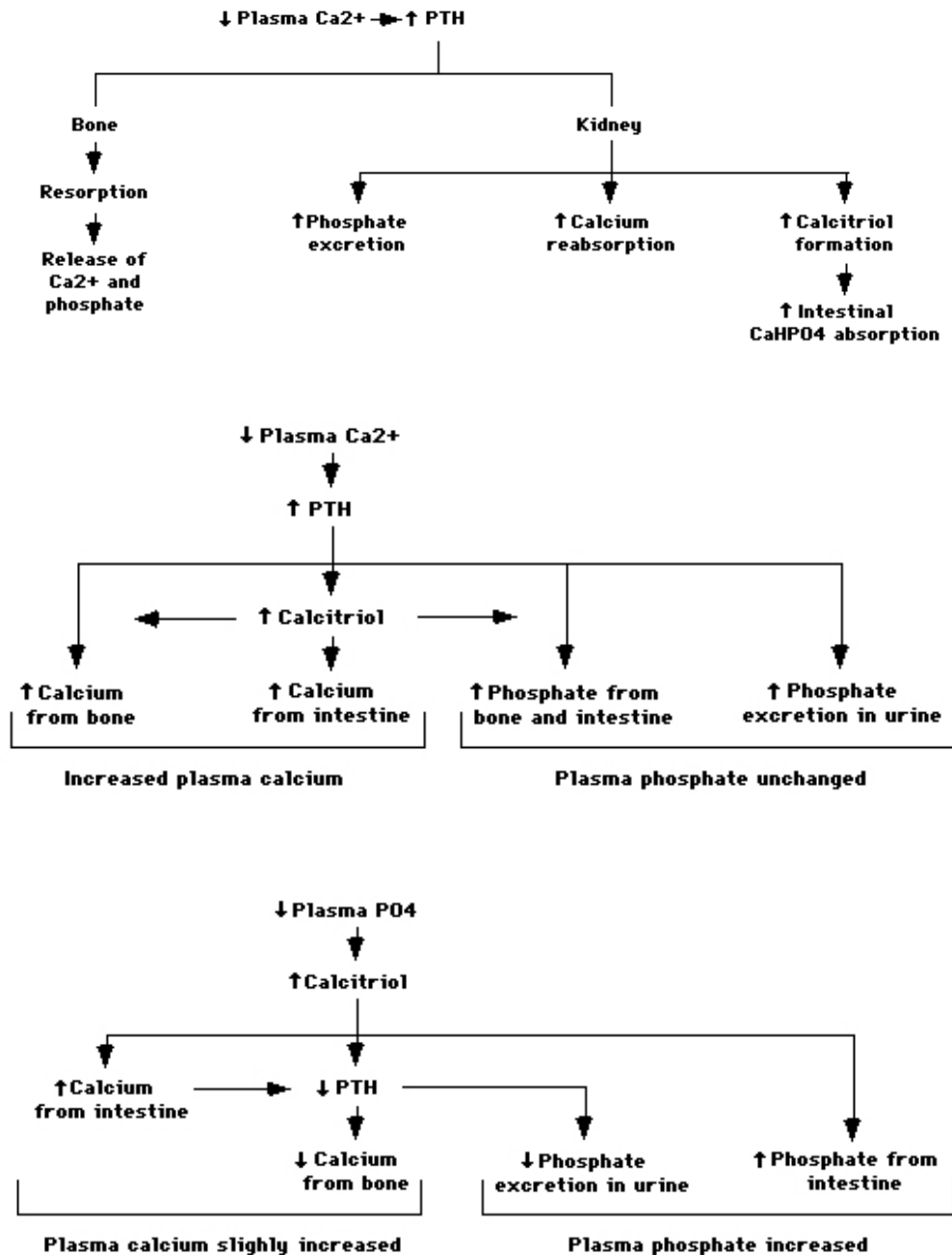


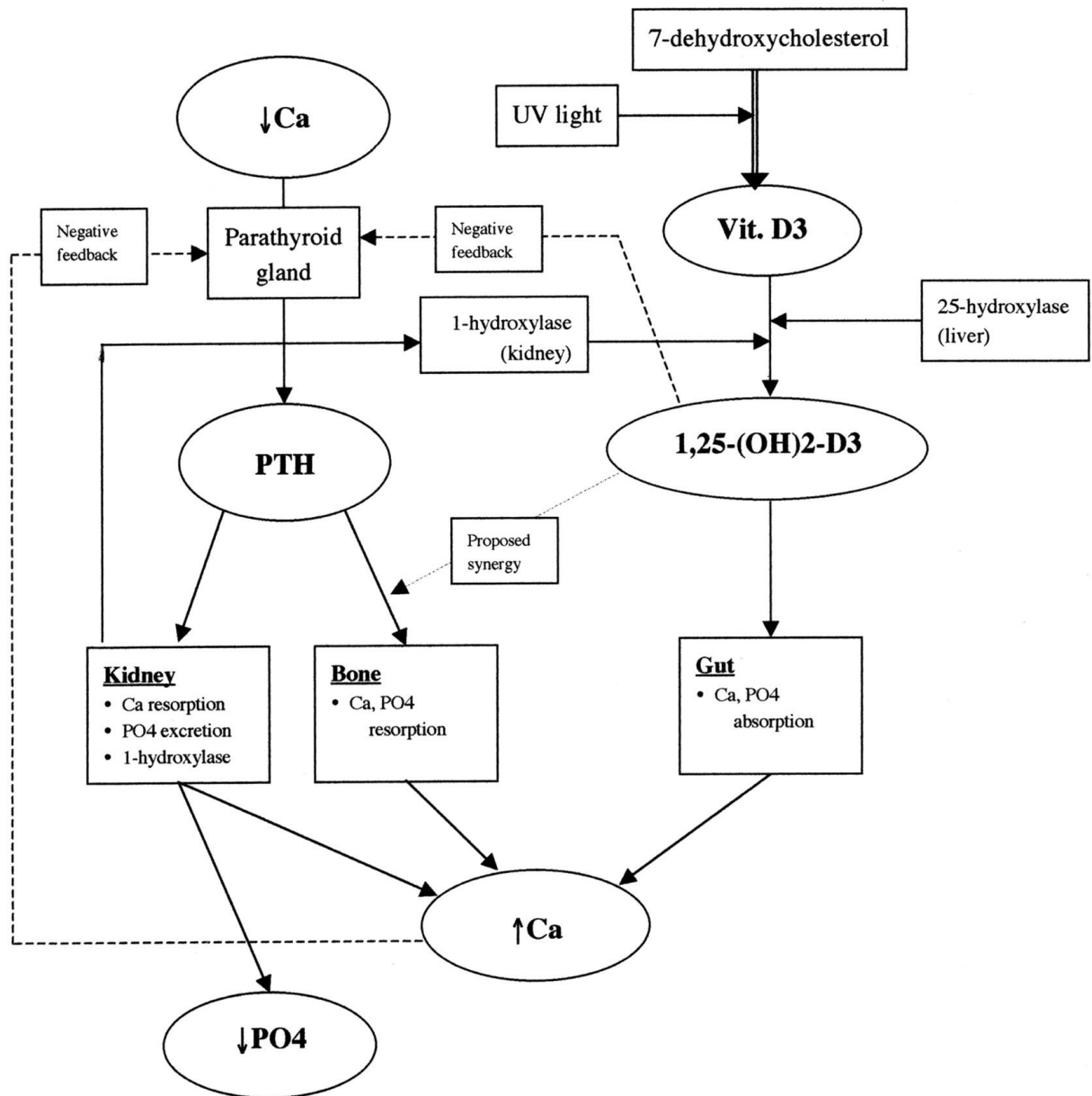
- Actions of Calcitriol (1,25-dihydroxyvitamin-D3):
 - 1. Intestine:**
 - o ↑ Calcium Reabsorption.
 - o ↑ Phosphate Reabsorption.
 - 2. Bone:**
 - o ↑ Calcium Resorption.
 - o ↑ Phosphate Resorption.
- Effects of Calcitriol (1,25-dihydroxyvitamin-D3):
 - 1. ↑ Serum Calcium.**
 - 2. ↑ Serum Phosphate.**

- **Calcitonin:**
- Produced by Parafollicular cells (C-cells) of Thyroid gland in response to $\uparrow$ free serum Calcium.
- Opposes actions of PTH.
- NOT important in normal calcium homeostasis.
- Actions of Calcitonin:

1. Bone:

- $\downarrow$ Calcium Resorption.





- **Diseases of Parathyroid Glands:**

- **Primary Hyperparathyroidism: ($\uparrow$ PTH $\uparrow$ Ca $\downarrow$ Phosphate)**

- Autonomous hypersecretion of one or more parathyroid glands.
- Unregulated overproduction of PTH results in abnormal calcium homeostasis:
 - o $\uparrow$ serum PTH
 - o $\uparrow$ serum Ca
 - o $\downarrow$ serum Phosphate
- Female-to-male ratio of 2:1.

- **Causes:**

1. Parathyroid Adenoma (85%):

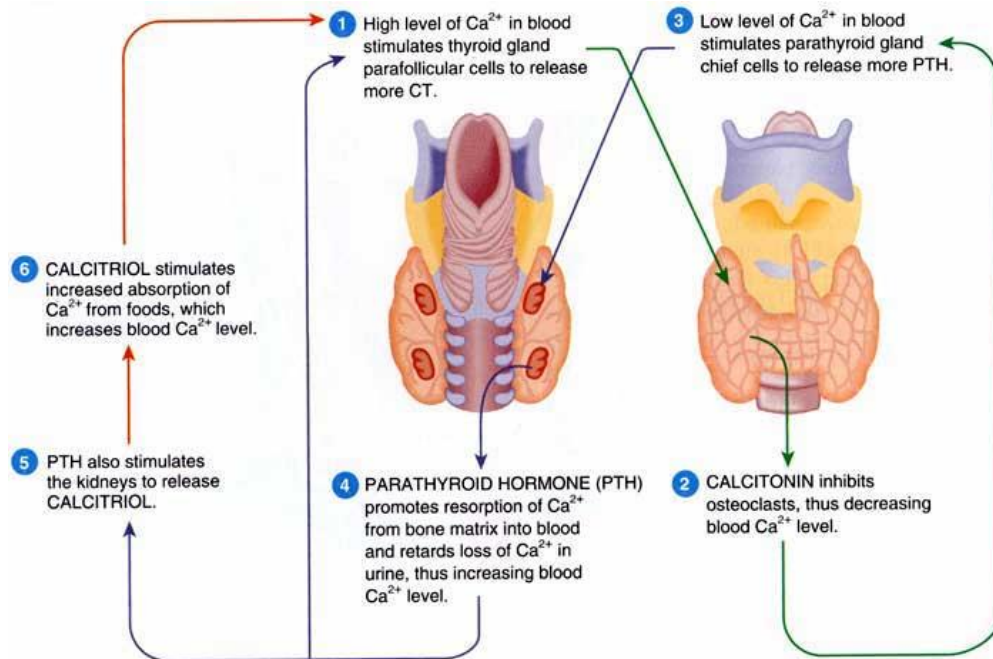
- Most common cause of Primary HPT.
- Benign and usually single.
 - 2-4% have multiple adenomas.
- More common in inferior glands.
- Risk factors:
 - Idiopathic (most common)
 - Neck Radiation
 - Chronic Lithium therapy
- Treated with Focused parathyroidectomy.

2. Parathyroid Hyperplasia (15%):

- Proliferation of parenchymal cells leading to an increase in gland weight in multiple parathyroid glands in the absence of a known stimulus for PTH secretion.
- Affecting all 4 glands.
- Associated with:
 - MEN I & IIA
- Treated with Subtotal parathyroidectomy.

3. Parathyroid Carcinoma (0.5-4%)

- Rare.
- Differentiation between adenoma and carcinoma is difficult clinically.
- Features that suggest Parathyroid cancer:
 1. Ca level more than 3.5 mmol.
 2. Increase PTH level 4 times more than normal.
 3. Palpable neck mass more than 2cm.
 4. Voice change.
- Treated with Parathyroidectomy + Hemithyroidectomy + central ND.

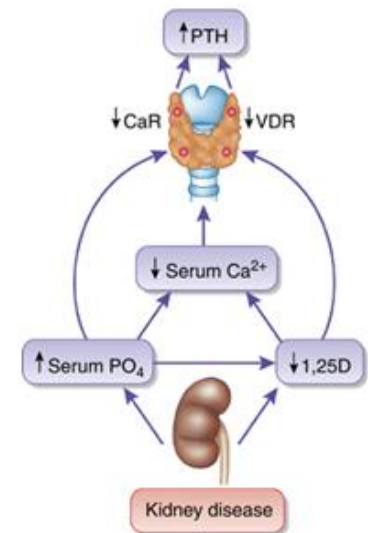


- **Secondary Hyperparathyroidism: ($\uparrow$ Phosphate $\downarrow$ Ca $\uparrow$ PTH)**

- Compensatory parathyroid hyperplasia secondary to chronic abnormal stimulus for its production (Hypocalcaemia).

- Causes:

- 1. Chronic Renal Failure (Most common)**
 - o Overproduction of PTH in response to hyperphosphatemia, hypocalcemia, and impaired 1,25-dihydroxyvitamin D production by diseased kidneys.
- 2. Chronic vit.D deficiency**
- 3. Multiple Myeloma**
- 4. Osteogenesis imperfecta**
- 5. Paget's disease**
- 6. Bone metastasis**
- 7. Pituitary Adenoma**



- **Tertiary Hyperparathyroidism: ($\uparrow\uparrow$ PTH N/ $\downarrow$ Ca)**

- Autonomous or irrepressible PTH production (Parathyroid hyperplasia from secondary parathyroidism, persistent hyperfunction despite correction).

- Persistent elevated serum PTH (normal or low calcium).

- Causes:

- 1. After successful renal transplantation.**
- 2. After long-standing hypocalcemia of any other cause is corrected.**

- **Clinical Presentation of HPT:**
"Bones, Stones, Abdominal groans, and Psychic moans"
- Skeletal manifestations:
 1. Bone and joint pain
 2. Osteopenia
 3. Osteoporosis
 4. Pseudogout
 5. Chondrocalcinosis
 6. Osteitis fibrosa cystica
 7. Renal osteodystrophy
- Renal manifestations:
 1. Polyuria
 2. Renal stones
 3. Hypercalciuria
 4. Nephrocalcinosis
- Gastrointestinal manifestations:
 1. Constipation
 2. Nausea and vomiting
 3. Abdominal pain
 4. Peptic ulcer disease
 5. Acute pancreatitis
- Neuromuscular and psychologic manifestations:
 1. Proximal myopathy
 2. Weakness and easy fatigability
 3. Depression
- Cardiovascular manifestations:
 1. Hypertension
 2. Bradycardia
 3. Shortened QT interval
 4. Left ventricular hypertrophy
- **Physical examination findings are usually noncontributory.**

- **Diagnosis of Hyperparathyroidism:**

- Lab works:

1. Serum Electrolytes:

- Primary HPT:
 - ↑ Ionized calcium.
 - ↓ Phosphate.
- Secondary HPT:
 - ↓ Ionized calcium.
 - ↑ Phosphate.
- Tertiary HPT:
 - ↑ Ionized calcium.
 - ↑ Phosphate.

2. ↑ Intact PTH levels:

- Allows differentiation of primary hyperparathyroidism from hypercalcemia of malignancy (tumors secrete a larger protein).

3. Alkaline phosphatase:

- Suggests bone disease.

4. BUN/Creatinine:

- Renal function.

5. 24-hour urine calcium:

- Allows calculation of calcium-creatinine clearance to distinguish primary hyperparathyroidism from Familial hypocalciuric hypercalcemia.

6. TFT, ACE levels:

- Sarcoidosis.

7. Serum prolactin and gastrin and urine catecholamines and metabolites:

- Evaluate for MEN syndromes.

	Primary	Secondary	Tertiary	FHH
Calcium	↑	↓ or N	↑	↑
PTH	↑	↑	↑	↑
Phosphate	↓	↑	↑	
Urine Calcium	↑	↓		↓

TABLE 4–4. Causes of Hypercalcemia: CHIMPANZEES Method

Calcium: exogenous

Hyperparathyroidism

Immobility

Metastasis to bone

Paget's disease

Addison's disease

Neoplasms: typically solid tumors (prostate, lung, colon, breast cancers)

Zollinger-Ellison syndrome (hypergastrinemia)

Excess: vitamin A or D, thiazides, lithium, estrogens, milk-alkali syndrome

Endocrine disorders: familial hypocalciuric hypercalcemia, hyperthyroidism, pheochromocytoma

Sarcoidosis: also other granulomatous diseases (tuberculosis and berylliosis)

- **Localization methods of Parathyroid Adenoma/Hyperplasia:**

1. Pre-op Localization
2. Intra-op Localization.

- **Pre-op localization (Radiological):**

- Imaging studies should be done after confirming diagnosis of primary Hyperparathyroidism on basis of biochemical findings.
- Preoperative localization studies are becoming the standard of care.
 - Enable minimally invasive surgical techniques.
 - Critical in patients with prior neck surgery or recurrent postoperative hypercalcemia.
- Most surgeons advocate for 2 concurrent examinations to definitively identify the site of disease.
- Almost all authors advocate for intraoperative parathyroid hormone level monitoring.
- The two most commonly used are ultrasound and technetium-99m-sestamibi scan.

1. **Ultrasound (Sensitivity 85%):**

- Homogenous hypoechoic and hypervascular.
- Less echogenic than thyroid.
- **Advantages:**
 - Non radiation emitting.
 - Non expensive.
- **Disadvantages:**
 - Highly operator dependent.
 - Low success rate in:
 - Intrathyroidal parathyroid lesions.
 - Deeply located lesions.
 - Ectopic especially mediastinal parathyroid lesions.
 - Associated thyroid nodules.

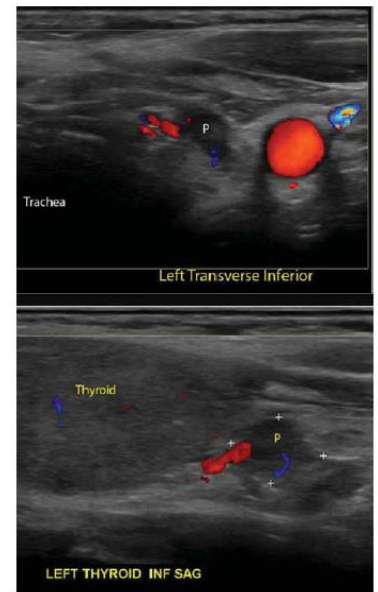
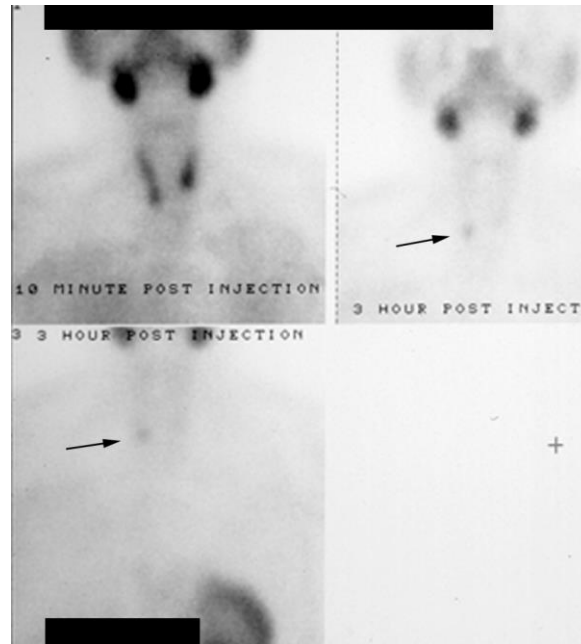


Figure 134.3 Ultrasound of left inferior parathyroid adenoma. **Top:** adenoma (P) between the trachea and carotid artery; **Bottom:** adenoma is at the inferior tip of the thyroid.

2. **Tc 99m SestaMIBI Scan (Sensitivity 90%):**

- **Current investigation of choice.**
- Tc 99m is taken up by both the thyroid and parathyroid adenoma but washes out of the thyroid faster than the parathyroid.
 - Parathyroid tissue has a large number of mitochondria in its oxyphil cells compared to thyroid tissue thus allowing Tc99m to enter mitochondria of parathyroid tissue more intensely.
- Early images at 20 minutes after injection are obtained, followed by delayed images typically at 2 hours "Double phase test".

- **If MIBI correlates with US results: 97% sensitivity**
- Decreased accuracy in:
 - Hyperplastic glands
 - Multinodular goiter
 - Hashimoto thyroiditis
 - Thyroid adenomas



3. CT scan (**Sensitivity 42-68%**):

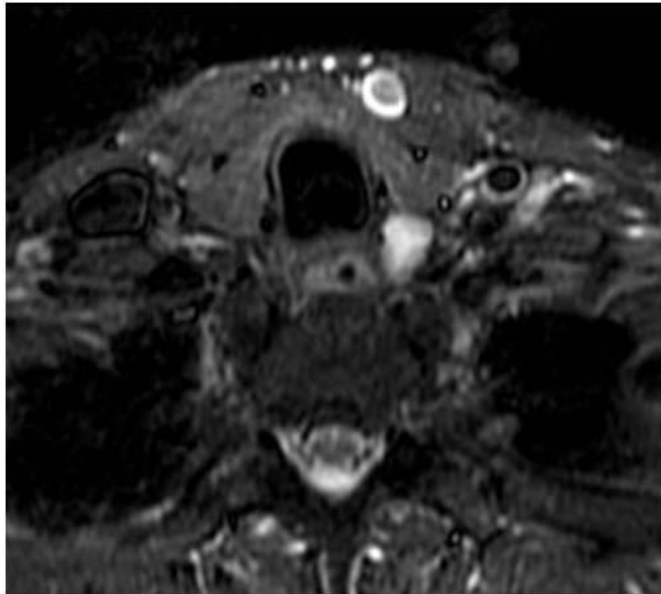
- Advantages:
 - Thin Cut CT Better in localizing Ectopic glands.
- Disadvantages:
 - Must use contrast.
 - Artifact w/ breathing/swallowing
 - Nodes and tortuous vessels look like adenomas



Figure 134.5 CT with contrast demonstrating a right paraesophageal adenoma (arrow).

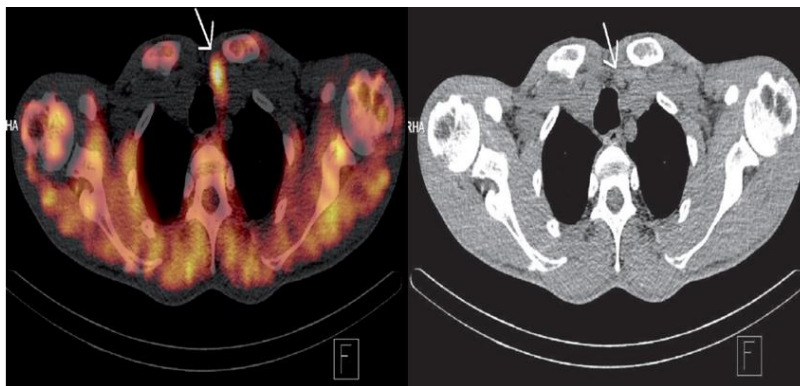
4. MRI (Sensitivity 57- 88%):

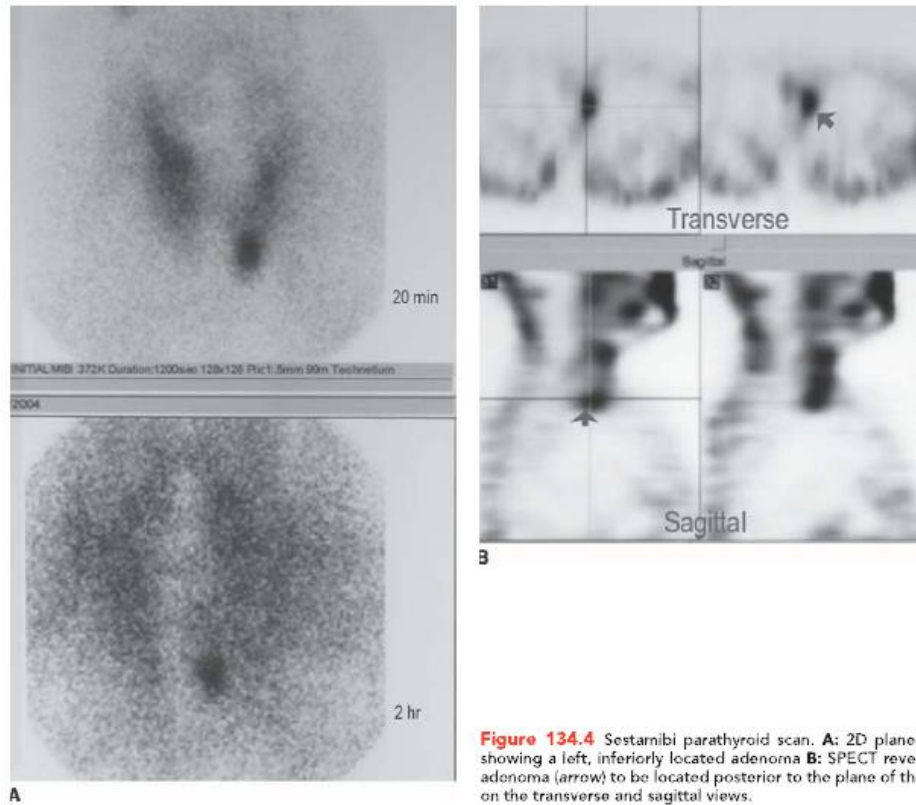
- Characteristics:
 - Bright on T2
 - Dark on T1
 - Enhance with gad
- Advantages:
 - Better than US for Ectopics
 - Little better than CT
- Disadvantages:
 - Adenomas look like nodes of ganglia



5. SPECT/CT (Sensitivity 95%):

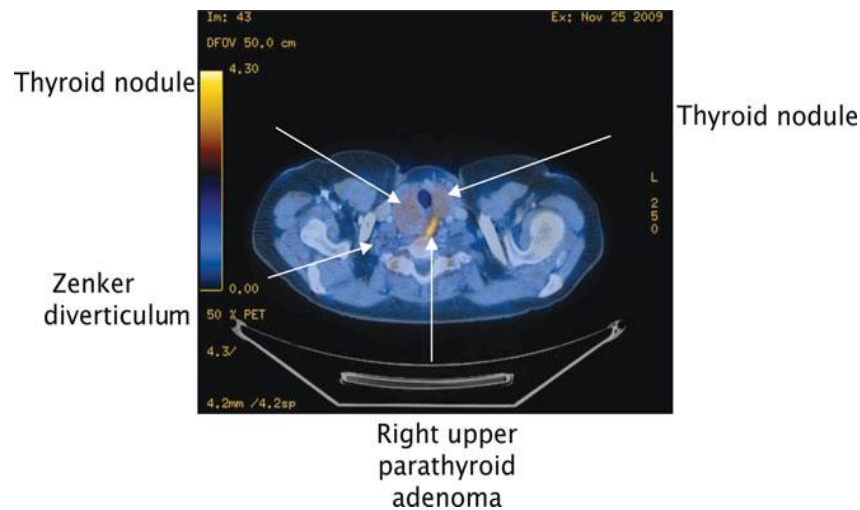
- Single photon emission CT
- When used with Sestamibi, has particular utility in the evaluation of:
 - Ectopic parathyroid adenomas, such as those located deep in the neck or in the mediastinum.





6. PET/CT (Sensitivity 100%):

- Highly sensitive tool.
- More helpful for lesion > 1.5 cm.
- Localize parathyroid adenomas even in atypical locations.
- Localize challenging anatomical positions in the neck, such as that occurs in concomitant multinodular goiter or reoperations.



- **Intra-op localization:**

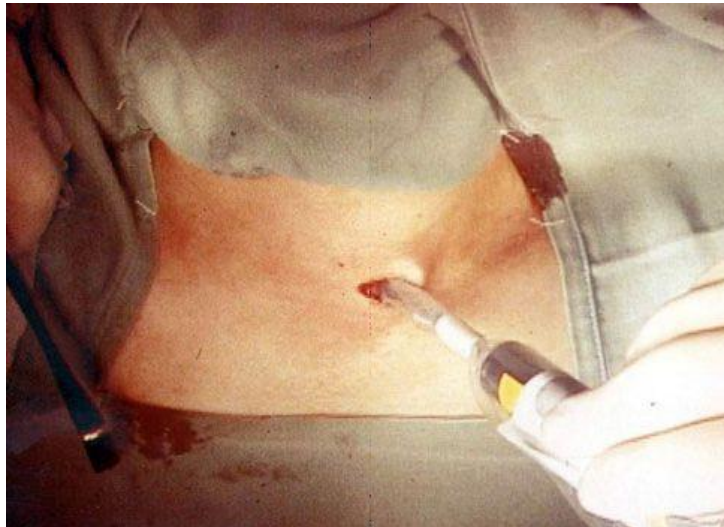
1. Intra-op PTH Assessment (IOPTH):

- Intact PTH has a very short half-life of 2-4 minute.
- Following successful removal of a single parathyroid adenoma, measurement of intact PTH level shortly after removal of an adenoma can be used to predict success of surgery accurately predicts presence or absence of other functioning glands.
- **Methods:**
 - Pre-excision level is drawn at the time of anesthetic induction.
 - Post-excision level is drawn 10 minutes after the adenoma is excised.
 - A drop > 50% and return to normal value from the pre-excision value is the most commonly used criteria for successful parathyroidectomy.
 - In hyperplasia each gland removal will correspond with decrease in PTH
- **Advantages:**
 - Avoid unnecessary exploration of multiple glands.
 - Reduce risk of missing multiglandular disease.
- **Disadvantages:**
 - False positive results up to 50% in some studies.
 - Post-excision level may fall below 50% of baseline yet one or more enlarged gland still remain.
 - Usually due to double adenoma when patient has 2 adenomas, one active and the other is smaller and not fully active.
 - After removal the active one, post-excision level will drop, then after sometime the other adenoma will start hyper-secreting.
 - False negative 2-13%

2. Gamma probe:

- Surgery is performed within 1.5-3 hours post-injection of Tc 99m sestamibi, so that the abnormal parathyroid gland is still radioactive.
- A hand-held gamma probe is used intra-op to guide the surgeon to the adenoma.
- All four quadrants of the neck are checked for radioactivity using the probe.
 - Difference of approximately 500 counts per second overlying the adenoma compared with the rest of the neck.
 - Once adenoma is excised, gamma counts decrease and equalizes in all four quadrants of the neck.

- Advantages:
 - Eliminate the possibility of inaccurate or false-positive scans.
 - Utilizes a small incision directed by gamma probe.
 - Improve cosmesis
 - Short operating time.
 - Allows procedure to be performed under LA.
- Disadvantages:
 - Require an experienced nuclear medicine physician and a close working relationship.
 - Timing of images is critical as there is a short operative window from 1.5 to 3 hours post-injection of the sestamibi.
 - A limited dissection does pose potential risk to the recurrent laryngeal nerve, as this is not always accurately identified.

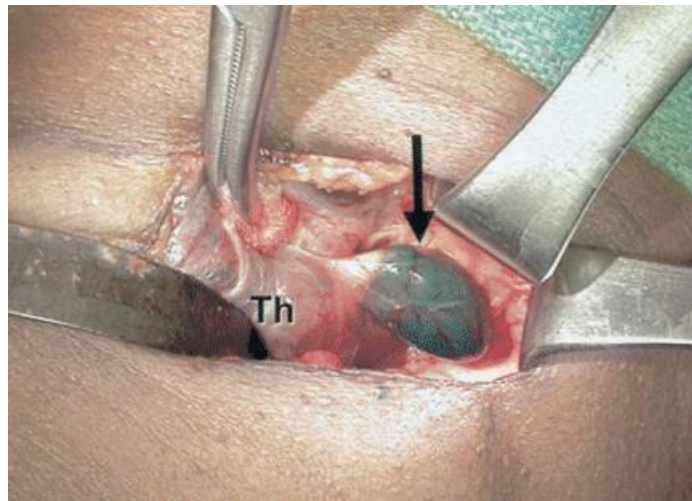


3. Intra-op Methylene blue infusion (IMBI):

- Methylene blue has the ability for selective staining of enlarged or hypercellular parathyroid tissue.
- Intensity of methylene blue staining can help identify all abnormal glands, since normal parathyroid tissue either does not stain or stains minimally.
- Dose is 4 mg/kg diluted in 250 to 500 cc of normal saline to be infused over 15-30 mins immediately after intubation.

▪ **Disadvantages:**

- Pseudo-cyanosis and pseudo-hypoxia.
 - Due to formation of low percent of methaemoglobin leading to erroneously low readings on pulse oximetry.
- Intensely blue-stained urine.
- Metabolic encephalopathy and neurotoxicity
 - Few reported cases with use of high dose of 7.5 mg/kg.
 - Mainly in patients on Anti-depressant drugs.
 - Complete recovery within 48 hours.



- **Management of HPT:**

- **Conservative management and Monitoring:**

- For asymptomatic patients with Primary HPT who do not undergo surgery.
- Recommendations:
 - Avoid factors that can aggravate hypercalcemia
 - Thiazide diuretic and lithium.
 - Volume depletion.
 - Prolonged bed rest or inactivity.
 - High calcium diet (>1000 mg/day).
 - Encourage physical activity to minimize bone resorption.
 - Encourage adequate hydration to minimize the risk of nephrolithiasis.
 - Maintain a moderate calcium intake (1000 mg/day).
 - Maintain moderate vitamin D intake (400-600 IU daily).
 - Serum calcium and Creatinine levels every 6 months.
 - Annual bone mineral density.

- **Medical management:**

- In primary HPT, it is reserve only for non-candidates of surgical management.
- Unlike primary HPT, medical management is mainstay of treatment for secondary HPT.
- Drugs:
 - **Elemental Calcium and Vit.D :**
 - Patients should maintain a daily intake of elemental calcium and vitamin D intake appropriate for their age and sex.
 - **Bisphosphonates:**
 - Alendronate.
 - Potent osteoclast inhibitor.
 - Useful in the long-term control of osteopenia associated with primary HPT.
 - Improves bone mineral density.
 - No effect on PTH, serum/urinary calcium.
 - **Calcimimetic Agents:**
 - Cinacalcet (Sensipar).
 - Act on parathyroid calcium sensing receptor (mimics calcium) to reduce PTH secretion.
 - Reduction and normalization of serum calcium.
 - **Reduces the need for parathyroidectomy by 90% in 2dary HPT.**

- **Surgery:**

- The only permanent and curative treatment for primary HPT.
- Indications of Parathyroidectomy in 1° HPT:
 - Patients with symptoms or complications of hyperparathyroidism.
 - Asymptomatic patients with:
 1. Serum Ca 1 mg/dL (0.25 mmol/L) > than upper limit of normal.
 2. Creatinine clearance < 60 ml/min.
 3. Reduced bone density (T score) < -2.5 OR previous fracture of fragility
 4. Age < 50 years
 5. Medical surveillance not desirable or possible
- Indications of Parathyroidectomy in 2° HPT:
 - Severe hyperparathyroidism associated with hypercalcemia and/or hyperphosphatemia that is refractory to medical therapy.

**TABLE
134.1****INDICATIONS FOR
PARATHYROIDECTOMY IN
ASYMPTOMATIC PATIENTS**

2002	2009
Ca ²⁺ > 1 mg/dL above upper limit of normal	Ca ²⁺ > 1 mg/dL above upper limit of normal
Creatinine clearance reduced by 30%	Creatinine clearance <60 mL/min
Reduction in bone density (hip, L Spine, Radius) T-score < -2.5	Reduction in bone density (hip, L Spine, Radius) T-score < -2.5; Previous fracture or bone fragility
Under 50 y of age	Under 50 y of age
Medical surveillance not feasible (e.g., coexistent illness) or not possible (e.g., poor follow-up)	Medical surveillance not feasible (e.g., coexistent illness) or not possible (e.g., poor follow-up)
24-h urine calcium >400 mg	

- **Surgical Approaches:**

1. Minimally invasive Parathyroidectomy:

- Standard procedure for a well-localized solitary adenoma.
- Adenoma is removed through a limited incision with direct exposure of the previously imaged parathyroid adenoma.
- Patients can be discharged on the same day, since their risk of hypocalcemia is low.



2. Bilateral neck exploration:

- Indicated in:
 - 1.** Failed pre-op localization of Parathyroid Adenoma.
 - 2.** Parathyroid Hyperplasia.
 - 3.** Secondary and Tertiary HPT.
- Methods:
 - Fully exploration of the neck.
 - Search for each gland systematically moving from
 - one quadrant to next in the expected locations of parathyroid glands.
 - If superior gland is missing, Paraesophageal and Retroesophageal areas should be explored.
 - If inferior parathyroid is missing, Thymus and Superior mediastinum should be explored.

3. Subtotal Parathyroidectomy (3 and ½):

- Indicated in:
 - 1.** Parathyroid Hyperplasia.
- Contraindicated in Secondary and Tertiary HPT.
 - Hypocalcaemia secondary to renal failure is a potent stimulus for hyperplasia of glandular tissue left behind which might needs surgical exploration.
- Methods:
 - Bilateral neck exploration as mentioned.
 - Identify all 4 parathyroid glands.
 - Biopsy part of the gland for histological confirmation.
 - Most normal-looking of residual glands should be selected for preservation either in situ or for re-implantation.
 - 30-40 g of parathyroid tissue is incised into small pieces to be reimplanted into the brachioradialis muscle of nondominant hand.
 - Reimplantation site should be marked with hemoclip and/or colored permanent suture for identification if the need arises to someday excise hyperplastic re-implanted tissue.
 - Excision of rest of the glands can be performed.
 - Some of excised parathyroid glands can be cryopreserved for possible future use.

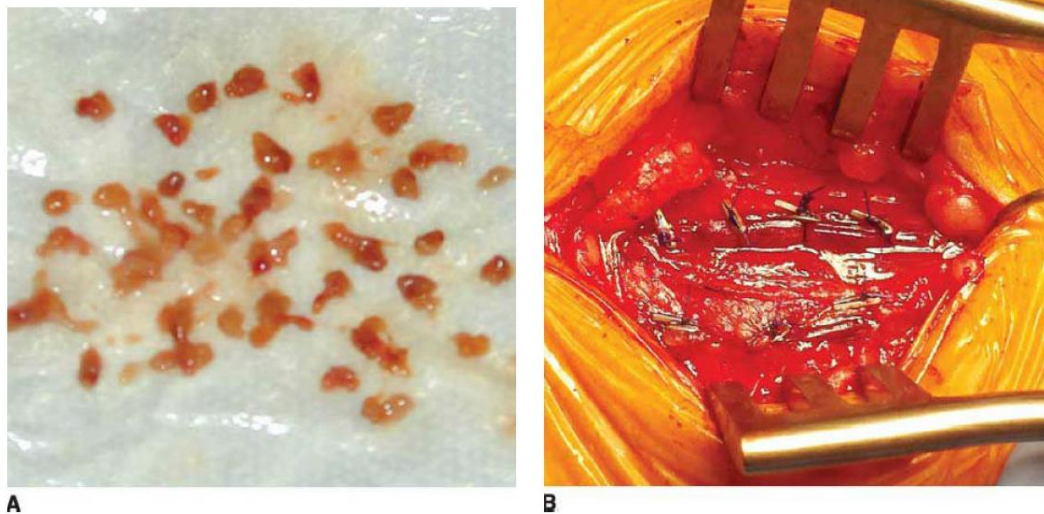


Figure 134.7 **A:** Parathyroid gland cut and prepared for reimplantation. **B:** Reimplanted sites in brachioradialis muscle marked with hemoclips.

4. Total Parathyroidectomy:

- Indicated in:

1. Secondary and Tertiary HPT.
2. Fully exploration of the neck.
 - Search for each gland systematically moving from one quadrant to next in the expected locations of parathyroid glands.
 - If superior gland is missing, Paraesophageal and Retroesophageal areas should be explored.
 - If inferior parathyroid is missing, Thymus and Superior mediastinum should be explored.

- **Complications of Parathyroidectomy:**

1. **Post-op Hypocalcemia:**

- Postoperative Hypocalcemia due to relative hypoparathyroidism, leads to reductions in bone reabsorption and increases in bone formation, leading to an increased influx of calcium into bone and increased calcium excretion.
- Causes:
 - Injury and devascularization of remaining glands.
 - Decreased PTH production from the remaining suppressed parathyroids.
 - Transient because the degree of bone disease is typically mild and normal parathyroid tissue recovers function quickly (usually within one week), even after long-term suppression.

- Hungry bones syndrome:
 - Severe and prolonged postoperative hypocalcemia despite normal or even elevated levels of PTH.
 - Occurs in patients who developed bone disease preoperatively due to a chronic increase in bone resorption induced by high PTH.
- Clinical picture:
 - Parenthesis of hands, feet, or perioral region as well as muscle spasms.
 - Severe hypocalcemia can induce muscle cramping, tetany, seizures, and cardiac arrhythmias.
 - Early signs of hypocalcemia include:
 - **Chovstek sign:**
 - Tapping preauricularly elicits twitching of the ipsilateral face.
 - Found in 10% of normal people.
 - **Trussou sign:**
 - Inflation of a blood pressure cuff elicits cramping in the extremity. Severe hypocalcemia.
 - More sensitive.
- Monitoring:
 - Patients undergoing bilateral exploration should be admitted for post-op overnight observation.
 - Calcium levels in morning of 1st day post-op and 6 hours after that since the calcium may not drop until well beyond 24 hours after surgery.
 - Discharge the patient without supplement if:
 - Post-op PTH is greater than 16 pg/mL.
 - Sequential calcium levels are stable or demonstrate an upward trend.
- Treatment:
 - If hypocalcaemia is severe or persistent, may require long-term calcium and vitamin D supplementation.

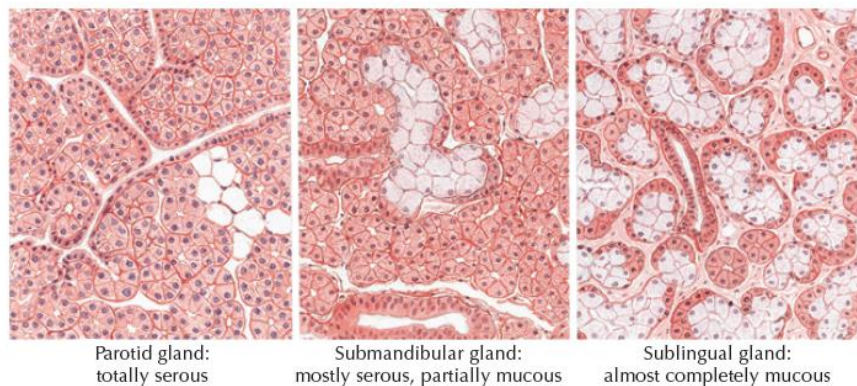
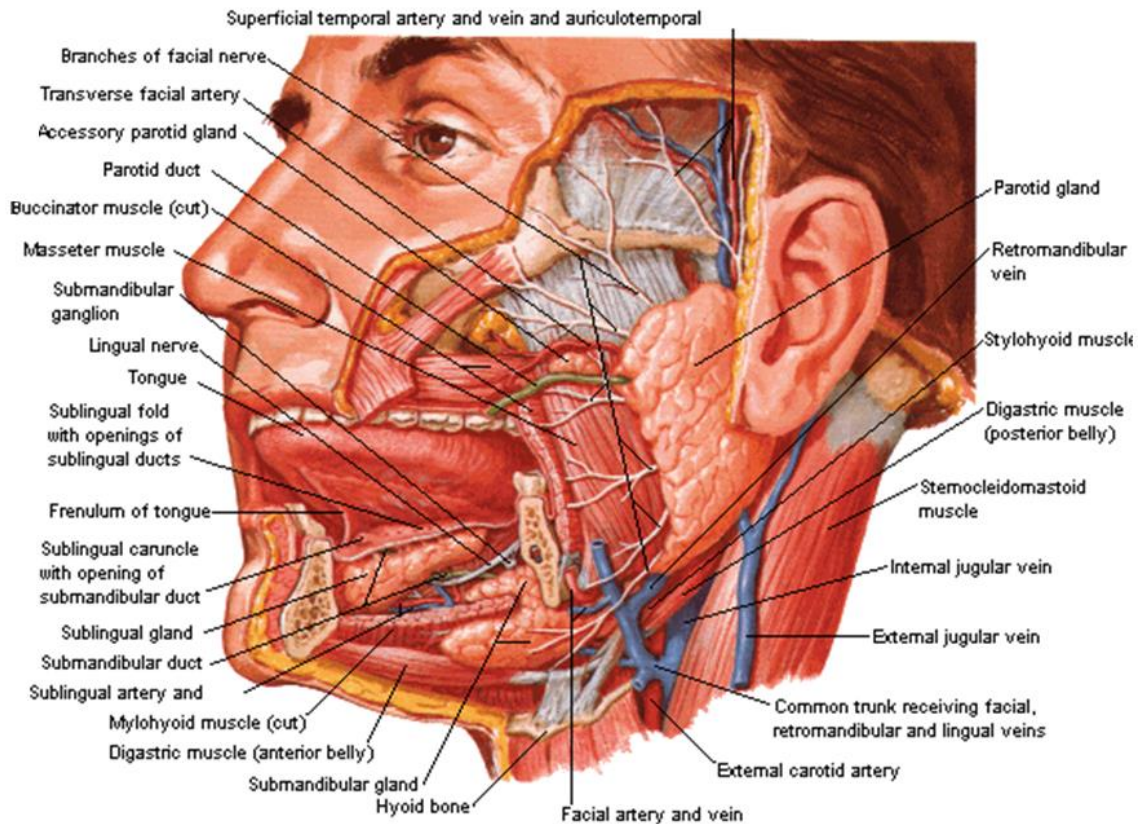
2. Persistent Hypercalcemia:

- Most commonly from missed adenoma.
 - The most commonly missed location is the posterior mediastinum.
- Also from supernumerary gland, second adenoma, failed recognition of parathyroid hyperplasia, incorrect diagnosis and residual adenoma.

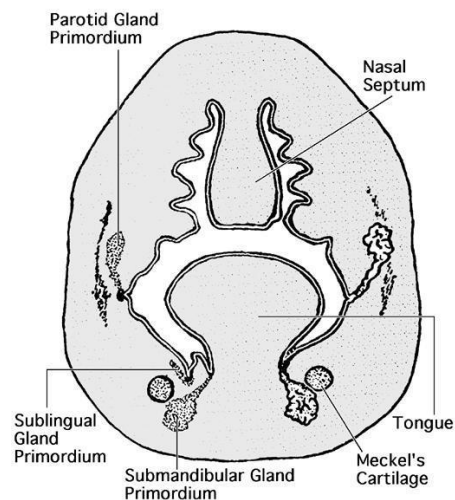
3. Nerve Injury and Hematoma

- **Salivary Glands:**

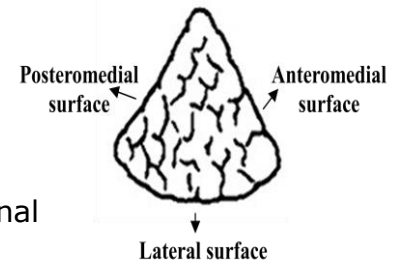
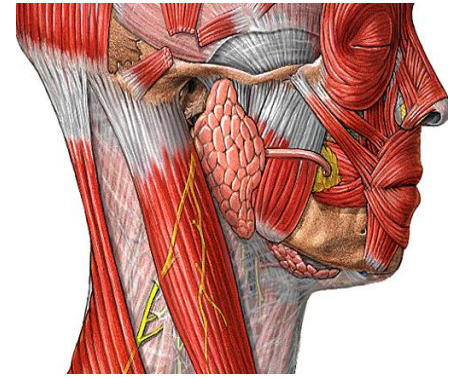
- Exocrine glands that produce saliva.
- Major salivary glands (produce majority of saliva):
 1. Parotid gland.
 2. Submandibular gland.
 3. Sublingual gland.
- Multiple minor salivary glands (600-1,000 glands) line the oral cavity and oropharynx, contributing a small portion of total salivary production.



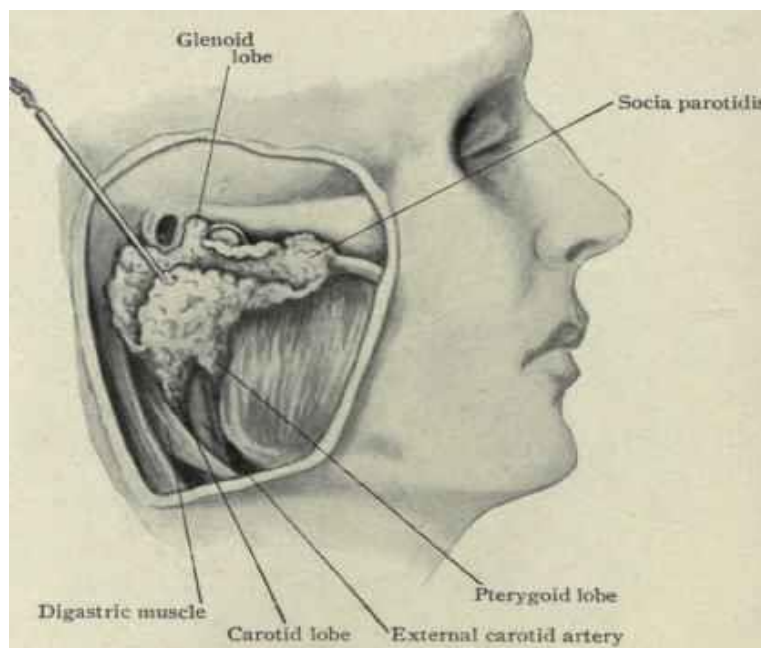
- **Embryology of Salivary glands:**
- Derived from **1st Pharyngeal POUCH.**
- **Parotid gland:**
 - **1st to develop.**
 - Develops during **5th** week.
 - From **Oral Ectoderm**
 - Growing in a posterior direction as Facial nerve advances Anteriorly.
 - Fully developed Parotid surrounds **CN-VII.**
 - **Last to be encapsulated** allowing entrapment of lymphoid tissue within the Parotid parenchyma.
- **Submandibular gland:**
 - **2nd to develop.**
 - Develops during **6th** week.
 - From **Oral Endoderm.**
 - No intraparenchymal lymph nodes
- **Sublingual gland:**
 - **3rd to develop.**
 - Develops during **7-8th** week.
 - From **Oral Endoderm.**
 - No intraparenchymal lymph nodes
- **Minor Salivary glands:**
 - **Last to develop.**
 - Develop during **3rd month.**
 - From **Oral Ectoderm** and **Nasopharyngeal Endoderm.**
- Sympathetic nerve stimulation leads to Acinar differentiation.
- Parasympathetic stimulation leads to overall glandular growth.



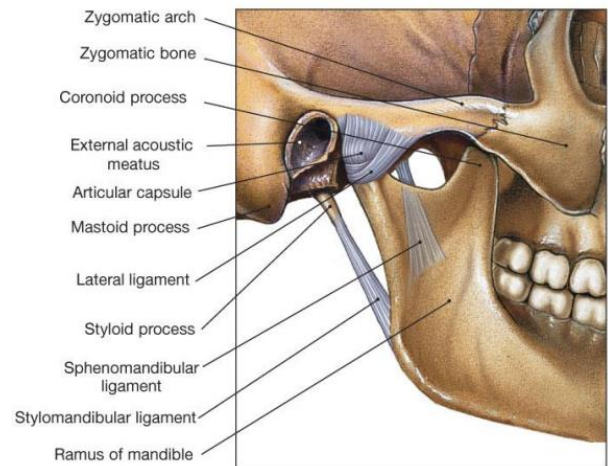
- **Parotid Gland:**
- Largest salivary gland.
- Irregular, wedge shaped, and unilobular.
- Located in Parotid compartment:
 - o between Ramus of Mandible and the External auditory canal and mastoid tip.
- Overlies Masseter muscle (Anteriorly) and SCM muscle (Posteriorly).
- Having 3 surfaces:
 1. **Lateral (Superficial).**
 2. **Anteromedial**
 3. **Posteromedial**
- Having 5 processes (3 superficial and 2 deep), thus making it very difficult to surgically removal all parotid tissue.



1. **Glenoid Process :**
 - Extends upward behind TMJ in front of External auditory meatus.
2. **Facial Process :**
 - Extends Anteriorly onto Masseter muscle.
3. **Accessory Process:**
 - Small part of Facial process lying along Parotid duct.
4. **Pterygoid Process:**
 - Extends forward from deeper part Lies between Medial pterygoid muscle and ramus of mandible.
5. **Carotid Process :**
 - that lies posterior to the external carotid artery



- Parotid gland is entirely **Serous** in secretion.
- **Stylomandibular Ligament:**
 - Formed by facial envelope between Styloid process and Mandible.
 - Separates Parotid gland from Submandibular gland.



- **Parotid Region:**

- Triangular space.

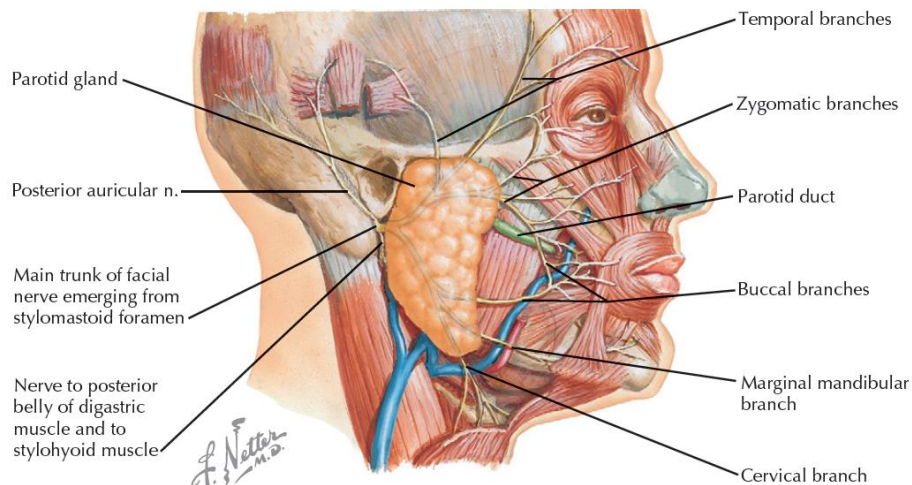
- **Boundaries:**

1. Superior border: Zygoma.

2. Inferior border: Styloid Process, Styloid Process musculature, Internal Carotid Artery, Jugular Veins.

3. Anterior border: Diagonal line drawn from the Zygomatic root to the EAC

4. Posterior border: External Auditory Canal



- **Contents of Parotid Region:**

- Parotid gland
- Facial nerve CN-VII
- Sensory and autonomic nerves
- External Carotid artery
- Retromandibular vein
- Parotid lymphatics.

- **80%** of Parotid gland overlies **Masseter and Mandible**.
- Retromandibular portion:
 - o Forms the remaining **20%** of Parotid gland.
 - o Extends medially through Stylomandibular tunnel formed by the posterior edge of the mandibular ramus (ventral), SCM and posterior belly of the Digastric (dorsal), and the stylomandibular ligament (deep and dorsal).
 - o Stylomandibular ligament separates Parotid from the Submandibular gland.
 - o Lies in **Prestyloid Compartment** of the Parapharyngeal space.
 - o Deep parotid tumor can push Tonsillar fossa and soft palate anteromedially.
 - o The isthmus of the Parotid gland runs between the mandibular ramus and the posterior belly of the Digastric to connect the retromandibular portion to the remainder of the gland.
- **Parapharyngeal space:**
- Inverted pyramid with its base at the skull base and its apex at the Greater Cornu of the Hyoid bone.
- Boundaries:
 - o Medially: Pharyngeal wall
 - o Laterally: Mandibular ramus and Medial Pterygoid muscle.
- Divided by Fascia between Styloid Process and Tensor Veli Palatini (Medial Pterygoid plate) into:
 1. **Pre-Styloid compartment:**
 - o Deep lobe parotid tumors (Dumbbell tumors) occupy Prestyloid Compartment
 - o Tend to push the Carotid sheath laterally.
 2. **Post- Styloid compartment:**
 - o Paragangliomas and nerve sheath tumors usually occupy the Poststyloid Compartment.
 - o Tend to push the Carotid sheath medially.

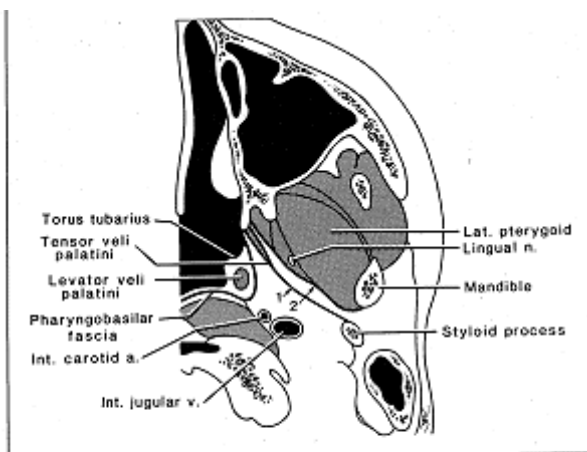


Figure 2: Parapharyngeal Space, Axial View—Axial view of the parapharyngeal space at the level of the nasopharynx. Arrow 1 indicates the fascia extending from the levator veli palatini muscle of the styloid process, which divides the parapharyngeal space into prestyloid and post-styloid compartments. Arrow 2 points to the fascia of the pterygoid muscles.

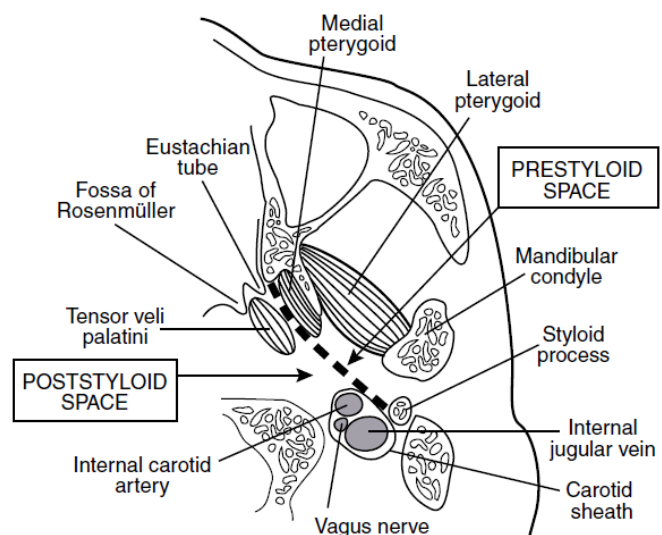


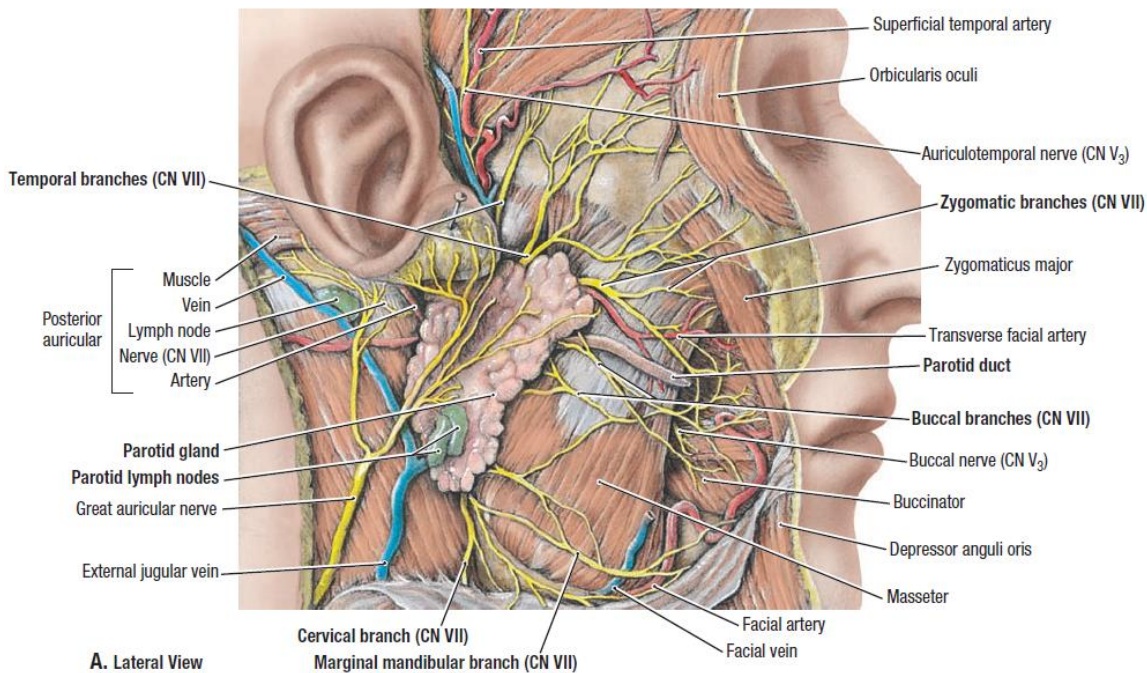
Figure 5-2. Cross-section at the level of the nasopharynx demonstrating the anatomy of the parapharyngeal space.

- **Tail of Parotid gland:**

- Overlies upper ¼th of SCM muscle.
- Extends toward the mastoid process.
- Patients with parotitis frequently have pain with mastication because the gland becomes trapped between the mandible and mastoid process upon opening of the mouth.

- **Stensen's duct.**

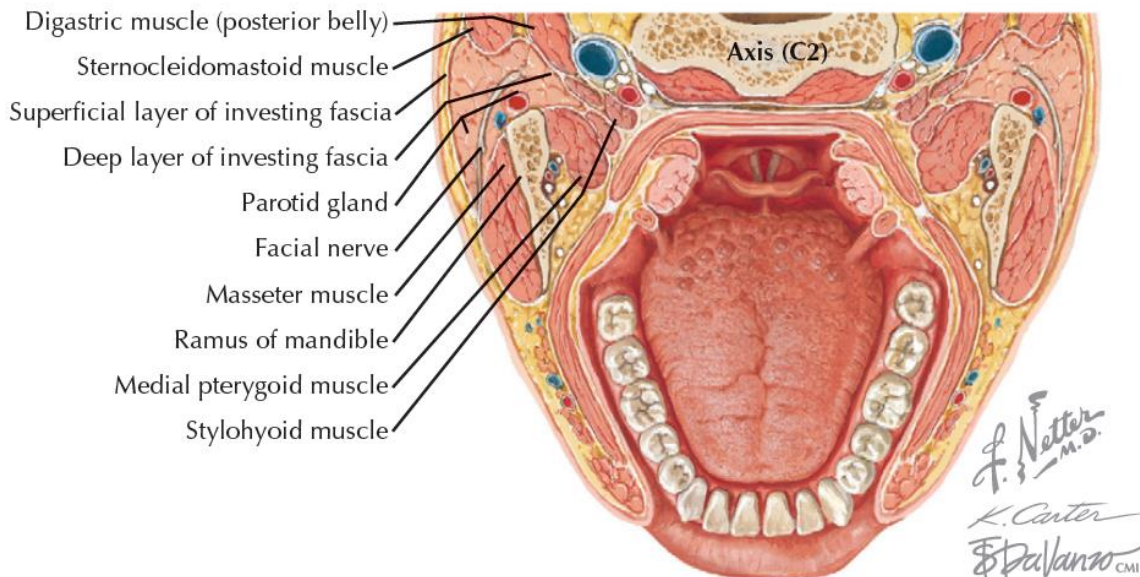
- Parotid's duct.
- 4-6 cm in length and 5 mm in diameter.
- Arises from Anterior border of Deep Parotid lobe.
- Parallel to Zygomatic arch (1.5 cm inferior to its inferior margin)
- Midway between Zygomatic arch and corner of mouth
- Along a line between the upper lip philtrum and the tragus.
- Runs superficial to Masseter muscle.
- Receives a small duct from the accessory parotid gland.
- Turns medially 90 degrees to pierce Buccinator muscle
- Opens onto oral cavity at the level of **2nd Upper molar**.
- Buccal branch of CN-VII runs with the parotid duct.



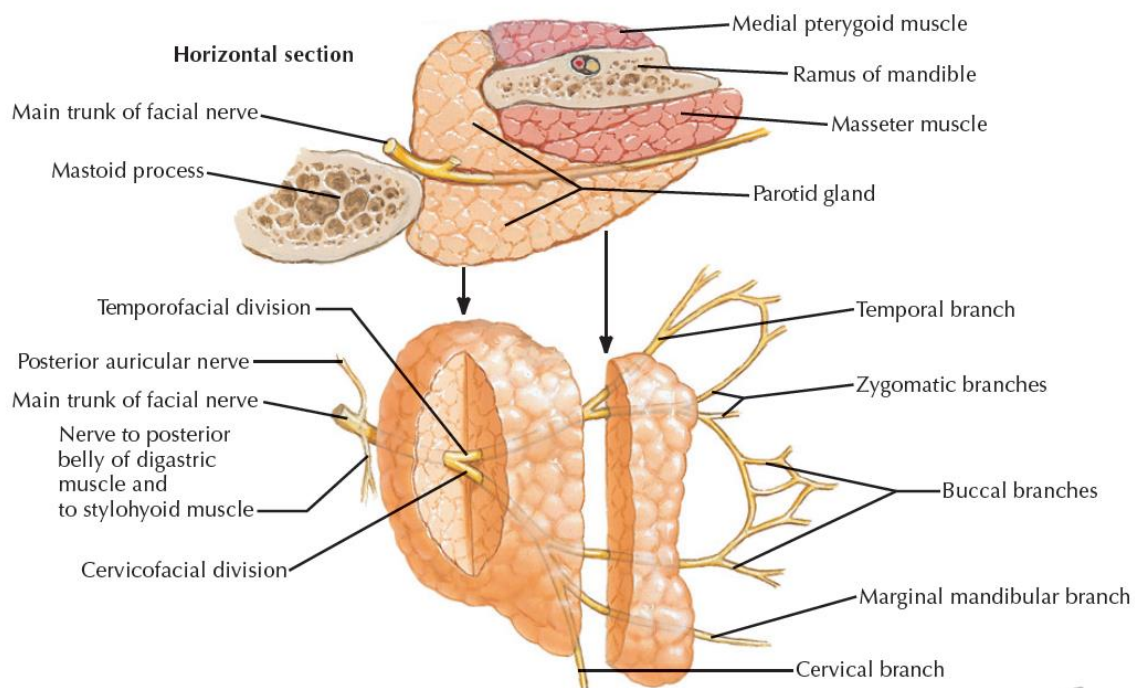
- **Accessory Parotid gland and duct:**

- 20% of people.
- Accessory gland is typically found overlying the masseter between zygomatic arch and parotid duct.
- Accessory glandular tissue is histologically distinct from parotid tissue in that it may contain mucinous acinar cells in addition to the serous acinar cells.
- Accessory duct typically lies cranial to Stensen's duct.

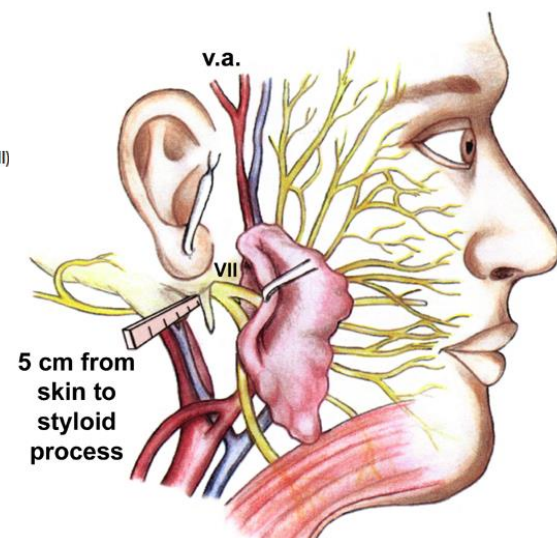
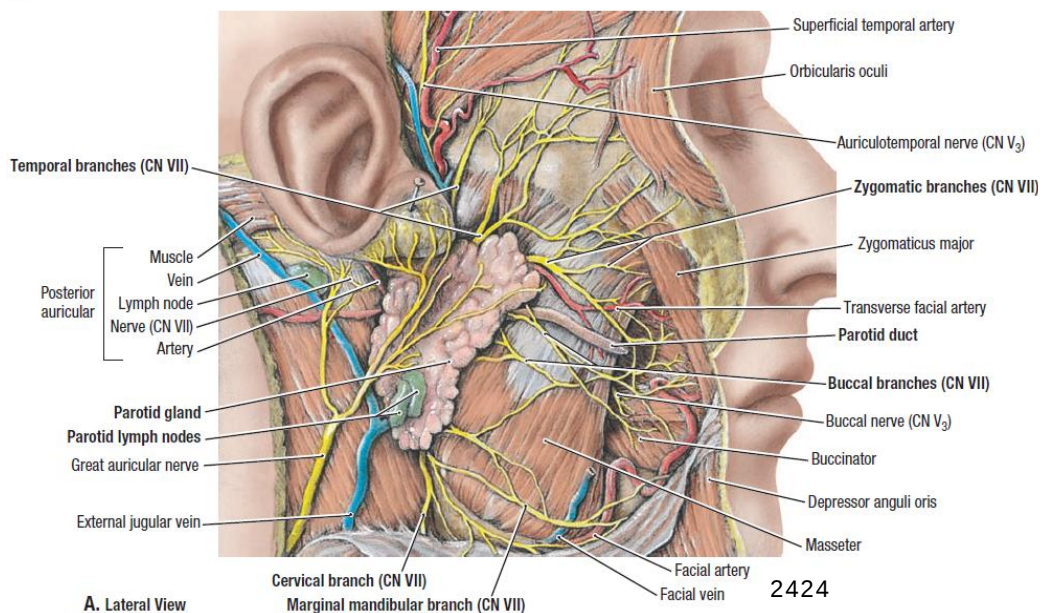
- **Parotid Fascia:**
- Parotid is invested (incompletely) in its own fascia (capsule).
- Parotid fascia is continuous with Superficial layer of deep cervical fascia.
- Parotid fascia consists of:
 1. Superficial layer:
 - Extends from Masseter and SCM to the Zygoma.
 2. Deep layer:
 - Extends from fascia of posterior belly of the Digastric muscle, and forms the Stylomandibular membrane separating Parotid and Submandibular glands.
- Parotid fascia sends septa into the glandular tissue, which prevents the possibility of separating the glandular tissue from its investing fascia.
- Attachments of Parotid fascia:
 - Anterior: Mandible
 - Posterior: Styloid process
 - Superior: Zygomatic arch
 - Inferior: Stylomandibular ligament
- Parotid tissue can herniate through the Stylomandibular membrane.
- Thus, deep parotid tumors can present as parapharyngeal masses.



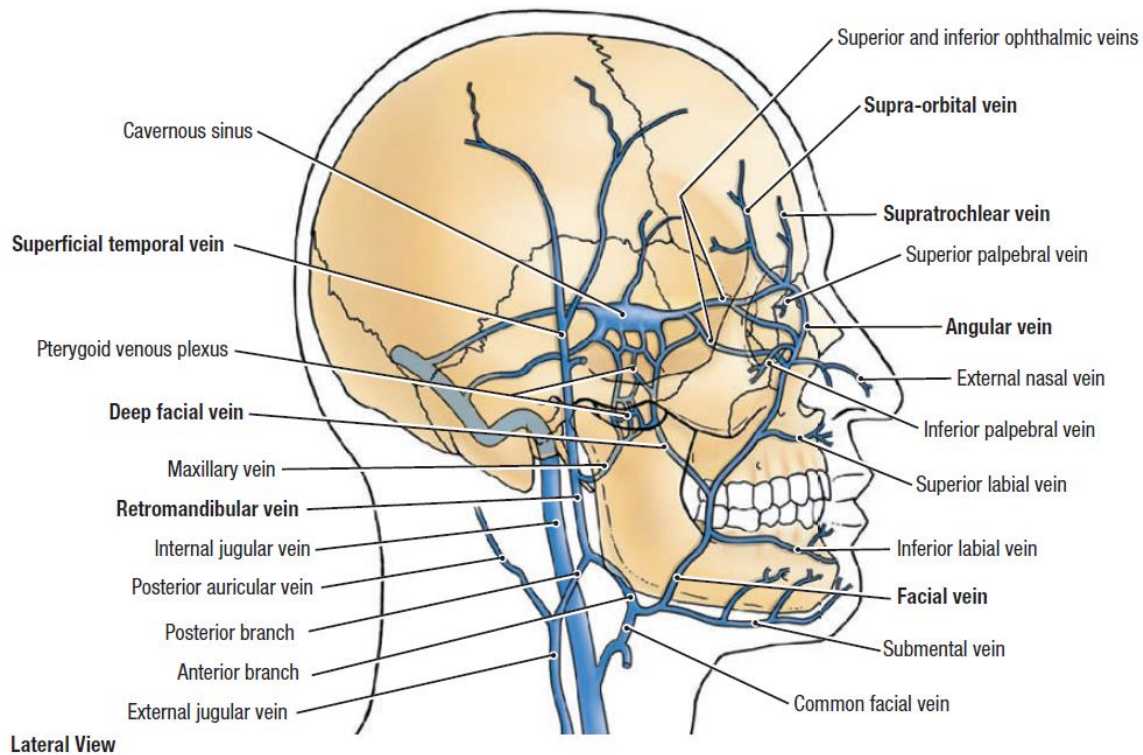
- **Facial nerve:**
- Facial nerve CN-VII is associated with the parotid gland.
- Divides it into 2 lobes connected by isthmus:
 1. Superficial lobe.
 2. Deep lobe.
- CN VII exits Skull base via Stylomastoid foramen (styloid process-medial, mastoid process-lateral).
- Immediately gives off 3 Motor branches upon exiting the foramen:
 1. Branch to Stylohyoid muscle
 2. Branch to Postauricular muscle
 3. Branch to posterior belly of Digastric
- After exiting the foramen, CN-VII turns laterally to enter the Parotid gland at its posterior margin.
- Then branches at the Pes Anserinus (Goose's foot) 1.3 cm from stylomastoid foramen.
- Gives rise to 2 divisions:
 1. Temporofacial (Upper)
 2. Cervicofacial (Lower)
- Followed by 5 terminal branches:
 1. Temporal
 2. Zygomatic
 3. Buccal
 4. Marginal Mandibular
 5. Cervical



- Branches of CN VII are more superficial at Anterior border of Parotid gland, and are therefore more prone to injury there.
- **Intra-operative Localization of CN-VII:**
- Stylomastoid foramen is the single most constant landmark.
- Important surgical landmarks:
 1. **Tragal pointer:**
 - Points to main trunk of CN-VII proximal to Pes Anserinus.
 - 1-1.5 cm deep and inferior to the pointer
 2. **Tympanomastoid suture:**
 - Traced medially.
 - Main trunk of VII is 6-8 mm deep to the suture line.
 3. **Posterior belly of Digastric muscle:**
 - Guide to the Stylomastoid foramen.
 - Trunk of VII is just superior and posterior to cephalic margin of the muscle
 4. **Styloid process:**
 - 5-8 mm deep to Tympanomastoid suture
 - Trunk of VII lies on Posterolateral aspect of Styloid near its base
 5. **Retrograde dissection:**
 - Employed to trace one of the terminal branches proximally.
 - **Buccal branch:**
 - Runs with the parotid duct either superiorly or inferiorly.
 - **Temporal branch:**
 - Crosses the zygomatic arch parallel with the superficial temporal artery and vein.
 - **Marginal Mandibular branch:**
 - Runs along inferior border of parotid superficial to the retromandibular vein.
 6. **Mastoidectomy:**
 - As a last resort
 - Performed to identify the nerve at the stylomastoid foramen or to identify the intratemporal mastoid course of VII.



- **Great Auricular nerve:**
 - Originates from the cervical plexus (**C2-C3**).
 - Passes from deep to superficial around posterior border of SCM.
 - Then travels superiorly, posterior to External Jugular vein.
 - Provides sensation to Posterior pinna and lobule.
 - **1st nerve encountered in surgery, lateral to the Parotid fascia and deep to the Platysma.**
 - Routinely divided during parotidectomy, and no benefit has been proven from preserving the posterior branch of the nerve.
- **Auriculotemporal nerve:**
 - Branch of Mandibular nerve (**V3**).
 - Runs anterior to the EAM.
 - Parallels to Superficial temporal artery and vein.
 - Provides sensory innervation to Parotid capsule, and skin of the auricle and temporal region (referred pain from parotitis can involve Auricle, EAM, TMJ, and temples).
 - Carries Parasympathetic postganglionic fibers from Otic ganglion to Parotid gland.
 - If injured intraoperatively, Aberrant parasympathetic innervation to the skin results in **Frey's Syndrome** (Gustatory sweating).
 - May be resected intentionally to avoid Frey's Syndrome.
- **Parotid compartments: (N → V → A)**
- **Neural compartment:**
 - Most superficial portion of the Parotid.
 - Contains:
 1. CN-VII.
 2. Auriculotemporal nerve
 3. Great Auricular nerve.
- **Venous compartment:**
 1. Located in the middle of the Parotid, deep to CN-VII.
 2. Venous drainage is provided by **Retromandibular vein:**
 - Formed from Superficial Temporal + Maxillary vein.
 - Lies deep to Facial nerve.
 - Runs lateral to the Carotid artery.
 - Emerges at the inferior pole of the Parotid gland.
 - **Posterior branch** + Postauricular vein → External Jugular.
 - **Anterior branch** + Facial vein → Common Facial vein → Internal Jugular vein.

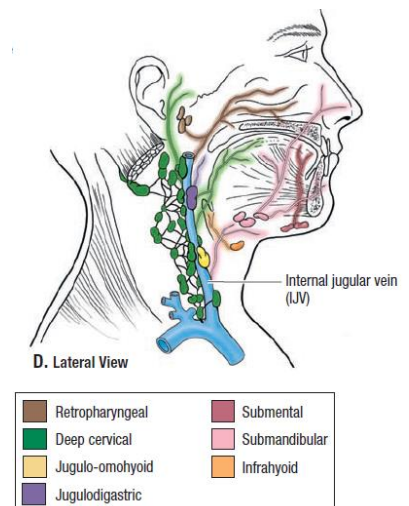


- **Arterial compartment:**

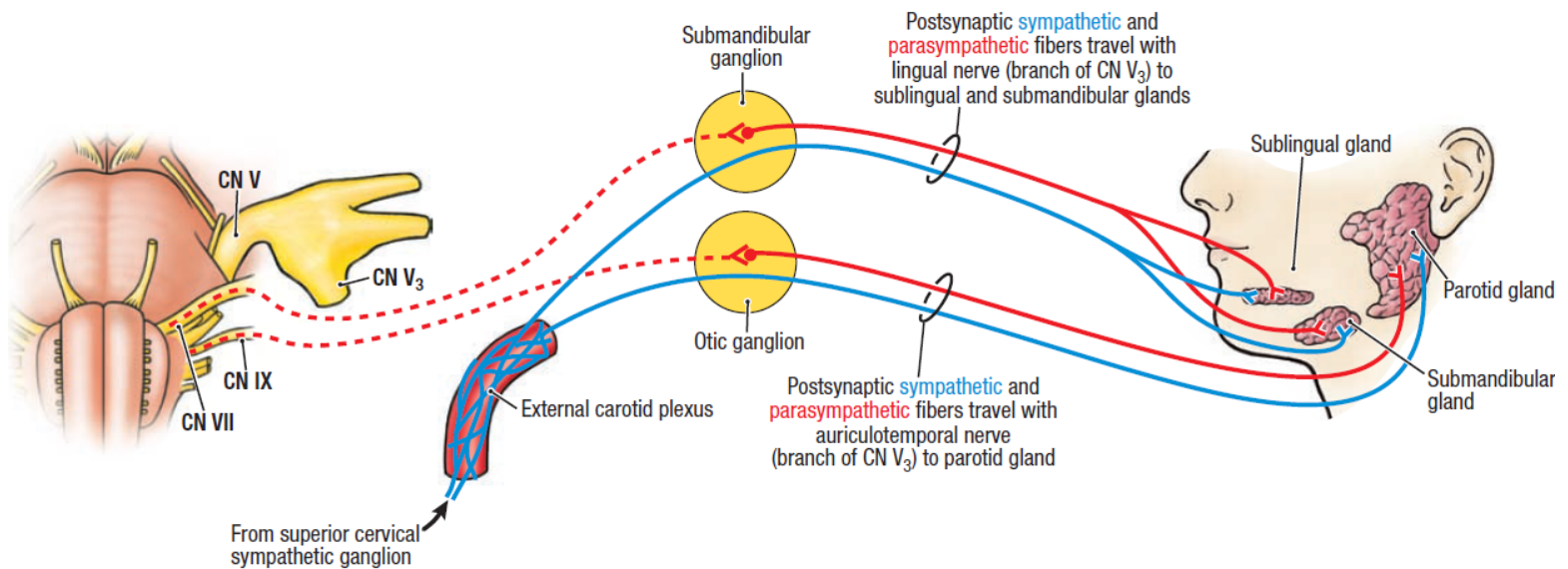
- Located in the deep portion of parotid gland.
- Contains:
 1. External Carotid Artery.
 2. Maxillary Artery.
 3. Superficial Temporal Artery:
 - Supplies the temperoparietal fascial flap which can be used to recontour defects left in the Parotid bed.
 - **Transverse Facial artery:** branch of the Superficial Temporal artery, providing blood supply to Parotid gland, Stensen's duct, and Masseter muscle, runs between the zygomatic arch and Stensen's duct.

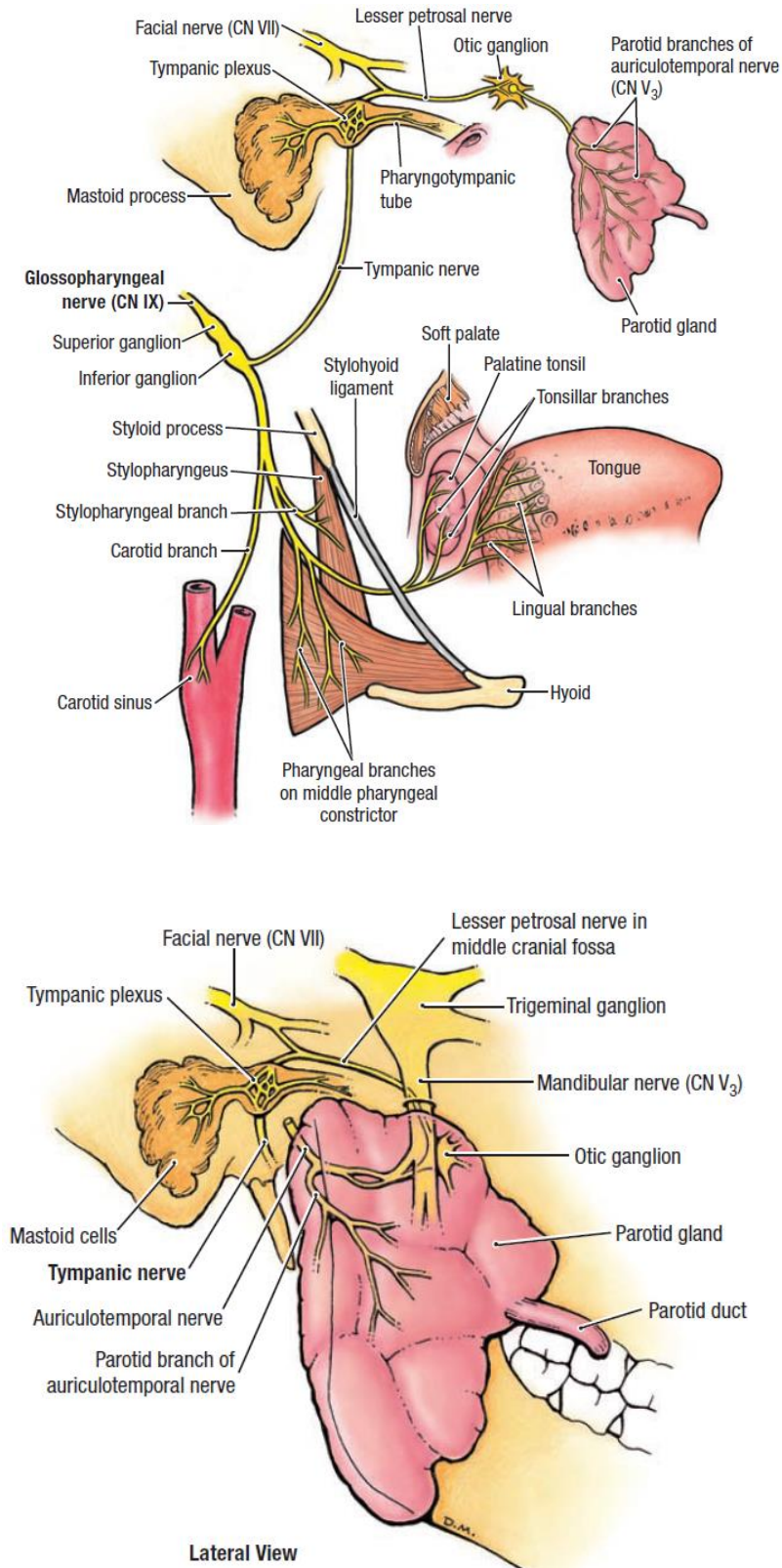
- **Lymphatic drainage:**

1. Paraparotid nodes:
 - More numerous
 - Drain Temporal region, scalp, and auricle.
2. Intraparotid nodes:
 - Drain posterior nasopharynx, soft palate, and ear.
 - Drain into the superficial and deep cervical lymph nodes.

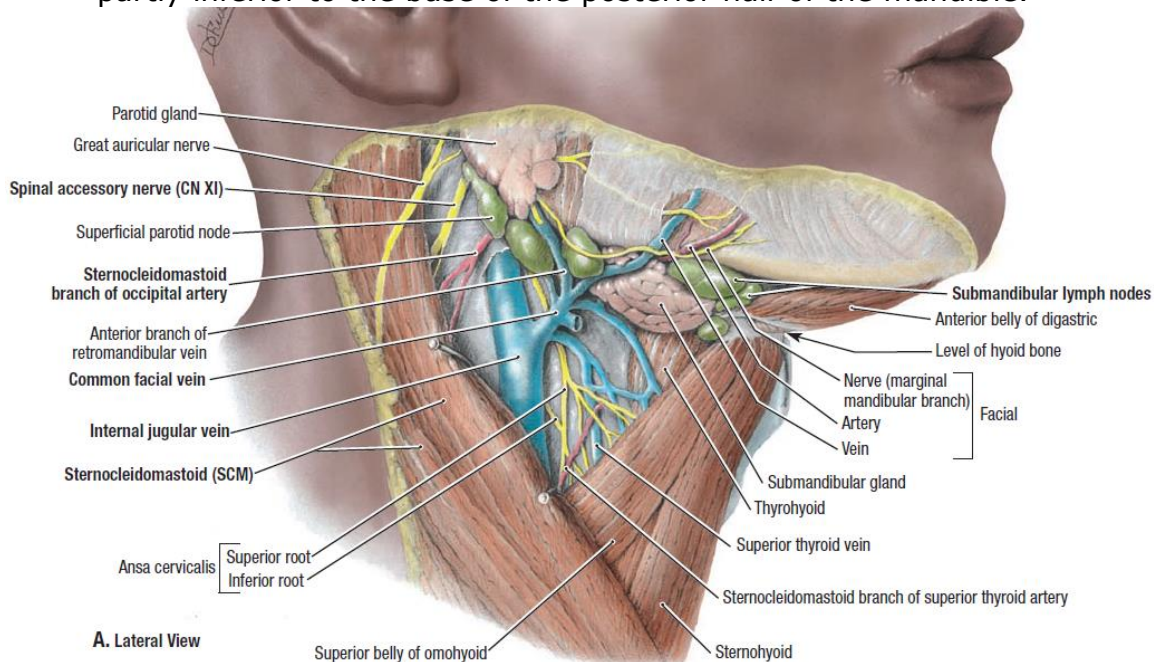


- **Parasympathetic's of Parotid:**
- Pre-ganglionic Parasympathetic fibers:
 - **Inferior Salivatory Nucleus** in Medulla.
 - → **Glossopharyngeal nerve (CN-IX).**
 - → Jugular foramen.
 - → **Tympanic Nerve (Jacobson Nerve).**
 - → Tympanic plexus.
 - → **Lesser Petrosal nerve.**
 - → Foramen Ovale.
 - → **Otic ganglion**
- Post-ganglionic Parasympathetic fibers:
 - **Otic ganglion.**
 - → **Auriculotemporal branch (V3)**
 - → Parotid gland.
- **Sympathetic's of Parotid:**
- Pre-ganglionic Sympathetic fibers:
 - **Intermediolateral horn nuclei from T1-T3(4)**
 - → Ventral root of Spinal cord
 - → Spinal nerve
 - → Sympathetic chain.
 - → **Superior cervical ganglion.**
- Post-ganglionic Sympathetic fibers:
 - **Superior cervical ganglion.**
 - → External carotid artery branches
 - → Parotid gland.

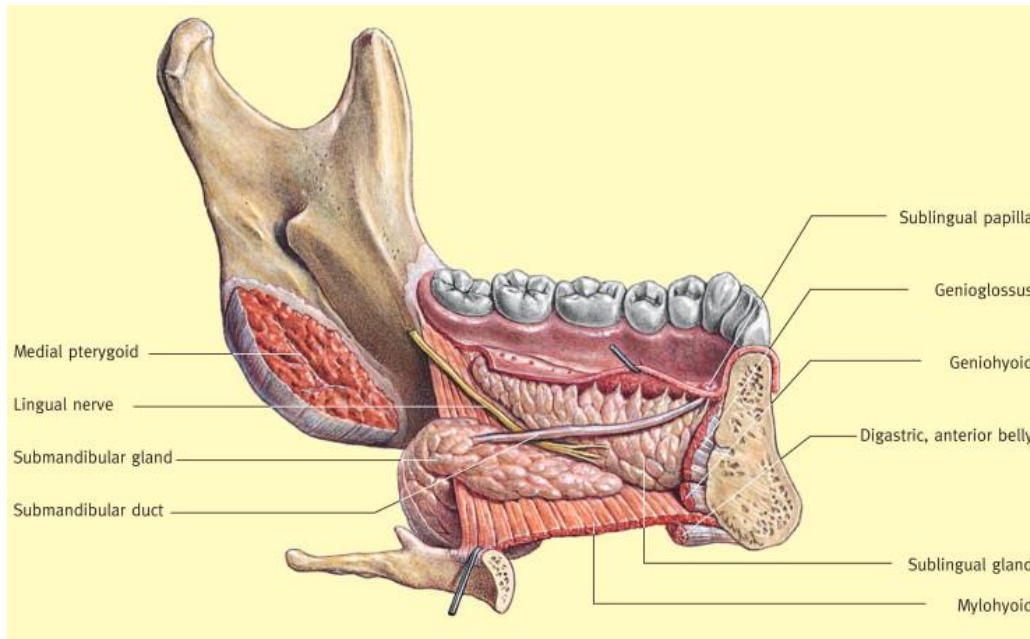




- **Submandibular Gland:**
- Submaxillary gland.
- 2nd largest salivary gland
- Weighs ½ weight of Parotid.
- Produce majority of saliva.
- **Mixed** serous and mucous secretions but **Predominantly Serous.**
- Lies in Submandibular triangle formed by:
 - o Anterior and Posterior bellies of Digastric.
 - o Inferior margin of the mandible.
- Positioned medial and inferior to Mandibular ramus partly superior and partly inferior to the base of the posterior half of the mandible.

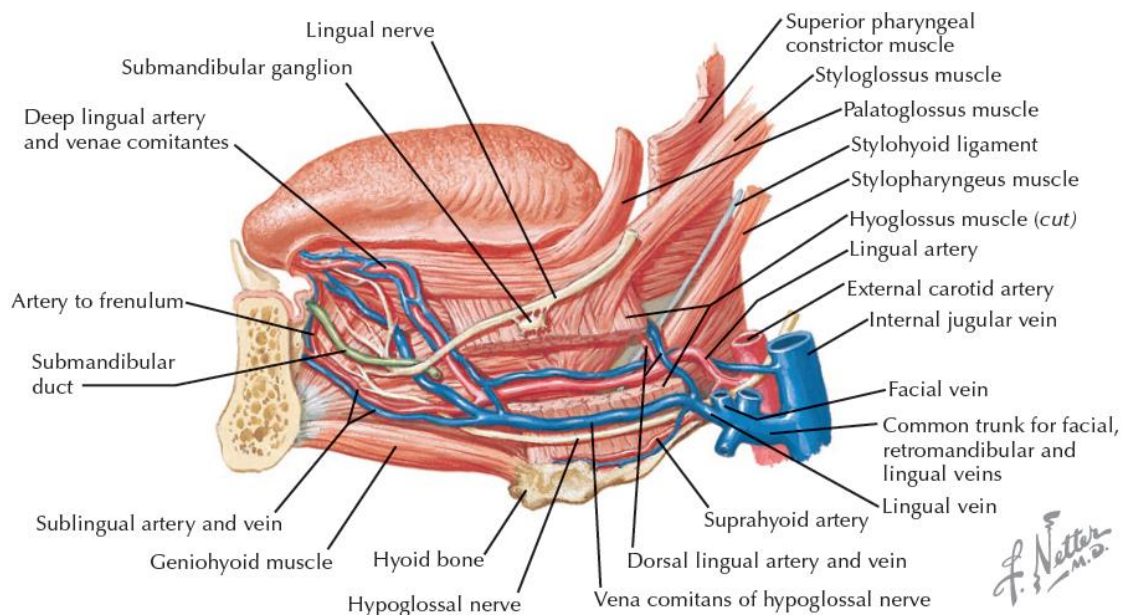


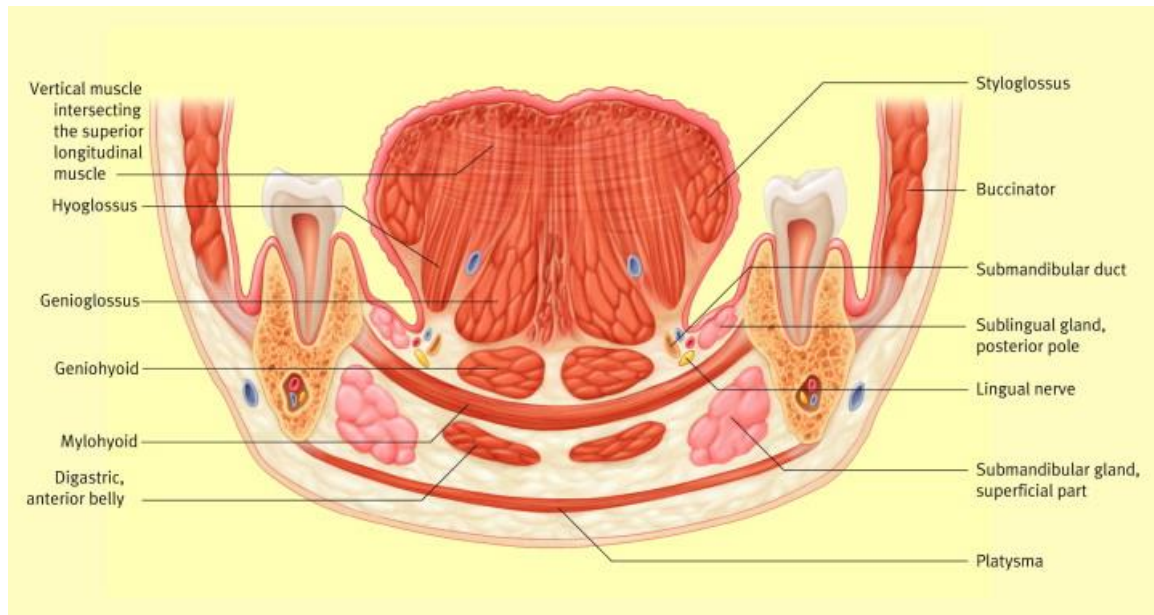
- Forms a "C" around Anterior margin of Mylohyoid muscle.
- **Mylohyoid muscle divides Submandibular gland into:**
 - o Superficial lobe:
 - Located in Submandibular Triangle.
 - Surrounded by Investing layer of deep cervical fascia.
 - o Deep lobe:
 - Major portion.
 - Lies in Oral cavity between Hyoglossus muscle. and Mandible.
 - Ends at the posterior border of Sublingual gland.
- **Marginal Mandibular branch of CN-VII courses:**
 - o Superficial to Submandibular gland.
 - o Deep to Platysma.
- Submandibular gland is invested in its own capsule which is also continuous with Superficial layer of deep cervical fascia.



- **Wharton's duct.**

- Submandibular gland duct.
- 5 cm in length
- Exits from medial surface of Submandibular gland.
- Runs on Genioglossus muscle
- Runs between Mylohyoid (lateral) and Hyoglossus muscle.
- Opens lateral to lingual frenulum on the anterior floor of mouth in **Sublingual cruncles**.
- **Lingual nerve** wraps around Wharton's duct, starting lateral and ending medial to the duct.
- **Hypoglossal nerve (CN XII)** runs parallel and inferior to Submandibular duct.

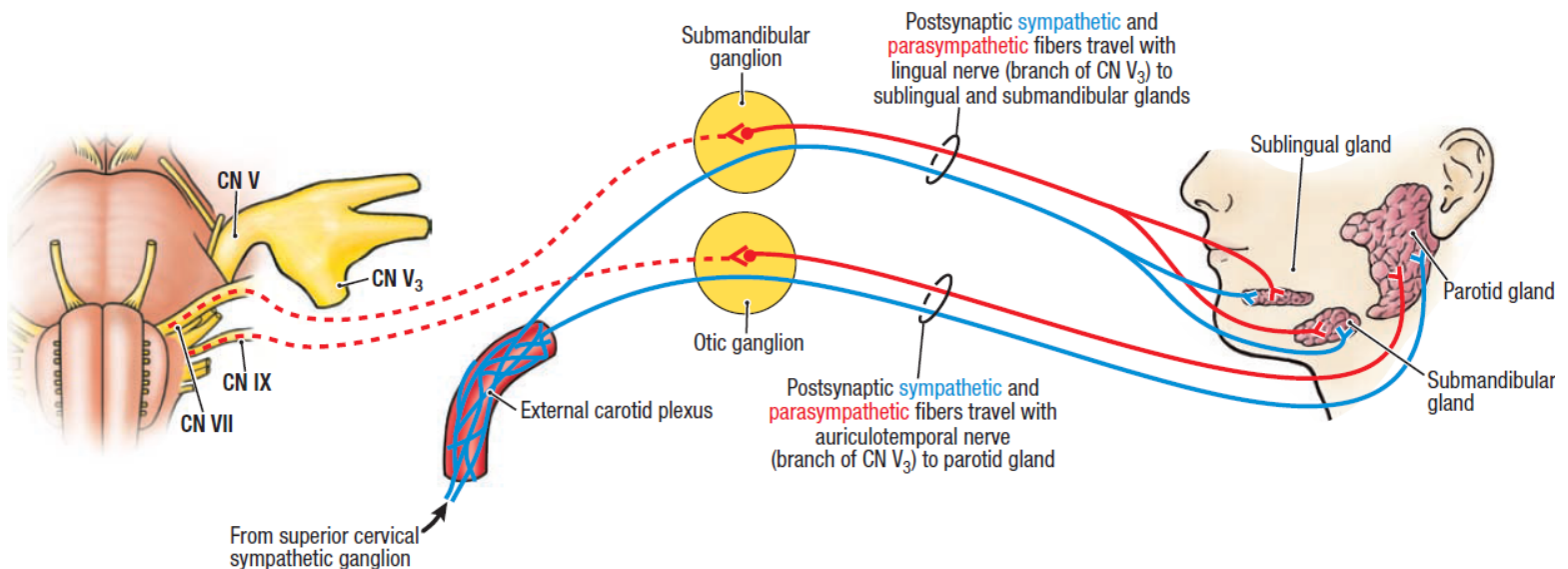




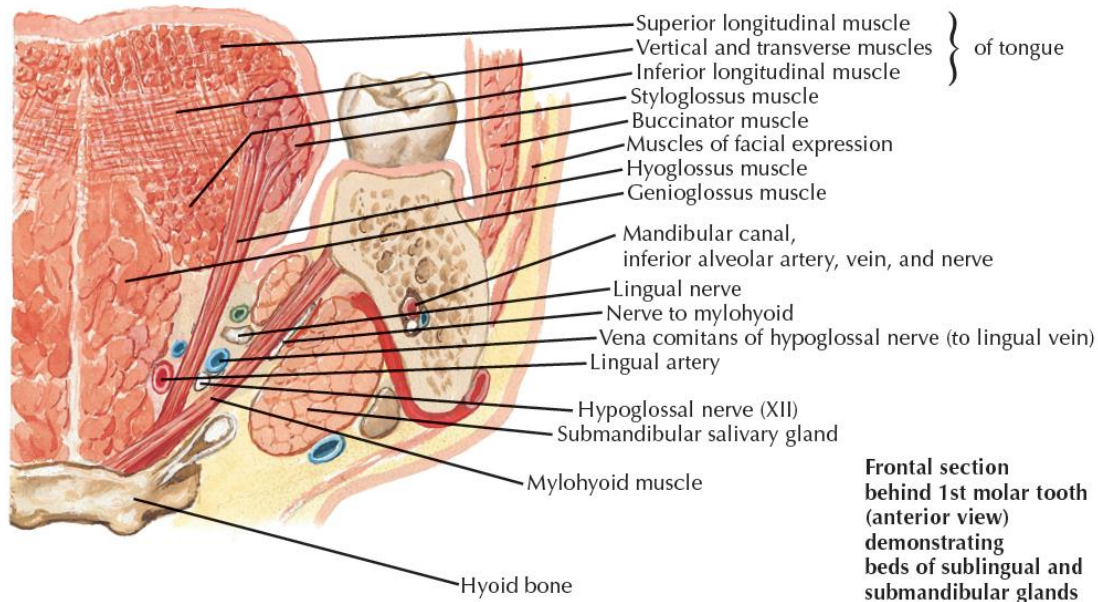
- **Blood Supply:**
- **Submental branch of Facial artery.**
- Facial artery arise from External Carotid artery and courses medial to Posterior Digastric muscle then hooks over the muscle to enter Submandibular gland and exits into the facial notch of the inferior mandible to supply the face.
- **Venous drainage:**
- **Facial vein** which lies deep to **Marginal Mandibular branch of CN-VII.**
- Courses over Lateral surface of Submandibular gland.
 - o One method of preserving the Marginal Mandibular nerve is to identify and ligate Facial vein 2-3 cm inferior to the mandible and elevate the vein and all superior tissue superiorly.
- Facial artery and vein are **1st blood vessels** encountered when resecting Submandibular glands as they cross superficially over the inferior border of the mandible.
- **Lymphatic drainage:**
- Drain to the deep cervical and jugular chains of nodes.

- **Parasympathetic's of Submandibular gland:**
- Pre-ganglionic Parasympathetic fibers:
 - **Superior Salivatory Nucleus** in Pons.
 - → **Facial nerve (CN-VII).**
 - → Nervus Intermedius.
 - → **Chorda Tympani nerve.**
 - → Petrotympanic fissure
 - → Lingual nerve.
 - → **Submandibular ganglion**
- Post-ganglionic Parasympathetic fibers:
 - **Submandibular ganglion**
 - → Submandibular and Sublingual glands.

- **Sympathetic's of Submandibular gland:**
- Pre-ganglionic Sympathetic fibers:
 - **Intermediolateral horn nuclei from T1-T3(4)**
 - → Ventral root of Spinal cord
 - → Spinal nerve
 - → Sympathetic chain.
 - → **Superior cervical ganglion.**
- Post-ganglionic Sympathetic fibers:
 - **Superior cervical ganglion.**
 - → External carotid artery branches
 - → Submandibular gland.

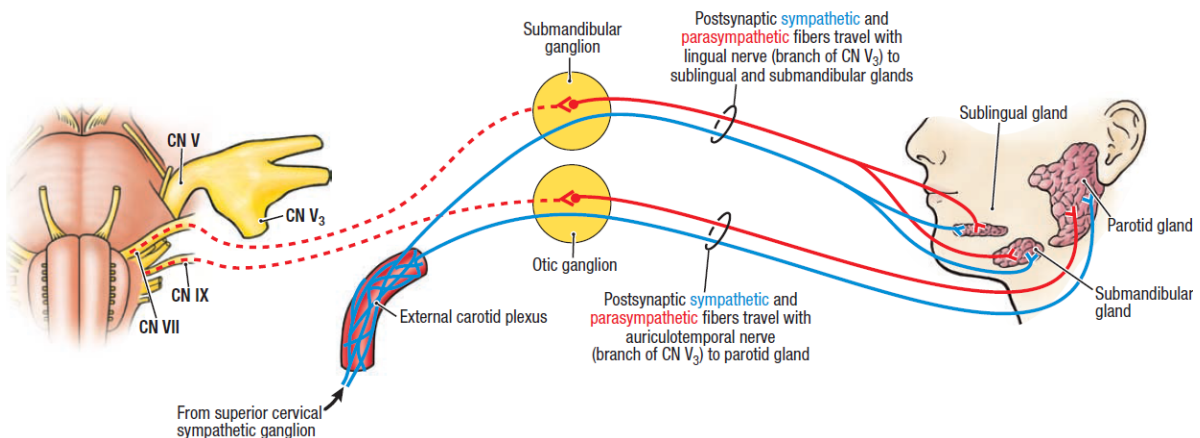


- **Sublingual Gland:**
- Smallest major salivary gland.
- Almond shaped.
- Located within Submucosal layer of Floor of Mouth.
 - o Between Mandible and Genioglossus muscle.
 - o Bounded inferiorly by Mylohyoid muscle.
 - o Wharton's duct and Lingual nerve pass between the Sublingual gland and Genioglossus muscle.
- **Mixed** serous and mucous secretions but **Predominantly Mucous**.



- **No True Fascial Capsule.**
- **No single dominant duct.**
- **Ducts of Rivinus:**
 - o Sublingual gland ducts.
 - o 10 small ducts.
 - o Exit superior aspect of Sublingual gland.
 - o Open along Sublingual fold on the floor of mouth.
 - o Ducts of the sublingual glands are too small for the injection of contrast, making a sialogram of this gland impossible.
 - o **Bartholin's duct:**
 - Common duct formed by union of several Anterior Sublingual ducts.
 - Opens into Wharton's duct.

- **Blood supply:**
 - Sublingual branch of Lingual artery.
 - Submental branch of Facial artery
- **Venous drainage:**
 - Reflects the arterial supply.
- **Lymphatic drainage:**
 - Drain into Submandibular nodes.
- **Parasympathetic's of Sublingual gland:**
- Pre-ganglionic Parasympathetic fibers:
 - **Superior Salivatory Nucleus** in Pons.
 - → **Facial nerve (CN-VII).**
 - → Nervus Intermedius.
 - → **Chorda Tympani nerve.**
 - → Petrotympanic fissure
 - → Lingual nerve.
 - → **Submandibular ganglion**
- Post-ganglionic Parasympathetic fibers:
 - **Submandibular ganglion**
 - → Submandibular and Sublingual glands.
- **Sympathetic's of Sublingual gland:**
- Pre-ganglionic Sympathetic fibers:
 - **Intermediolateral horn nuclei from T1-T3(4)**
 - → Ventral root of Spinal cord
 - → Spinal nerve
 - → Sympathetic chain.
 - → **Superior cervical ganglion.**
- Post-ganglionic Sympathetic fibers:
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 - → External carotid artery branches
 - → Sublingual gland.

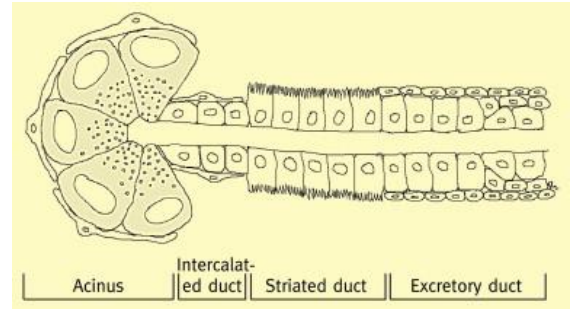


- **Minor Salivary Glands:**
- Several minor glands within the submucosal layer of Oral cavity.
- **Concentrated in:**
 - Buccal region.
 - Labial region.
 - Palatal region.
 - Lingual region.
- **Can be found in:**
 - Superior pole of Tonsils (**Weber's glands**).
 - Tonsillar pillars.
 - Base of tongue (**von Ebner's glands**).
 - Paranasal sinuses
 - Larynx
 - Trachea
 - Bronchi.
- **Anterior hard palate** and **Gingivae** are devoid of Minor salivary glands.
- Minor salivary glands Lack a branching network of draining ducts.
- Each salivary unit has its own simple duct.
- Minor salivary glands are **Mixed Predominantly Mucous**.
 - **Except** in Posterior Hard Palate → Mucous
- **Most common tumor sites derived from Minor salivary glands:**
 - Palate.
 - Upper lip
 - Cheek.
- Most of Minor glands receive parasympathetic innervation from the **Lingual nerve**.
 - **Except** Minor glands of **Palate** → **Palatine nerves**, fed by the Sphenopalatine ganglion.

- **Histology of Salivary Glands**

- Secretory unit consists of:

1. Acinus
2. Myoepithelial cells
3. Intercalated duct
4. Striated duct
5. Excretory duct.



- **Acini:**

- Responsible for producing Primary secretion.
- Contain secretory granules, Endoplasmic reticulum, Golgi Apparatus.
- Produce Saliva.
- Divided into 3 types:

1. **Serous:**

- Protein-secreting.
- Spherical cells rich in zymogen granules.
- Well developed Intercalated and Striated ducts.
- Secretion is heavily modified.
- Secretory granules contain Amylase.

2. **Mucous:**

- Mucin-secreting.
- Tubular shaped cells.
- Intercalated duct is short and poorly developed.
- No Striated duct.
- No significant modification in secretion.
- Secretory granules contain Mucin.

3. **Mixed:**

- Serous demilunes
- or predominantly mucous acinar cells capped by a few serous acinar cells

- **Myoepithelial cells:**

- Surrounds Acini and Intercalated duct.
- Moving secretions toward Excretory duct.

- **Ductal system:**

- Made up of (from proximal to distal):

1. **Intercalated duct:**

- Lined by Low Cuboidal epithelial cells.
- Secrete Bicarbonate (**CHO⁻³**) into Ductal lumen.
- Absorb Chloride (**Cl⁻**) from the lumen.

2. **Striated duct:**

- Lined by Simple cuboidal epithelial cells.
- Secrete Potassium (**K⁺**) into the lumen.
- Absorb Sodium (**Na⁺**) from the lumen.
- Produce **HYPOTonic** fluid.
- The Faster the salivary flow rate, the less time allowed for these cells to act, resulting in a Less HYPOTonic product.

3. Excretory duct:

- Lined proximally by Simple Cuboidal epithelium.
- Lined distally by Stratified Cuboidal or Pseudostratified columnar epithelium.
- NOT perform any modification of the saliva.

- **Parotid gland:**

- Purely serous salivary gland.

- **Submandibular gland:**

- Mixed salivary gland
- Predominantly Serous.
- 10% of its acini are Mucinous.

- **Sublingual gland:**

- Mixed salivary gland
- Predominantly mucous.

- Salivary gland stroma is rich in lymphocytes and plasma cells, which are responsible for the production of IgA.

- **Function of Saliva**

1. Moistens oral mucosa:

- Mucin layer on Oral mucosa is the most important non-immune defense mechanism in the oral cavity.

2. Moistens dry food and cools hot food.

3. Provides a medium for dissolved foods to stimulate the taste buds.

4. Buffers oral cavity contents:

- Saliva has a high concentration of bicarbonate ions.

5. Digestion:

- Alpha-amylase, contained in saliva, breaks 1-4 glycoside bonds.
- Lingual lipase helps break down fats.

6. Controls bacterial flora of the oral cavity.

7. Mineralization of new teeth and repair of precarious enamel lesions:

- Saliva is high in calcium and phosphate.

8. Protects the teeth by forming a "Protective Pellicle".

- Problems with the salivary glands generally result in rampant dental caries.

9. Antibacterial:

1. Lysozyme, Secretory IgA, and Salivary Peroxidase.

2. Salivary flow rate may play a more important role in oral hygiene than any of these factors.

- **Intraoral complications of salivary hypofunction:**
 1. Candidiasis
 2. Oral Lichen Planus (Painful)
 3. Burning Mouth Syndrome (normal appearing oral mucosa with a subjective sensation of burning)
 4. Recurrent Aphthous ulcers
 5. Dental caries
- **Evaluation of Salivary Function**
- Best way by Measurement of Salivary flow rate in stimulated (e.g., by using a parasympathomimetic such as pilocarpine) and unstimulated states.
- Xerostomia is NOT a reliable indicator of salivary hypofunction.
- **Production of Saliva:**
- **Active process** occurring in 2 phases:
 1. Primary secretion:
 - Occurs in Acinar cells.
 - Results in a **Isotonic** salivary fluid
 2. Ductal secretion:
 - Results in a **Hypotonic** salivary fluid.
 - **Low Na** concentration.
 - **High K** concentration.
- Degree of modification of saliva in Ducts turns heavily on salivary flow rate:
 - **Fast** Flow Rate → Less Hypotonic.
 - **Slow** Flow Rate → More Hypotonic.
- **Normal saliva composition:**
 - Water: **99.5%**
 - Proteins
 - Glycoproteins
 - Potassium (7x plasma) **High**
 - Bicarbonate (3x plasma) **High**
 - Sodium (1/10 x plasma). **Low**
 - Calcium
 - Phosphorous
 - Chloride
 - Thiocyanate
 - Urea.
 - **Normal pH of 5.6-7**

- **Autonomic Innervation**
- Parasympathetic nervous system is the primary instigator of salivary secretion.
 - If interruption of Parasympathetics → Atrophy.
 - If interruption of Sympathetics → No significant change.
- Stimulation by Parasympathetic:
 - Results in Abundant, watery saliva.
 - Decrease in [Amylase] in saliva.
 - Increase in [Amylase] in serum.
 - Acetylcholine is Active neurotransmitter.
 - Parasympathetic fibers from CN-IX → Lesser Superficial Petrosal nerve → Otic ganglion → Auriculotemporal Nerve → Parotid.
 - Parasympathetic fibers from CN-VII → Chorda Tympani → Submandibular ganglion → Submandibular and Sublingual.
- Stimulation by Sympathetic:
 - Result in Scant, viscous saliva.
 - Rich in organic and inorganic solutes.
 - Increase in [Amylase] in saliva
 - No change in [Amylase] in serum.
 - Sympathetic fibers from Superior Cervical ganglion
 - → Travel with External Carotid artery → Parotid
 - → Travel with Lingual artery → Submandibular
 - → Travel with Facial artery → Sublingual.
- **Salivary Flow:**
- Average volume of saliva secreted in a 24 hour period is 1-1.5 liters.
- 1 cc/minute)
- Most of saliva is secreted during meals.
- Basal salivary flow rate=0.001-0.2 ml/minute/gland.
- With stimulation, salivary flow rate=0.18-1.7 ml/min/gland.
- In **UNSTIMULATED** state the relative contribution of the major salivary glands is as follows:
 - **Submandibular gland= 69%**
 - Parotid gland= 26%
 - Sublingual gland= 5%
- In **STIMULATED** state the relative contribution of the major salivary glands is as follows:
 - **Parotid gland= 69%**
 - Submandibular gland=26%
 - Sublingual gland=5%
- Minor salivary glands flow rate is **independent of stimulation.**
 - 7-8% of total salivary output.

- **Sublingual glands and Minor salivary glands:**
 - Contribute only about 10% of all saliva
 - Produce the majority of mucous.
 - Critical in maintaining Mucin layer over the oral mucosa.
- Ptyalism, or drooling, may be secondary to salivary hypersecretion.
- This is caused either by excessive salivary flow, or a salivary flow rate which surpasses the ability to swallow the saliva. Possible surgical treatments for Ptyalism are tympanic neurectomies (eliminating parasympathetic innervation to the Parotid gland) or Parotid duct rerouting.
- 80-90% of salivary gland stones occur in the Submandibular gland, and of those, 85% occur in Wharton's duct. Complete ductal obstruction generally results in atrophy of the gland, while partial obstruction usually results in glandular mucocele.
- **Effects of Aging**
- Acinar cells do degenerate with age.
- Total salivary flow rate is independent of age.
- Xerostomia, or the subjective complaint of dry mouth, must be distinguished from the objective finding of decreased salivary flow. Xerostomia in the elderly is generally either secondary to medications or to systemic disease.

Approach to Salivary Gland Pathology:

- Character of Parotid Mass:
 - Rapid onset (inflammatory) vs slow growing (neoplastic)
 - Diffuse versus discrete mass (tumor)
 - Unilateral vs bilateral involvement (sialadenosis, mumps)
 - Pain associated with malignant lesion can imply neural invasion
 - Association with food ingestion (sialadenitis)
- Contributing Factors:
 - Smoking or alcohol abuse
 - Exposure to radiation or toxins (lead or mercury)
 - History of sarcoidosis, Sjögren's disease, tuberculosis, gout, amyloidosis
 - Recent facial trauma or surgery, dental work, immunization (specifically MMR vaccine).
- Associated Symptoms:
 - Xerostomia
 - Sialorrhea
 - Weight loss
 - Fever
 - Trismus
- Physical Exam:
 - Palpation (mobility, size, consistency)
 - Bimanual palpation with duct inspection for saliva expression (or purulence).
 - Tenderness (inflammatory process)
 - Facial nerve function (malignancy)
 - Minors salivary gland tumors may present as nonulcerated painless masses of oral cavity (palate); can cause hoarseness, respiratory complaints or dysphagia.
 - Parapharyngeal space involvement (examine intraorally)
 - Cervical lymphadenopathy
 - Complete head and neck physical exam
- Signs and symptoms of possible malignant tumors:
 1. Facial nerve paralysis/paresis
 2. Fixation to overlying/underlying structures
 3. Overlying skin ulceration/infection
 4. Localized pain or trismus
 5. Cervical adenopathy
 6. Metastatic disease
 7. Large size (>5cm)

- Imaging and Ancillary Tests
 - **US:**
 - Helpful in differentiating cystic lesions.
 - Part of evaluation in children.
 - **CT/MRI:**
 - Indicated if suspect a tumor or for preoperative evaluation.
 - MRI is indicated for parapharyngeal involvement.
 - Don't tell benign from malignant
 - Peri-parotid fat strip separates deep lobe from parapharyngeal space; important landmark.
 - **Fine Needle Aspirate (FNA):**
 - 90% sensitivity.
 - Need for repeat aspirations may delay diagnosis and treatment.
 - Widely practiced although controversial (may not change management).
 - Indicated for pre-op surgical planning and patient counseling.
 - Utilized to counsel patient of risk of facial nerve involvement and malignancy confirmation must be determined by a superficial parotidectomy.
 - Incisional biopsies are contraindicated due to the possibility of tumor seeding and violation of tumor margins, high recurrence; CN VII injury, EXCEPT in lymphoma).
 - **Technetium-99m Isotope Scan:**
 - Rarely utilized.
 - May differentiate a Warthin's tumor or oncocytoma from other salivary gland neoplasms.
 - **Sialography:**
 - Rarely utilized.
 - Visualizes ductal anatomy.
 - Indications:
 - Suspicion of chronic, recurrent or nonspecific sialoadenitis
 - Autoimmune disease (Sjogren's, Mikulicz's syndromes)
 - Sialolithiasis
 - Postop or posttraumatic fistula/stricture/cyst
 - Contraindications:
 - Iodine allergy
 - Acute sialoadenitis
 - Findings for chronic inflammation on sialography:
 - Saccular dilatation of terminal ducts/acini
 - Segmental strictures & dilatation
 - Pseudocyst formation
 - **Lab Work:**
 - May consider mumps titers, complete blood count, autoimmune and Sjögren's Profile (SS-A, SS-B, ANA, ESR).

TABLE 2–1. Computed Tomography versus Magnetic Resonance Imaging of the Parotid Gland

Computed Tomography	Magnetic Resonance Imaging
• better for bone imaging • less expensive • quicker image • less sensitive to patient motion • may differentiate deep tumors by identifying a fat strip • identifies calcific stones • distinguishes cystic nature of Warthin's tumors • contrast allows differentiation of vascular channels and abnormal lymph nodes	• better for soft tissue imaging (distinguish parotid tumors from parapharyngeal lesions, identifies capsule) • multiplanar views • no radiation required (less invasive) • facial nerve or retromandibular vessels may be used to distinguish deep and superficial lobes • cannot be used with pacemakers and metallic implants (aneurysm clips, cochlear implants) • better determines involvement of the facial nerve and parapharyngeal masses • <u>Recall</u> T_1 weighted: enhances fat, water appears dark, TR <1000, TE <25 ms T_2 weighted: enhances water, fat appears dark, TR >1000, TE >40 ms Spin Density: $T_R >1000$, $T_E <25$ ms

- **Types of Parotid surgery:**

- 1. Enucleation:**

- Excision of pathology while preserving the capsule.

- 2. Partial Parotidectomy:**

- Excision the pathology with cuff of normal parotid tissue.

- 3. Superficial Parotidectomy:**

- Excision of entire superficial lobe of parotid with preservation of FN.

- 4. Total Parotidectomy:**

- Excision of entire superficial and deep lobes of parotid with preservation of FN.

- 5. Radical Parotidectomy:**

- Excision of entire superficial and deep lobes of parotid with FN sacrifice.

- **ALL salivary gland tumors should be surgically excised due to:**
 1. FNAB is not very accurate.
 2. Risk of malignant transformation
 3. Cosmesis
- **EXCEPT:**
 1. Patients with co-morbidities and unfit for surgery → Observation.
 2. Lymphoma → Open biopsy.
- **ALL Benign parotid tumors should undergo Superficial Parotidectomy**
EXCEPT:
 - Involvement of Deep lobe → Total Parotidectomy.
- **ALL Malignant salivary tumors should be surgically excised:**
 - **Low Grade (Treated like benign tumors):**
 - Superficial Parotidectomy only.
 - Supraomohyoid ND → If Advance Stage (T3-T4) only.
 - No Post-op XRT.
 - **High Grade:**
 1. Total Excision.
 2. Neck Dissection:
 - **NO** → Supraomohyoid ND.
 - **N+** → MND.
 3. Post-op XRT.
- **Metastasis to lung is NOT a contraindication for Surgical Excision.**
- **If there is a pre-op Facial Nerve paralysis, consent the patient for:**
 - Total parotidectomy.
 - Neck Dissection.
 - Facial nerve sacrifice + intraoperative Frozen section +/- Mastoidectomy (to obtain negative proximal margin).
 - Cable graft.
- **Indications of Post-op Adjuvant Radiation in Salivary glands tumors:**
 - High grade tumors.
 - Advance Stage (III -IV).
 - Peri-neural invasion.
 - Vascular invasion
 - Positive margin.
 - Nodal extra-capsular invasion.
- **No rule of Chemotherapy in Salivary glands tumors.**

Benign Salivary Gland Diseases

- **Sialorrhea and Ptyalism:**
- Sialorrhea:
 - Excessive saliva production.
- Ptyalism:
 - Drooling of saliva.
- Causes:
 - Central neurological (Parkinson's disease, epilepsy, stroke)
 - Psychogenic
 - Parasympathomimetic medications (pilocarpine)
 - Diseased gland (tumor, inflammation)
 - GERD
 - Anatomic (macroglossia, orthodontic problem, oral incompetamce)
 - Swallowing disorders (relative ptyalism)
- Diagnosis:
 - Clinical exam.
- Management:
 - **Speech therapist and orthodontist:**
 - Evaluate swallowing, techniques to improve motor functions, orthodontic appliance.
 - **Anticholinergic drugs:**
 - Block parasympathetic.
 - Glycopyrrolate, Scopolamine.
 - **Botox injection:**
 - Into major salivary glands.
 - **Ductal rerouting procedure to the posterior cavity:**
 - Rarely done.
 - Often combined with bilateral sublingual gland excision.
 - **Parotid duct ligation:**
 - Risk of sialocele, dental caries.
 - Often combined with bilateral submandibuler duct excision.
 - **Submandibuler gland excision:**
 - Risk on marginal branch of facial nerve, external scar.
 - **Chorda tympani transection (Jacobson's nerve neurectomy):**
 - Transtympanic approach.
 - **Radiation.**

- **Xerostomia:**

- Causes:

- Central (rare)
- Primary salivary disorders (Sjögren's disease, radiation sialadenosis)
- Dehydration
- Medications (psychotropics, general anesthesia, beta blockers)
- Mouth breathing from nasal obstruction
- Stress
- Anxiety

- Complications:

- Risk of dental caries
- Impaired taste
- Malnutrition

- Treatment:

- Artificial saliva
- Frequent small drinks
- Pilocarpine Hydrochlorate drops
- Aggressive dental care

- **Acute Sialadenitis:**

- Pathogen:

- S. aureus (most common), strep. viridans, H. influenzae, strep. Pyogens and E. coli (bacteria = sapurative).
- Must also consider virus such as HIV, mumps, coxsacki, influenza.

- Pathophysiology:

- Salivary stasis or obstruction and retrograde migration of bacteria from oral cavity.
- May be postoperative parotiditis day3-7 due to dehydration and NPO status.
- Most commonly from parotid gland.
- Less in submandibular gland because more lysozyme, IgA and saialicacide bacteriostatic.

- Risks:

- Dehydration
- Elderly
- Postsurgical (GI procedures)
- Radiation and chemotherapy
- Sjögren's syndrome

- Clinical picture:

- Sudden onset of erythema, tenderness, warmth, and purulence at ductal orifice.
- 20% are bilateral.
- Auricle may protrude (parotitis)
- Trismus (movement of jaw cause pain)

- Diagnosis:

- Clinical history and exam
- Leukocytosis
- Cultures
- FNA is not required (unwise to be done in infection!)
- Sialography is contraindicated.

- Complication:

- Ductal fistula (cutaneous)
- Abscess (toxemia)
- Deep neck space invasion (Ludwig's angina)

- Treatment:

- Oral hygiene
- Rehydration
- Warm compresses
- Antimicrobial therapy (may require IV antibiotics for severe cases)
- Sialogogues
- Parotid massage
- Oral irrigations
- **If no resolution after 2–3 days then consider CT or U/S to evaluate for abscess (may require I&D).**

- **Mumps:**
- Most common cause of parotid swelling.
- Presents in pediatric population (4-6 years old).
- Reduced after MMR vaccination.
- Caused by paramyxoviruses.
 - Contracted by droplet infection and fomites.
 - Incubation period 2-3 weeks.
 - Patient is infective even before the symptoms and remains infective 7-10 days after parotid swelling subside.
- Clinical picture:
 - 75% bilateral (simultaneously or after some time).
 - Painful parotid swelling (may involve submandibular [rare isolated] and sublingual glands).
 - Initially may present malaise, fever, trismus and sore throat.
- Diagnosis:
 - Mumps titers
 - Hemoagglutination antigens
 - Virus may be cultured from saliva, urine, or CSF; high amylase
 - IgG indicate past exposure, but if ≥ 4 times titer it indicate recent infection.
 - IgM indicate recent infection
- Complications:
 - Sudden SNHL (unilateral) due to CN VIII involvement
 - Infertility (with orchitis)
 - Encephalitis
 - Pancreatitis
 - Nephritis
 - Aseptic meningitis
 - Others thyroiditis, myocarditis and arthritis
- Treatment:
 - Self-limiting (in about a week).
 - Requires only supportive care (hydration, analgesics).
 - Audiological evaluation.
 - Vaccine is available (mumps immunoglobulin is of no value).
- **Other Viral Infections:**
 - CMV (higher mortality in neonates)
 - Coxsackievirus, influenza A

- **Chronic Inflammation:**

- Types:

- 1. **Granulomatous:**

- Tuberculosis [acute inflammatory or chronic tumourous], atypical mycobacterium, actinomycosis (poor oral hygiene, dental caries), cat scratch fever, sarcoidosis [20%] (Uveoparotid Fever (Heerfordt's Syndrome).

- 2. **Secondary:**

- Progression from acute sialadenitis.

- Clinical picture:

- Xerostomia (perminant in 80%)
 - Slow painless enlargement
 - May be recurrent

- Diagnosis:

- Sialography (Tree-leaf appearance)
 - CT
 - PPD
 - Serum markers, VDRL, FTA-ABS, cat-scratch antigen test, etc

- Complications:

- Abscess, fistula, ductal destruction

- Treatment:

- **Medical:**

- Sialogogues, antibiotics, warm compress, massage.

- **Conservative Surgical Management:**

- Ductal dilatation

- **Destructive Surgical Management:**

- Gland excision, ductal ligation, gland irradiation, or neuronectomy

NOTE:

- Incisional biopsy should be avoided because of high risk of fistula formation (especially with mycobacterial infections).
- Secondary TB tend to affect SMG/SL, and associated with pulmonary TB.

What is recurrent parotitis of childhood?

- Recurrent uni or bilat swelling
- Stops in adolescence (usually). No xerostomia

Differential diagnosis of recurrent parotid swelling (Bailey)

- I) Autoimmune "pseudosialectasis"
 - i) Mikulicz's disease – localized to parotid gland (1) Adult and pediatric
 - ii) Primary Sjogren's
 - iii) Secondary Sjogren's
- II) Nonautoimmune (Mikulicz's syndrome)
 - i) Recurrent sialoadenitis
 - ii) Sialosis
 - iii) Multinodular gland

- **Uveoparotid Fever (Heerfordt's Syndrome):**

- More common in women.
- Pathophysiology:
 - Extra-pulmonary form of sarcoidosis.
 - Nerve dysfunction may occur secondary to local ischemia\infarct from granulomatous infiltration.
- Clinical picture:
 - Self-limited uveitis, parotid enlargement, facial palsy (50%), fever.
- Diagnosis:
 - Based on clinical history and exam and evidence of sarcoidosis in salivary gland tissue.
- Treatment:
 - Corticosteroids, ocular care (artificial tears, ocular ointment at night, eye patch at night).

- **Kuettner's Tumor (Chronic Sclerosing Sialadenitis of the Submandibular Gland):**

- Pathophysiology:
 - May be autoimmune mediated.
- Clinical picture:
 - Firm, enlargement of submandibular gland (similar to tumor or malignancy), may be painful.
- Diagnosis:
 - Biopsy (submandibular excision).
- Histopathology:
 - Chronic inflammation with destruction of acinar cells, sclerosis, "cirrhotic" changes.
- Treatment:
 - Submandibular excision for diagnosis and treatment.

- **Radiation Sialadenitis**

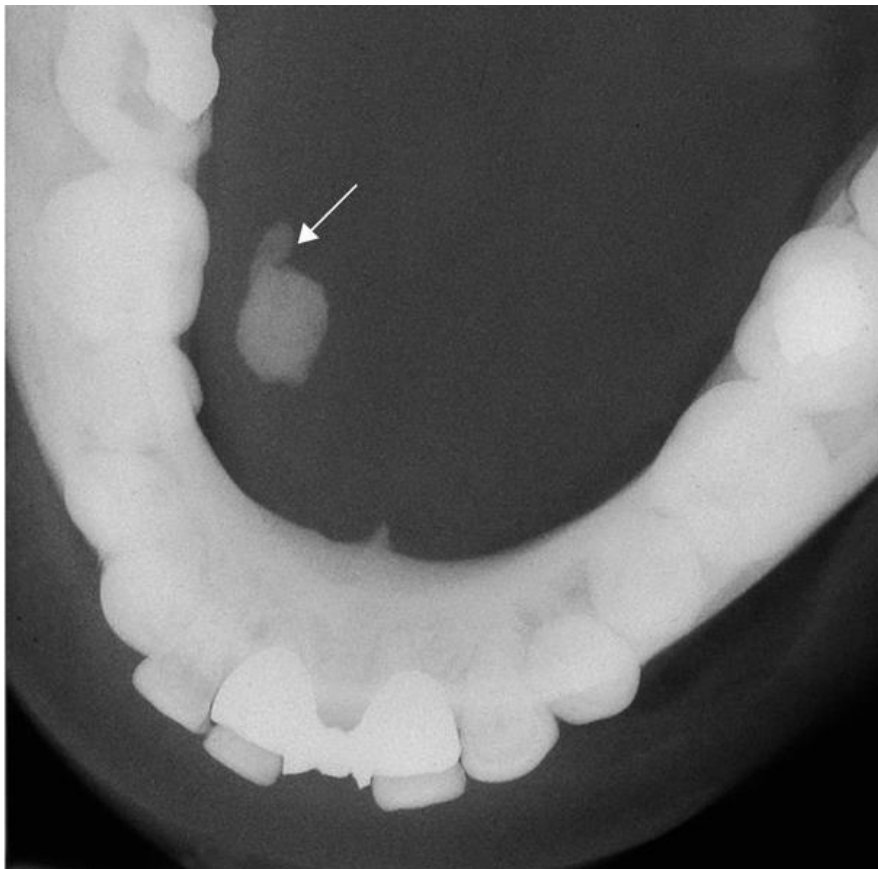
- Often permanent if exposed to >40–50 Gy
- Clinical picture:
 - Xerostomia, hypogeusia, ageusia.
- Diagnosis:
 - Clinical exam and history of radiation exposure
- Histopathology:
 - Interstitial fibrosis.
- Treatment:
 - Symptomatic (pilocarpine drops, artificial saliva, frequent drinks), dental care (fluoride rinses).

- **Salivary Calculi (Sialolithiasis):**

- It is deposition of concentration within the ductal system of the gland [75% single stone].
- Associated with gout (uric acid calculi) [only systemic disease cause stones].
- Most common in submandibular gland (80%).
 - Longer duct
 - saliva alkali
 - More calcium and phosphate
 - More mucus viscosity
 - antigravity flow.
- The rest is 19% in parotid and 1% in SLG.
- Pathophysiology:
 - Change in the viscosity of the saliva may cause mucous obstruction, injury to epithelial ductal system, salivary stagnation (dehydration), calcium phosphate and calcium carbonate precipitation causing obstruction.
- Clinical picture:
 - Recurrent pain, swelling, worse with meals (salivary colic), sometimes stone is visible or can be palpated.
- Diagnosis:
 - Stone may be palpable.
 - Sialography (100% sensitivity).
 - X-ray (90% of submandibular calculi are radiopaque, 90% of parotid calculi are radiolucent).
 - CT (100% sensitivity).
- Complications:
 - Fistulas
 - Acute suppurative sialoadenitis
 - Ductal strictures
- Treatment:
 - Gland massage
 - Bimanual expression
 - Transoral incision
 - Sialodochoplasty (reconstruct duct)
 - Gland excision:
 - If recurrent or if stone is lodged within substance of the gland.
 - Extracorporeal lithotripsy (controversial)
 - Sialoscopy

El Deeb's predisposing factors for submandibular gland calculi formation

- I) Submandibular saliva
 - i) High mucin content
 - ii) Alkaline pH
 - iii) Higher percentage of organic matter
 - iv) Concentration of calcium & phosphate salts
 - v) Low carbon dioxide level
 - vi) High phosphatase enzyme content
- II) Anatomy
 - i) Length & irregular course of Wharton's duct
 - ii) Dependent position of gland & duct system
 - iii) Position of ductal orifice
 - iv) Size of orifice smaller than duct lumen



- **Sjögren's Syndrome (Myoepithelial Sialadenitis, Benign Lymphopithelial Lesion):**
- Pathophysiology:
 - Systemic autoimmune destruction of exocrine glands.
 - B-cell hyperactivity (lymphocytic infiltration).
 - Most common in middle-aged women
 - Associated with Non-Hodgkin's Lymphoma
- Types
 - **Primary:**
 - Exocrine gland involvement only.
 - **Secondary:**
 - Associated with other connective tissue disorders (most commonly rheumatoid arthritis) with keratoconjunctivitis sicca and xerostomia.
 - Other Sicca-like Causes:
 - Aging
 - Medications (diuretics, anticholinergics, antihistamines, antidepressants)
 - Dehydration
 - Hepatitis
- Clinical picture:
 - Keratoconjunctiva sicca (filamentary keratitis, sandy sensations in eyes).
 - Xerostomia (dental caries, dry mucosa).
 - Intermittent bilateral parotid swelling (atrophy at end-stage of disease).
 - Achlorhydria.
 - Raynaud's phenomenon (excessively reduced blood flow in response to cold or emotional stress, causing discoloration of the fingers, toes, and occasionally other areas).
 - Pancreatitis, Myositis, Glomerulonephritis, Hepatosplenomegaly.
- Diagnosis
 - Clinical History (must have 2 of 3):
 - Keratoconjunctiva sicca, xerostomia, or other connective tissue disease.
 - Biopsy:
 - Lip biopsy or minor salivary glands reveals lymphocytic infiltration with glandular atrophy.
 - Serology:
 - ANA, RF, ESR, **SS-A and SS-B** (Antibodies specific for primary Sjögren's syndrome), Decreased IgM (suggest higher risk of progression to malignancy).

- Sialography:
 - Globular, multiple contrast collections throughout gland (Pine tree appearance).
- Schirmer test:
 - Evaluates tear production.
- Treatment:
 - Artificial saliva and tears.
 - Frequent small drinks.
 - Consider pilocarpine hydrochlorate drops and oral corticosteroids for severe acute exacerbation.
- **Mikulicz's Syndrome (Sialadenosis)**
- Recurrent non-tender non-inflammatory non-neoplastic salivary gland swelling.
- Pathophysiology:
 - Secondary to underlying:
 - Nutritional pathology (anorexia & bulimia)
 - Endocrine or metabolic pathology (cirrhosis, diabetes, malnutrition, ovarian, thyroid, or pancreatic insufficiency)
 - Medications (hypertensive medications, catecholamines, iodine-containing compounds)
 - Pregnancy and lactation
- Clinical picture:
 - Recurrent bilateral non-tender parotid swelling.
 - Painful sialadenosis is associated with antihypertensive medications.
- Treat the underlying cause.
- **Sialectasis:**
- Dilatation of the ductal system, leading to stasis of secretions, which predisposes to infection.
- Clinically, sialectasis resembles chronic recurrent sialadenitis, but can be differentiated from it by sialography.
- Different degrees (punctuate, globular or cavitary type) may be seen.
- Sialectasis may be congenital, associated with granulomatous disease or autoimmune disease such as Sjogren's syndrome

Salivary Gland Cysts

- Most common mass in parotid.
- Risks:
 - Sialoadenitis
 - Sialolithiasis
 - Trauma
- Diagnosis:
 - CT scan or U/S to confirm cystic nature
 - FNA to evaluate for malignant cells
- Treatment:
 - May observe or inject with sclerosing agents (tetracycline).
 - Surgical excision for cosmesis, recurrent lesions, solid lesions, abnormal cytology.

What are cysts of salivary glands?

- Congenital
- Branchial cleft
- Dermmoid
- Epidermoid
- Acquired
- Post-traumatic sialocele
- Retention mucocele
- Benign lymphocytic lesions (BLL) of HIV

Differential diagnosis of salivary gland cysts

- I) Congenital
 - i) Branchial cleft
 - ii) Epidermoid
 - iii) Dermoid
- II) Acquired
 - i) Benign lymphoepithelial lesion (AIDS associated)
 - ii) Mucus retention cyst/mucocele
 - iii) Sialocele (pseudocyst), associated with trauma

- **Benign Lymphoepithelial Cysts:**
 - Increased incidence since HIV epidemic.
 - May progress to pseudolymphoma.
 - Clinical picture:
 - Multiple, maybe bilateral parotid cysts, asymptomatic.
 - Diagnosis:
 - Clinical history and exam, FNA.
 - Histopathology:
 - Lymphoreticular infiltrate, clusters of lymphoid tissue (germinal centers), acinar atrophy, ductal metaplasia.
 - Treatment:
 - Antiviral therapy in HIV may cause regression of cyst
 - Aspiration (more common especially in HIV) or excision (superficial parotidectomy, less common).
 - May also consider sclerosing agents (doxycycline).
- **Mucous Retention Cysts and Ranulas**
 - Pathophysiology:
 - Obstruction of minor salivary glands (may be from trauma).
 - **Mucous Retention Cysts:**
 - True cysts of the minor salivary glands (lined with epithelial layer).
 - **Ranulas:**
 - Mucous retention cyst of the floor of mouth, usually from the sublingual gland.
 - **Plunging Ranula:**
 - Extension into cervical tissue (mylohyoid muscles), may present as a neck mass.
 - **Mucocele:**
 - Not a true cyst, extravasation of mucus into soft tissue, usually from trauma to duct (Post-traumatic Sialocele) [most common in lower lip].
 - Clinical picture:
 - Cystic mass on floor of mouth, lip, buccal mucosa, or minor salivary gland.
 - Diagnosis:
 - clinical history and exam, excisional biopsy
 - Treatment:
 - Excision or marsupialization (cutting a slit into the cyst and suturing the edges of the slit to form a continuous surface from the exterior surface to the interior surface of the cyst).

- **Congenital Cysts:**

- Dermoid cysts:
 - Keratinizing squamous epithelium.
 - Contains skin appendages.
- Ductal cyst:
 - Manifest in infancy.
 - Requires sialography for diagnosis.
 - No therapy needed unless repeated infections.
- First arch branchial cleft cyst:
 - Accounts for less than 1% of all branchial cleft anomalies.
 - Type 1 is ectodermal only, duplication of membranous/cartilaginous EAC.
 - Type 2 is ectoderm and mesoderm of 1st and 2nd arches.

- **Necrotizing Sialometaplasia:**

- Non-neoplastic, (inflammatory) self-healing process of unknown etiology of the salivary glands
- Pathophysiology:
 - May be secondary to vascular ischemia.
- Clinical picture:
 - Asymptomatic mucosal ulceration or nodular lesion of the minor salivary gland.
 - Most common site is hard palate.
- Diagnosis:
 - Excisional biopsy
- Histopathology:
 - Lobular necrosis, ductal and acinar squamous metaplasia, pseudoepitheliomatous hyperplasia may be present.
 - May be mistaken as malignant, SCC, or mucoepidermoid Ca.
- Treatment:
 - Self-limiting.

- **Parotid Trauma:**

- Penetrating parotid injury may damage duct or nerve.
- Any penetrating injury posterior to the anterior border of the masseter muscle should be suspected of causing a ductal injury.
- Probe duct to confirm its condition:
 - If cut, end to end anastomosis with 9-0 sutures over a catheter is the treatment of choice.
 - The catheter is then sutured in place to the buccal mucosa and to be removed in 2 weeks.
 - If not enough length, can suture the duct directly into oral cavity through puncture.
 - Always ligate the duct if all else fails.
- For parenchymal lacerations:
 - Just close it up in layers.
- For fistula/sialocele:
 - Repeated aspiration and compression may be required.
 - Persistent fistula means the duct is probably obstructed.
 - Sialography may be performed.
- Blunt trauma:
 - Edema or hemorrhage which is usually transient.
 - Large hematomas should be drained before they cause scarring.
- If the injury is medial to a vertical line from lateral canthus to the mental foramen, facial nerve repair is unnecessary (even with clear dysfunction) because recovery is likely.

- **Other Disorders:**

- Pneumoparotitis:
 - Glass blowers and after intubation and endoscopy or idiopathic.
- Cheilitis glandularis:
 - Enlargement of labial salivary glands.
 - Secretion of thick, sticky mucous.
- Kussmaul disease (sialodochitis fibrinosa):
 - Mucous plug obstructs collecting duct
 - Dehydration; recurrent swelling and pain.
- Drugs that cause salivary gland enlargement:
 - Isoproterenol
 - Ethambutol
 - Phenbutazone
 - Phenothiazine
 - Iodine compounds
 - Heavy metals

TABLE 39.1**SYSTEMIC AND EXOGENOUS SOURCES OF SALIVARY DYSFUNCTION**

Alzheimer disease	Dehydration
Cystic fibrosis	Radiation therapy
Diabetes	Various medications
HIV/AIDS	Chemotherapy
SLE	
Parkinson disease	
RA	
Sarcoidosis	
Scleroderma	
Sjögren syndrome	

DISEASES OF THE SALIVARY GLAND

Disease	Features	Management
Viral	Acute onset; associated symptoms	Symptomatic
Acute suppurative sialadenitis	Acute onset; postoperative or debilitated patient; purulent saliva from duct; diffusely swollen and painful gland	Hydration; massage; antistaphylococcal antibiotic
Chronic sialadenitis	Middle painful recurrent swelling; decreased saliva production	Sialagogues; massage; antibiotics for acute exacerbations
Tuberculosis	Acute inflammatory lesion or chronic swelling	Antituberculous drugs if organism is susceptible—excision otherwise
Actinomycosis	Painless swelling, <u>often with drainage</u>	Long-term antibiotics
Sarcoidosis	Usually asymptomatic swelling	Symptomatic
Sjögren syndrome	Painless swelling of multiple salivary glands; may be associated with another autoimmune disease	Symptomatic
Sialolithiasis	Painful swelling, usually associated with eating	Removal of calculus
First branchial cleft cyst	Repeated acute suppurative swelling	Excision, usually with parotidectomy
Penetrating injury	Obvious defect with appropriate history	Careful evaluation repair
Sialadenosis	Asymptomatic swelling	Workup to find underlying cause

Salivary Glands Tumors:

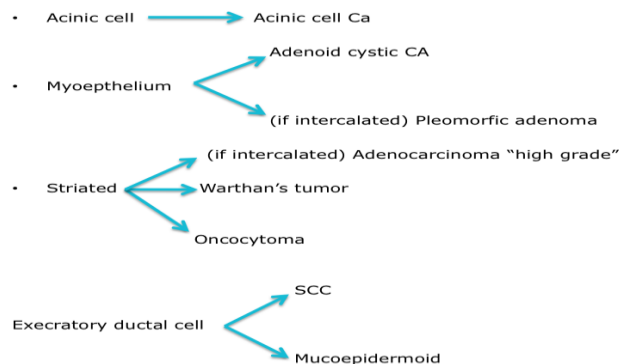
- 2% of Head and Neck tumors (relatively uncommon).
- 95% of salivary glands are found in Adults.
 - o 80% are located in parotid.
 - o 15% are located in Submandibular.
 - o 5% in Sublingual/Minor salivary glands.
- The smaller the gland the more likely malignant.
 - o 80% of parotid tumors are benign.
 - o 50% of submandibular tumors are benign.
 - o 40% of sublingual/minor glands are benign.
- Rule of 80:
 - o 80% of Salivary tumors are in Parotid.
 - o 80% of Parotid tumors are Benign.
 - o 80% of Benign Parotid tumors are Pleomorphic Adenoma.
- **In Adults:**
- **Benign Tumors:**
 - o Most common is Pleomorphic Adenoma.
 - o 2nd most common is Warthin's Tumor.
- **Malignant Tumors:**
 - o **Parotid:**
 - Most common is Mucoepidermoid Ca.
 - 2nd most common is Adenoid Cystic Ca.
 - o **Submandibular and Minor salivary glands:**
 - Most common is Adenoid Cystic Ca.
 - 2nd most common is Mucoepidermoid Ca.
- **In Pediatrics:**
- **Benign Tumors:**
 - o Most common is Hemangioma.
 - o 2nd most common is Pleomorphic Adenoma.
- **Malignant Tumors (85% in Parotid):**
 - o Most common is Mucoepidermoid Ca.
 - o 2nd most common is Acinic Cell Ca.

	ALL	Adults	Children
ALL	1. Pleomorphic Adenoma (Benign Mixed Tumor) 2. Mucoepidermoid Ca.	1. Pleomorphic Adenoma (Benign Mixed Tumor) 2. Mucoepidermoid Ca.	1. Vascular Tumors (Hemangioma) 2. Pleomorphic Adenoma
Malignant	1. Mucoepidermoid Ca. 2. Adenoid Cystic Ca. 3. Adenocarcinoma	1. Mucoepidermoid Ca. 2. Adenoid Cystic Ca. 3. Adenocarcinoma	1. Mucoepidermoid Ca. 2. Acinic Cell Ca.
Benign	1. Pleomorphic Adenoma (Benign Mixed Tumor) 2. Warthin's Tumor	1. Pleomorphic Adenoma (Benign Mixed Tumor) 2. Warthin's Tumor	1. Vascular Tumors (Hemangioma) 2. Pleomorphic Adenoma

- **Multicellular Theory:**

- Neoplastic cells originate from secretory units counterparts:

- Acinic cells:
 - Acinic cell ca.
- Myoepithelial cells only:
 - Adenoid cystic.
- Intercalated ducts and Myoepithelial cells:
 - Pleomorphic Tumors
- Intercalated ducts and Striated ducts:
 - Adenocarcinoma
- Striated ducts only:
 - Warthin's Tumors.
 - Oncocytic Tumors
 - Adenocarcinoma.
- Excretory ducts:
 - Mucoepidermoid ca.
 - Squamous cell ca.



- **Bicellular Theory:**

- All neoplastic cells differentiate from basal cells of excretory and intercalated ducts.

- Intercalated ducts:
 - Pleomorphic adenoma
 - Warthin's tumor
 - Oncocytoma
 - Acinic cell
 - Adenoid cystic
- Excretory ducts:
 - Mucoepidermoid.
 - Squamous cell

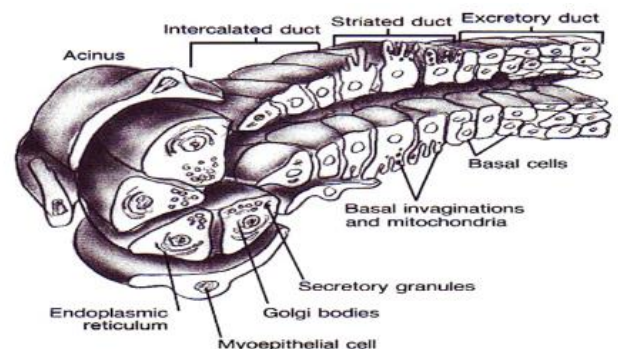
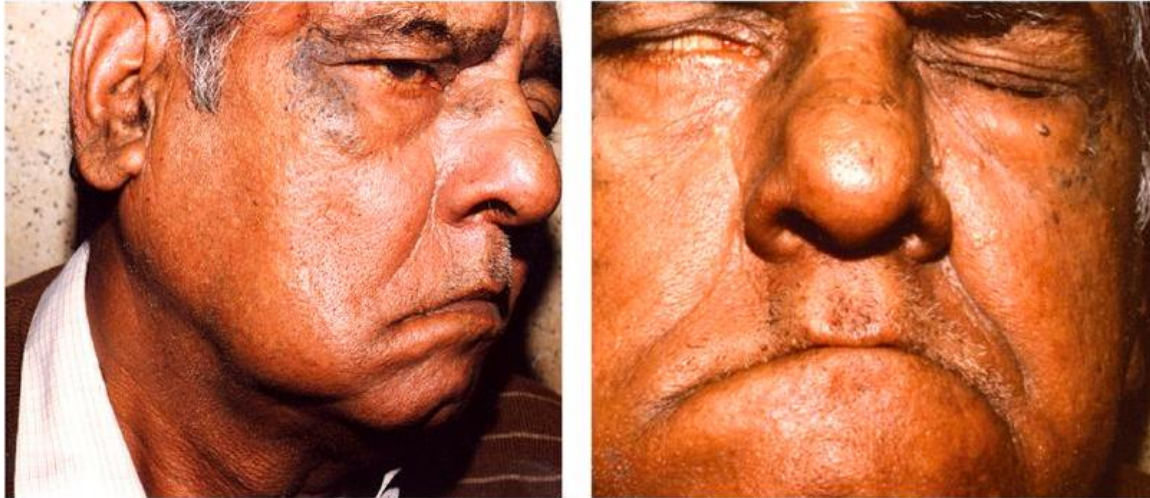


Table 45-1. Tumours of salivary glands

Benign	Malignant
Epithelial • Pleomorphic adenoma • Adenolymphoma (Warthin's tumour) • Oncocytoma • Other adenomas Mesenchymal • Haemangioma • Lymphangioma • Lipoma • Neurofibroma	Epithelial • Mucoepidermoid carcinoma ○ Low grade ○ High grade • Adenoid cystic carcinoma (cylindroma) • Acinic cell carcinoma • Adenocarcinoma • Malignant mixed tumour • Squamous cell carcinoma • Undifferentiated carcinoma Mesenchymal • Lymphoma • Sarcoma

- **Risk Factors for Salivary Gland Tumors:**
 - 1. Radiation:**
 - Mainly with Pleomorphic adenoma and Mucoepidermoid.
 - Latency of 7–30 years.
 - 2. Smoking:**
 - Only associated with Warthin's tumors.
 - 3. EBV:**
 - Only associated with undifferentiated carcinoma
- Alcohol is **NOT** a risk factor for salivary gland tumors.
- Most common bilateral salivary gland tumors:
 - Warthin's tumor (10%)
 - Oncocytoma
 - Acinic Cell (3%)

- **Benign Salivary Tumors:**
 1. Pleomorphic Adenoma
 2. Warthin's Tumor
 3. Oncocytoma
 4. Monomorphic Adenoma
 5. Hemangioma
- **Pleomorphic Adenoma (Benign Mixed Tumor):**
- Benign heterogeneous tumor composed of epithelial and myoepithelial components.
- Most common of all salivary gland tumors:
 - 70% of Parotid tumors.
 - 50% of Submandibular tumors.
 - 45% of Minor salivary gland tumors.
 - 6% of sublingual tumors
- More common in women.
- Clinical picture:
 - Slow-growing, Painless, Unilateral, Firm.
 - Rarely:
 - Dysphagia (Pharyngeal extension).
 - Dyspnea and Hoarseness (laryngeal involvement),
 - Facial nerve palsy.
 - **Malignant Transformation:**
 - Carcinoma ex-pleomorphic adenoma.
 - Sarcoma.
 - Location:
 - Parotid:
 - 90% in Superficial lobe (Mainly the tail).
 - 10% in Deep lobe with intraoral swelling.
 - Minor salivary gland:
 - Lateral palate.
 - Submucosal mass.
- Diagnosis:
 - U/S
 - CT with contrast
 - MRI
 - FNA
- Histopathology
 - Myoepithelial component:
 - Spindle shaped with hyperchromatic nuclei, may be more than one cell layer thick.
 - Epithelial components:
 - Varied growth patterns (trabecular, solid, cystic, papillary).
 - Stromal components:
 - Product of myoepithelial cells: myxoid, chondroid, fibroid, or osteoid components



- Management:
- Surgical excision with Superficial Parotidectomy or submandibular gland excision.
 - o Total Parotidectomy for deep lobe involvement.
 - o Facial nerve preservation.
 - o Wide surgical margin for pseudopod extensions to prevent recurrence.
- >90% 10-year cure rate.
- **Radioresistant.**
- **Warthin's Tumor (Adenolymphoma):**
- Papillary Cystadenoma Lymphomatosum.
- 2nd most common salivary gland tumor.
- Mainly found in parotid gland.
- Almost exclusively found in middle-aged to elderly men.
- Associated with smoking.
- 10% bilateral (Synchronous or Metachronous).
- 10% multicentric.
- Rare malignant transformation.
- Pathophysiology:
 - o Entrapped lymphoid tissue (parotid is the last gland embryologically to be encapsulated).
 - o Ectopic ductal epithelium that develops within intraparotid lymph nodes.
 - o Hypersensitivity disease resulting in metaplasia of the duct
- Clinical picture:
 - o Slow-growing
 - o Painless
 - o Unilateral
 - o Cystic
 - o Compressible mass.
 - o Mostly involve the tail of the parotid

Diagnosis:

- U/S
- CT with contrast
- MRI
- FNA
- **Techmetium 99m scan:**
 - o Concentrates technetium-99m due to the presence of high mitochondrial content of oncocytes.
- **Histopathology**
 - o Epithelial components:
 - lines papillary projections; double lining of oncocytes; inner or luminal cells, nonciliated, tall columnar nuclei at luminal aspect; outer or basal cells are round, cuboidal with vesicular nuclei.
 - o Lymphoid components:
 - Mature lymphocytes with germinal centers.
 - o Mucous secreting cells
- **Management:**
- Surgical excision with Superficial Parotidectomy or submandibular gland excision.
 - o Total Parotidectomy for deep lobe involvement.
 - o Facial nerve preservation.
- **Oncocytoma (Oxyphilic Adenoma):**
- Rare benign tumor exclusively of oncocytic cells.
 - o 1% of salivary gland tumors.
- Rare malignant low-grade tumor transformation.
- Mainly found in parotid gland.
- Concentrates technetium-99m due to the presence of high mitochondrial content of oncocytes.
- **Histopathology**
 - o Encapsulated (cystic rather than solid)
 - o Oncocytic cells
 - o Granularity from mitochondria (large number of mitochondria)
- **Monomorphic Adenoma:**
- Similar to pleomorphic except no mesenchymal stromal component.
- Predominantly an epithelial component or (rarely) the myoepithelial component.
- More common in the minor salivary glands (upper lip).
- 10% bilateral.
- Rare malignant potential
- **Types:**
 - o Basal Cell Adenoma
 - o Clear Cell Adenoma
 - o Glycogen-Rich Adenoma

- **Hemangiomas:**
 - Benign tumor of endothelial origin.
 - Most common benign parotid tumor in kids.
 - Usually discovered few weeks after birth, enlarge for 6-12 months, but typically regress by 2nd year of life.
 - More common in females
 - 50% association with cutaneous hemangiomas
 - Cutaneous hemangioma can present anywhere in H&N (also common in parotid, lip [oral cavity] and larynx).
 - Complications of H&N hemangioma:
 - o Cosmetic
 - o Ulceration, infection and bleeding
 - o Airway compromise (laryngeal)
 - o Thrombocytopenia and high output cardiac failure (rare)
- **Lymphangiomas:**
 - They are less common and may involve parotid and submandibular glands.
 - On palpation, they feel soft and cystic.
 - They do not regress spontaneously and are surgically excised.
- **Lipoma** and **neurofibroma** are rare

- **Malignant Salivary Tumors:**

1. Mucoepidermoid Carcinoma
2. Adenoid Cystic Carcinoma (Perineural invasion)
3. Acinic Cell Carcinoma
4. Adenocarcinoma
5. Malignant Mixed Tumour
6. Squamous Cell Carcinoma
7. Undifferentiated Carcinoma
8. Lymphoma
9. Sarcoma

Table 1 General categories of management of primary salivary gland carcinomas [3]

Surgery alone	Surgery and radiotherapy	Additional neck dissection	Systemic chemotherapy
Negative margins	Close (<2 mm) or positive margins	All cN+	Metastatic or unresectable disease
Low grade histology	High grade histology	cN0 but high grade histology	
Low risk (non angioinvasive, non infiltrative) histologic subtype	High risk (highly infiltrative) histologic subtype	cN0 but high risk (angioinvasive) histologic subtype	
Low T stage (T1 or T2)	High T stage (T3 or T4) pN+ Perineural invasion ^a	cN0 but high T stage (T3 or T4)	

T = tumor stage in TNM classification, cN+ = clinically node positive, cN0 = clinically node negative, pN+ = pathologically node positive

^a Somewhat controversial depending on tumor type

Table 2 Risk stratification of WHO [1] recognized salivary gland malignancies

Low risk	High Risk
Acinic cell carcinoma	Sebaceous carcinoma and lymphadenocarcinoma
Low grade mucoepidermoid carcinoma ^a	High grade mucoepidermoid carcinoma ^a
Epithelial-myoepithelial carcinoma	Adenoid cystic carcinoma ^b
Polymorphous low grade adenocarcinoma	Mucinous adenocarcinoma
Clear cell carcinoma	Squamous cell carcinoma
Basal cell adenocarcinoma	Small cell carcinoma
Low grade salivary duct carcinoma (low grade cribriform cystadenocarcinoma)	Large cell carcinoma
Myoepithelial carcinoma	Lymphoepithelial carcinoma
Oncocytic carcinoma	Metastasizing pleomorphic adenoma
Carcinoma ex pleomorphic adenoma (intracapsular/minimally invasive or with low grade histology)	Carcinoma ex pleomorphic adenoma (widely invasive or high grade histology)
Sialoblastoma	Carcinosarcoma
Adenocarcinoma NOS and Cystadenocarcinoma, low grade ^a	Adenocarcinoma and cystadenocarcinoma, NOS, high grade ^a

^a Intermediate grade variants of these tumors are controversial in the assignment of risk. For mucoepidermoid carcinoma this may depend on grading scheme used. For adenocarcinoma NOS, there is little data, but what is present suggests that intermediate grade should be placed in the high risk group

^b Adenoid cystic carcinomas are all considered high risk in terms of local recurrence, but only solid adenoid cystic carcinoma (i.e. high pattern grade) is considered high risk for nodal metastasis

- **TNM Staging for Salivary Glands:**
- **PRIMARY TUMOR (T):**
- **TX:** Primary tumor cannot be assessed.
- **T0:** No evidence of primary tumor.
- **T1:** Tumor 2 cm or less without extraparenchymal extension.
- **T2:** Tumor greater than 2 cm but not more than 4 cm without extraparenchymal extension.
- **T3:** Tumor more than 4 cm and/or tumor having extraparenchymal extension
- **T4a:** Moderately advanced local disease, Tumor invades the skin, mandible, ear canal, and/or facial nerve.
- **T4b:** Very advanced local disease, Tumor invades the skull base and/or pterygoid plates and/or encases the carotid artery.
- Extraparenchymal extension is a clinical macroscopic evidence of invasion of soft tissues.
- Microscopic evidence alone does not constitute extraparenchymal extension for classification purposes.
- **Regional lymph nodes (N) and Distant metastasis (M) are similar to mucosal SCC.**

H. TNM Staging for the Larynx, Oropharynx, Hypopharynx, Oral Cavity, Salivary Glands, and Paranasal Sinuses

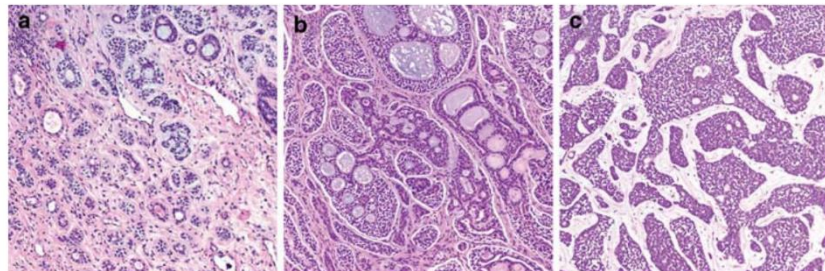
Stage Grouping			
Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
	T1	N1	M0
	T2	N1	M0
	T3	N1	M0
Stage IVA	T4a	N0	M0
	T4a	N1	M0
	T1	N2	M0
	T2	N2	M0
	T3	N2	M0
	T4a	N2	M0
Stage IVB	Any T	N3	M
	T4b	Any N	M0
Stage IVC	Any T	Any N	M1

- **Mucoepidermoid Carcinoma:**
- Most common parotid salivary gland malignancy in children and adults
- 30% of all salivary malignancies.
- Some pathologists do not consider it to be malignant and call it mucoepidermoid tumor and not cancer, but it is known to metastasize and kill.
- Most commonly found in the parotid > minor salivary > submandible.
- Commonly induced by radiation.
- 30–70% overall regional metastatic potential.
- Slow-growing but can invade the facial nerve.
- Histology:
 - o Mucin-producing cells and squamous cells "mucoepidermoid".
 - o Greater the epidermoid element, more malignant is the behavior of the tumor (Higher grade tumor).
- Behavior of mucoepidermoid tumors:
 - o More aggressive in minor salivary glands.
 - o Behave like pleomorphic adenoma in major salivary glands.

Low grade tumors (Well-Differentiated)	High grade tumors (Poorly-Differentiated)
- Good prognosis (70-90% 5-years survival rate). - More common in children. - Can invade and metastasize. - Slow growing, painless mass. - <u>Histopathology:</u> - More mucinous elements - Less epidermoid cells - Positive keratin staining. - <u>Gross:</u> - Small and partially encapsulated. - <u>Treatment (As benign):</u> - Superficial or total parotidectomy depending on the location of the tumor. - Facial nerve is preserved. - N0 and ≥T3 → Supraomohyoid ND. - N+ve → MND.	- More aggressive - Poor prognosis (30-50% 5-years survival rate). - More likely to metastasize - Rapidly enlarging, +/- pain - <u>Histopathology:</u> - More epidermoid cells - Less mucinous elements. - Requires <u>mucin staining</u> to differentiate from SCC. - Positive keratin staining - <u>Stains:</u> - Mucicarmine, PAS, alcian blue - <u>Gross :</u> - Larger and locally invasive - <u>Treatment:</u> - Total parotidectomy. - N0 and ≥T1 → Supraomohyoid ND. - N+ve → MND. - Facial nerve may be sacrificed if invaded by tumor. - Post-op XRT.

- **Adenoid Cystic Carcinoma (Cylindroma):**
- M=F; 5th decade
- Insidious slow-growing tumor but infiltrates widely into the tissue planes and muscles.
- Monolobular, either no capsule or partially.
- Invades perineural spaces and lymphatics and thus causes pain and VIIth nerve paralysis.
- Most common in submandibular and minor gland malignancy.
- Local recurrences after surgical excision are common and can occur as late as 10-20 years after surgery.
- Distant metastases go to the lung, brain and bone.
- Histopathological types: **STC**
 - **Solid:**
 - High-grade tumor.
 - Dense cellular pattern with few spaces.
 - Higher risk of metastasis.
 - **Tubular:**
 - Low-grade tumor.
 - Best prognosis.
 - **Cribiform:**
 - Low-grade tumor.
 - Most common type.
 - Nests of cells with round spaces.
 - "Swiss cheese" appearance.

Fig. 2 The various patterns/grades of adenoid cystic carcinoma. **a** Tubular, **b** cribriform, **c** solid. All grades are cytologically monomorphic and retain small dark angulated nuclear features



- Treatment:
- Total parotidectomy with facial nerve resection if involved.
- **N0 and $\geq T1$ $\rightarrow$ Supraomohyoid ND.**
- **N+ve $\rightarrow$ MND.**
- Post-operative radiation for high grade type.
- Long-term follow-up required because of indolent course and possible distant metastasis.
- Prognosis:
- High-grade associated with poor prognosis (<20% 5-year survival).
- Low-grade up to a 100% 5-year survival.
- Local recurrence: 42%.

- **Acinic Cell Carcinoma:**
- Low grade tumor which appears similar to a benign mixed tumor (pleomorphic adenoma).
- F>M; 4th-6th decade; multi-centric.
- Most commonly found in the parotid (serous acinar cells)
- Second most common salivary gland cancer in pediatrics.
- It presents as a small, firm, movable and encapsulated tumor.
- Well circumscribed, fibrous capsule
- 3% are bilateral.
- Metastases are rare.
- Histopathology:
 - o 2 types serous acinar cells or clear cytoplasm cells.
 - o Amyloid is a hallmark pathologic finding.
 - o 4 configurations (microcystic, papillary, solid, follicular).
 - o Lymphoid infiltrate.
 - o Positive on periodic acid-Schiff staining
- Treatment:
- Superficial or total parotidectomy is adopted.
- Neck dissection: " low grade "
- **N0 and ≥T3 → Supraomohyoid ND.**
- **N+ve → MND**
- Adjuvant radiation therapy may be considered for advanced disease:
 - o CN VII involvement
 - o N+
 - o Skin involvement
 - o Positive margin.
- Good prognosis (63–87% 10-year survival).

- **Adenocarcinoma:**
- Highly aggressive locally and sends distant metastasis.
- Strong propensity for recurrence
- Most often it arises in minor salivary glands (**palate**; buccal mucosa; upper lip) then parotid gland.
- M=F
- Gross:
 - o Firm/hard, attached to surrounding tissue.
- Histology:
 - o Cylindric cells of various height; forms papillae, acini or solid.
 - o Produces mucous (mucicarmine stain).
 - o No keratin like mucoepidemoid,
 - o Histologic classification "Depends on degree of glandular structure cellular differentiation":
 - Grade I – Well-formed ductal structures
 - Grade III – Solid growth pattern with few glandular characteristics

- **Malignant Mixed Tumor:**
- High-grade tumor, aggressive, explosive growth rate.
- Poor prognosis (<50% 5-year survival).
- Malignant transformation of pleomorphic adenoma is 1.5% in the first 5 years and increases to 10% in more than 15 years.
- There are varieties of this tumor:

Carcinoma Ex-Pleomorphic Adenoma	Metastasizing Mixed Tumor	Carcinosarcoma	Noninvasive Carcinoma
2–3% malignant transformation from pleomorphic adenomas. Carcinoma components only (Epithelial component). <u>Histology:</u> Lesion in background of pleomorphic adenomas. Neurovascular invasion and necrosis. Difficult diagnosis due to mixed histology, look for infiltrative growth.	Metastasizing Mixed Tumor Distinct from carcinoma expleomorphic Remains histologically benign.	Contains components of both carcinomas epithelial and sarcomas stromal.	Carcinoma in situ within a pleomorphic adenoma

- Treatment:
- Total parotidectomy.
- Facial nerve if sacrificed during operation should be grafted immediately.
- N0 and $\geq T1 \rightarrow$ Supraomohyoid ND.
- N+ve $\rightarrow$ MND
- Postoperative radiation therapy

- **Squamous Cell Carcinoma**
- Rare; 0.3-1.5% of all tumors.
- M>F; 7th decade.
- It is a rapidly growing tumor that infiltrates, causes pain, and ulcerates through the skin.
- It can metastasise to neck nodes.
- Gross: firm, indurated mass
- Micro: intracellular keratinization; intercellular bridges; keratin pearls.
- Must rule out the following in parotid SCC:
 1. High grade Mucoepidermoid ca
 2. Metastatic SCC to intraglandular nodes
 3. Direct extension of Cutaneous SCC to parotid
- Treatment
- Total parotidectomy which may include cuff of muscle or even a portion of mandible, temporal bone and the involved skin.
- N0 and $\geq T1 \rightarrow$ Supraomohyoid ND.
- N+ve $\rightarrow$ MND.
- Post-operative radiation to primary site and the neck.
- **Undifferentiated Carcinoma:**
- It is a rare, but aggressive tumor.
- Parotid is common, linked to EBV.
- Subdivided:
 - o Small cell
 - o Large cell
 - o Lymphoepithelial "good prognosis"
- It has a tendency to spread rapidly, causes pain, becomes fixed to skin and ulcerates.
- It causes facial paralysis and cervical nodal metastasis.
- Treatment is wide excision, MRND "high grade" and post-operative radiation.
- **Lymphoma:**
- It is a rare tumor.
- Parotid most common
- Associated with systemic disease " Sjögren's Syndrome"
- Occasionally be a primary tumor.
- Criteria
 - o No known extra-salivary lymphoma
 - o Histologic proof that it comes from gland and not a node
 - o Architectural and cytologic confirmation of malignant nature
- Treatment is same as for other lymphomas.
- **Sarcoma:**
- Rhabdomyosarcoma & fibrosarcoma may arise from the parotid.
- Enlarging painless mass.
- Must exclude invasion or metastasis.
- Behaves like soft tissue sarcomas.

- **Survival rates for parotid cancer:**

Type	5-yr Survival (%)	10-yr Survival (%)
Low-grade tumors		
Acinic cell carcinoma	80–90	80
Low-grade mucoepidermoid carcinoma	92–97	90–97
High-grade tumors		
Adenocarcinoma	49–75	41–60
Adenoid cystic carcinoma	45–82	28–77
Carcinoma ex-pleomorphic adenoma	50–77	30–39
Squamous cell carcinoma	42–57	57
High-grade mucoepidermoid carcinoma	35–56	28–54
Undifferentiated carcinoma	30–40	22–25

- **Poor Prognostic Indicators:**

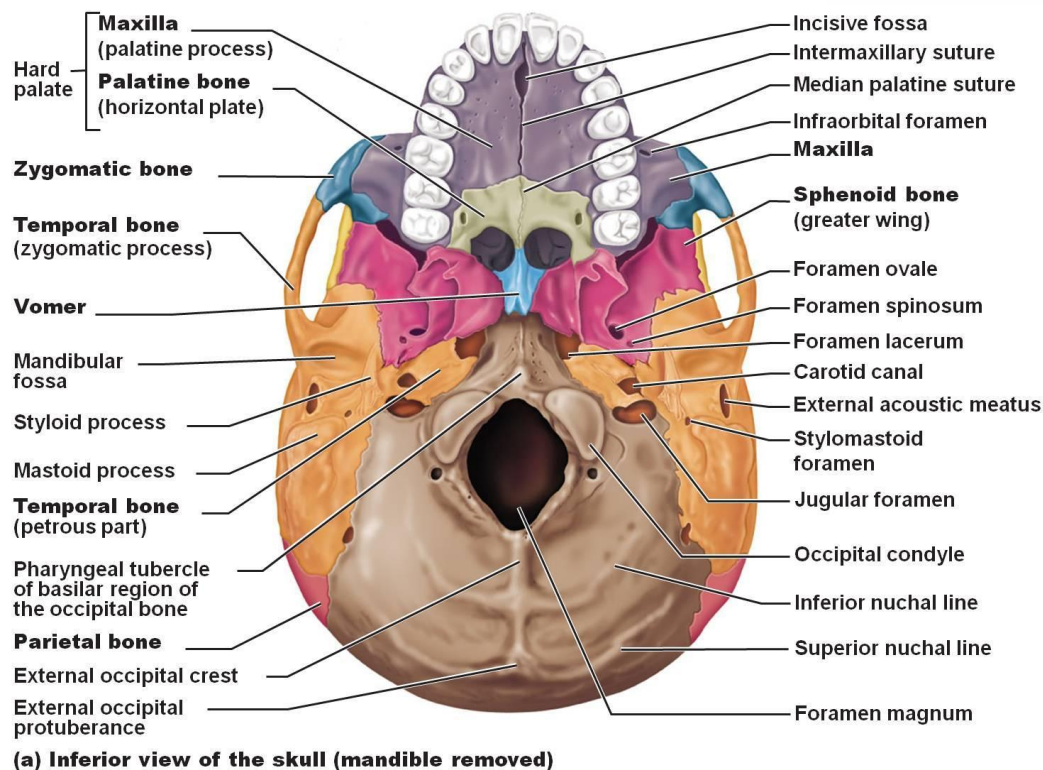
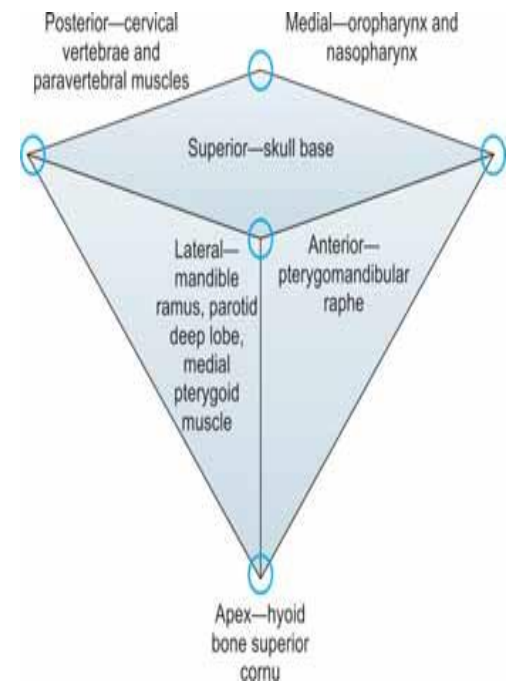
1. Submandibular gland involvement (parotid gland more favorable).
2. Parapharyngeal space involvement.
3. High-grade tumors.
4. Larger size (refer to T-staging; bigger is bad).
5. Facial nerve or skin involvement (refer to T-staging; skin involvement is bad (decrease survival)).
6. Painful tumors (ominous sign; decreased survival).
7. Recurrence
8. Regional lymph nodes
9. Distant metastasis (more common in adenoid cystic and undifferentiated tumors).

Parapharyngeal Space Tumors

- Parapharyngeal space is an area of complex anatomy, which is difficult to examine clinically
- Approximately 0.5% of all head and neck tumors
- 80% are benign and 20% are Malignant.
- Surgical excision is the primary treatment

❖ **Anatomy:**

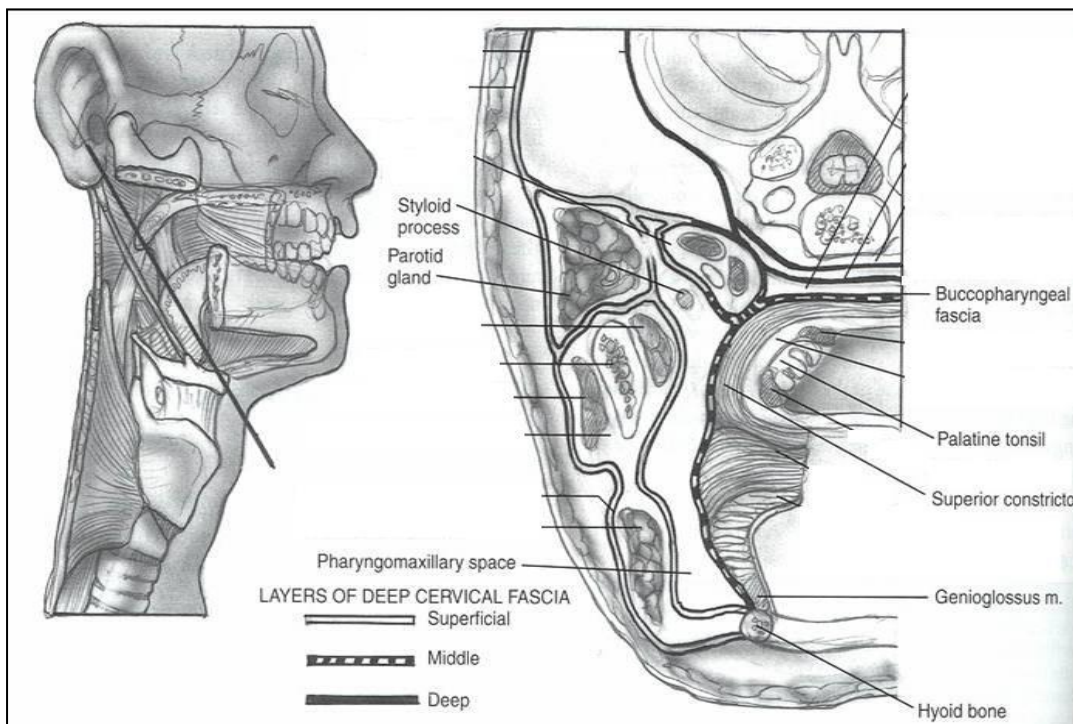
- A potential deep neck space.
- Inverted pyramid with its floor at the Skull base and the tip at the greater cornu of hyoid.
- Two compartments:
 - Pre-styloid compartment (Anterolateral)
 - Post-styloid compartment (Posteromedial)
- **Superior limit** of the parapharyngeal space is a small portion of the temporal and bone and the sphenoid bone.
- It includes:
 - Carotid canal
 - Jugular foramen
 - Hypoglossal foramen



- **Inferior border** of the parapharyngeal space is the junction of posterior belly of digastric and the Greater cornu of hyoid bone

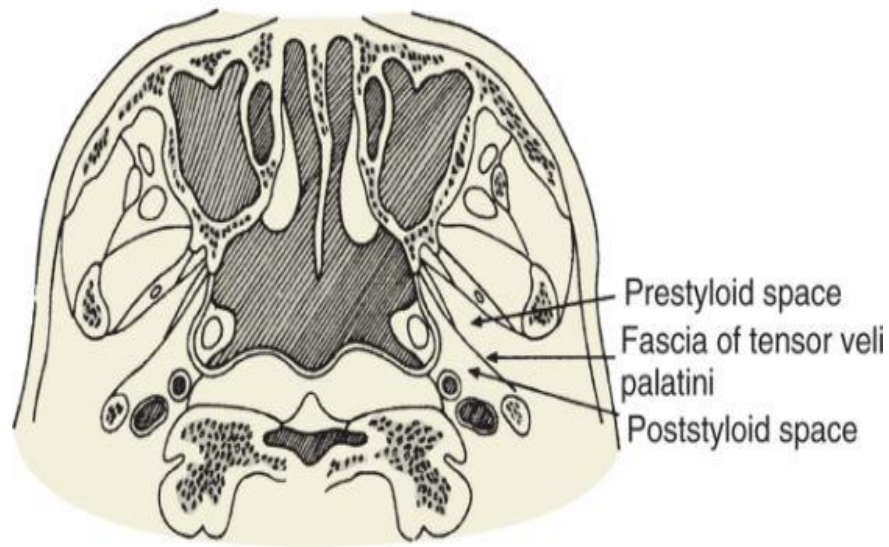


- **Medial boundary** is made up of the buccopharyngeal or visceral fascia overlying the superior pharyngeal constrictors.
- **Lateral boundary** is made up of the fascia over :
 - Medial pterygoid muscle
 - Ramus of the mandible
 - Posterior belly of the digastric muscle
 - Fascia over the retromandibular deep portion of the parotid gland



- **Anterior border** is the Pterygomandibular raphe.
- **Posterior limit** is the Prevertebral fascia covering prevertebral muscles.

- Parapharyngeal space is divided by Fascia which extends from the styloid process to the tensor veli palatini muscle, called the tensor-vascular-styloid fascia, because it also contains the ascending palatine artery and vein.



❖ **Contents of Pre-styloid compartment (Anterolateral):**

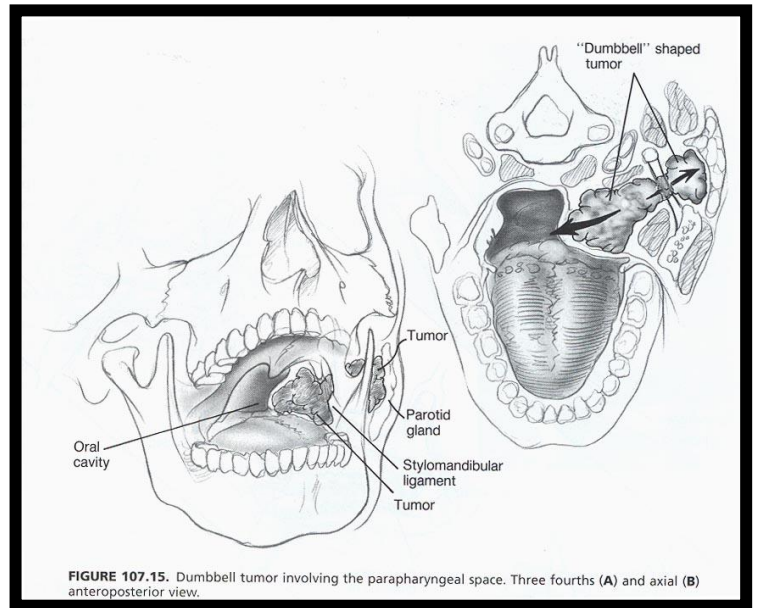
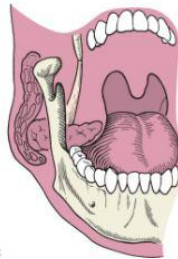
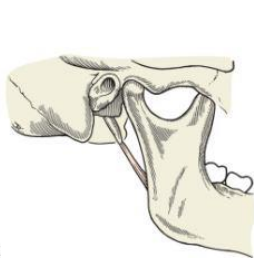
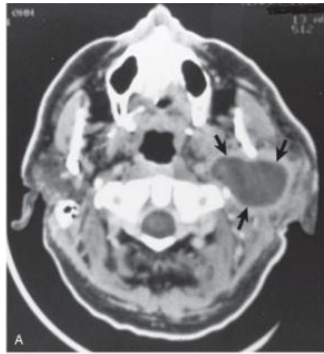
- Mainly adipose tissues
- Retromandibular part of deep lobe of parotid
- Minor or ectopic salivary glands
- A small branch of CN V to Tensor palatine muscle
- Ascending pharyngeal artery and venous plexus
- Lymph nodes
- Internal maxillary Artery
- Lingual nerve
- Auricotemporal nerve

❖ **Contents of Post-styloid compartment (Posteromedial):**

- Internal carotid artery
- Internal jugular vein
- CN IX to XII
- Cervical sympathetic chain
- Lymph nodes

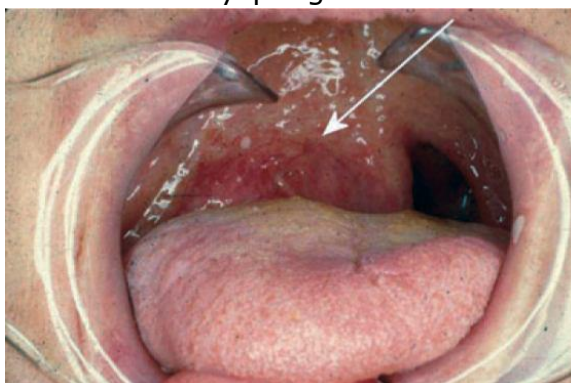
❖ **Stylomandibular tunnel:**

- **Bounded by:**
 - Posterior ramus of the mandible
 - Skull base
 - Stylomandibular ligament
- Deep parotid tumors can grow into parapharyngeal through this tunnel
- The constriction of this tunnel on tumor growth gives a characteristic "dumbbell" shape on CT scan.



❖ **Clinical Presentation:**

- Clinical detection can be difficult.
- Sign and symptoms are numerous and depends on the location and histopathology of the tumor.
- Need to be as large as 2.5-3 cm to be detected clinically.
- **Prestyloid Symptoms:**
 - Asymptomatic mass (most commonly)
 - OME
 - Voice change
 - Nasal obstruction
 - Dyspnoea
 - Dysphagia



▪ **Poststyloid Symptoms:**

- May present as a mass in the neck.
- Hoarseness of Voice.
- Deviation of tongue.
- Jugular foramen syndrome (Vernet's 9-11th CN palsy)
 - Dysphonia/hoarseness
 - Soft palate dropping
 - Deviation of the uvula towards the normal side
 - Dysphagia
 - Loss of sensory function from the posterior 1/3 of the tongue
 - Decrease in the parotid gland secretion
 - Loss of gag reflex
 - Sternocleidomastoid and trapezius muscles paresis
- Horner's syndrome (sympathetic chain injury):
 - Ptosis
 - Miosis
 - Anhidrosis
- Pain, CNs involvement and trismus Usually suggest malignant tumor.

Sign/Symptoms	No.	(%)
Neck mass	76	47.5
Hoarseness or vocal cord paralysis	16	10.0
Hearing loss	13	8.1
Oropharyngeal mass	11	6.9
Dysphagia	10	6.3
Pain	9	5.6
Tinnitus	9	5.6
Otalgia/aural fullness	7	4.4
CN XII dysfunction	6	3.8
Dyspnea/postnasal drip	5	3.1
Headache	4	2.5
Facial weakness/paralysis	3	1.9
Dizziness	2	1.3
Trismus	2	1.3
Bleeding	1	0.6
Horner's syndrome	1	0.6
Sleep apnea	1	0.6
Trigeminal dysesthesia	1	0.6
Weight loss	1	0.6

Table 19.3 Presenting Symptoms and Signs of Parapharyngeal Space Tumors (1)

Symptoms	Frequency (%)
Intraoral/neck mass	85
Ear pressure or pain	36
Dysphagia	13
Hearing loss	11
Hoarseness	10
Facial or jaw pain	6
Facial nerve weakness, throat pain, pulsatile tinnitus, tongue paraesthesia, aspiration, headache, hypertension, syncope	<5
Signs	
Intraoral displacement of tonsil/soft palate	65
Neck mass (11% pulsatile)	58
Vocal cord paralysis/paresis	8
Hearing loss/OME	9
Palatal paralysis	5
Atrophic or fasciculating tongue	5
Horner's syndrome	2
Trismus	2

Abbreviation: OME, Otitis media with effusion.

❖ **Differential diagnosis of Parapharyngeal tumors:**

- Primary Parapharyngeal tumors:
 - 80% benign and 20% malignant.
 - Location of parapharyngeal tumor can be helpful for differential diagnosis:
 - **Pre-styloid space tumors:**
 - Salivary gland tumor.
 - Lipoma
 - Atypical 2nd branchial cleft cyst
 - Neurogenic tumors
 - **Post-styloid space tumors:**
 - All contents are potential sources for post-styloid tumors.
- Direct extension from adjacent structures
 - Mandible
 - Maxilla
 - Nasopharynx
 - Neck
 - Oral cavity
 - Oropharynx
 - Temporal bone tumors
- Metastatic:
 - Thyroid Ca
 - Squamous Cell Ca
 - Osteogenic sarcoma

❖ **Primary Para-Pharyngeal Space tumors**

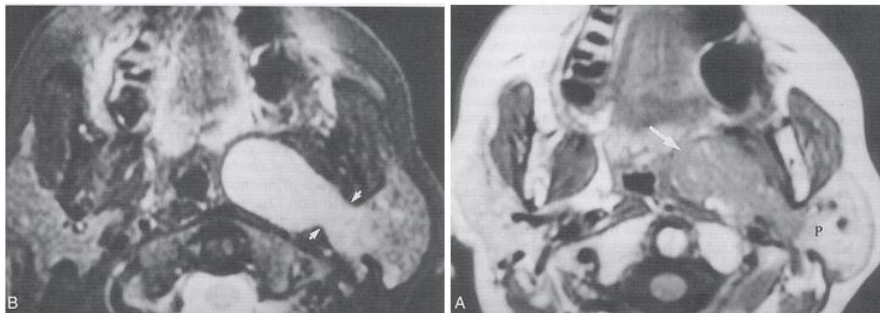
1. Salivary Gland Tumors
 - Most common PPS tumors.
 - Most common Pre-styloid space tumors.
2. Neurogenic Tumors:
 - 2nd most common PPS tumors.
 - Most common Post-styloid space tumors.
3. Miscellaneous Tumors

Salivary gland Tumors

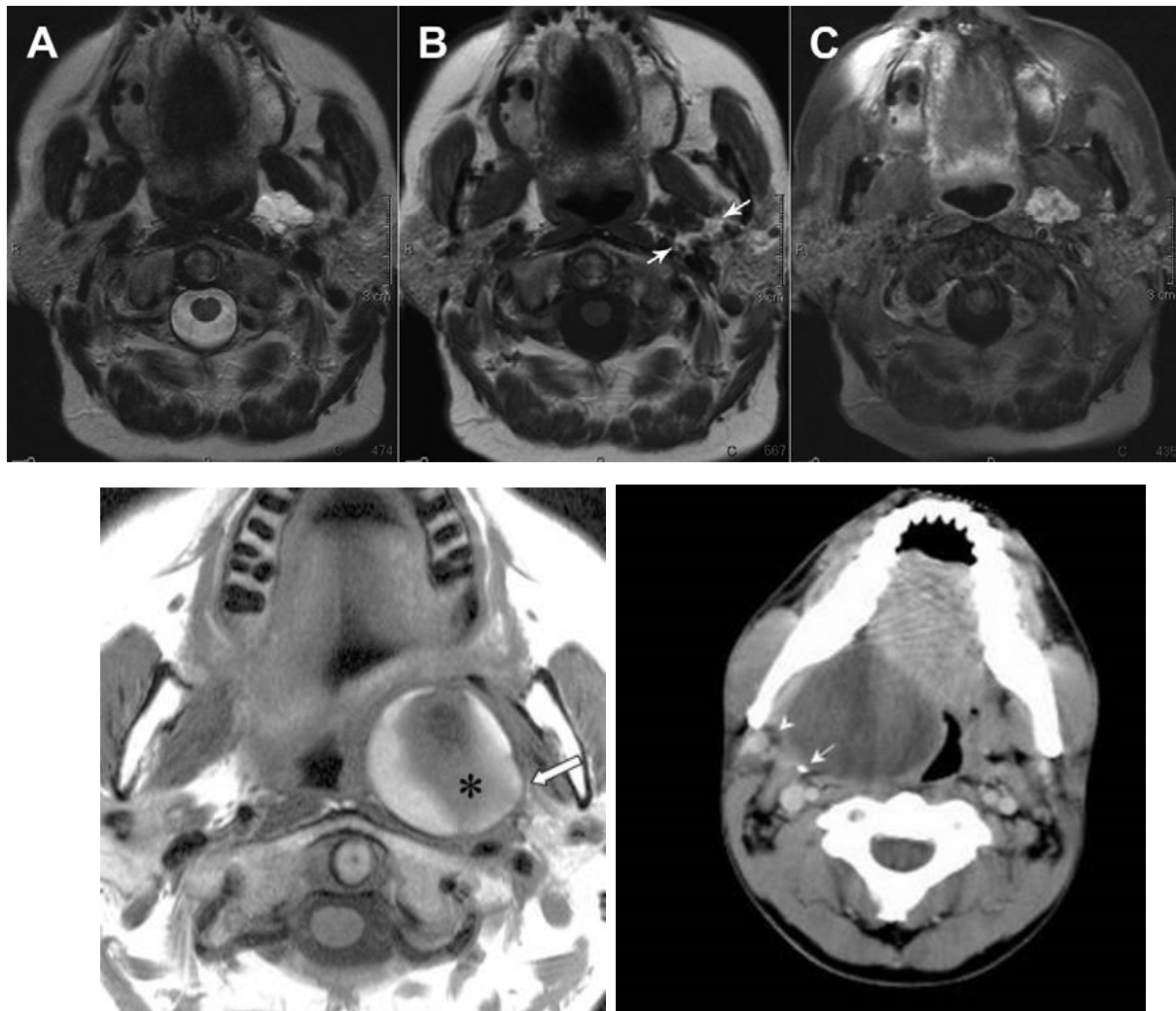
- In general, salivary gland tumors are the most common primary parapharyngeal tumor.
- 40-50% of parapharyngeal lesions
- Rises in the prestyloid compartment from :
 - Deep lobe of parotid
 - Ectopic salivary
 - Minor salivary glands of lateral pharyngeal wall
- Most common prestyloid PPS lesion is **Pleomorphic adenoma** (80-90%).
- 20% of all salivary lesions in PPS are malignant.
- **Benign lesions:**
 - **Pleomorphic adenoma (Most common)**
 - Warthin tumors
 - Oncocytomas
- **Malignant Lesions:**
 - **Mucoepidermoid (Most common)**
 - Carcinoma ex pleomorphic adenoma
 - Adenoid cystic carcinoma

❖ **Pleomorphic Adenoma:**

- Originate from either:
 - Deep lobe of the parotid gland
 - Minor salivary glands in pre-styloid space.
- A deep lobe parotid gland tumor can extend through the stylomandibular tunnel into the parapharyngeal space.
- This kind of tumor will give a characteristic “dumbbell” appearance on CT and MRI.



- Also tumor can arise from the retromandibular portion of the parotid gland, then expands into the parapharyngeal space and displace tonsil and soft palate and cause obstruction of the nasopharynx.
- Extension of tumours from the deep lobe of a parotid gland is distinguishable from tumour arising de novo in the parapharyngeal space **by a fine lucent line of fibroadipose tissue** between the tumour and deep lobe of parotid.



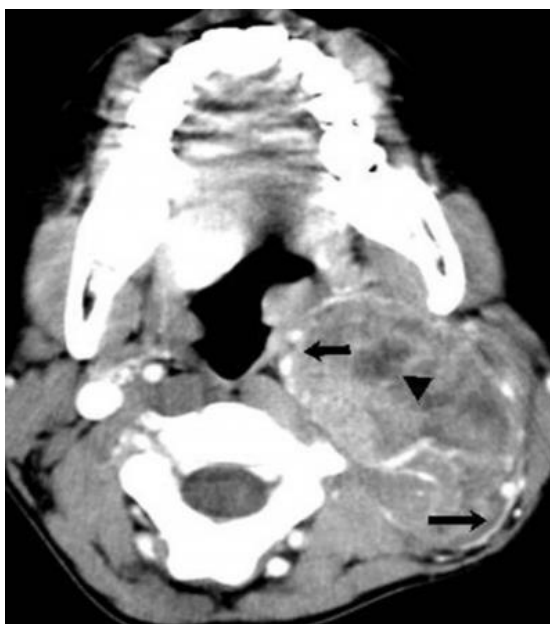
- On CT or MRI these lesions displace the carotid artery posteriorly

Neurogenic tumors:

- **Second** most common PPS tumors (25-30%).
- Most common **Poststyloid** tumors.
- Types :
 - Schwannoma
 - Paraganglioma
 - Neurofibroma

❖ **Schwannoma:**

- **Most common** type of neurogenic tumors.
- Benign tumors of the peripheral nerve sheath due to proliferation of the myelin-producing Schwann cells.
- **Rarely** undergo malignant transformation.
- Can rise from :
 - CN IX
 - CN X (Most Common, as high as 50%)
 - CN XI
 - CN XII
 - Sympathetic chain (2nd most common)
- **Slow growing tumors** and **don't effect the nerve of origin.**
- By CT or MRI these tumors displace the carotid artery depending on the nerve of origin.
- In CN X Schwannoma:
 - Anteromedial displacement of ICA
 - Posterolateral displacement of IJV
- In Sympathetic chain Schwannoma:
 - Anterior displacement of BOTH ICA and IJV



❖ **Paraganglioma:**

- Benign vascular neoplasms arise from the paraganglia (part of system of baroreceptive and chemoreceptive).
- 2nd most common neurogenic tumors in PPS.
- In PPS, vagal paraganglioma is the most common (60%).
- Carotid body tumors rarely extend above the posterior belly of the digastric muscle.
- 1-3% of paragangliomas secrete catecholamines, A fivefold increase in catecholamines is sufficient to produce symptoms.
- Malignancy
 - Vagal paragangliomas have a rate of 10%.
 - Carotid body tumors being at 3% to 6%.
 - Sporadic have a higher rate of malignancy than familial-type.
 - No histologic criteria for malignancy
 - Confirmed by tumor present in lymph nodes or distant metastatic sites.
- Multiple paraganglioma:
 - Present in up to 20% of patients.
 - Mostly hereditary or familial.
 - 10% of sporadic cases will have concurrent second paraganglioma.
 - Multiple tumors maybe metachronous.
 - Most common pattern of a synchronous 2nd paraganglioma is a second carotid body tumor (20%).
 - Other than Genetics, the only known predisposing factor is **high altitude living associated with chronic hypoxemia**
 - 9-fold increase for those living between 2 and 3 Jan.
 - 12-fold increase for those living between 3 and 4.5 km above sea level.

❖ **Neurofibroma:**

- 3rd most common neurogenic tumor.
- Neurofibromas are benign nerve sheath **Unencapsulated** tumors that can develop as solitary lesions or as multiple lesions as part of type-1 Neurofibromatosis.
- Intimately involved with the nerve of origin.

Miscellaneous tumors:

- Account for 20% of total Parapharyngeal space tumors
- Include :
 - Lymphomas
 - Lipomas
 - Hemangiomas
 - AVM
 - IC aneurysm
 - Branchial cleft cyst
 - Teratoma

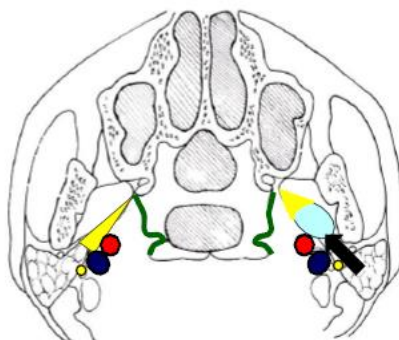
❖ **Work-Up:**

- Thorough History (family Hx, Symptoms of catecholamine secretions).
- Complete H&N Examination With Cranial nerves.
- Screen for Catecholamines in urine (if paraganglioma suspected).
- **Imaging:**
 - CT with contrast
 - +/-MRI/MRA
 - +/-Angiography
- FNA (currently no role)

❖ **Imaging**

- Essential in the evaluation of a patient with a suspected PPS tumor.
- **Imaging studies should answer the following questions:**
 - Prestyloid vs Poststyloid tumors
 - Relationship to the parotid gland
 - Relationship to the great vessels
 - Soft tissue characteristics of the tumor and enhancements
 - Bone erosions
- **CT SCAN:**
- **Advantages :**
 - Can locate the compartment involved
 - Fat plane between the parotid and the mass Suggest an Extra-parotid origin
 - Presence of calcifications and bony defects
 - When contrasted can show relations with great vessels
 - CT can be the only study required because most parapharyngeal space (PPS) masses are prestyloid lesions arising from the parotid gland
- **Disadvantages :**
 - In Enhanced lesions difficult to assess the relation to the great vessel
 - CT scanning is inferior to MRI in delineating soft tissue characteristics of the lesion
 - Can be difficult to assess the fat lucent line in comparison to MRI

Prestyloid tumours displace the PPS fat anteromedially (*Figures 4, 5*).



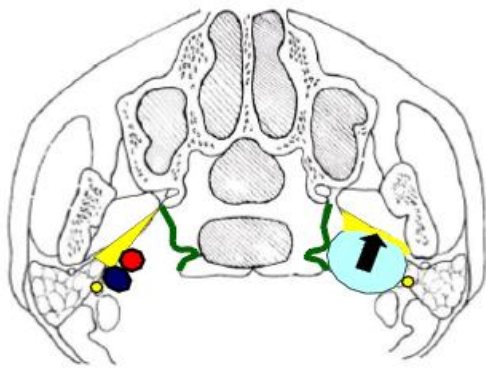
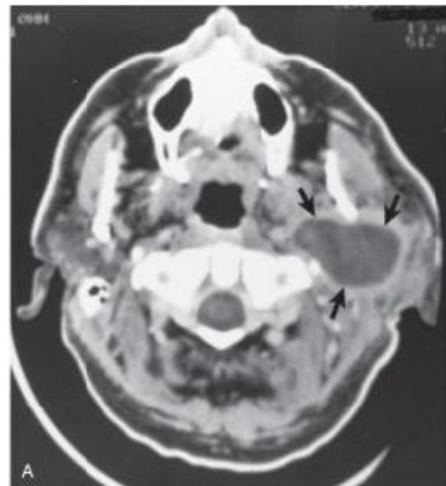
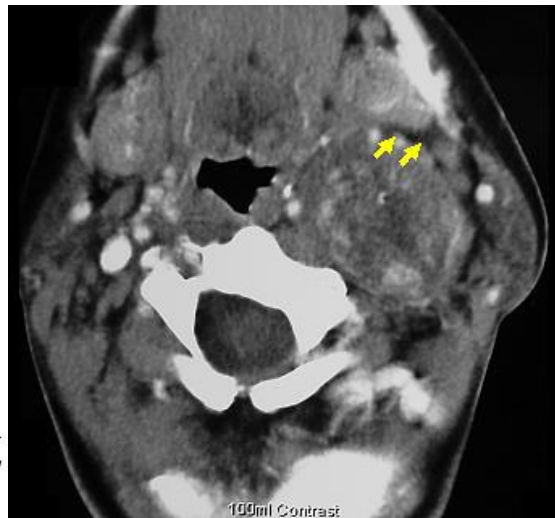
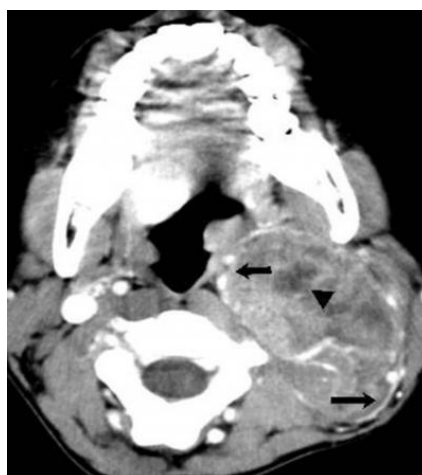


Figure 8: Direction of displacement of fat as seen on CT or MRI with poststyloid mass (light blue)



A- Pleomorphic adenoma, dumbbell sign.

B- Mucoepidermoid, ICA posteriorly displaced, fat line pushed medially.

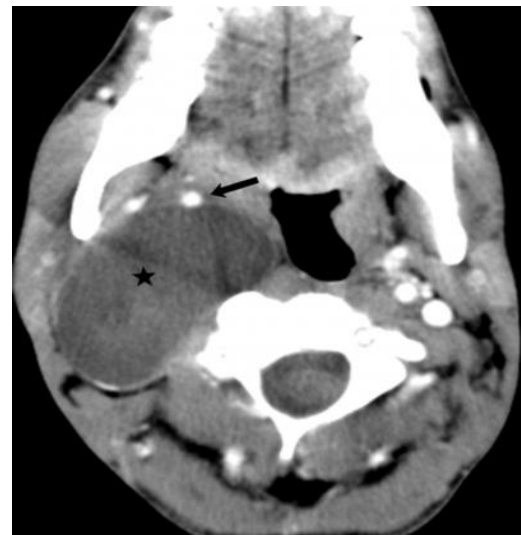


- Vagal schwannoma:
- CT scan shows a heterogeneously enhancing Schwannoma with areas of necrosis displacing the internal carotid artery anteromedially.

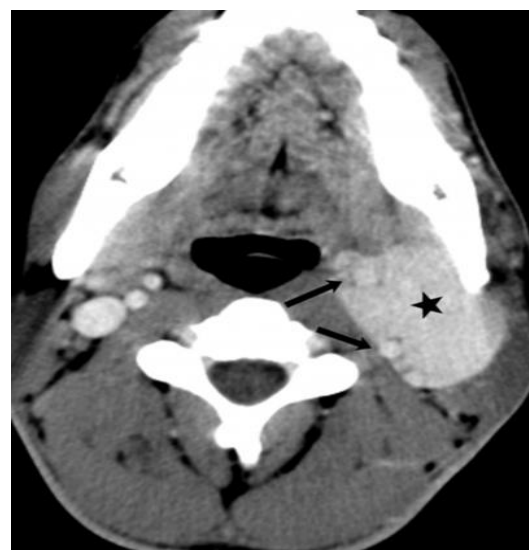
- Pleomorphic adenoma:
- CT scan shows a minimally enhancing water attenuation well-defined mass (star) extending into the prestyloid parapharyngeal space with widening of the stylomandibular tunnel (arrows).



- Neurofibroma:
- CT scan shows the minimally enhancing tumor in the post-styloid space (star) causing anterior displacement of ICA

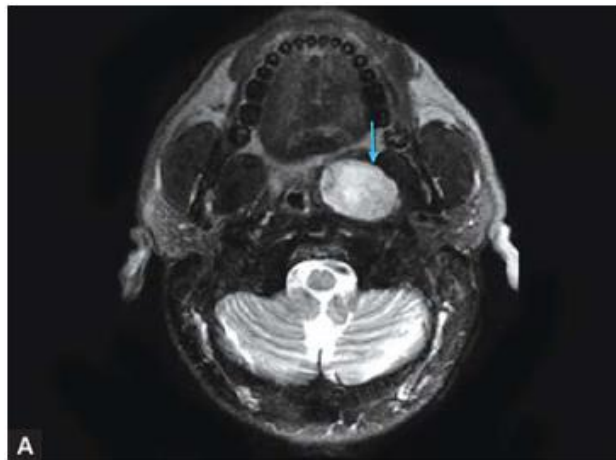


- Carotid body tumor:
- Post contrast axial CT shows intensely enhancing tumor in the post-styloid space (star) causing splaying of internal and external carotid artery

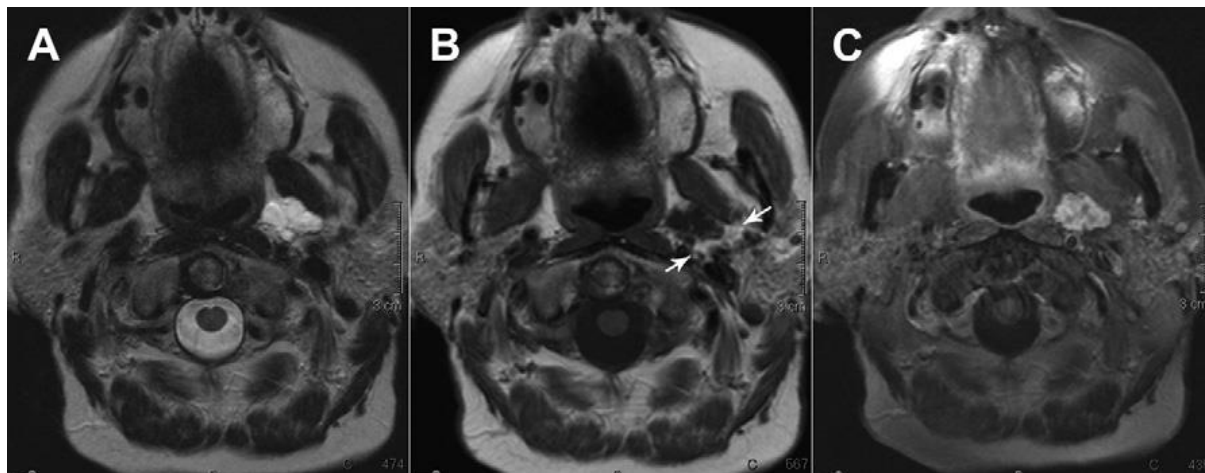


❖ MRI

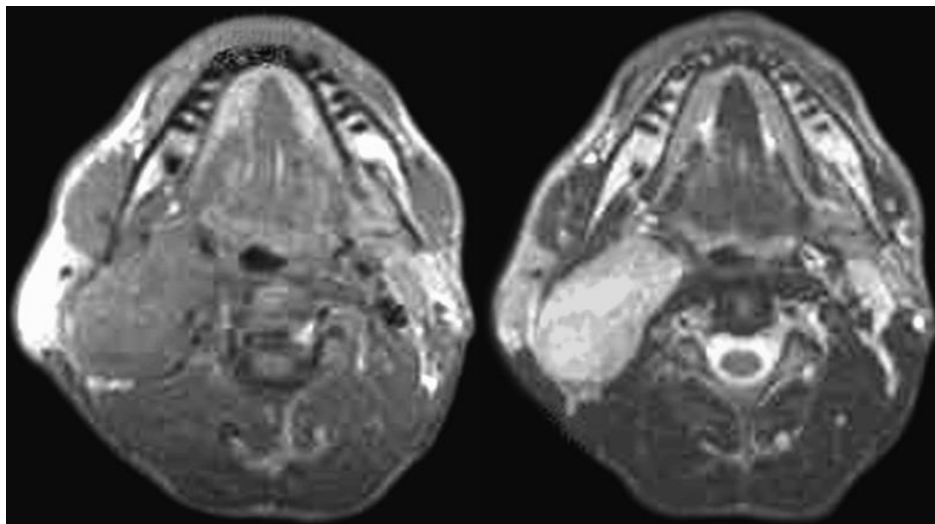
- Can easily differentiate minor salivary gland lesion from deep lobe lesions
- Relation to the Carotid easily assessed.
- Patterns of carotid artery displacement can be accurately determined distinguishing carotid body tumors from vagal paragangliomas.
- If carotid involvement suspected angiogram should be done.
- Tumors with intracranial extension, intradural vs. extradural involvement can be appreciated
- Relationship of the tumor to important intracranial structures can be delineated.
- Classic "**salt and pepper**" appearance of **paragangliomas** on **T2**-weighted images is due to the high-flow vascular voids that are essentially pathognomonic.



- Pleomorphic adenoma:
 - T1- isotense to intermediate
 - T2- High intensity
 - Salivary gland tumors always anterior to ICA
 - If large enough might displace it posteriorly
 - Can show patchy, heterogeneous enhancement
 - appearance of benign mixed tumors is fairly characteristic on MRI and more nonspecific on contrast-enhanced CT

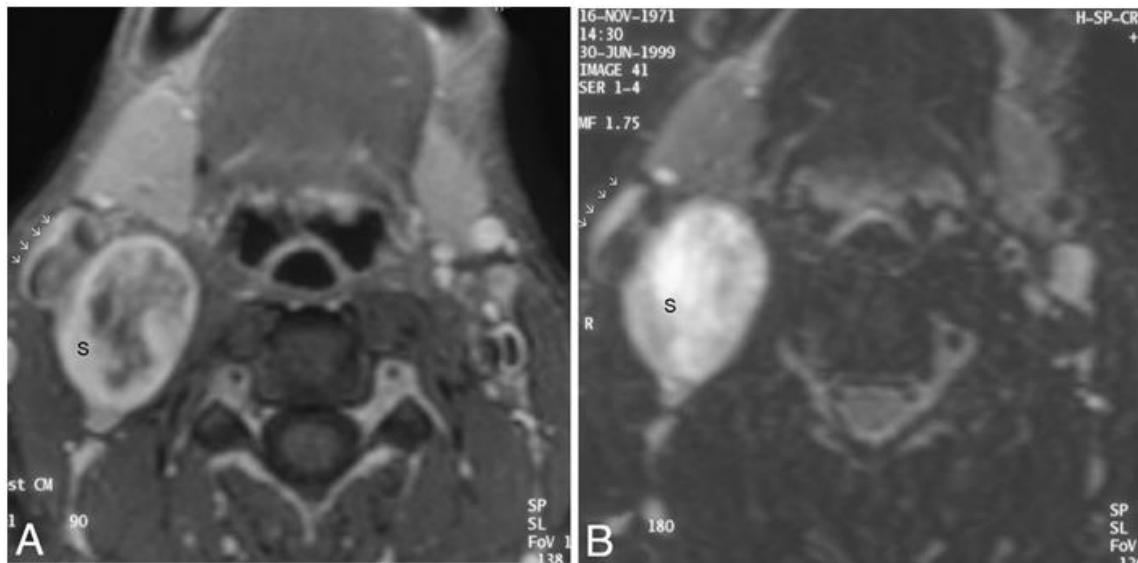


Note the preserved fat plane from the along the margin of deep lobe the left parotid gland (arrows) demonstrating that this lesion is not primary to parotid space.



- **Schwannoma**

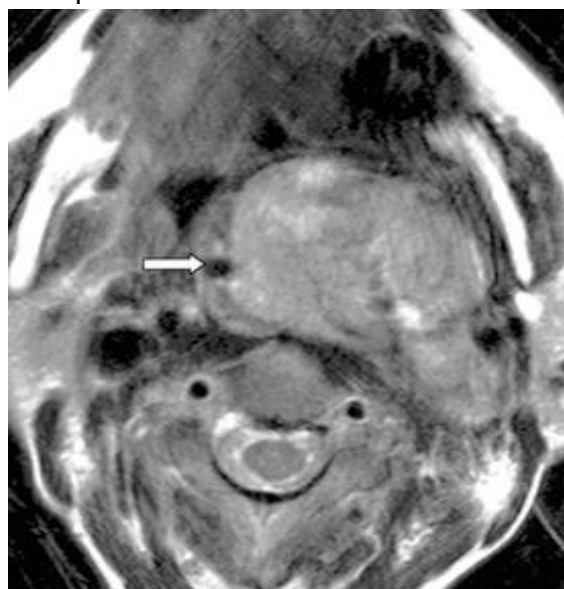
- Heterogenous
- T1 isointense to hypointense
- T2 Hyperintense
- **NO FLOW VOID** (differentiated from paraganglioma)
- Hypovascular (show slow enhancement on delayed scans, incontrast to paraganglioma)
- Causes **scalloping or smooth enlargement** of involved skull base foramen (incontrast to paragangliomas which causes irregular erosins).



- Fig: symphatic Schwannoma displacing the whole carotid sheet anteriorly

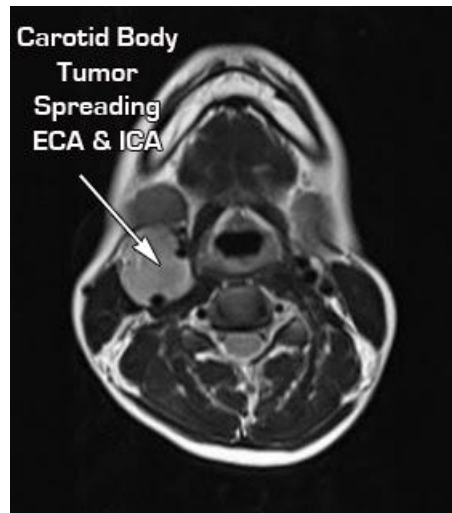
- **Schwannoma**

- Displacement of the great vessels depends on the nerve of origin
- Vagal schwannoma causes :
 - Anterolateral displacement of ICA
 - Posterolateral on IJV
- Sympthatic chain scwannoma :
 - Anterior displacement of BOTH ICA and IJV

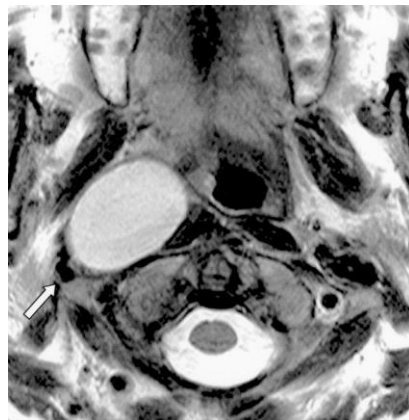


- Fig: T2 axial Cut vagal paraganglioma ICA displaced anteromedially

- Paraganglioma:
 - **Smoothly** contoured
 - Multiple vascular **FLOW VOID**
 - CLASSIC "**Salt and Pepper**" on T2 (due to flow voids ,due to high fast flow)
 - Hypervascular rapid enhancement
 - **Irregular erosions** at base of skull foramens (better on CT) called moth bite



- Carotid body tumors:
 - Cause characteristic splaying of both ICA and ECA at the bifurcation called the Lyre's sign



- 2nd Branchial cleft cyst:
- T2 images shows mass is slightly hypointense to cerebrospinal fluid. Displacement of internal carotid artery and internal jugular vein to its posterolateral side

Surgical approaches

- Transoral +/- Mandibulotomy
- Transcervical submandibular
- Transparotid
- Transcervical +/- mandibulotomy
- Infratemporal fossa
- Transoral robotics

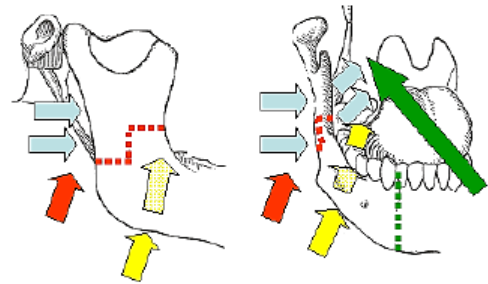


Figure 11: Approaches to PPS: transoral $\pm$ mandibulotomy (green); transcervical submandibular (yellow), transparotid (blue), and transcervical $\pm$ mandibulotomy (red)

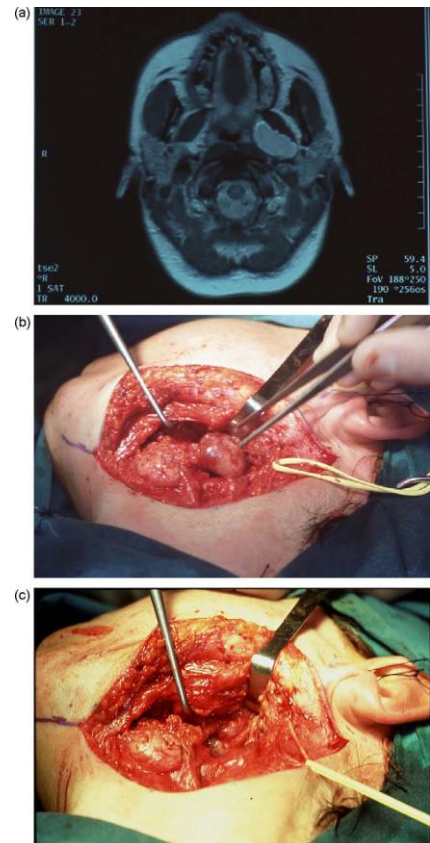
Transoral Approach:

- For Prestyloid tumors
- Multiple disadvantages:
 - Usually Doesn't provide enough exposure
 - compromise the wound through contamination by oral secretions or seed the tumor into the mucosa of the palate
 - Neck isn't exposed incase of major vessel injury



❖ **Transcervical Approach:**

- Ideal approach to Prestyloid tumors
- A transverse incision at the level of the hyoid bone, 2 fingerbreadths below the mandible
- The carotid artery and internal jugular vein are identified
- The digastric and stylohyoid muscles are retracted to allow access to the parapharyngeal space (PPS).
- The submandibular gland can be retracted anteriorly for exposure, or it can be removed if necessary.

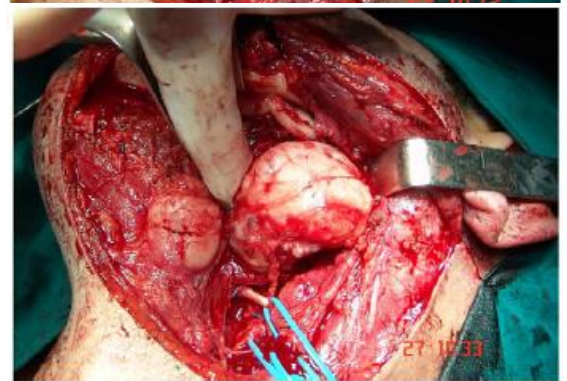
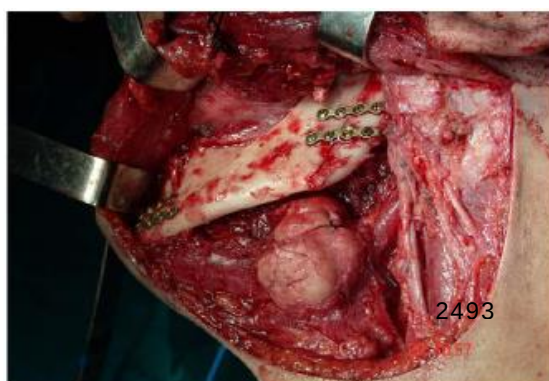


Transparotid Approach:

- Can be used For deep lobe parotid tumors
- the transcervical approach can be combined with a transparotid approach by extending the incision superiorly as for parotidectomy
- The facial nerve is identified and dissected, superficial parotidectomy is performed
- the deep lobe portion of the tumor is identified
- The cervical incision allows access to the parapharyngeal space (PPS) component of the tumor.
- If necessary, mandibulotomy may be used posterior to the entrance of the inferior alveolar nerve.

Transcervical-transmandibular Approach:

- For Removal of very large tumors, vascular tumors or superior extension with skull base invasion
- The advantages are better exposure and gives distal carotid control at the skull base



Paranglioma:

❖ Anatomy and physiology of paraganglia :

- Paraganglia are part of a system of cell clusters that facilitate the baroreceptive and chemoreceptive reflexes of the cardiovascular system.
- Paraganglia are found in:
 - Adrenal medulla
 - Extra-adrenal paraganglia (diffusely distributed) and those are divided:

➤ **Branchiomic paraganglia:**

- Distributed along arteries and cranial nerves of the head neck:
 1. Jugulotympanic paraganglia arise from the 2nd branchial arch.
 2. Carotid paraganglia arise from the 3rd branchial arch.

➤ **Vagal paraganglia**

❖ Jugulotympanic paraganglia:

- Distributed within the temporal bone in close association with:
 1. **Jacobson nerve** (Tympanic branch of the glossopharyngeal nerve)
 2. **Arnold nerve** (Auricular branch of the vagus nerve).
- Locations:
 1. Jugular fossa (most common)
 2. Jacobson bony canal
 3. Submucosa of tympanic cavity

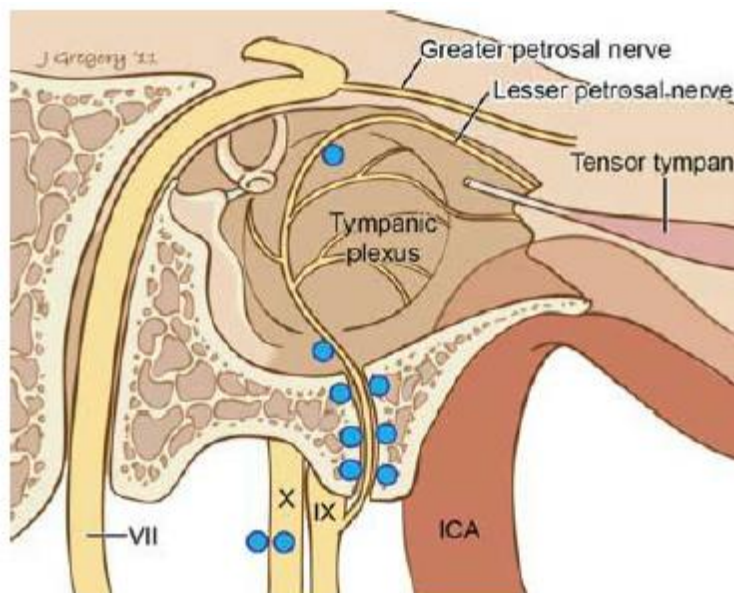


Figure 127.3 Distribution of frequent locations for glomus bodies in the temporal bone.

- ❖ There is a marked difference in the clinical behavior of tumors arising from the jugular paraganglia versus the tympanic paraganglia which has significant therapeutic implications.

❖ Carotid body:

- A chemoreceptor that senses changes in arterial oxygen pressure, pH, and carbon dioxide.
- A discrete oval structure situated behind the carotid bifurcation.
- Receives its blood supply directly from the carotid bifurcation via the glomic arteries.
- Afferent reflex is mediated by a branch of the glossopharyngeal nerve (nerve of Hering)

❖ Vagal paraganglia:

- Do not form discrete bodies (incorporated within the vagus nerve underneath the perineurium).
- Interspersed within the vagal nerve fibers in the pars nervosa of the jugular foramen.

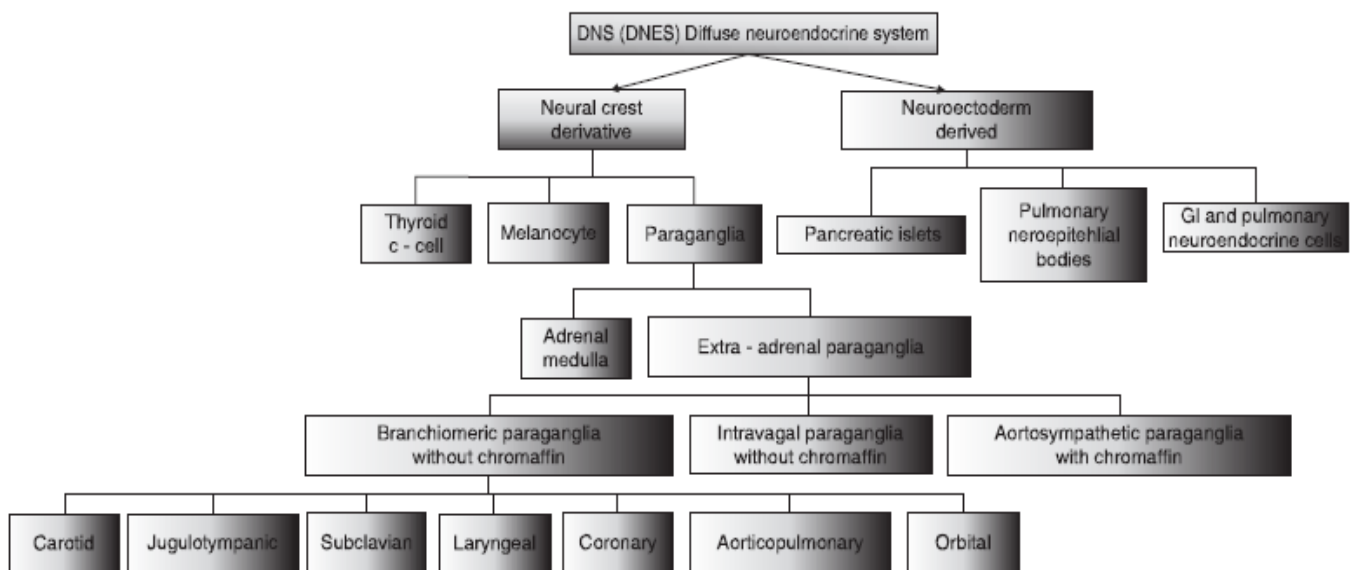
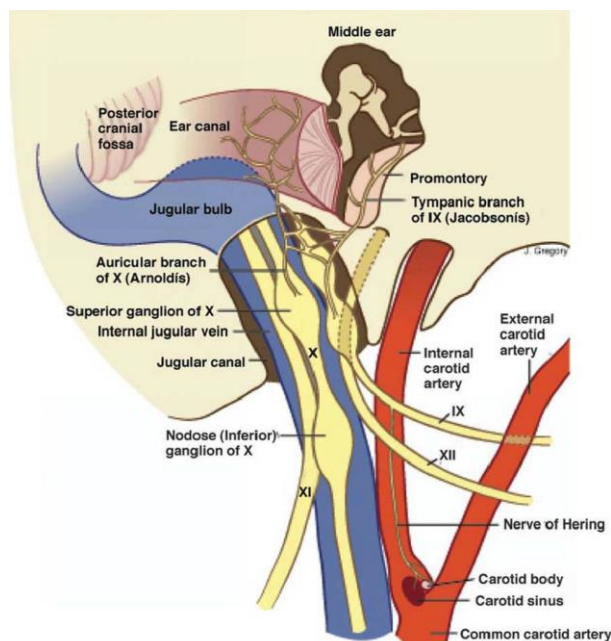


Figure 127.1 Classification of the diffuse neuroendocrine system.



▪ **Pathophysiology and Epidemiology**

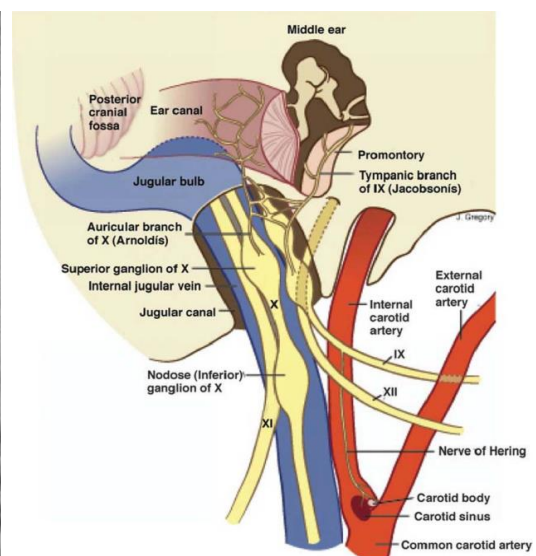
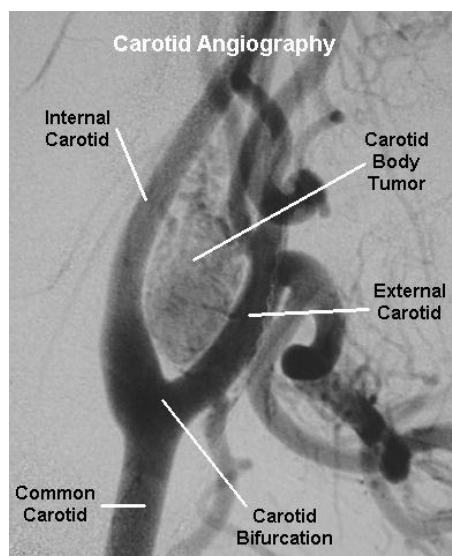
- Benign vascular neoplasms rises from the paraganglia (part of system of baroreceptive and chemoreceptive).
- Derived from neural crest origin cells.
- **2nd most common** neurogenic tumors in PPS
- Carotid body tumors and jugulotympanic tumors account 80%
- Vagal ganglioma 5%
- Multiple paraganglioma are present in up to 20% of patients (mostly hereditary or familial).
- **10** % of sporadic cases will have concurrent second paraganglioma
- Multiple tumors maybe metachronous.
- Most common pattern of a synchronous 2nd paraganglioma is a second carotid body tumor (20%).
- Other than Genetics, the only known predisposing factor is **high altitude** living associated with chronic hypoxemia
- Who is at risk:
 - 9-fold increase for those living between 2 and 3 km above sea level.
 - 12-fold increase for those living between 3 and 4.5 km above sea level.
- 1-3% of paragangliomas secrete catecholamines.
- A fivefold increase in catecholamines is sufficient to produce symptoms.
- **In Para-Pharyngeal Space, vagal paraganglioma is more common (60%).**
- In contrast, carotid body tumors rarely extend above the posterior belly of the digastric muscle.
- Malignant potential :
 - **Orbital and laryngeal** highest rate 25%
 - Vagal paragangliomas up to 10%,
 - Jugulotympanic paraganglioma 5%
 - Carotid body tumors being at 3-6%.
 - No histologic criteria by which primary tumor malignancy can be diagnosed.
 - Malignancy is confirmed by tumor present in lymph nodes or distant metastatic sites.

❖ **Clinical Presentation :**

- Clinical presentation of paragangliomas is **location specific**.
- As these tumors enlarge, they tend to produce cranial neuropathies.
- Additionally, secreting tumors can produce:
 - Hypertension
 - Tachycardia
 - Sweating
 - Nervousness secondary to catecholamine release.

❖ **Carotid body tumors presentation:**

- **Deep neck mass:**
 - Slowly enlarging asymptomatic at the level of the carotid bifurcation.
- **Pain:**
 - Present around the tumor (small minority of patients).
- **Cranial nerve paralysis symptoms:**
 - Unusual and present in very large tumors with superior extension toward the jugular foramen.
- Because of the association with the carotid artery they are **laterally mobile, but rostrocaudally fixed on examination**
- **Bruit** may be auscultated As the tumor enlarges.
- **Shamblin classification:**
 - Commonly used to stage carotid body tumors, and though this is based on intraoperative findings, it can be applied to radiologic findings prior to treatment.



❖ Shamblin classification

- **Type 1:**
 - Small tumor with minimal attachment to ICA.
- **Type 2:**
 - Larger tumor with moderate attachment to ICA but respectable with preservation of the carotid vessels.
- **Type 3:**
 - Tumor encase ICA requiring arterial sacrifice with reconstruction.

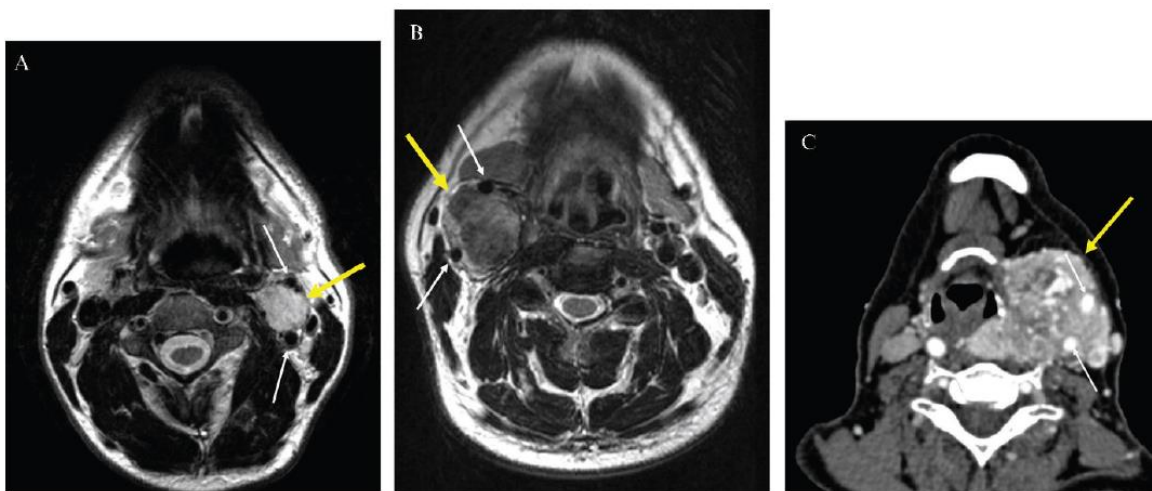
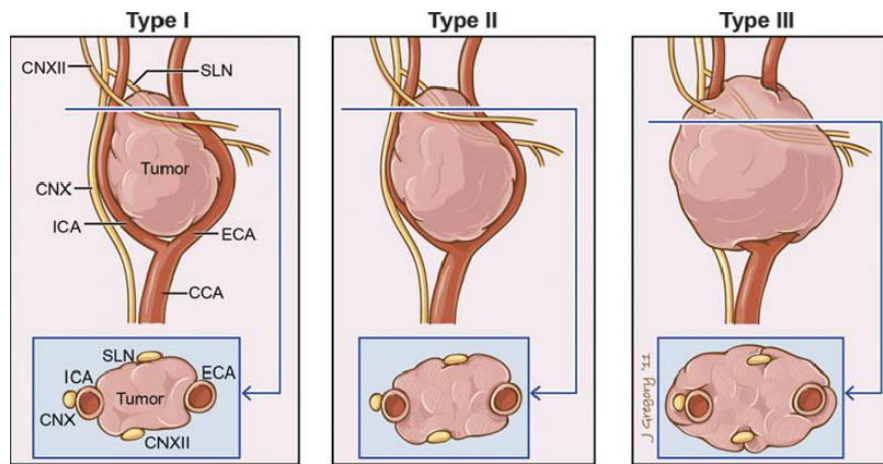


Figure 1 - **Shamblin classification**. The Shamblin classification is based on involvement of the carotid arteries, which are shown by white arrows in A–C. The tumors are indicated by yellow arrows.

(A) CBT (class I): the axial T2-weighted MRI shows a CBT in a typical location. Bulging of the left carotid bifurcation without any encasement of the carotid vessels.

(B) CBT (class II): axial T2-weighted MRI showing a right-sided CBT with a partial encasement of the internal and external carotid artery.

(C) CBT (class III): an axial slice of CT-angiography reveals a huge left-sided CBT with a complete encasement of the carotid vessels. In this case, bone destruction of the skull base, attributed to the enormous CBT, was also detected by CT (not shown).

❖ **Presentation of Jugulotympanic Paragangliomas:**

- Mostly 5th-6th decade
- Female predominance
- Slow growing
- pattern of growth follows the pathways of least resistance in the temporal bone and surrounding skull base structures
- In their later stages, both types produce similar symptoms including cranial nerve deficits
- **Tympanic paragangliomas :**
 - Present early with pulsatile tinnitus and CHL.
 - Red-blue middle ear mass that blanches with positive pressure on pneumatic otoscopy is appreciated (Brown sign)
 - With subsequent growth, there is ossicular erosion and filling of the middle ear deft.
 - Ear canal polypoid mass produce bloody otorrhea.
 - Facial paralysis in mastoid involvement.
 - Extend anteriorly toward carotid or Eustachian tube or intracranially.
- **Jugular paragangliomas:**
 - Due to their origin location within the jugular bulb, they can:
 - Early extensive **skull base invasion**
 - **Intracranial involvement** through the jugular foramen is frequent.
 - **Involvement of CN 9 - 12.**
 - **CHL, tinnitus, and facial paralysis** by (Growth through Jacobson nerve canal and the hypotympanic air cells leads to involvement of the middle ear and mastoid bone)
 - **Petrous carotid artery involvement** by Erosion of the jugulocarotid spine leads to involvement of the vertical petrous carotid artery then horizontal portion of the petrous carotid artery.
 - **Clivus and cavernous sinus** by Further medial extension.
 - Around 10% will have either :
 - **Vernet syndrome:**
 - Paralysis of CN 9, 10, and 11.
 - **Collet-Sicard syndrome:**
 - Paralysis CN 9, 10, 11, and 12.
- **Classifications :**
 - Fisch (no distinction between tympanic and jugular)
 - Glasscock-Jackson (treat each type differently)

**TABLE
127.4****GLASSCOCK-JACKSON
CLASSIFICATION OF JUGULOTYMPANIC
PARAGANGLIOMAS****Glomus tympanicum**

Type 1	Small mass limited to the promontory
Type 2	Tumor completely filling the middle ear
Type 3	Tumor filling the middle ear and mastoid
Type 4	Tumor completely filling the middle ear, extending into the mastoid or through the external auditory canal. May also extend anteriorly to involve the carotid artery

Glomus jugulare

Type 1	Small tumor involving the jugular bulb, middle ear, and mastoid
Type 2	Tumor extending under the internal auditory canal. May have intracranial extension
Type 3	Tumor extending to the petrous apex. May have intracranial extension
Type 4	Tumor extending beyond the petrous apex into the clivus or infratemporal fossa. May have intracranial extension.

**TABLE
127.3****FISCH CLASSIFICATION
OF JUGULOTYMPANIC
PARAGANGLIOMAS**

Class A	Tumors arising on the tympanic plexus confined to the middle ear
Class B	Tumors arising from the inferior tympanic canal in the hypotympanum with middle ear and/or mastoid invasion, but jugular bulb and carotid canal intact
Class C	Tumors arising in the dome of the jugular bulb and involving the overlying cortical bone
C1	Tumors eroding the carotid canal but not involving the carotid artery
C2	Tumors involving the vertical petrous carotid artery
C3	Tumors involving the horizontal carotid canal but not foramen lacerum
C4	Tumors involving the foramen lacerum and cavernous sinus
Class D	Tumors with intracranial extension
De1	Extradural extension of <2 cm medial dural displacement
De2	Extradural extension of >2 cm medial dural displacement
Di1	Intradural extension of <2 cm
Di2	Intradural extension of >2 cm
Di3	Neurosurgically unresectable tumors

❖ **Vagal paraganglioma**

- Account for 5% of all head and neck paragangliomas.
- Originate in vagus nerve where the paraganglia are diffusely distributed in the Superior, middle, and inferior vagal ganglia.
- Female preponderance of 3:1
- Tumors originating superiorly often present with a dumbbell appearance with an intracranial component in the posterior cranial fossa.
- Tumors originating inferiorly will extend into the poststyloid compartment of the parapharyngeal space
- Vagal paragangliomas are frequently associated with multiple cranial neuropathies at presentation in up to 50% of patients
- Most affected CN (in order) 10,12,11
 - **Vagus nerve**
 - **Hypoglossal**
 - **spinal accessory**

Diagnostic Imaging Studies of Paragangliomas:

❖ **CT with contrast:**

- Excellent imaging choice for the diagnosis and delineation of paragangliomas.
- Since these tumor are highly vascular, early and intense contrast enhancement is seen.
- Relation with carotid and the jugular makes it diagnostic.
- Carotid body tumors:
 - Has characteristic splaying of the internal and external carotid arteries by a well-circumscribed mass occupying the carotid bifurcation, and will displace the internal carotid posterolaterally.
- Vagal paraganglioma:
 - Typically will displace the internal carotid *artery* anteriorly and occupy the high parapharyngeal space, with or without involvement of the skull base.
- Jugulotympanic paragangliomas:
 - Can be distinguished in their early phases, especially when a tympanic paraganglioma is confined to the tympanic cavity.
 - Other characteristic findings is erosions of:
 - Jugulotympanic spine.
 - Jugular foramen enlargement with bone destruction
 - Middle ear and mastoid involvement

❖ **MRI with contrast:**

- Complementary and equally important imaging modality in the evaluation and treatment of head and neck paragangliomas.
- Serves three purposes:
 - **Delineation** of the tumor
 - **surveillance** and **detection of concurrent paragangliomas** of the head and neck.
 - **confirmation** of the diagnosis of paraganglioma.
- The classic "***salt and pepper***" appearance of paragangliomas on **T'2-weighted** images is **due to the high-flow vascular** voids that are essentially **pathognomonic**

❖ **Angiography:**

- **Limited role after the evolution of detailed CT and MRI.**
- Angiography is essential in providing a detailed map of tumor blood supply and venous drainage.
- Contraindicated in patients with severe atherosclerosis.
- Preoperative preparation through superselective embolization of the feeding arterial supply to the tumor decreases the risk of intraoperative blood loss.
- Carotid body tumors:
 - Takes blood supply directly from feeding vessel to the carotid body.
- Jugulotympanic tumors:
 - In early development, received blood supply from ascending pharyngeal *artery*.
 - As they grow they recruit feeding vessels from the internal carotid system via caroticotympanic arteries.
 - With invasion of the posterior fossa more supply from the posterior circulation (vertebral arteries).
- **Angiographic balloon occlusion test:**
 - Involves the threading of a trans-femoral catheter to the internal carotid and involves temporarily occluding the internal carotid artery, usually at the carotid siphon within the cavernous sinus to determine whether there will be neurologic deficit
- **Xenon CT angiography** is a precise quantitative study with the **best sensitivity** but is cumbersome and rarely available.

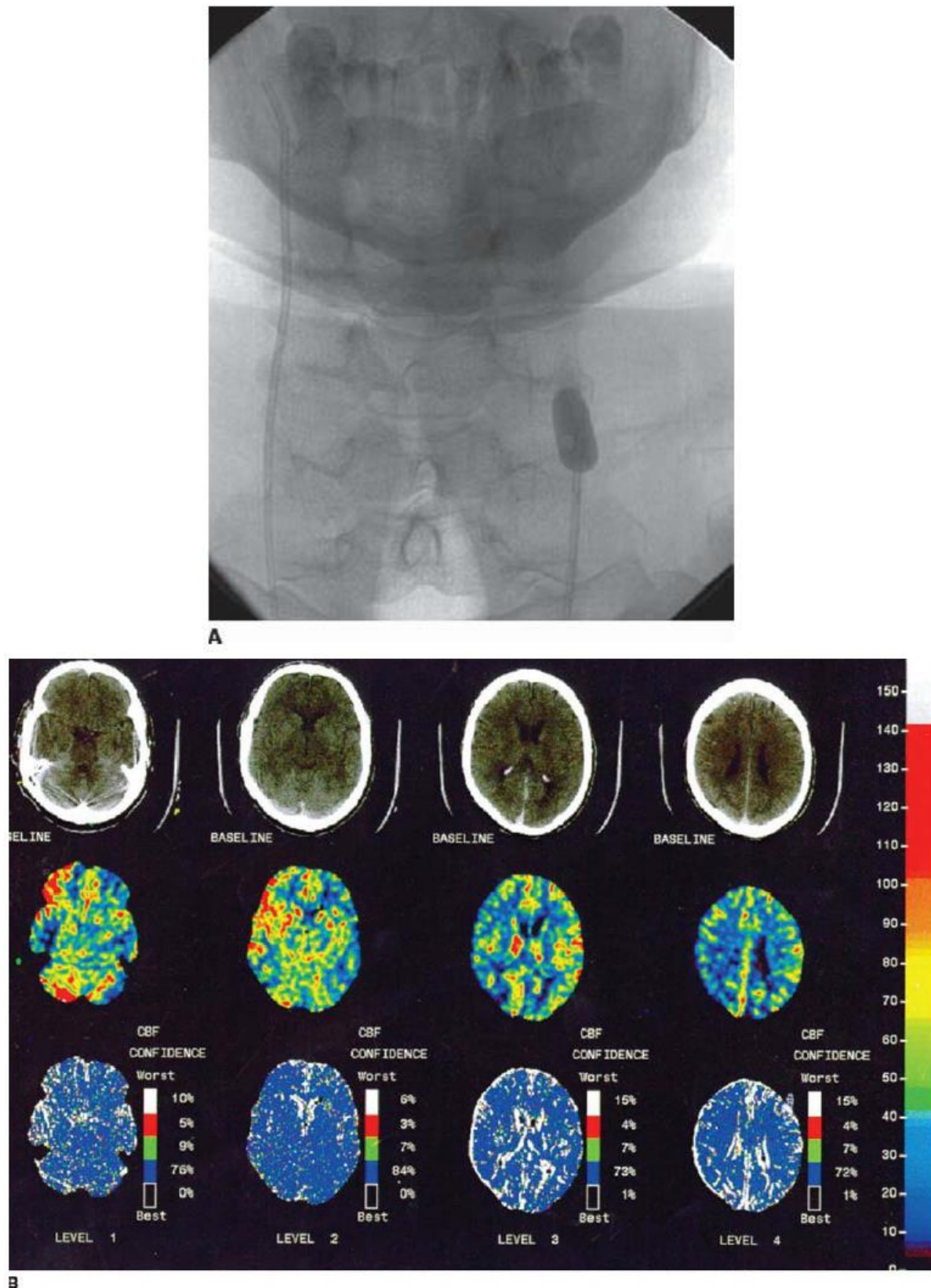


Figure 127.11 Balloon occlusion testing. **A:** Frontal angiographic image shows the inflated radiopaque balloon in the proximal left internal carotid artery, occluding flow. **B:** Quantitative cerebral blood flow images created by inhaling xenon gas during dynamic CT. The color images reveal reduced flow (blue instead of red) in the left MCA distribution.

❖ **Radionuclide Imaging:**

- Targets the biochemical pathways of catecholamine synthesis, storage, and secretion by chromaffin tumor cells.
- A variety of different imaging techniques exist like:
 - MIBG
 - ^{18}F -FDA-PET
 - Octertide scanning

Treatment of Paragangliomas:

❖ Approaches:

1. Observations (Unfit or old patients with slow growing tumor)
 2. Embolization (Pre-op)
 3. Surgical resection (Gold standard)
 4. Radiation (Recurrence or unresectable tumor)
- ❖ Surgical Resection has been the traditional treatment for paragangliomas of the head and neck.
- ❖ Microsurgical skull base techniques such as Fisch infratemporal fossa approaches were developed to specifically resect temporal bone paragangliomas.

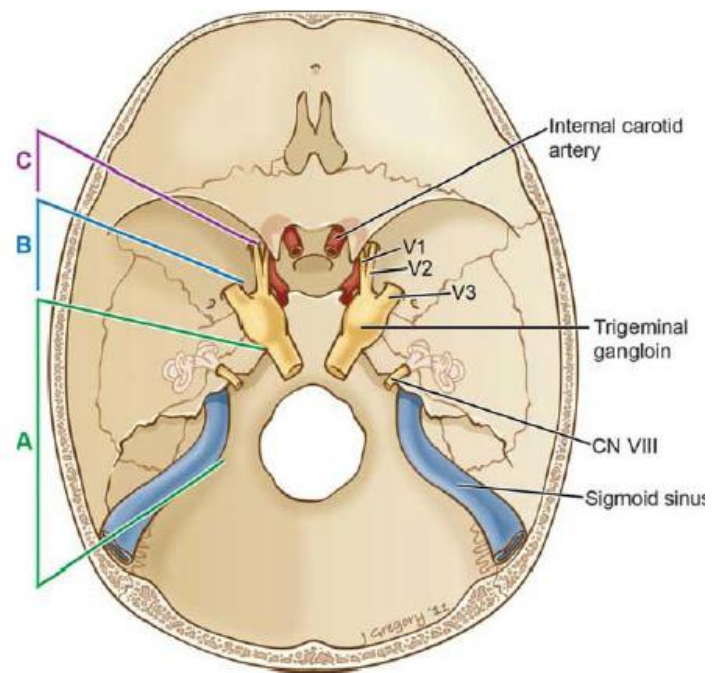


Figure 127.12 Fisch classification of infratemporal fossa approaches. Type A, exposure to the level of the anterior eustachian tube, middle meningeal artery/foramen spinosum. Type B, exposure to the foramen ovale, V3. Type C, exposure to the foramen rotundum, V2.

❖ Surgical considerations

- Age
- Co-morbidities
- Anesthesia risk (active catecholamine metabolites)
- Pre op angiography
- Pre op embolization
- Respectability

❖ **Embolization:**

- Advantages of embolization are:
 - **Tumor shrinkage** and decreased blood flow
 - **Less intraoperative bleeding** diminishes the requirement of potential transfusions
 - Provides for a much **easier dissection**
- Surgery should be performed **within 48 hours** of embolization to avoid reinitiation of collateral circulation
- Administration of **steroids** is useful in reducing the post-embolization inflammatory response.

❖ **Surgical Resection:**

❖ **Carotid Body Tumors:**

- Smaller carotid body tumors can be approached through a transverse neck incision.
- Larger ones through an oblique vertical incision along the anterior border of the sternocleidomastoid muscle.
- Proximal and distal dissection of tumor identify:
 - Internal carotid
 - External carotid
 - Jugular vein
- Vessel loop control applied.
- Carotid **Bypass** precautions should be ready
- Entire course of internal carotid should be exposed
- **The dissection of the tumor is in the subadventitial plane and must be done with extreme care.**
- Tumor may have to be split as it encases the internal carotid artery.
- The dissection of the tumor at **the bifurcation of the carotid is left last** since this is the most vulnerable point for breaching the artery as it is intimately associated with the arterial wall where the tumor originates from the carotid body.

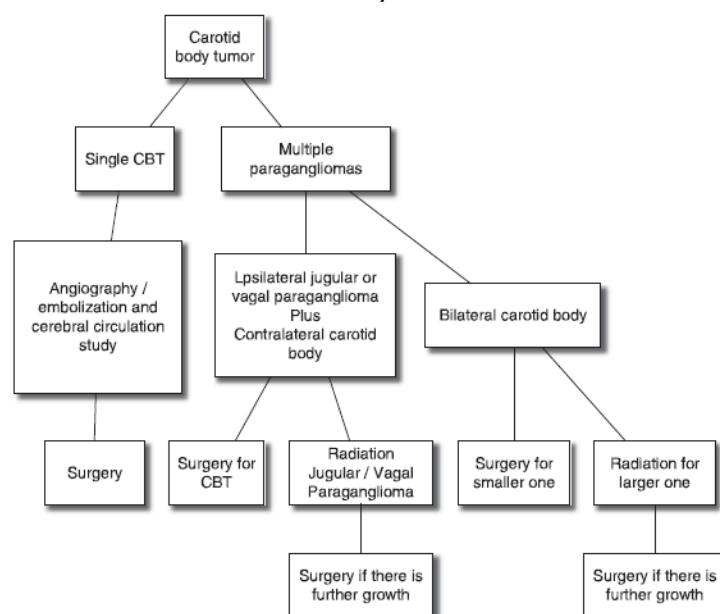


Figure 127.16 Decision analysis tree for the treatment of carotid body tumors.

❖ **Jugulotympanic Paragangliomas:**

- **Small tympanic paraganglioma:** (Glucoc-Jackson Type I or FISCH class A):
 - Can be approached through **transcanal**/inferiorly based tympanomeatal flap.
 - **Embolization is not required.**
 - Bipolar electrocautery microforceps can be used to shrink the tumor and resect it
- **Larger tympanic paraganglioma confined to the middle ear and /or mastoid:**
 - Through postauricular/endaural approach.
 - With a canal wall up mastoidectomy and extended facial recess approach inferiorly that sacrifices the chorda tympani.
- **With jugular bulb involvement:**
 - Combined temporal/cervical approach is required.
 - An extended mastoidectomy and extended facial recess approach is performed with skeletonization of the sigmoid sinus and exposure of the jugular bulb through the retrofacial air cells
 - The internal carotid artery is dissected superiorly and controlled
 - Cranial nerves 9, 10, and 11 are identified and traced proximally and distally
 - The internal jugular vein is then ligated superiorly and dissected toward the jugular bulb in the neck.
 - The sigmoid sinus is occluded and ligated above the tumor extension.
 - Contralateral venous outflow should be verified through preoperative studies.
 - Check Bailey part2 page 373 for full details
- More advanced tumors that involve the vertical and for horizontal petrous carotid artery with possible intracranial extension.
 - Require a postauricular infratemporal fossa approach

❖ **Vagal Paragangliomas:**

- Most vagal paragangliomas originate in the nodose (inferior) vagal ganglion, which is situated 2 cm below the jugular foramen.
- Their bidirectional growth along the vagus results:
 - Involvement of the jugular bulb superiorly
 - Poststyloid parapharyngeal space inferiorly.
- Vagal paragangliomas originating in the superior and middle ganglia, which are situated within the jugular foramen cause early skull base invasion with intracranial extension.
 - The surgical techniques to approach these tumors are identical to those for jugulotympanic paragangliomas when the skull base is involved.

- **Surgical Complications:**

- **Vascular injury (more with carotid body tumors)**
- **Stroke rate (0-2% was around 20%)**
- **Baroreflex Failure** (when bilateral carotid body tumors removed , bilateral denervation of the carotid sinus will happen leading to **in the postoperative period:**
 - Labile hypertension
 - Diaphoresis
 - Headache
 - Tachycardia
 - May be managed with sodium nitroprusside
 - Long term control with clonidine
- **Cranial nerve injuries** its location and site specific (in order) :
 - vagal
 - jugultympanicum
 - carotid body tumors

- ❖ **External Beam Radiation**

- Paragangliomas are responsive to radiation treatment.
- By obliterative endarteritis at the capillary level, followed by fibrosis.
- Indications:
 - Unresectable tumors.
 - Tumors in elderly and medically infirm patients deemed
- Optimal dose appears to be **45 Gy/over 5 weeks**.

- ❖ **Stereotactic Radiation**

- Deliver a **one time high dose of radiation** to the target in question, which is highly focused and has a sharp drop off to **spare surrounding normal tissue**.
- Sparing Cranial nerves
- Cannot be used in :
 - Tumor larger than 4 cm
 - Carotid body tumors
 - Tumors extending caudal to the skull base as they are out of the stereotactic frame field.

Treatment Comparisons

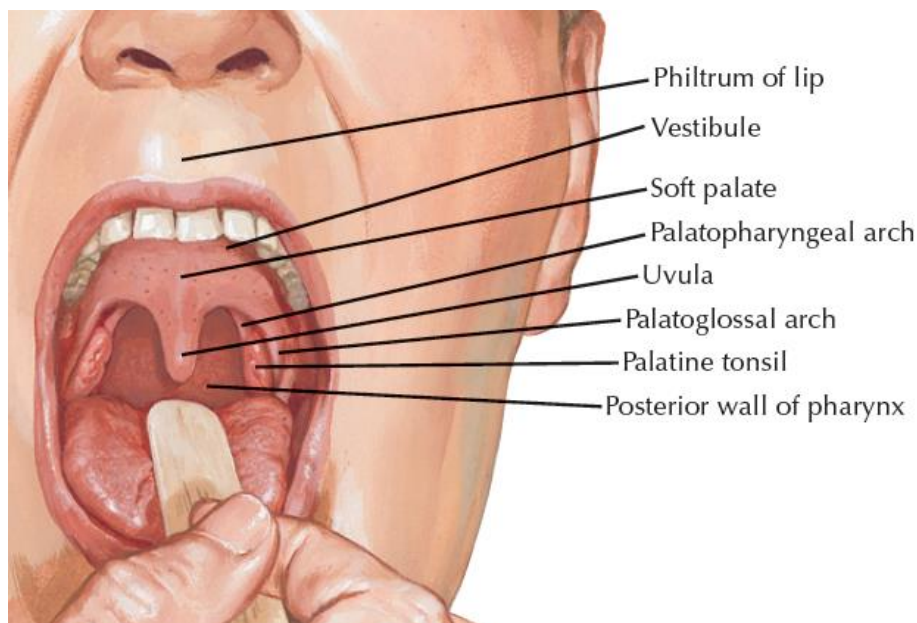
- Results of surgery, external beam radiation, and stereotactic radiation show similar efficacy in affecting cure **around 90%**.

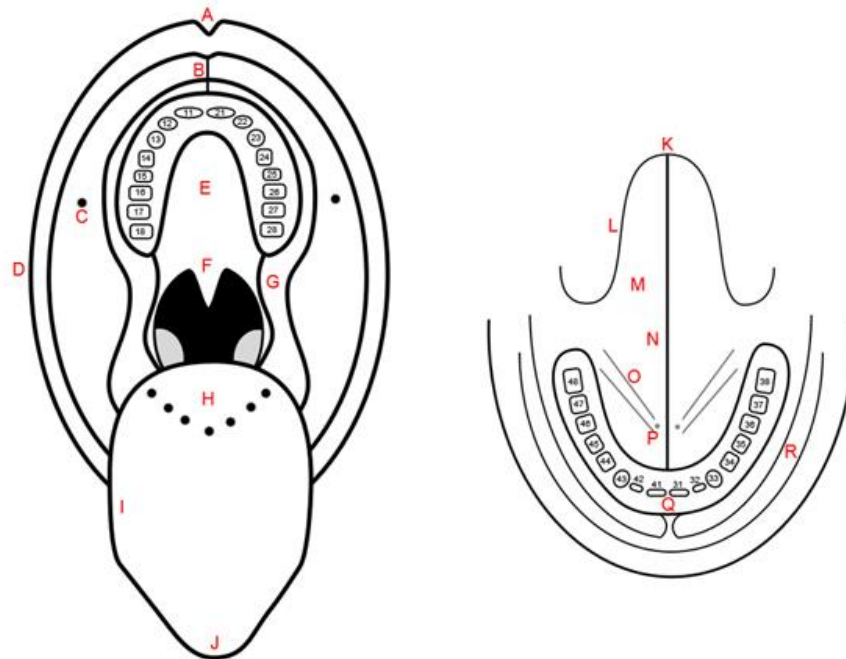
Treatment Strategies:

- Primary surgery is the first line of choice for:
 - Unilateral small- to moderate-sized carotid body tumors
 - Tympanic paragangliomas without cranial nerve dysfunction
- In bilateral carotid body tumors, surgery on both sides should be avoided because of labile hypertension.
- Radiation therapy is a highly effective treatment for jugulotympanic and vagal paragangliomas without cranial nerve dysfunction.
- For patients with jugulotympanic as well as vagal paragangliomas, surgery should be the primary modality when lower cranial nerve dysfunction is present especially vagal.
- Radiation therapy as a primary modality is best suited for those who have medical comorbidities and those who have no lower cranial deficits at presentation.
- Elderly patients can either be observed or treated with radiation as the primary treatment.
- Salvage therapy for recurrent tumors should be offered.
- Paragangliomas suspected or proved to be malignant, either by:
 - Fine needle biopsy of suspicious lymph nodes
 - Positive PET-FDG scans
 - Should also be managed by surgery as the primary modality, given the unknown response rate to radiation alone

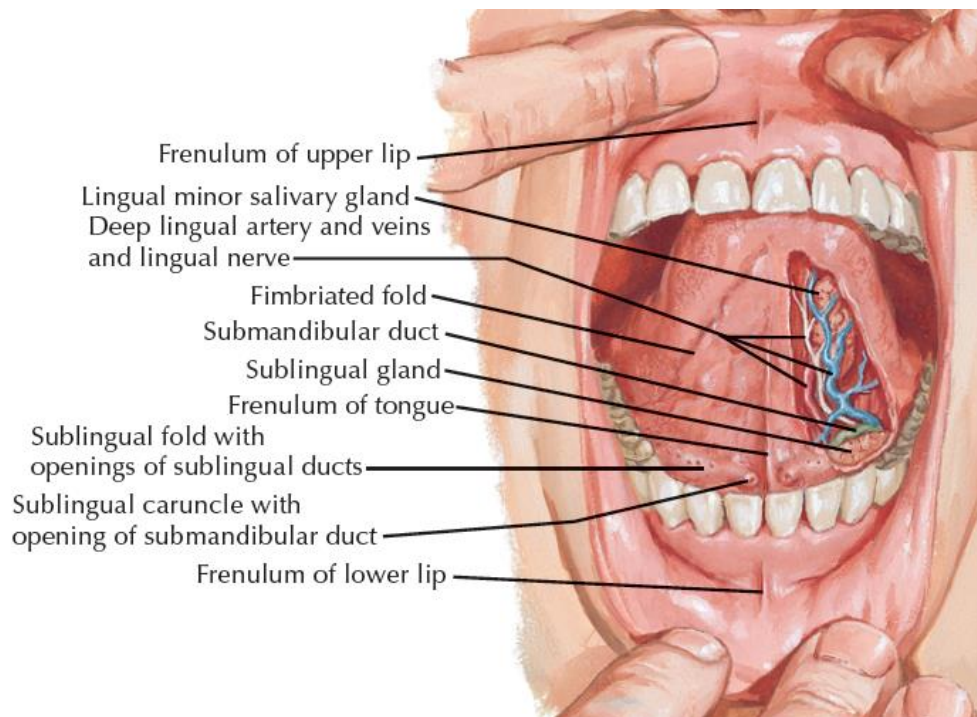
Anatomy of Oral Cavity:

- Space bounded by:
 - **Anteriorly:** Lips.
 - **Posteriorly:** Palatoglossal Fold and Oropharynx.
 - **Laterally:** Cheeks
 - **Superiorly:** Hard Palate.
 - **Inferiorly:** Floor of mouth and Anterior 2/3 of tongue.
- Oral cavity can be divided into:
 - **Oral Vestibule:**
 - Area between Alveolar ridges and Lips or Cheek.
 - **Oral Cavity proper:**
 - Area located internal to Teeth.
- Important in mastication, tasting, and talking
- All of the major salivary glands drain into Oral cavity.
- Lining Mucosa of Oral Cavity:
 - 1. Lining mucosa:**
 - Non-keratinized stratified squamous epithelium.
 - Found in most of oral cavity.
 - 2. Masticatory mucosa:**
 - Keratinized stratified squamous epithelium.
 - Found in:
 - 1.** Dorsum of Tongue.
 - 2.** Hard palate.
 - 3.** Gingiva.
 - 3. Specialized mucosa:**
 - Found in regions of taste buds on lingual papillae on dorsal surface of Tongue.



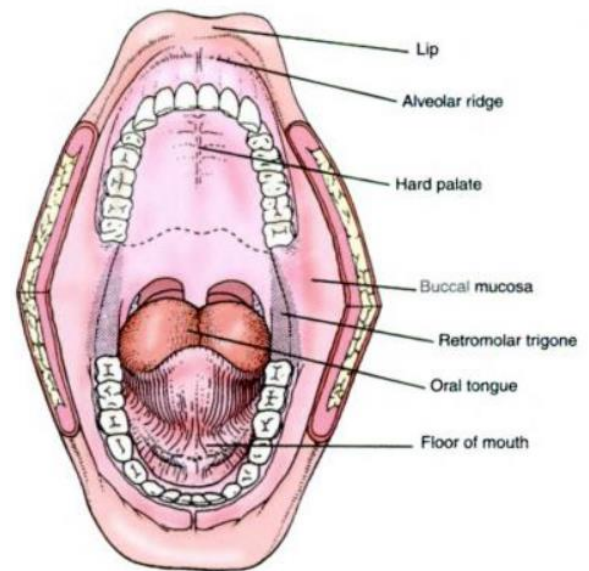


A: Philtrum; **B:** Upper Labial Frenulum; **C:** Opening of Stensen's Duct
D: Labial Commissure; **E:** Hard Palate; **F:** Soft Palate
G: Intermaxillary Commissure; **H:** Base of Tongue;
I: Lateral Border of Tongue; **J:** Tip of Tongue; **K:** Tip of Tongue
L: Lateral Border of Tongue; **M:** Ventral Surface of Tongue
N: Lingual Frenulum; **O:** Floor of Mouth; **P:** Opening of Wharton's Duct
Q: Vestibular Gingiva; **R:** Vestibule.



- **Oral Cavity Sub-sites:**

1. Lips
2. Buccal
3. Alveolar ridges
4. Retro-Molar Trigone
5. Hard Palate
6. Anterior 2/3 of Tongue
7. Floor of mouth



- **Oral Vestibule:**

- Area between Alveolar ridges and Lips.
- Fold of tissue created by vestibule between Lips and Teeth is called **Vestibular or Mucolabial Fold**.
 - o As vestibular fold reflects on alveolar bone holding the teeth, the mucous membrane abruptly changes into **Gingiva**.
 - o Within vestibular fold are bands of tissue known as **Labial Frenula**.
 - Labial frenula are pronounced at Maxillary and Mandibular midline as Upper and Lower Frenula.
 - Other accessory frenula also are located in the vestibule
- When teeth are in occlusion, Vestibule communicates with Oral Cavity Proper via Intermaxillary Commissure behind Last Molar teeth.

- **Lips:**

- Divided into upper and lower lips that surround opening of oral cavity.
- Longer upper lip and Shorter lower lip are connected to each other by Labial Commissures at corners of Mouth.
- Both lips have a muscular composed of **Orbicularis Oris muscle**.
- Upper lip is separated from cheek by Nasolabial groove
- Lower lip is separated from chin by Labiomental groove.
- **Vermilion** is Red area of lip that is clearly demarcated from the skin.



- Each Lip has the following parts:

1. Outer Surface:

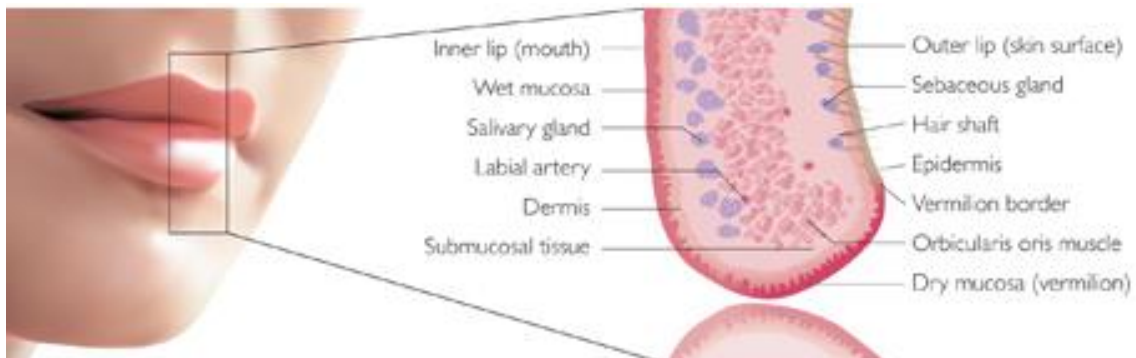
- Dry vermillion.
- Covered by Skin ([Keratinized stratified squamous epithelium](#)).
- Rich in Nerve endings, Blood and Lymphatic capillaries.
- Lip skin contains hair follicles, sebaceous and sweat glands.

2. Inner Surface:

- Wet vermillion.
- Covered by Mucous membrane ([Non-keratinized stratified squamous epithelium](#)).
- Rich in Nerve endings, Blood and Lymphatic capillaries and Minor salivary glands.

3. Red Margin:

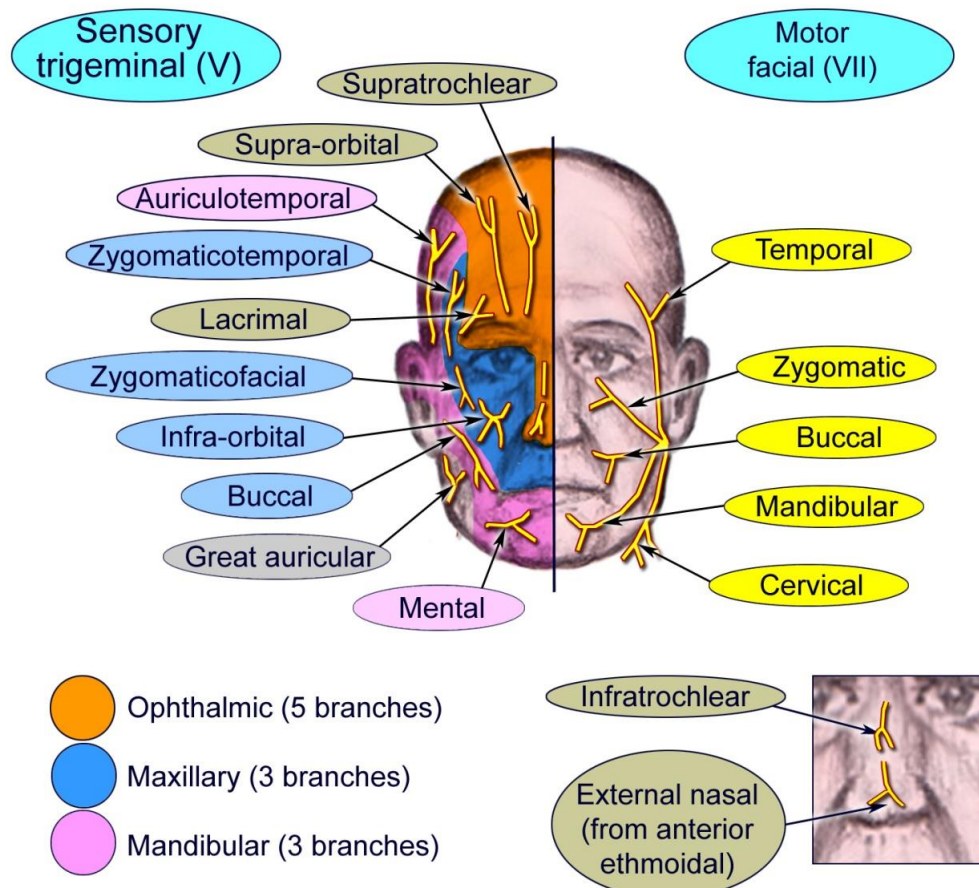
- Transitional zone between wet and dry vermillion.
- Found in humans only.
- Covered with partially keratinized stratified squamous epithelium.
- Rich in blood capillaries and sensory nerve endings, which render the lip highly vascular as well as highly sensitive.
- No sweat glands or hair follicles are present.



- **Philtrum** is the depressed area located between base of Nose and Vermilion border of upper lip.
- **Motor Nerve Supply to Lips:**
- Upper Lip:
 - Facial Nerve (CN-VII) → **Buccal Nerve.**
- Lower Lip:
 - Facial Nerve (CN-VII) → **Marginal Mandibular Nerve.**

- **Sensory Nerve Supply to Lips:**
- Upper Lip:
 - o Trigeminal Nerve (CN-V) → Maxillary Nerve → Infraorbital Nerve → **Superior Labial Nerve**.
- Lower Lip:
 - o Trigeminal Nerve (CN-V) → Mandibular Nerve → Inferior alveolar nerve → **Mental Nerve**.

Face: motor and sensory supply



Facial nerve branches

Temporal: frontalis and procerus

Zygomatic 1: eye and around orbit

Zygomatic 2: mid face and smile

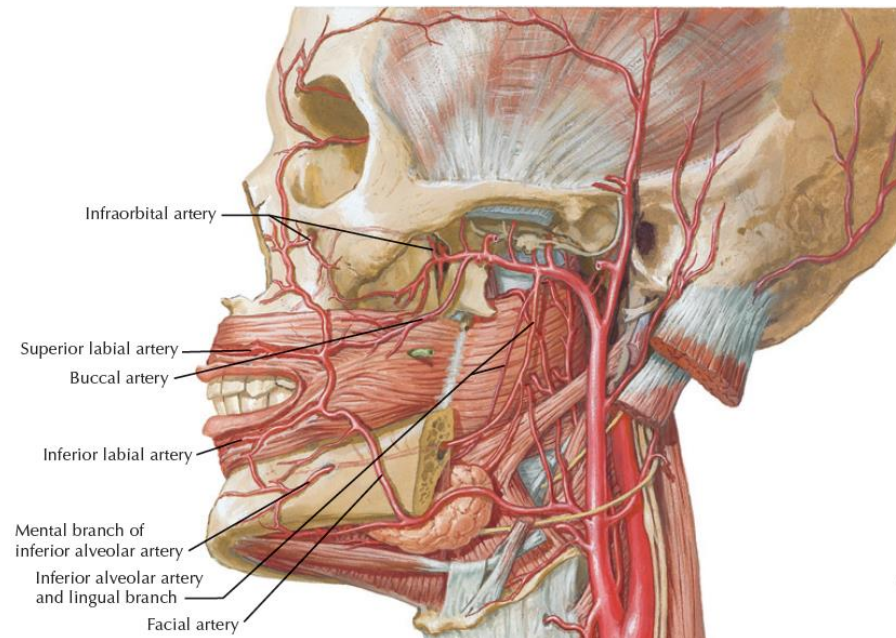
Buccal: buccinator and upper lip

Mandibular: lower lip and orbicularis oris

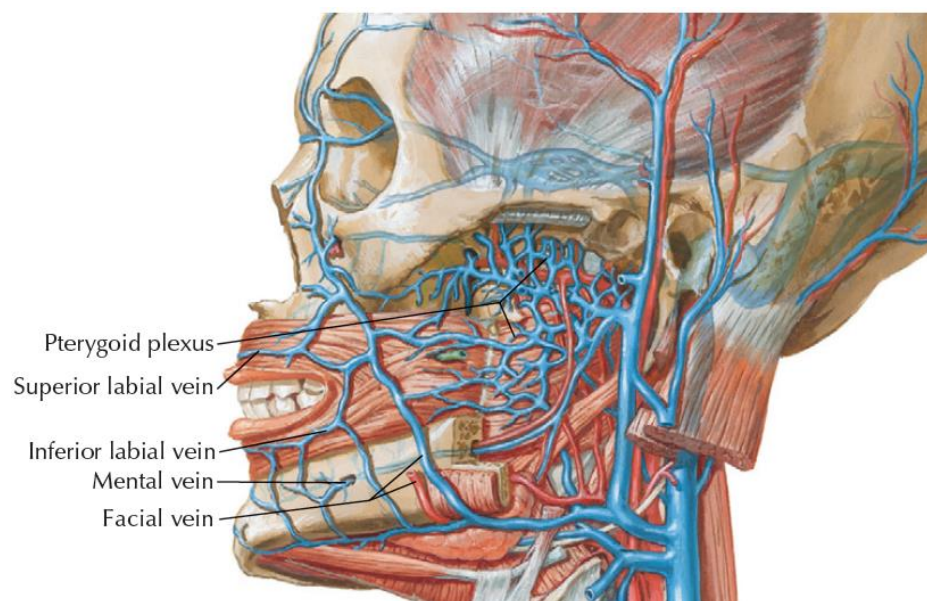
Cervical: platysma

(Note: proprioception is supplied by trigeminal)

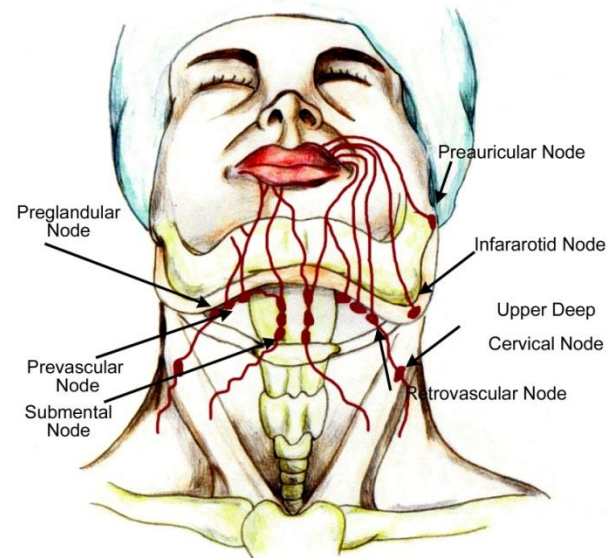
- **Arterial Blood Supply to Lips:**
- External carotid Artery
- → Facial Artery.
- → **1- Superior Labial Artery** → Upper Lip.
- → **2- Inferior Labial Artery** → Lower Lip.



- **Venous Drainage of Lips:**
- Upper Lip → **Superior Labial Vein.**
- Lower Lip → **Inferior Labial Vein.**
- → Facial Vein



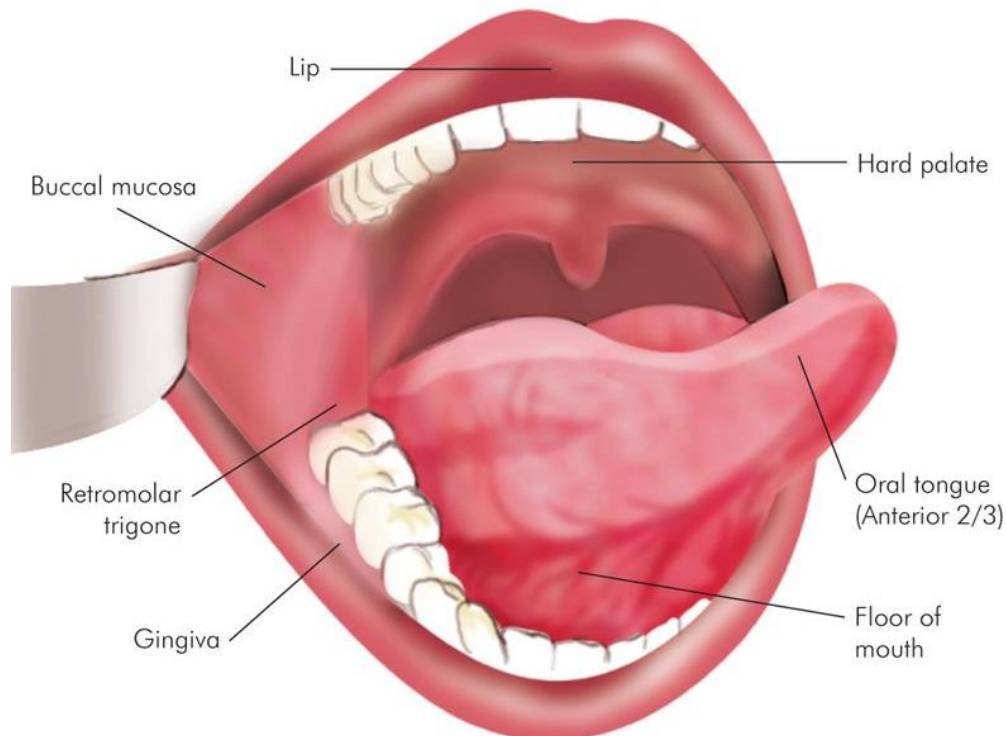
- **Lymphatic Drainage of Lips:**
- Upper Lip:
 - o Mainly to Submandibular LN.
 - o Some drainage also going to Submental and Periparotid LN.
 - o Bilateral involvement of LN if in Midline.
- Lower Lip:
 - o Midline:
 - Bilateral Submental LN.
 - o Lateral:
 - Unilateral Submandibular LN.
- Submental LN secondarily drain to ipsilateral submandibular LN, and both submandibular and parotid nodes secondarily drain to ipsilateral jugulodigastric LN.



- **Cheeks:**
- Musculomembranous structure limited:
 - o Superiorly and Inferiorly: by Upper and Lower Vestibules
 - o Anteriorly: by Labial Commissure.
 - o Posteriorly: by Retromolar trigone and Intermaxillary commissure.
- Lined with **Non-keratinized stratified squamous epithelium**.
- Contains many Minor salivary glands.
- **Buccinator muscle** forms the muscular framework of Cheeks.
 - o Covered by Buccal fat pad.
 - o Masseter muscle covers the buccinator.
- **Retromolar region** is the only area in which vestibule and oral cavity proper communicate.
- **Stensen's duct** (drainage duct of Parotid) runs through Buccinator muscle and opens opposite to 2nd upper molar.



- **Blood Vessels of Cheeks:**
- External carotid Artery → Maxillary Artery → **Buccal Artery.**
- **Motor Nerve Supply to Cheeks:**
- Facial Nerve (CN-VII) → **Buccal Nerve.**
- **Sensory Nerve Supply to Cheeks:**
- Trigeminal Nerve (CN-V) → Mandibular Nerve → **Buccal Nerve.**
- **Lymphatic Drainage of Cheeks:**
- Skin of Cheeks → Parotid and Submandibular LN.
- Mucous membrane of cheeks → Submandibular and Upper Deep Cervical LN.
- **Retromolar Trigone:**
- Small triangular-shaped subsite of oral cavity.
- Mucosa lies behind 3rd molar tooth, covering Anterior ramus of Mandible.
- Bounded laterally by Gingival buccal sulcus
- Bounded medially by Anterior tonsillar pillar.
- Base of the triangle is posterior to last inferior molar tooth.
- Apex of the triangle is in continuity with Tuberosity of Maxilla behind last upper molar tooth.
- Contains many Minor salivary glands.

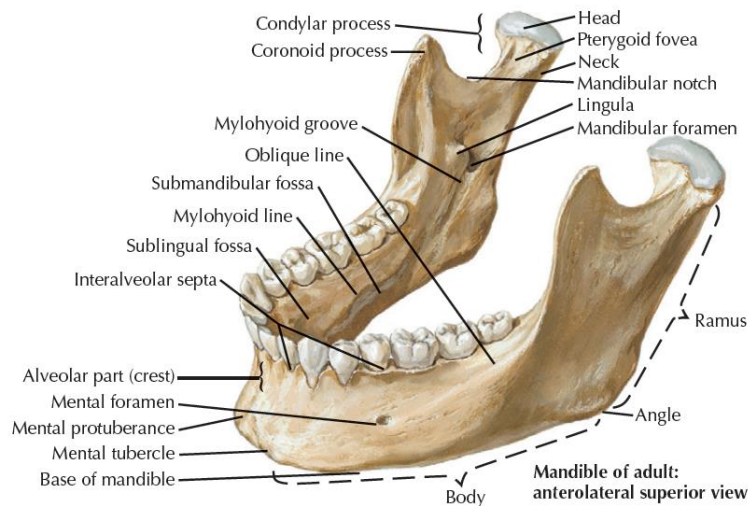


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Figure 41.1 Various sites in oral cavity.

- **Gums and Alveolar Process:**
- Gingiva (Gum) is a fibroepithelial mucosal tissue that surrounds teeth and covers the alveolar jawbone.
- Alveolar process is the tooth-bearing area of Jaws.
- Lined with **Keratinized stratified squamous epithelium**.
- **Devoid of Minor Salivary glands.**

- **Lymphatic Drainage of Gums:**
- Outer aspect of Upper alveolus → Submandibular LN.
- Inner aspect of Upper alveolus → Upper Deep Cervical LN.
- Both aspects of Lower alveolus → Submandibular LN.



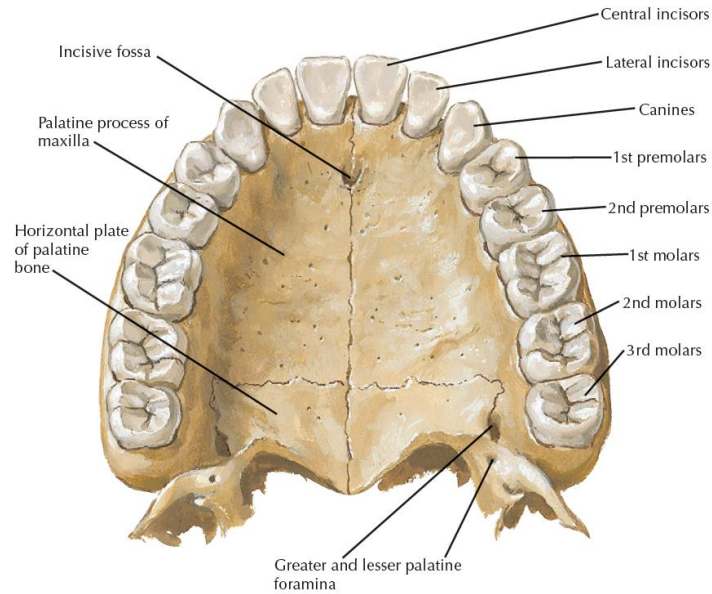
- **Hard Palate:**
- Horseshoe-shaped, domed roof of the oral cavity.
- Separates Oral cavity from Nasal cavities.
- Hard palate is concave, occupied mostly by Tongue at rest.
- Subdivided into:

1. Primary Palate (Premaxilla):

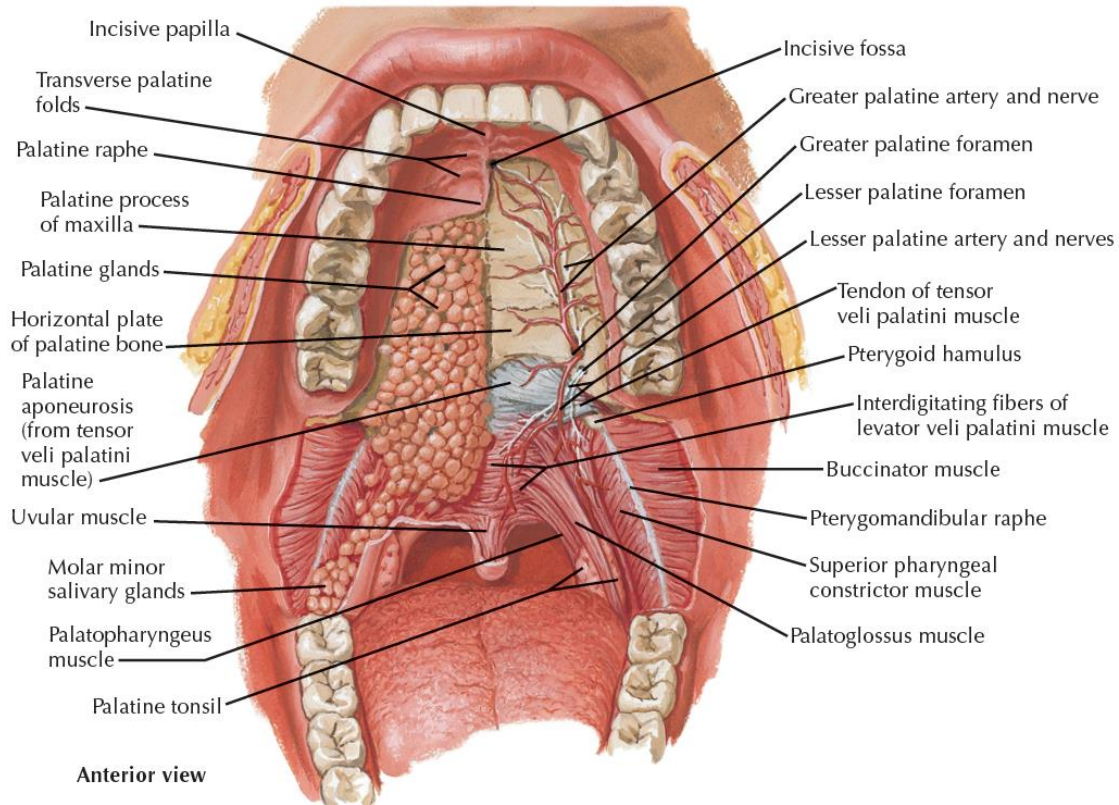
- Anterior to Incisive Foramen.
- Formed First.
- Wedge shaped.
- Houses 4 incisors.
- **Devoid of Minor Salivary glands.**

2. Secondary Palate :

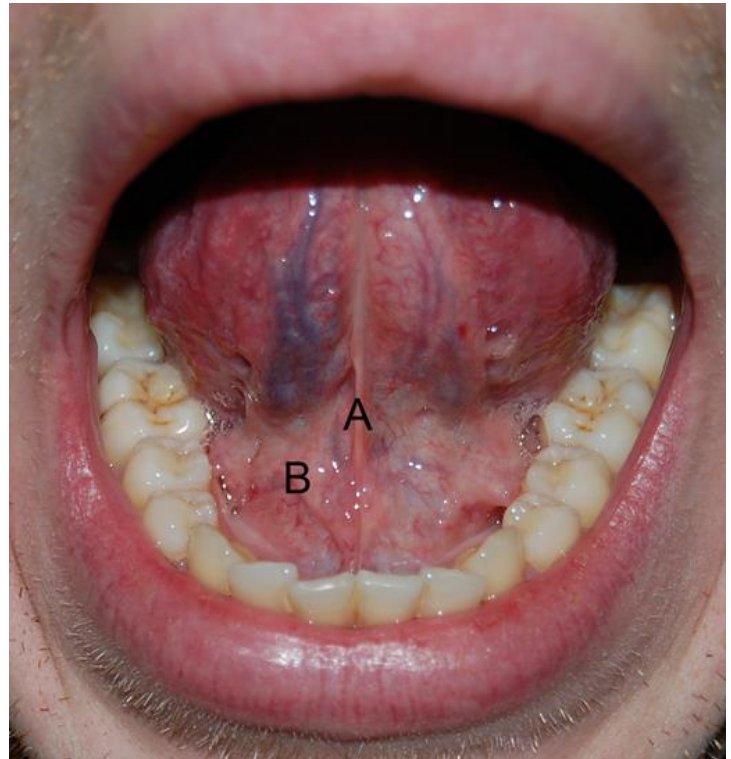
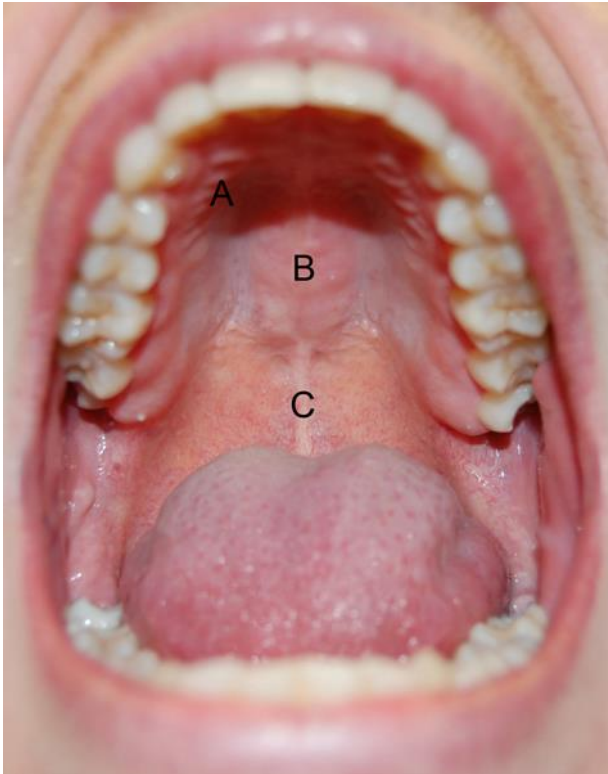
- Non-premaxilla portion of hard palate
- Posterior to Incisive Foramen.
- Anterior 2/3 is formed by Palatine processes of Maxilla.
- Posterior 1/3 is formed by Horizontal Plates of Palatine bone.
- Midline suture line called Median or Palatine raphe.
- Transverse ridge serves to retain food bolus.
- Contains many Minor salivary glands.



- Hard palate is lined with [Keratinized stratified squamous epithelium](#).
- Mucosa adheres firmly to the periosteum.
- Incisive foramen transmits terminal branches of Nasopalatine Nerve and Sphenopalatine Vessels.
- Greater and lesser palatine foramina transmit Greater and Lesser Palatine nerve and vessels.

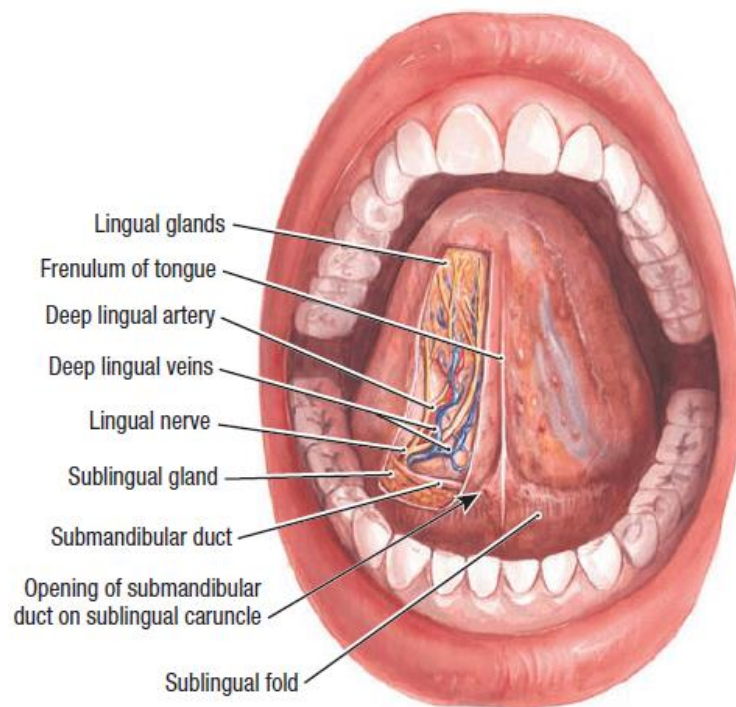


- **Lymphatic Drainage of Hard Palate:**
- Anterior part of Palate → Submandibular LN.
- Rest of Palate → Upper deep cervical and Lateral Retropharyngeal LN.



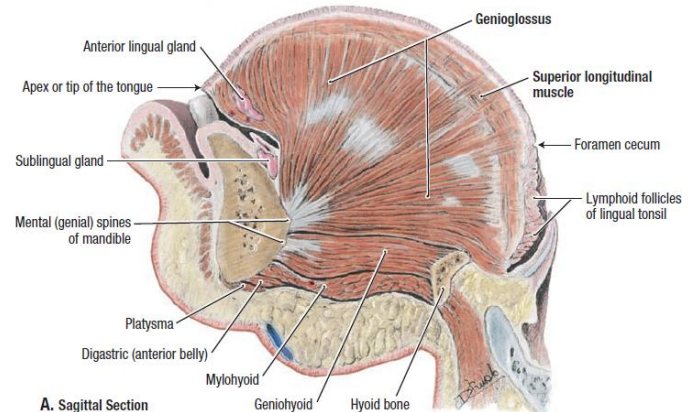
- **Floor of Mouth:**
- Inferior limit of oral cavity.
- Semilunar space over Mylohyoid and Hyoglossus muscles.
- Lined with **Non-keratinized stratified squamous epithelium**.
- Extends from Inner surface of lower alveolar ridge to undersurface of Anterior 2/3 of Tongue.
- Its posterior boundaries are the bases of Anterior pillars of tonsils.
- Floor of oral cavity is divided into two sides by Frenulum of Tongue.
- Sublingual papillae (Caruncles or Folds) can be identified on both sides of Frenulum in Anterior part of the floor of mouth when tip of Tongue is raised.
- Excretory duct of Submandibular gland (Wharton's duct) runs in the floor of the mouth along medial border of Sublingual gland to pierce surface of mouth at Paramedian sublingual caruncle.
- Sublingual glands have multiple small ducts that drain directly into Floor of the mouth
- Contains many Minor salivary glands.

- **Sensory Nerve Supply to Floor of mouth:**
- Trigeminal Nerve (CN-V) → Mandibular Nerve → **Lingual Nerve.**
- **Lymphatic Drainage of Floor of mouth:**
- Anterior portion of floor of mouth → Submental LN.
- Posterior portion → Submandibular and upper deep cervical LN.



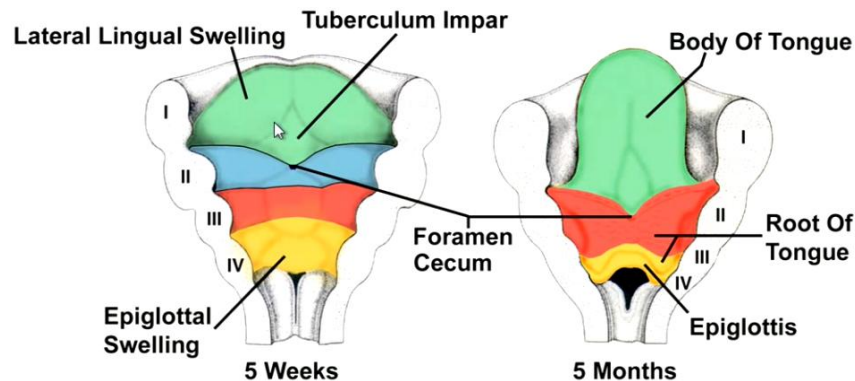
- **Tongue:**

- Mass of muscle that is almost completely covered by Stratified squamous epithelium.
- Occupies most of Oral cavity and oropharynx.
- Role in taste, Mastication, Deglutition, Articulation and oral cleaning.
- Five cranial nerves contribute to the complex innervation of this multifunctional organ.



- **Embryology:**

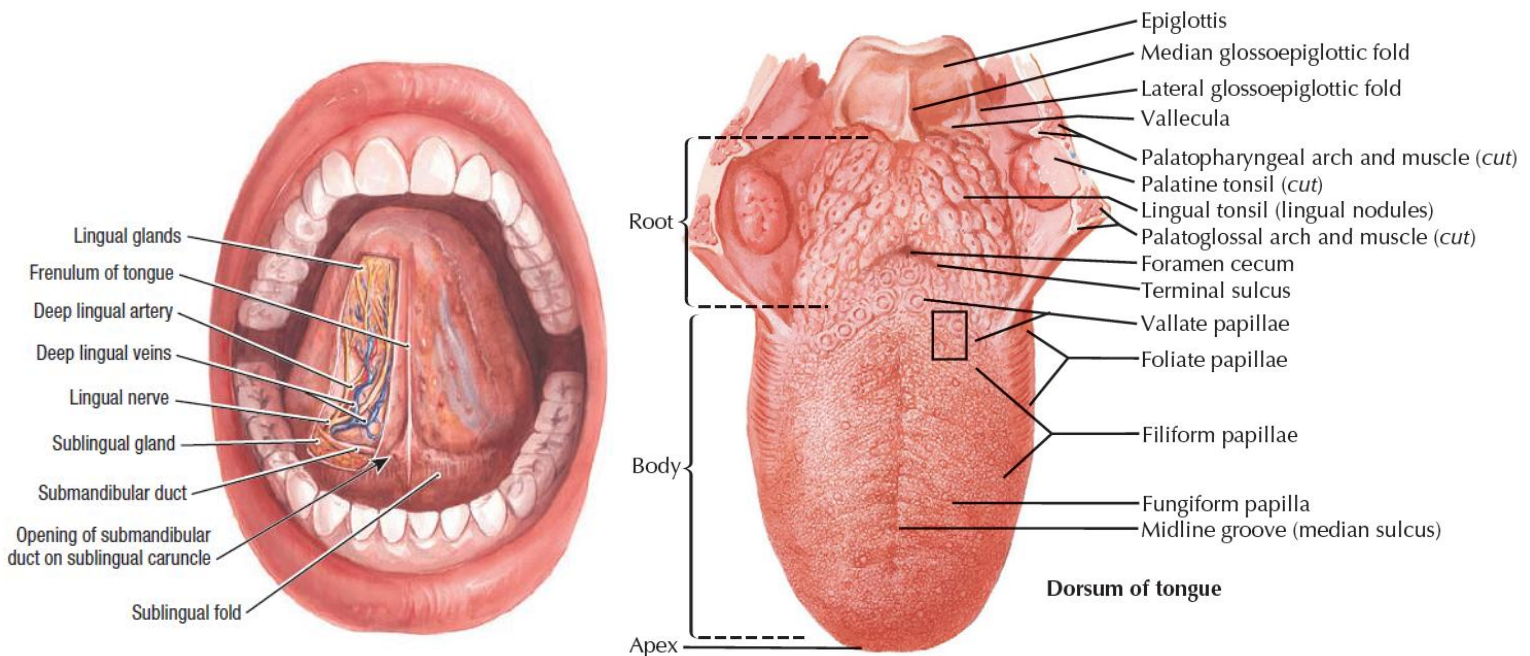
- Appear at 4 weeks' gestation.
- **1st Pharyngeal Arch gives rise to:**
 1. One median swelling (Tuberculum impar).
 2. Two Lateral lingual swellings
 - o Slowly grow over Tuberculum impar and merge forming **Anterior 2/3 of Tongue.**



- **3rd and 4th Pharyngeal Arches** give rise to **Base of the tongue.**
- **2nd Pharyngeal Arch** doesn't contribute to tongue formation.

Pharyngeal Arch	Embryonic Structure(s)	Adult Structure	Innervation
1	2 lateral lingual swellings Tuberculum impar	Anterior 2/3 of the tongue	GSA: Lingual branch of the mandibular division of the trigeminal n. SVA: Chorda tympani of the facial n.
2	Is overgrown by the 3rd arch; does not contribute to the adult tongue Very little contributes to the hypobranchial eminence	Does not contribute to the adult tongue	
3	Hypobranchial eminence	Posterior 1/3 of the tongue	GSA: Glossopharyngeal n. SVA: Glossopharyngeal n.
4	Hypobranchial eminence Epiglottic swelling Arytenoid swelling Laryngotracheal groove	Root of the tongue	GSA: Internal laryngeal branch of the vagus n. SVA: Internal laryngeal branch of the vagus n.

- **Anatomy:**
- Tongue has 3 surfaces:
 1. **Tip:**
 - Pointed Anterior portion of Tongue.
 - Highly mobile.
 2. **Body:**
 - Anterior 2/3, posterior to the tip.
 - Dorsal (Superior) surface:
 - **Epithelium:** Keratinized stratified squamous epithelium.
 - **Median sulcus:** separates the body into left and right halves.
 - **Terminal sulcus:** separates body from the base of the tongue.
 - **Foramen cecum:** remnant of the proximal thyroglossal duct located at the tip of Terminal sulcus.
 - Ventral (Inferior) surfaces:
 - **Epithelium:** Non-keratinized stratified squamous epithelium.
 - **Lingual Frenulum:** Midline fold connects ventral surface of the tongue to floor of oral cavity.
 - **Sublingual Caruncle:** A swelling on both sides of lingual frenulum marks openings of Submandibular glands ducts.
 - **Sublingual Folds:** Overlies Sublingual glands on floor of oral cavity
 3. **Base:**
 - Posterior 1/3.
 - Contains Lingual tonsils, Inferiormost portion of **Waldeyer's ring**.
 - **Epithelium:** Non-keratinized stratified squamous epithelium.
 - **Glossoepiglottic folds:** Mucous membranes connect base of tongue to with Epiglottis.



- **Lingual Papillae:**

- Projections of lamina propria covered with epithelium.
- Limited only to Anterior 2/3 of dorsal surface of tongue.

- Types of lingual papillae:

1. **Fungiform:**

- Mushroom shaped papillae.
- More numerous at Tip of the tongue
- Rich in blood supply that gives them a bright-red color.
- Involved in taste sensation.

2. **Filiform:**

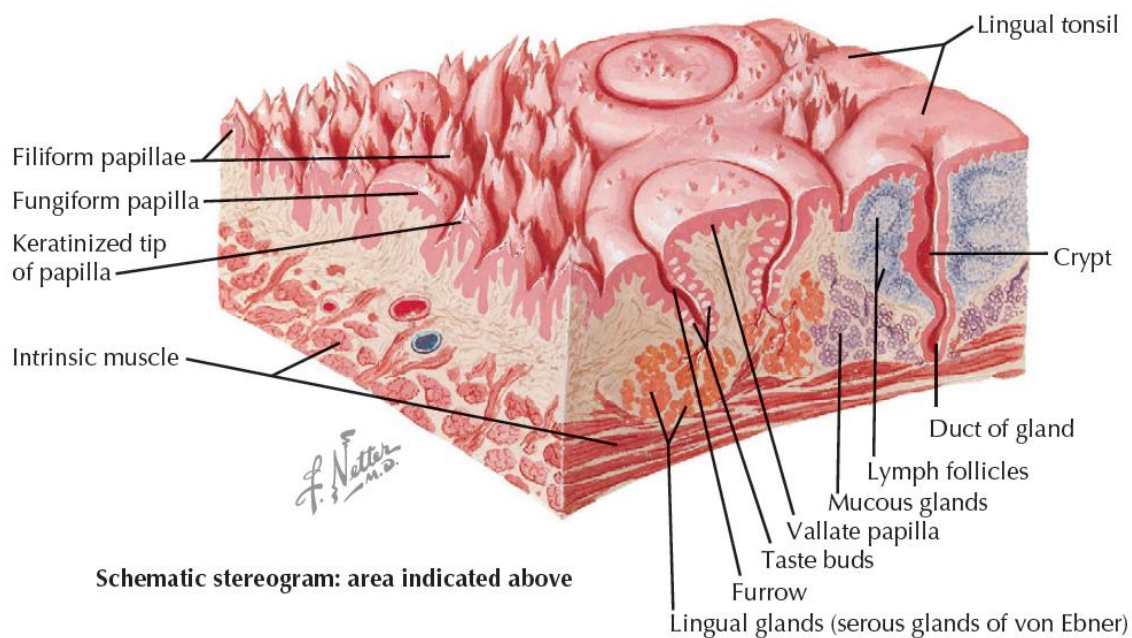
- Thread-like papillae.
- Most numerous papillae
- Located along entire Dorsum of tongue
- **Not involved in taste sensation.**

3. **Foliate:**

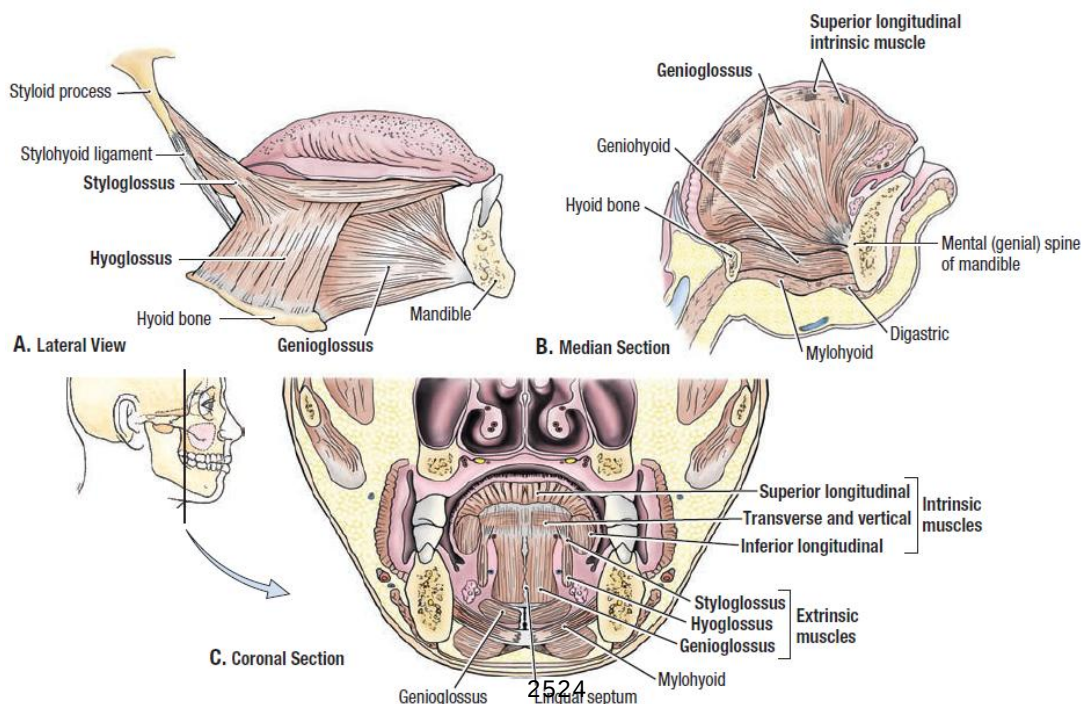
- Folds of mucosa separated from each other by deep grooves.
- Located along Lateral surface of Tongue.
- Rudimentary in humans
- Involved in taste sensation.

4. **Circumvallate:**

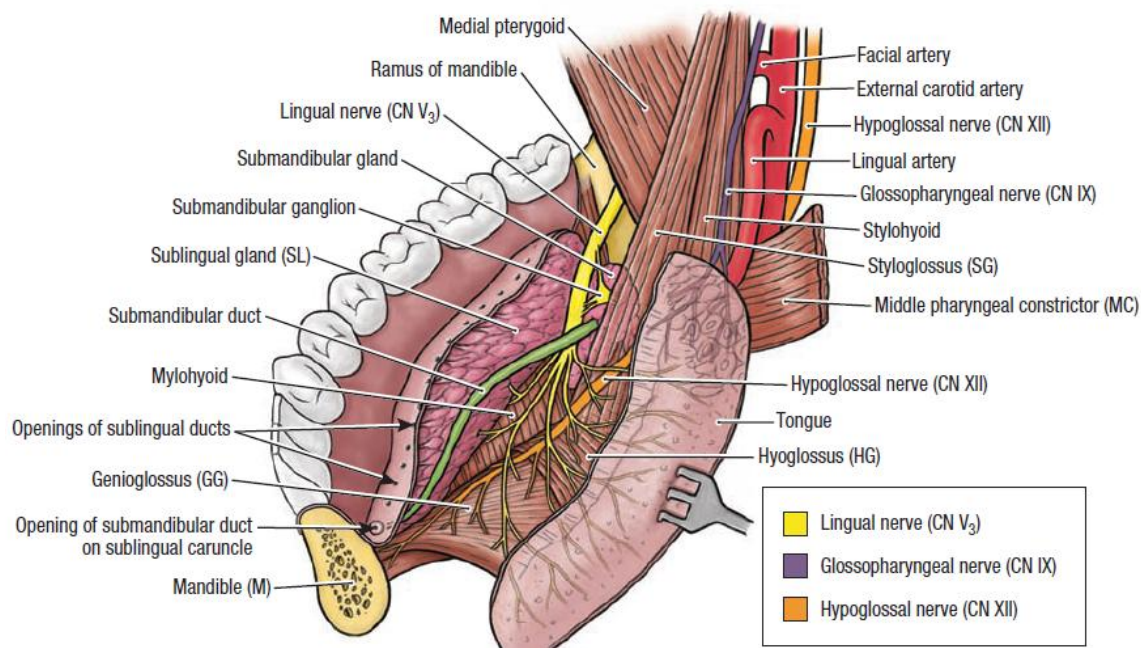
- Large, flat papillae surrounded by troughs.
- 8-12 Circumvallate papillae.
- Located directly Anterior to Terminal sulcus.
- Involved in taste sensation.
- Contains ducts of Von-Ebner glands which secrete lingual lipase into the surrounding troughs to begin the process of lipolysis.



- Taste buds presents on the tongue, mainly along the sides of the Circumvallate, Fungiform and Foliate papillae **(Not in Filiform)**.
- All taste buds can perceive 5 different taste qualities:
 1. Salt
 2. Sweet
 3. Bitter
 4. Acid
 5. Umami.
- **Minor salivary glands:**
- Dorsal surface of tongue contains both serous and mucous salivary glands.
- Serous glands are present in relation to Circumvallate papillae:
 - o Known as **von-Ebner's glands**.
- **Tongue Musculature:**
- Intrinsic muscles:
 - o Originated within tongue and Not attached to bone.
 - o Change tongue's shape.
 - 1. **Superior longitudinal:** (CN-XII)
 - 2. **Superior longitudinal:** (CN-XII)
 - 3. **Transverse:** (CN-XII)
 - 4. **Vertical:** (CN-XII)
- Extrinsic muscles:
 - o Originated outside tongue and attached to it.
 - o Move the tongue in the oral cavity.
 - 1. **Genioglossus:** (CN-XII)
 - 2. **Hyoglossus:** (CN-XII)
 - 3. **Styloglossus:** (CN-XII)
 - 4. **Palatoglossus:** (CN-X)



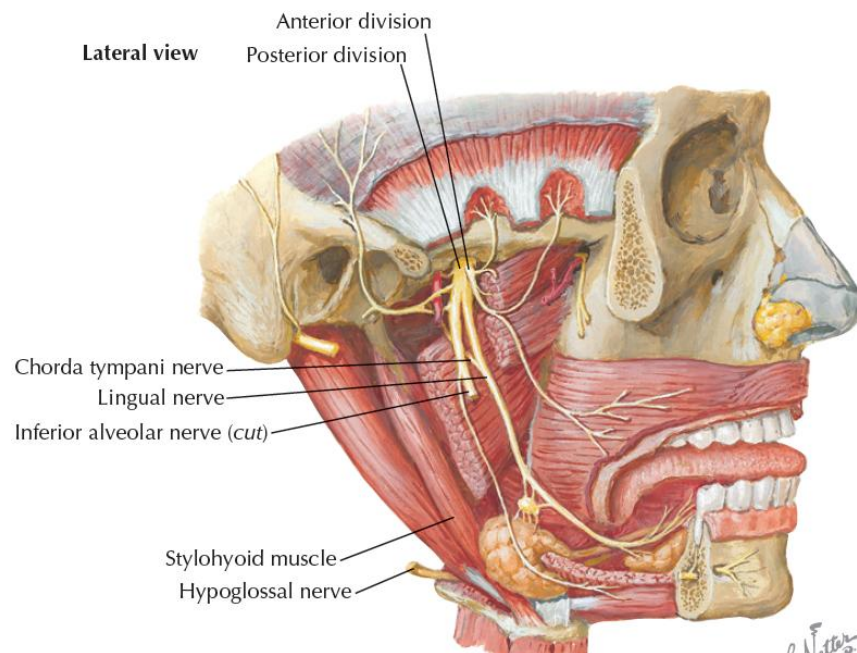
- **Nerve supply:**
- **Lingual Nerve:**
- Terminal branch of Mandibular nerve (**V3**).
- Chorda tympani branch of Facial nerve (**CN-VII**) joins its posterior part.
- Lingual nerve passes between Medial pterygoid and Ramus of the mandible.
- Enters oral cavity bounded by Superior pharyngeal constrictor muscle, Medial pterygoid, Mandible.
- Submandibular ganglion is suspended from Lingual nerve. at Posterior border of Hyoglossus muscle.
- Passes from lateral side inferiorly and medial to Submandibular duct.
- Supplies:
 - General sensation to epithelium and papillae of the tongue's anterior 2/3.
 - Mucosa along the floor of oral cavity (lingualveolar ridge).
 - Gingiva on lingual aspect of the mandibular teeth.



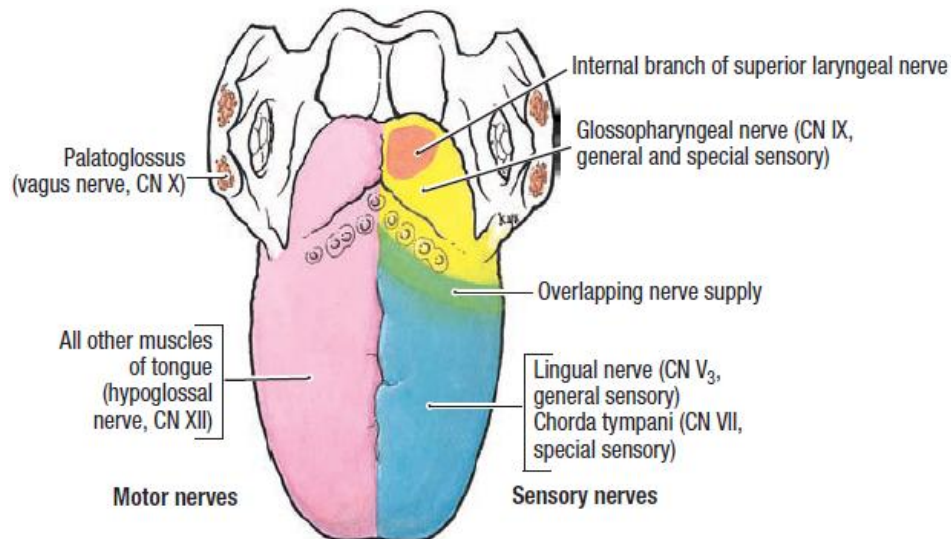
- **Chorda tympani nerve:**
- Branch of Facial nerve (CN-VII).
- Passes Anteriorly to enter tympanic cavity and lies along tympanic membrane and malleus until exiting Petrotympanic fissure.
- Joins the posterior border of Lingual nerve.
- Supplies:
 - Carries preganglionic parasympathetic fibers to tympanic cavity submandibular ganglion.
 - Taste fibers to Anterior 2/3 of the tongue

- **Glossopharyngeal nerve (CN-IX):**
- Passes through Jugular foramen with the Vagus (CN-X) and Accessory nerve (CN-XI).
- Passes between Internal carotid and Internal jugular within the Jugular foramen.
- Continues inferiorly and posteriorly relative to Stylopharyngeus muscle.
- Passes Anteriorly with Stylopharyngeus and travels between the superior and middle constrictor muscles to be located by Palatine tonsils.
- Supplies:
 - Lingual branches distribute Taste sensation to posterior 1/3 and Circumvallate papilla.
 - Lingual branches distribute General sensation posterior 1/3, in addition to fauces.

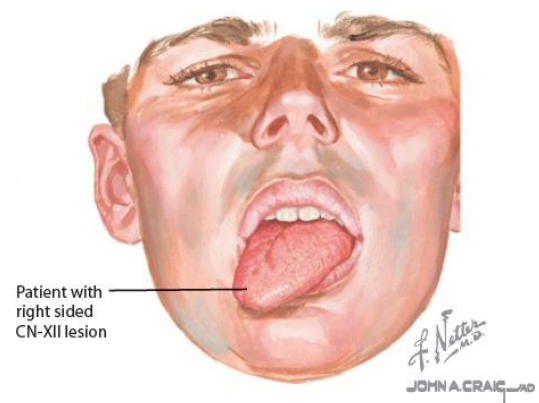
- **Internal laryngeal nerve:**
- Branch of Superior laryngeal nerve of Vagus nerve (CN-X).
- Vagus nerve (CN-X) pass through Jugular foramen between Internal Carotid and Internal jugular vein.
- A series of nerves branch from the vagus in the neck.
- Superior laryngeal nerve travels inferiorly posterior to Internal carotid artery on side of Pharynx and divides into Internal and External laryngeal nerves.
- Internal laryngeal nerve passes inferior to the larynx and passes through the thyrohyoid membrane with Superior Laryngeal vessels.
- Supplies:
 - General sensation fibers to tongue's base at the epiglottic region and mucous membranes of larynx as far inferiorly as the false vocal folds.
 - Taste sensation fibers to tongue's base at the epiglottic region.



- **Motor Innervation:**
- **All of the muscles of tongue (Except Palatoglossus):**
 - o Hypoglossal nerve (CN-XII)
- **Palatoglossus:**
 - o Pharyngeal plexus (CN-X).
- **General Sensation:**
- **Anterior 2/3 of tongue:**
 - o Lingual nerve, terminal branch Mandibular nerve (V3).
- **Posterior 1/3 of tongue:**
 - o Lingual-tonsillar branch of Glossopharyngeal nerve (CN-IX).
- **Base of tongue (Anterior to Epiglottis):**
 - o Internal laryngeal branch of Superior laryngeal nerve (CN-X).
- **Taste Sensation:**
- **Anterior 2/3 of tongue:**
 - o Chorda tympani branch of Facial nerve (CN-VII).
- **Posterior 1/3 of tongue:**
 - o Lingual-tonsillar branch of Glossopharyngeal nerve (CN-IX).
- **Base of tongue (Anterior to Epiglottis):**
 - o Internal laryngeal branch of Superior laryngeal nerve (CN-X).



- **Hypoglossal nerve (CN-XII) Paralysis:**
- Deviation of tongue toward Paralyzed side during protrusion.
- Eventually atrophies on the paralyzed side.

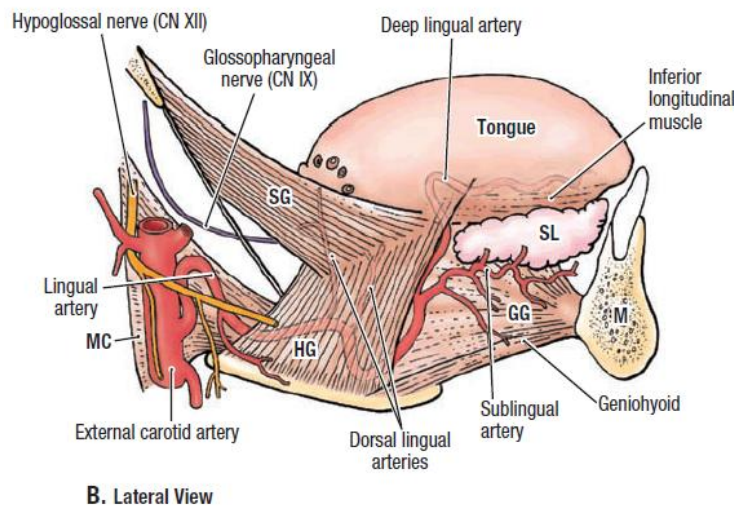


If hypoglossal nerve is affected on one side, the tongue often deviates toward the side of the lesion on protrusion (due to imbalance of genioglossus contraction).

- **Blood Supply:**

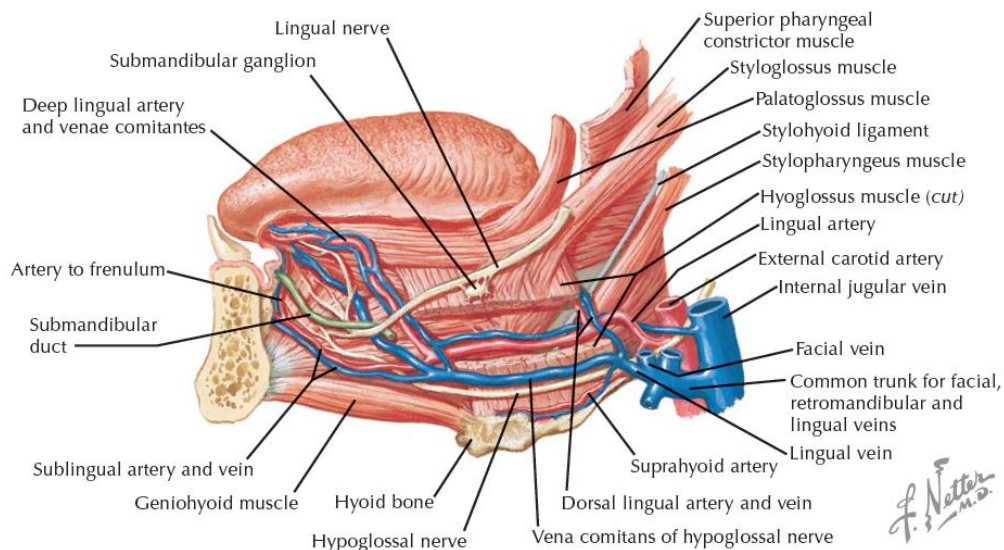
- **Lingual Artery:**

- Branches off **External carotid artery** deep to stylohyoid muscle.
- Travels superomedially then anteroinferiorly.
- Hypoglossal nerve (CN-XII) crosses over it laterally before it enters the tongue deep to Hyoglossus muscle.
- Gives rise to its 3 main branches within the tongue:
 - 1. Dorsal lingual artery:** Supplies the base of the tongue.
 - 2. Deep lingual artery:** Travels on lower surface of tongue to tip.
 - 3. Sublingual artery:** Branch to Sublingual gland and floor of mouth.

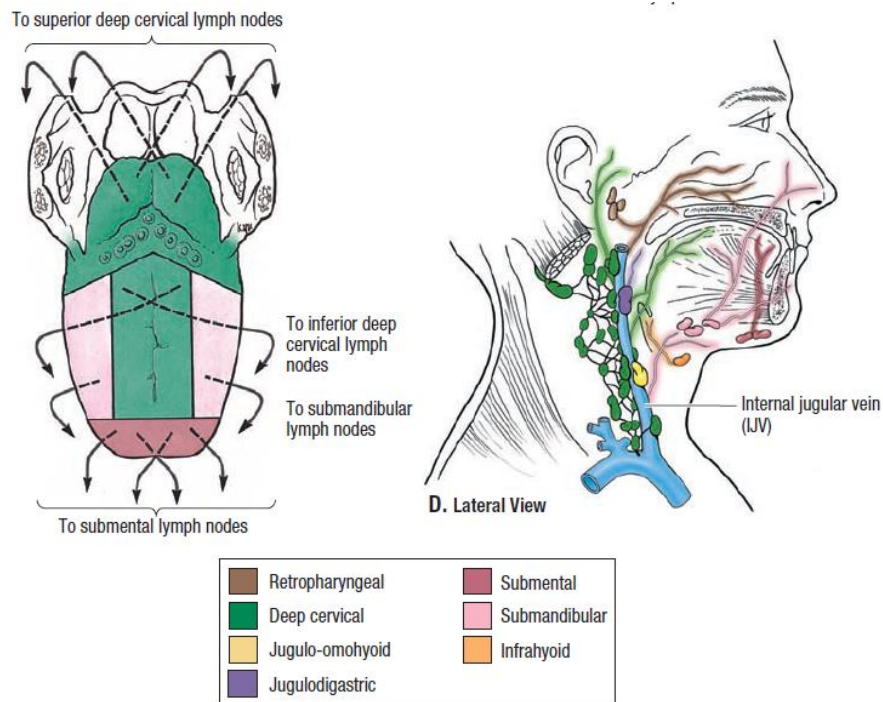


- **Venous Drainage:**

- **Deep lingual vein:** begins at tip of tongue and travels posteriorly to join **Sublingual vein** and drains into **Dorsal lingual vein**, which accompanies Lingual artery to empties into **Internal jugular vein**.



- **Lymphatic Drainage:**
- **Tongue tip:**
 - o Drains into bilateral Submental LN (**Level Ia**)
- **Medial Anterior 2/3 of tongue:**
 - o Drains into Deep cervical LN (**Level II,III,IV**)
- **Lateral Anterior 2/3 of tongue:**
 - o Drains into Submandibular LN (**Level Ib**).
- **Tongue base:**
 - o Drains into bilateral Deep cervical LN (**Level II,III,IV**)



- **Taste System Anatomy:**

- **Taste Buds:**

- Sensory end organs for gustation.
- Flask-shaped structures with a life span of 10-12 days.
- Present on:

1. Tongue:

- Anterior 2/3: Chorda tympani of Facial (**CN-VII**).
- Posterior 1/3: Lingual branch of Glossopharyngeal (**CN-IX**)

2. Palate (soft and hard Palate junction):

- Greater Petrosal nerve of Facial (**CN-VII**).

3. Epiglottis (Laryngeal surface):

- Internal branch of Superior laryngeal branch of Vagus (**CN-X**)

- **Taste receptors:**

- **Ionic channels:**

- Salt Taste.
- Acid Taste.

- **Protein receptors:**

- Sweet Taste.
- Bitter Taste.

- **Peripheral Pathway:**

- Taste is mediated by 3 cranial nerves:

1. Facial (CN-VII)

2. Glossopharyngeal (CN-IX)

3. Vagus (CN-X)

- **Central pathway**

- Taste fibers from CN-VII, IX terminate at different levels in **Nucleus solitarius** at Medulla oblongata:

- Rostral part: **CN-VII + CN-IX**
- Caudal part: **CN-X**

- From this nucleus, 2 sets of fibers project:

- To preganglionic parasympathetic neurons in Superior and inferior salivary nuclei and the dorsal vagal nuclei.

- A reflex response to taste includes an increase in salivary secretion, as well as gastric and pancreatic juice.

- **2nd-order Neurons:**

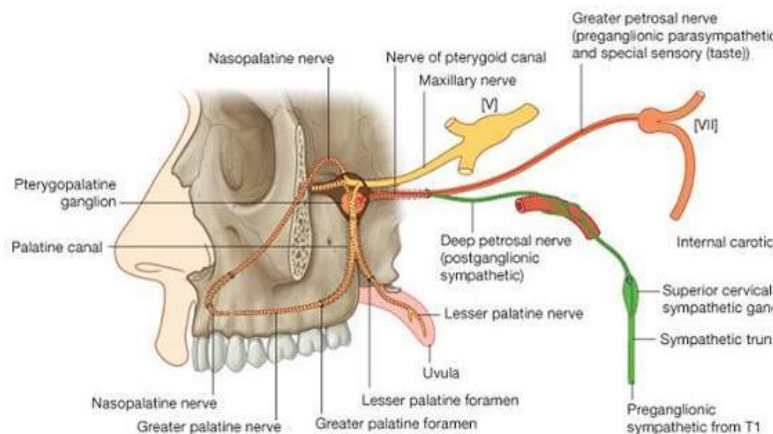
- Bulbo-thalamic pathway joins Medial Lemniscus of opposite side.

- Relay in Thalamus.

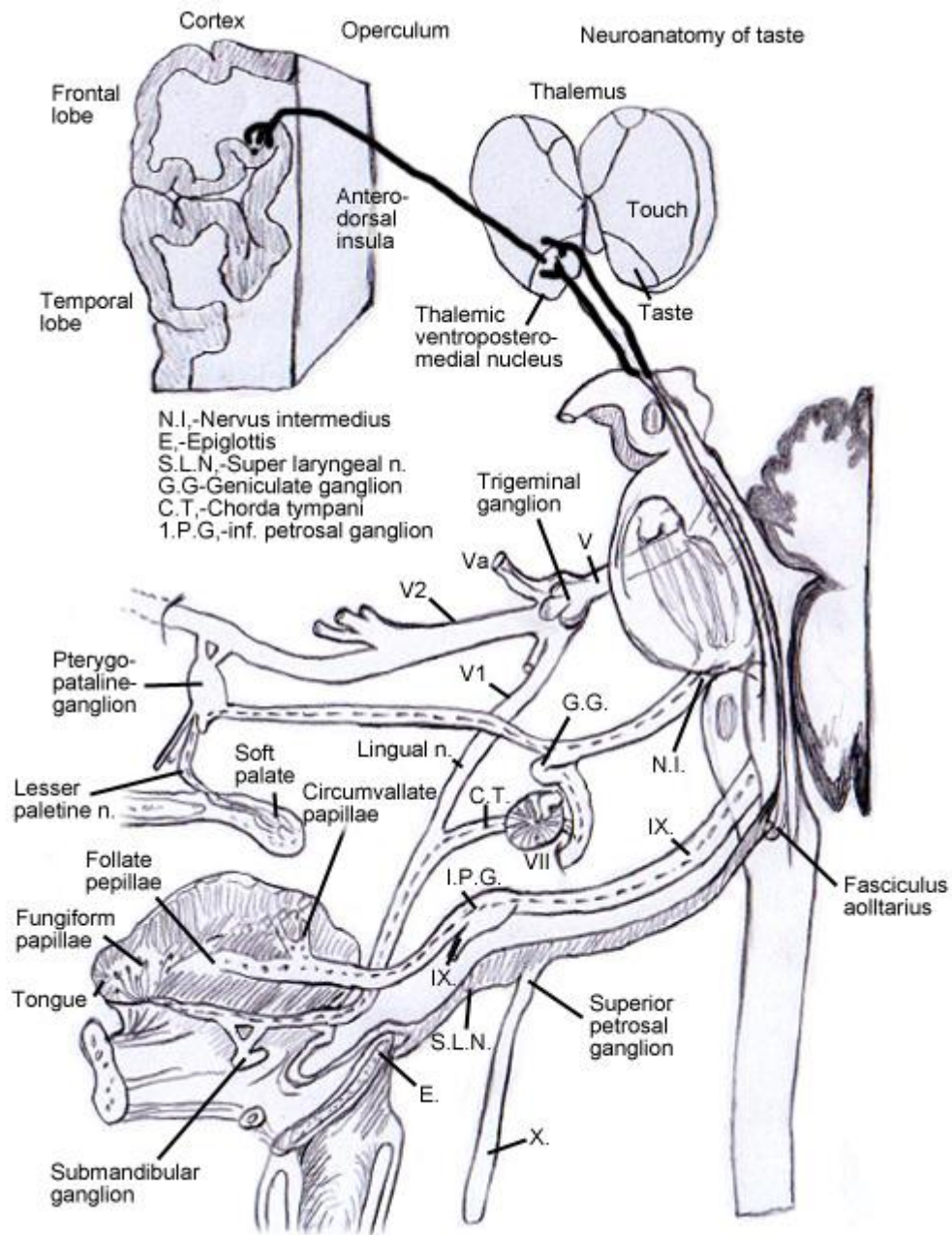
- **3rd-order Neuron:**

- Projects from Arcuate thalamic nucleus to conduct taste to the cortical taste center in **Inferior part of Parietal Cortex, Lateral fissure and Insula.**

- **Taste fibers pathway for Anterior 2/3 third of the tongue:**
 - Lingual nerve
 - → Chorda tympani nerve
 - → Facial nerve (**CN-VII**)
 - → Geniculate ganglion
 - → Nervus intermedius
 - → Nucleus Solitarius in Medulla oblongata.
- **Taste fibers pathway for Posterior 1/3 third of the tongue:**
 - Lingual branch of Glossopharyngeal nerve (**CN-IX**)
 - → Inferior Glossopharyngeal ganglion
 - → Nucleus Solitarius in Medulla oblongata.
- **Taste fibers pathway for Palate (Soft and Hard plate junction):**
 - Lesser Palatine Nerve of Maxillary nerve (**V2**).
 - → Synapse at Pterygopalatine ganglion.
 - → Nerve of Pterygoid canal (**Vidian nerve**).
 - → Greater Petrosal nerve.
 - → Geniculate ganglion
 - → Nervus intermedius
 - → Nucleus Solitarius in Medulla oblongata.



- **Taste fibers pathway for Epiglottis (Laryngeal surface):**
 - Internal laryngeal nerve.
 - → Superior laryngeal nerve.
 - → Vagus nerve (**CN-X**).
 - → Inferior Vagus ganglion
 - → Nucleus Solitarius in Medulla oblongata.



Common Disorders of Oral Cavity

❖ Ulcers of Oral Cavity

✓ The causes of the ulcers of oral cavity are

1. Infections

- (i) **Viral**: Herpangina; herpes simplex (primary and secondary); hand, foot and mouth disease
- (ii) **Bacterial**: Vincent's infection, TB, syphilis
- (iii) **Fungal**: Candidiasis

2. Immune disorders: Aphthous ulcer, Behcet's syndrome

3. Trauma

- (i) **Physical**: Cheek bite, jagged tooth, ill-fitting denture
- (ii) **Chemical**: Silver nitrate, phenol, aspirin burns
- (iii) **Thermal**: Hot food or fluid, reverse smoking

4. Neoplasms

5. Skin disorders: Erythema multiforme, lichen planus, BMMP, bullous pemphigoid, lupus erythematosus

6. Blood disorders: Leukaemia, agranulocytosis, pancytopenia, cyclic neutropenia, sickle cell anaemia

7. Drug allergy: Mouth washes, tooth paste, etc. Reactions to systemic drugs

8. Vitamin deficiencies

9. Miscellaneous: Radiation mucositis, cancer chemotherapy, diabetes mellitus, uraemia

1. Infection

✓ **Viral**

Herpangina	Herpetic gingivostomatitis (orolabial herpes)	Hand, foot and mouth disease
• coxsackie viral infection • mostly affecting children. • begin with, multiple small vesicles appear on the faucial pillars, tonsils, soft palate and uvula. • They rupture to form ulcers which are usually 2-4 mm in size, have a yellow base and red areola around them. • They seldom persist beyond one week.	• herpes simplex virus • two types: primary and secondary. • primary infection :- • affects children and is characterised by clusters of multiple small painful vesicles which soon rupture to form ulcers with erythematous base with a gray cover, heals without scar • Any part of the oral cavity may be affected.	• caused by various strains of coxsackie or enterovirus • affecting children. • Oral lesions are seen on the palate, tongue and buccal mucosa. Vesicles also develop on the skin of hands, feet and sometimes buttocks.

	• Constitutional symptoms like fever, malaise and headache may accompany sore throat and lymphadenopathy. • usually resolves in 1–2 weeks • Secondary or recurrent herpes :- • chiefly affects adults. • milder in form as adults have some immunity to this virus. • Most commonly, it involves the border of the lip (herpes labialis) but less often lesions appear intraorally on the hard palate and gingiva. • In recurrent herpes, it is presumed that virus lies dormant in the trigeminal ganglion and, when reactivated, travels along peripheral sensory nerves to involve oropharyngeal mucosa. • Precipitating factors include emotional stress, fatigue, fever, pregnancy or immune deficiency states. • Dx: history and clinical exam, viral culture in early stages, monoclonal antibodies and DNA hybridization		
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	• Treatment is mostly symptomatic • Acyclovir, helps to cut down the course of recurrent herpes labialis (oral acyclovir for primary infections or for prophylaxis (immunocompromised patient), topical acyclovir for active lesions		
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✓ **Bacterial**

1. Vincent's infection (Acute necrotising ulcerative gingivitis) (Trench Mouth)

- ✓ It is similar to **Vincent's angina**.
- ✓ Causative organisms are the same (a **fusiform bacillus** and a **spirochaete-Borrelia vincentii**).
- ✓ More often the disease affects young adults and middle-aged persons.
- ✓ Risks: malnutrition, degenerative diseases, immunocompromised, debilitating diseases
- ✓ It starts as ulcerative ("punched out") create the interdental papillae and then spreads to free margins of the gingivae which get covered with necrotic slough (gray pseudomembranous cover)
- ✓ Gingivae also become red and oedematous.
- ✓ Ass with malaise, fever, cervical adenopathy, halitosis



- ✓ Similar **ulcer** and necrotic membrane may also form over the **tonsil** (Vincent's angina).
- ✓ Diagnosis is made by clinical exam and history & **smear** from the affected area.
- ✓ Treatment:-
 - systemic antibiotics (penicillin or erythromycin and metronidazole)
 - frequent mouth washes
 - attention to dental hygiene.

2. Specific bacterial infections. Tuberculosis, syphilis and actinomycosis may present as chronic ulcers.

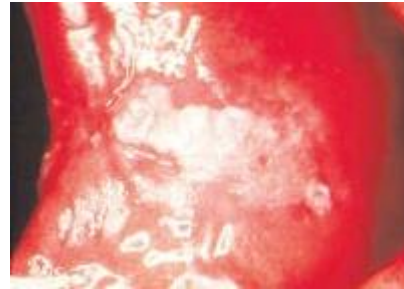
- ✓
- ✓ **Fungal** : opportunistic infection Moniliasis (candidiasis) It is caused by *Candida albicans* , and occurs in two forms: *Aspergillus* may also be cultured

Thrush	Chronic hypertrophic candidiasis
• appears as white grey patches on the oral mucosa and tongue. • When wiped off, they leave an erythematous mucosa. • Also cause odynophagia, taste disturbances • The condition is seen in infants and children. Adults are also affected when they are suffering from systemic malignancy and diabetes ,poor nutrition status or taking broad spectrum antibiotics, cytotoxic drugs, steroids or radiation.	• called candidal leukoplakia. • appears as white patch which cannot be wiped off. • Mostly affects anterior buccal mucosa just behind the angle of mouth. ✓ Hypertrophic form usually requires excisional surgery. ✓ have increased frequency of epithelial dysplasia ✓ Rx :- ○ Polyene agent (nystatin , amphotericinB)

- treated by topical application of nystatin (swish and swallow) or clotrimazole.



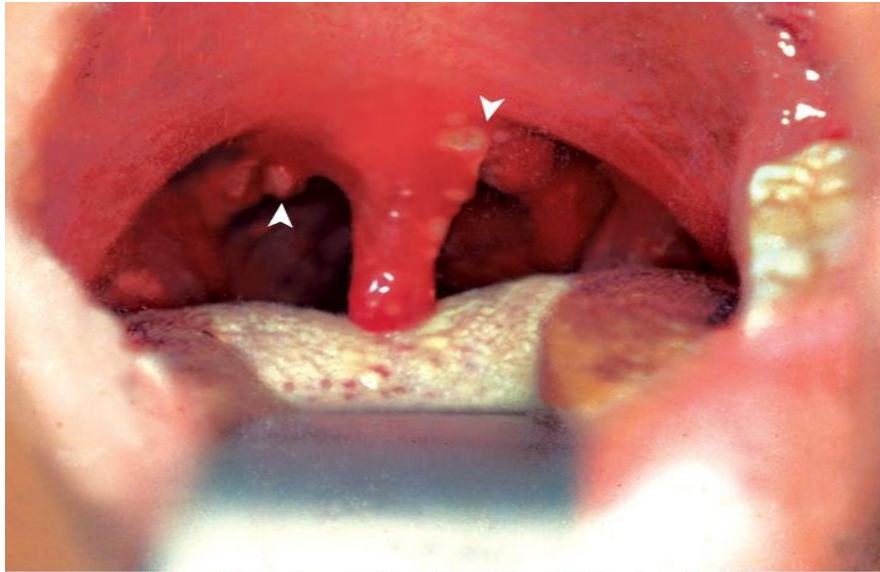
- Imidazole agent (clotrimazole and ketoconazole)
- Triazole (fluconazole, itraconazole)



2. Immune Disorders

Aphthous ulcers	Behcet's syndrome (Oculo-oro-genital syndrome)
• recurrent and superficial • most common oral ulcer • usually involving movable mucosa, i.e. inner surfaces of lips, buccal mucosa, tongue, floor of mouth and soft palate, while sparing mucosa of the hard palate and gingivae. 1. minor form, which is more common, <u>painful</u> ulcers are 2-10 mm in size and multiple with a central necrotic area and a red halo. They heal in about 2 weeks without leaving a scar.  2. major form, painful ulcer is very big, 2-4 cm in size, and heals with a scar but is soon followed by another ulcer.	• Pathophysiology: idiopathic vasculitis • It is characterised by a triad of 1. aphthous-like ulcers in the oral cavity 2. genital ulcerations 3. ocular inflammation (uveitis, iritis, papilledema, blindness) • The edge of the ulcer is characteristically punched out. • There may also be :- ○ progressive SNHL, tinnitus, vertigo ○ Lesions of the skin cutaneous vasculitis, ○ joints ○ central nervous system. • Dx: clinical symptomatology • Rx: no proven cure, may consider immunosuppressives, corticosteroids, or gamma globulin

3. Herpetiform: numerous small ulcers 1–3 mm in diameter, lasts >1 month, risk of scar formation 4. Sutton's Disease: recurrent aphthous ulcers (major type) • Aetiology is unknown It may be an autoimmune process, nutritional deficiency (vitamin B₁₂, folic acid and iron), viral or bacterial infection, food allergies or due to hormonal changes or stress. • Aphthous ulcers can be differentiated from viral ulcers by their frequent recurrence, involvement of movable mucosa as on the soft palate or cheek, and the absence of constitutional symptoms like fever, malaise and enlargement of cervical nodes. • Treatment ○ observation (self-limiting), ○ topical application of steroids ○ cauterisation with 10% silver nitrate. ○ In severe cases, 250 mg of tetracycline dissolved in 50 ml of water is given as mouth rinse and then to be swallowed, four times a day. ○ Local pain can be relieved with lignocaine viscous.	
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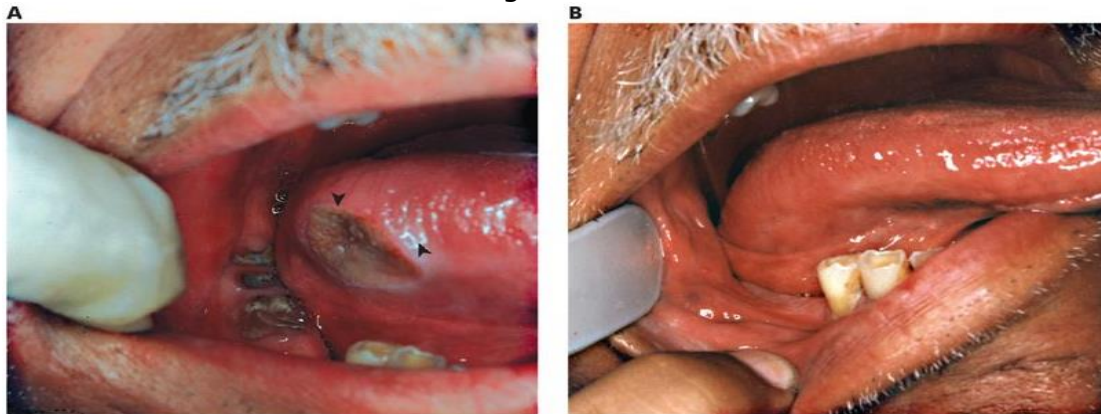


Multiple aphthous ulcers on the uvula and faucial pillars (arrowheads).

3. Trauma

✓ Traumatic ulcer

- on the lateral border of tongue may be due to jagged tooth or ill-fitting denture
- on the buccal mucosa due to cheek bite
- on the palate due to injury with a foreign object such as pencil or tooth brush
- acute ulcerative lesions of oral and oropharyngeal mucosa can result from accidental ingestion of acids or alkalies or hot fluids.



Ulcer on lateral border of tongue simulating carcinoma (arrowheads). It was caused by a sharp jagged tooth (A) and healed completely following tooth extraction (B).

- Aspirin burn is seen in the buccal sulcus when a tablet of aspirin is kept against a painful tooth to get relief from toothache.
-

4. Neoplasms

- ✓ Malignancies of the oral cavity or oropharynx may present as chronic ulcers.
- ✓ Though most commonly it is squamous cell carcinoma,
- ✓ it could be carcinoma of minor salivary glands or non-Hodgkin's lymphoma.

5. Skin Disorders

(i) Erythema multiforme

- ✓ It is a disease of rapid onset involving the **skin** and **mucous membranes**, either of which may be involved alone
 - ✓ Pathophysiology: antigen-antibody complex deposit in small vessels of the dermis and submucosa
 - ✓ The aetiology is **unknown (50%)** but may be associated with drug allergy (**sulphonamides**) or recent herpes simplex infection.
 - ✓ Oral mucosal lesions consist of **vesicles** or bullae which soon rupture to form ulcers covered with **pseudomembrane**.
 - ✓ Any area of oral mucosa is involved but the common sites are **lips, buccal mucosa** and **tongue** "lesions bleed easily"
 - ✓ The distinctive feature is to form haemorrhagic crusts on the lips (may occur without skin involvement in 25%)
 - ✓ Skin lesions consist of erythematous target lesion patches on the **palms, soles** and **extensor surfaces of the extremities**
 - ✓ May associate with fever, regional adenopathy
 - ✓ Dx: positive direct and indirect immunofluorescence (nonspecific presentation)
 - ✓ Management: - "self-limiting" mainly supportive. Steroids are used to treat the severe form
- 
- 
- ✓ **Stevens-Johnson Syndrome:-**
 - ✓ severe life-threatening form of **Erythema Multiforme**
 - ✓ SSx: widespread lesions (mouth, eyes, genitalia, respiratory tract), photophobia, blindness, fever
 - ✓ Rx: supportive care (hydration, analgesics, antipyretics), airway management, high-dose corticosteroids

(ii) Pemphigus vulgaris

- ✓ Pathophysiology: **autoantibodies** against desmosome-tonofilament complexes (**intracellular bridges**) results in **acantholysis** (**loss of cellular cohesiveness**)
- ✓ affecting older age group (50-70).
- ✓ Oral lesions are seen in 50% of the cases and may precede skin lesions.

✓ Oral lesions: -

- **painful ulcerations** are superficial and involve palate, buccal mucosa and tongue.
- **Desquamative gingivitis**,
- **Nikolsky's sign** (rubbing or trauma of uninvolved mucosa produces an ulcer)



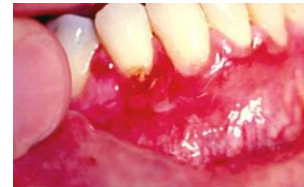
✓ Dx:

- direct immunofluorescence of **intracellular substance** (intraepithelial blistering)
- **positive** serum antibodies (indirect immunofluorescence)

✓ Treatment consists of **systemic steroids** and **cytotoxic drugs**.

(iii) **Benign mucous membrane pemphigoid (BMMP) (Cicatricial Pemphigoid ,, Ocular Pemphigus)**

- ✓ It is also an autoimmune disorder.
- ✓ Mucosal lesions involve **cheek, gingivae** and **palate**.
- ✓ **Conjunctiva** is the next important site (50–70% incidence; conjunctivitis, blindness)
- ✓ Lesion starts as a bulla filled with clear or haemorrhagic fluid which ruptures to form superficial ulceration covered with shaggy collapsed mucosa with **Nikolsky's sign**.
- ✓ Skin lesions may be absent.



✓ Dx:

- direct immunofluorescence in **basement membrane (subepithelial clefting)**
- **negative** indirect immunofluorescence (too localized)

✓ Treatment consists of **steroids**.

(iv) **Lichen planus**

- ✓ Pathophysiology: autoimmune disease in which the basal layer is destroyed by activated lymphocytes, may be familial, may be induced by medication (eg, penicillamine, methyldopa, phenothiazide, antimalarials)
- ✓ Oral lesions are seen with or without skin lesions.
- ✓ Skin lesions are
 - **pruritic, purple, polygonal papules**.
 - seen on the forearms and medial side of thigh.

✓ Oral lesions occurs in two forms:

1. **Reticular:**

- White striae forming lace-like pattern (**Wickham's striae**), are seen on the buccal mucosa on both sides.
- They are asymptomatic and require no treatment.

2. **Erosive:**

- characterised by **painful** ulceration on the buccal mucosa, gingiva or lateral tongue.
- Each ulcer is surrounded by a keratotic periphery.
- Some data shown that there is an association between erosive lichen planus and oral squamouscell carcinoma
- Biopsy is recommended
- Treatment consists of topical steroids.



3. **Plaque:** appears like leukoplakia, may manifest on the lips or palate

4. **Atrophic:** atrophy in the center of the papule

5. **Ulcerative:** may involve buccal mucosa (more common of the feet and toes), painful



6. **Annular:** more common on the lips (may also be found on the penis), ringed edges composed of small papules



✓ Dx:

- clinical exam, biopsy
- **Kobner Isomorphic Phenomenon:** lesions may be provoked by physical trauma (eg, itching, scratching)

✓ **Management**

- no cure, treat painful, erythematous, and erosive lesions
- Identify Reversible Contributing Factors: medications, dental restoration, improve oral hygiene (frequent teeth cleaning),
- Avoid tobacco, alcohol, and smoking abuse
- Medical Therapy: may consider oral or topical corticosteroids and retinoids; may also consider cryotherapy, UV light, and laser surgery

(v) **Chronic discoid lupus erythematosus**

- ✓ Oral lesions are almost always associated with skin lesions. Oral lesions are similar to those of erosive form of lichen planus.

6. Blood Disorders

- ✓ Blood dyscrasias cause ulcerations in the oral cavity and pharynx.
- ✓ Due to lack of defence mechanism, e.g. granulocytes, infections quickly supervene causing ulcers.
- ✓ **Acute leukaemia** is mainly of 2 types-acute
 - **lymphoblastic type**, which occurs in young children and acute
 - **myeloid type**, occurring in the middle-aged or the elderly.
- ✓ Both cause hypertrophy of gums with ulceration and bleeding.
- ✓ **Agranulocytosis** is characterised by ulcerations in throat with severe neutropenia.
- ✓ Cyclical neutropenia is a condition with periodic falls in neutrophil count when the person becomes prone to infections and oral ulceration.
- ✓ In pancytopenia, there is a drop in RBC count, white cell count and platelets.
- ✓ When suspected, blood dyscrasias are investigated by peripheral blood film, blood counts, and bone marrow aspiration

7. Drug Allergy

- ✓ Systemic administration of drugs like penicillin, tetracycline, sulpha drugs, barbiturates, phenytoin, etc.
- ✓ may cause erosive, vesicular or bullous lesions in the oral cavity.
- ✓ Contact stomatitis may occur due to local reaction to mouth washes, lozenges, chewing gum, tooth pastes or to prosthetic dental materials.
- ✓ Oral lesions may vary from erythema to vesicles and bullae formation.

8. Vitamin Deficiencies

9. Miscellaneous

- ✓ **Radiation mucositis.**
 - It follows radiation of oral cavity or oropharynx for cancer.
 - At first, the mucosa becomes red and then forms spotty areas of mucositis which coalesce to form large ulcerated areas covered by slough.
 - Mucositis of cancer chemotherapy can be caused by drugs like methotrexate, 5-FU and bleomycin. It manifests as erythema, oedema and ulceration.

Premalignant Oral ulcers & lesions

- ✓ **Oral Premalignancy are groups of disorders or conditions that associated with increase risk for developing oral cancer and it can be divided to :**

1. Premalignant **Lesions**
2. Premalignant **Conditions**

- ✓ **WHO Classifications PMD**

- ❖ **Group 1**

- A precancerous **lesion** (a morphologically altered tissue in which oral **cancer** is more likely to occur than its apparently normal counterpart)
- **leukoplaia**, **erythroplakia** and **palatal lesions** of reverse smokers

- ❖ **Group 2**

- A precancerous **condition** (a generalized state associated with significantly increased risk of cancer)
- **submucous fibrosis**, **lichen planus**, **epidermolysis bullosa** and **discoid lupus erythematosus**

- ✓ **Premalignant Lesions** In these lesions histopathological examination demonstrate
 - **Epithelial dysplasia** (Atypia) (**abnormality of development**)
 - Or
 - **growth abnormality**

- About **5-18%** of epithelial dysplasias become malignant
- A greater risk of malignant change in an epithelial dysplasia has been associated with the following factors:
 - (1) Erythroplakia within a leukoplakia
 - (2) A proliferative verrucous appearance
 - (3) Location at a high-risk anatomic site such as the **tongue** or **floor of mouth**
 - (4) The presence of multiple lesions, and paradoxically
 - (5) A history of not smoking cigarettes

✓ **Histopathologic Diagnosis**

- The histomorphologic changes of epithelial **dysplasia** consist of the following:

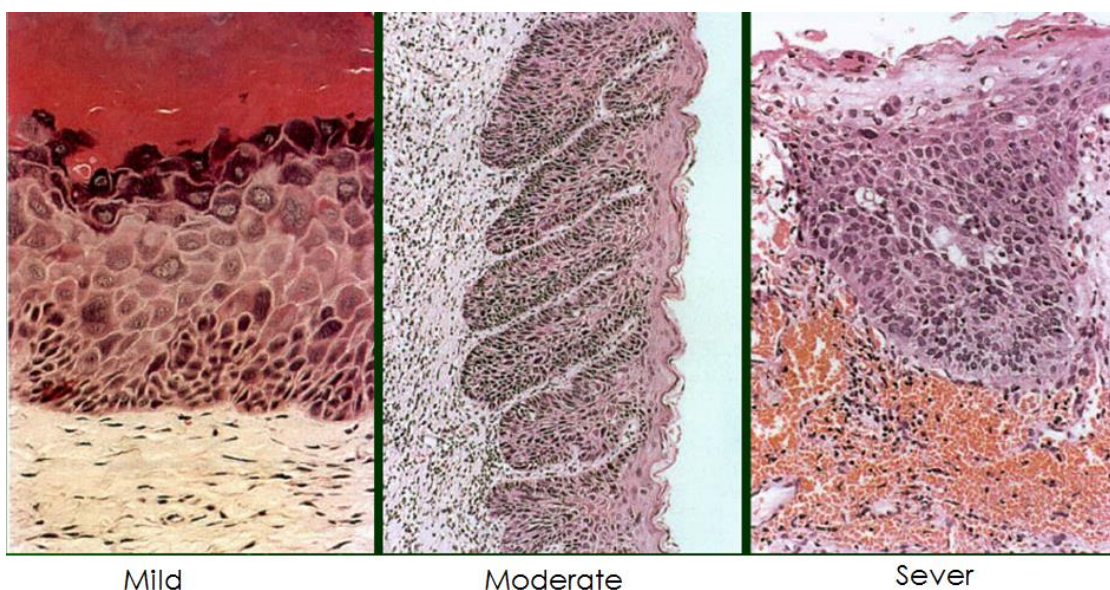
• Proliferation $\Longrightarrow$ Maturation $\Longrightarrow$ Differentiation

✓ **Dysplasia :-**

Loss of basal cell polarity
Parabasilar **hyperplasia**
Increased nuclear:cytoplasmic ratio
Drop-shaped rete ridges
Abnormal epithelial maturation
Increased mitotic activity
Mitoses in the superficial half of the surface epithelium
Cellular pleomorphism
Nuclear hyperchromaticity
Enlarged nucleoli
Loss of cellular cohesiveness
Individual cell keratinization in the spinous cell layer.

✓ **The histologic grade reflects the degree of involvement**

- ❖ **Mild** cases of epithelial dysplasia are those in which changes are seen within the **lower third** of the epithelium
- ❖ **Moderate** cases, those in which **at least half the epithelium** is involved
- ❖ **Severe** cases, those in which **most of the epithelium is affected**.
Carcinoma in situ is similar in appearance to severe epithelial dysplasia, and some authorities do not attempt to distinguish between the two.

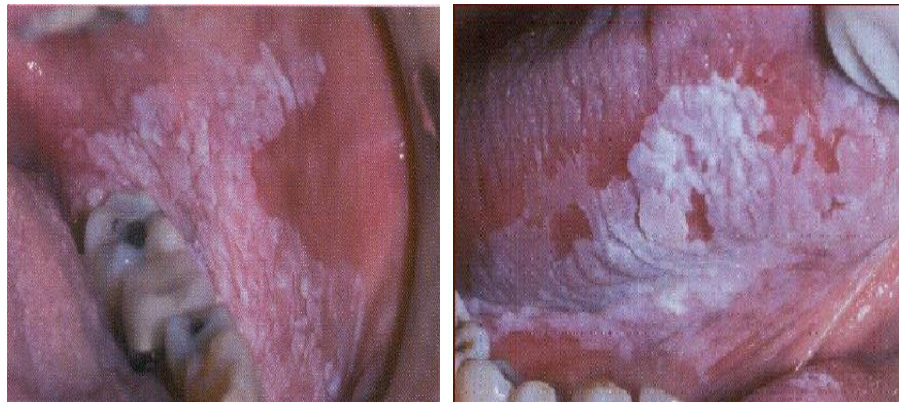


✓ **Premalignant Lesions**

- Leukoplakia
- Erythroplakia
- proliferative verrucous Leukoplakia (PVL)
- Palatal lesions of reverse smoker

❖ **Leukoplakia**

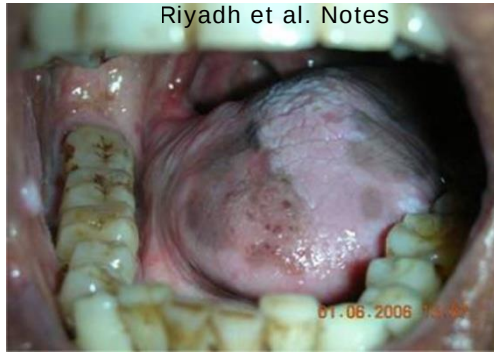
- ✓ a white patch or plaque that cannot be characterized clinically or pathologically as any other disease & will **not rub off**
- ✓ leukoplakia remains a clinical diagnosis of **exclusion** other white lesions of oral mucosa, i.e. lichen planus, discoid lupus erythematosus, white spongy nevus and candidiasis are excluded



- ✓ Leukoplakia occurs most often in middle-aged and older **men** and arises most frequently on the **buccal mucosa**, **alveolar mucosa**, and **lower lip**.
- ✓ However, note that lesions arising on the **floor of mouth**, **lateral tongue**, and **lower lip** are the most likely to harbor dysplasia or progress to malignancy.
- ✓ it is commonest premalignant lesions 85%
- ✓ The rate of progression to malignancy is low ,has been reported to be between **3.6%** and **17.5%** , On an average about **5%** become malignant. **Malignant potential** varies according to the site and type of leukoplakia, and the duration of follow up
- ✓ **25%** of leukoplakic lesions may demonstrate some degree of **dysplasia**

☒ **Types (dr. alyaa)**

- ✓ **Thin** leukoplakia
- ✓ **Thick or homogenous** leukoplakia
- ✓ **Granular or nodular** leukoplakia

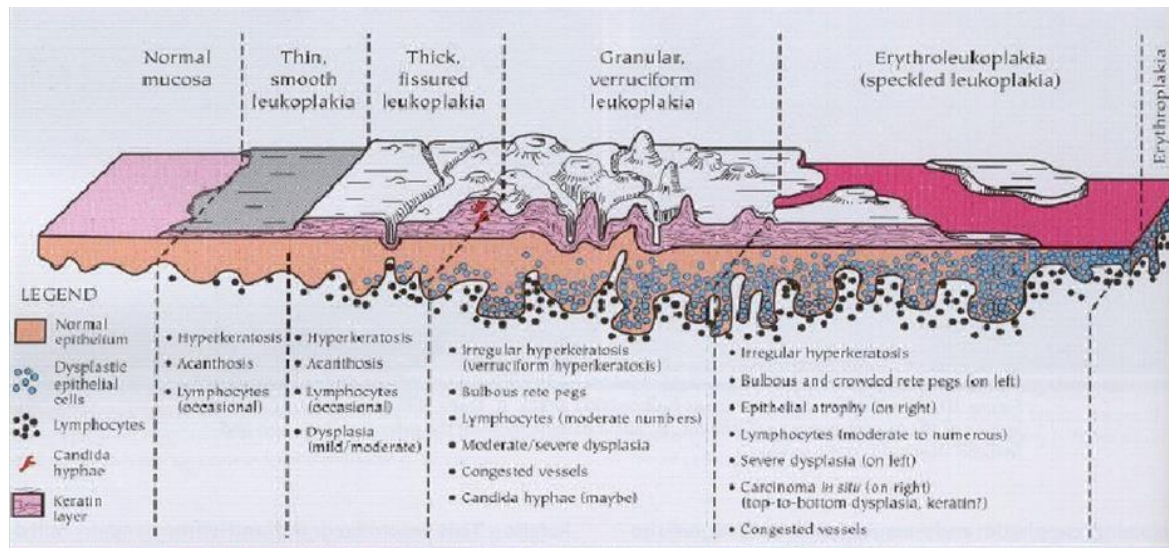


- ✓ Verruciform leukoplakia
- ✓ Proliferative verruciform leukoplakia
- ✓ Erythroleukoplakia or speckled leukoplakia

☒ Types (Dhingra)

- ✓ Homogenous variety presents with a smooth or wrinkled white patch. It is less often associated with malignancy
- ✓ Nodular (speckled) variety presents as white patches or nodules on erythematous base;
- ✓ Erosive (erythroleukoplakia) variety where leukoplakia is interspersed with erythroplakia and has erosions and fissures.

- The latter two varieties have higher incidence of malignant transformation.



☒ Types (Pasha)

- ✓ **Keratotic**: adherent, insidious development, protracted course, nonerosive surface (**higher risk of carcinoma**)
- ✓ **Nonkeratotic**: nonadherent, acute onset, erosive and ulcerative features (**higher risk of acute infections**)

☒ **Histology** About 25% of leukoplakias may show some form of epithelial

- dysplasia "mild to severe" " see above"
- hyperkeratosis
- acantosis " hyperplasia"

☒ **Etiology**

- (1) tobacco chewing
- (2) smoking "more than 80% are smoking" " Most leukoplakic patches either improve or disappear within a year after stopping smoking"
- (3) alcohol
- (4) sanguinaria 'toothpaste containing herbal
- (5) ultraviolet Lower lip vermillion
- (6) microorganism ,like treponema pallidum ,candida albicans , HPV (16, 18)
- (7) trauma " ill-fitting dentures or cheek bites"
- (8) may also be associated with
 - submucous fibrosis
 - hyperplastic candidiasis
 - Plummer-Vinson syndrome

☒ **Treatment**

- ✓ Stop the causative habit
- ✓ Biopsy is mandatory
- ✓ If dysplasia is mild 6 monthly follow up
- ✓ If moderate to severe dysplasia complete removal is recommended either by surgical excision , laser ablation , electrocautery and cryosurgery
- ✓ Follow-up is necessary; recurrences are frequent
- ✓ Chemo-preventive is experimental
 - retinoids (derivatives of vitamin A; effective; may cause side effects) are the most commonly used
 - Beta-carotene , an antioxidant that's converted to vitamin A in the body, also may completely or partially reduce leukoplakic patches
 - Vitamin E (alpha-tocopherol) may shrink the lesions associated with leukoplakia (more research is needed)

❖ **Oral hairy leukoplakia**

- benign mucosal hyperplasia results from infection with the **EBV**; normally, the virus is dormant, but if the immune system is weakened (HIV) it can become reactivated, leading to hairy leukoplakia (corrugated or hairy appearance).
- Although the use of antiretroviral drugs has reduced the number of cases, hairy leukoplakia still may affect as many as 25% of HIV-positive people, and may be one of the first signs of HIV infection

- locate in lateral tongue lesions bilaterally
- isn't painful and isn't likely to lead to cancer
- Histopathology: hyperkeratosis, acanthosis, atypia
- Confirmation can be achieved by **in situ hybridization**, Southern blot procedure, PCR, or ultrastructural demonstration of virus
- Rx: observation, may consider high-dose acyclovir



❖ **Erythroplakia**

- ✓ is a clinical term used to describe a **fiery red** patch that cannot be clinically or pathologically distinguished as any other definable disease
- ✓ the erythroplakic lesion is considered as a **diagnosis of exclusion**
- ✓ The clinical appearance of erythroplakia is described as a red macule or patch with a soft, velvety texture
- ✓ most often occurring on the **floor of mouth**, **lateral tongue**, **retromolar pad**, **gingivobuccal sulcus** and **soft palate**.
- ✓ Although far less common than leukoplakia, erythroplakia is a worrisome clinical condition that **often harbors dysplasia**.
- ✓ potential is 17 times higher than in leukoplakia
- ✓ Upon histological analysis
 - **51%** of erythroplakic lesions have been shown to demonstrate **invasive squamous cell carcinoma (SCC)**
 - **40%** demonstrating **carcinoma in situ**
 - **9%** exhibiting **mild-moderate dysplasia**

☒ **Clinical feature**

- ✓ disease of **old age**
- ✓ There is no sex predilection
- ✓ usually **asymptomatic**
- ✓ usually **irregular in outline**
- ✓ typically **not plaque** instead its a **flat or depressed** , **red color** result of the red vascular connective tissue of the submucosa shines is du to atrophied mucosal lining "decreased keratinisation" as a through

- ✓ Grossly, the lesion may be of **three varieties**:-
 - homogenous
 - speckled or granular
 - erythroplakia, interspersed with areas of leukoplakia (often indistinguishable from erythroleukoplakia, type of leukoplakia)
- ✓ Risk of malignant transformation is very high (may reach 100%) , and often already malignant on first biopsy

☒ **Treatment**

- ✓ Biopsy is mandatory
- ✓ Removal regarding to degree of dysplasia recommended for leukoplakias



❖ **Proliferative verrucous leukoplakia (PVL)**

- ✓ is a unique form of aggressive disease considered to be within the continuum of **leukoplakia** and **erythroplakia**.
- ✓ Most patients with PVL are **women**,
- ✓ many do not have a history of tobacco use.
- ✓ PVL generally appears on the oral mucosa as an irregular white patch or plaque with a **varying surface**.
- ✓ The disease is often characterized by **resistance to treatment**, **recurrence**, **multifocal proliferation**, and a **progression to carcinoma** in up to **87%** of patients.



❖ **The palatal lesion of reverse smokers**

- ✓ is unique to individuals who place the lit end of a cigarette inside the mouth.
- ✓ The resulting palatal lesion may appear clinically as a red, white, melanotic patch or papule.
- ✓ Up to 84% of palatal lesions have been demonstrated to harbor dysplasia upon histologic analysis

❖ Chronic hyperplastic candidiasis

- ✓ Chronic candidial infection cause white plaque or lesion that can not be removed by scraping
- ✓ specially it affects **oral commissures**
- ✓ the immunological condition of the patient may be affected to cause candidial infection
- ✓ Its least common form of candidasis
- ✓ have increased frequency of epithelial **dysplasia**
- ✓ Dx :-
 - Polyene agent (nystatin , amphotericinB)
 - Imidazole agent (clotrimazole and ketoconazole)
 - Triazole (fluconazole , itraconazole)



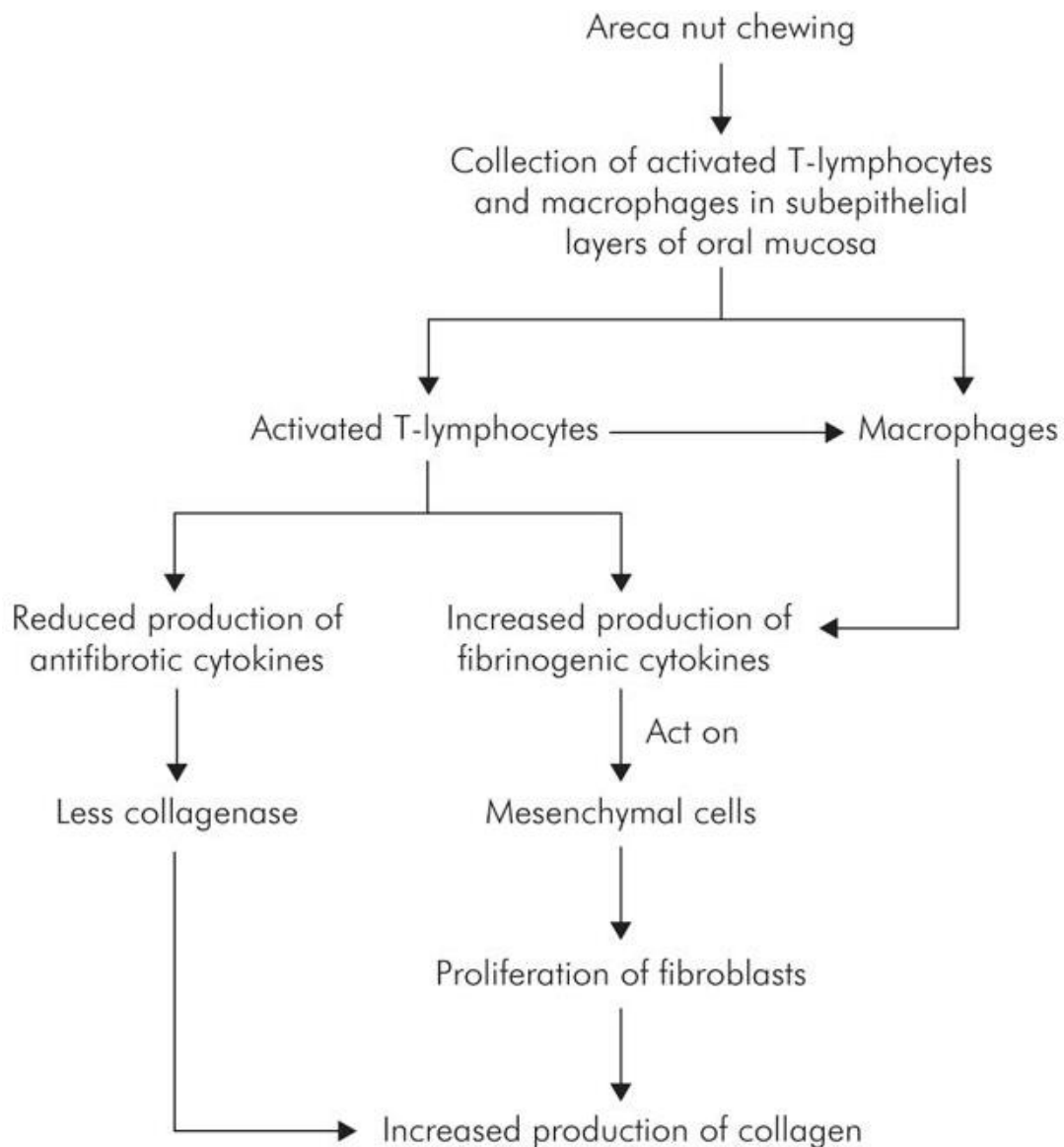
• Premalignant conditions:-

- Are generalized state or conditions associated with significantly increase risk for **cancer development**
 - OR
 - Are groups of conditions which affect epithelium of oral mucosa makes it atrophied and more susceptible to environmental carcinogens
- ✓ Premalignant Conditions
1. Oral Submucous Fibrosis
 2. Erosive Lichen Planus
 3. Discoid Lupus Erythematosus
 4. Epidermolysis Bullosa
 5. Achilachal Dysphagia
 6. Syphilitic Glossitis
 7. Actinic Cheilitis
 8. Previous Oral Malignancy
 9. Immunosuppression

❖ Oral Submucous Fibrosis (OSF)

- ✓ Female > Male
- ✓ Is a chronic ,progressive scarring In which there is a fibrous band formation beneath the epithelium and it will cause limitation of mouth opening and tongue movement
- ✓ Is a chronic progressive condition found predominantly in people of Asian decent. (India, Pakistan, Taiwan, Sri Lanka, Nepal and Thailand)
- ✓ Considered to be the result of the use of the [Areca nut](#)(betel-nut) [product](#) with resultant disruption of the extracellular matrix
- ✓ The disease often manifests with diffuse involvement of the [oral cavity](#), [pharynx](#), and [upper esophagus](#) that appears clinically as [whitish mucosa lacking elasticity](#).
- ✓ Epithelial dysplasia has been described in [7-26%](#)
- ✓ A [malignant transformation](#) rate in approximately [7% of these lesions](#).

☒ Pathogenesis



☒ **Pathology**

- ✓ The basic change is fibroelastotic transformation of connective tissues in lamina propria associated with epithelial atrophy, sometimes preceded by vesicle formation.
- ✓ In later stages, when fibrosis is marked, there is progressive trismus and difficulty to protrude the tongue.
- ✓ Leukoplakia and squamous cell carcinoma may be associated with submucous fibrosis possibly because of common aetiological factors involved.

☒ **Symptoms** :- Patient often presents with:

1. Intolerance to chillies and spicy food.
2. Soreness of mouth with constant burning sensation; worsened during meals particularly of pungent spicy type.
3. Repeated vesicular eruption on the palate and pillars.
4. Difficulty to open the mouth fully.
5. Difficulty to protrude the tongue.

☒ **Findings**

- ✓ Changes of submucous fibrosis are most marked over
- ✓ (i) soft palate
- ✓ (ii) faucial pillars
- ✓ (iii) buccal mucosa



Submucous fibrosis. (A) Note the blanched appearance of the soft palate and faucial pillars. (B) Marked trismus due to submucous fibrosis.

- ✓ In **initial stages**, there is **patchy redness** of mucous membrane with formation of **vesicles** which rupture to form superficial ulcers.
- ✓ In **later stages**, when fibrosis develops in the submucosal layers, there is blanching of mucosa with loss of suppleness. Fibrotic bands can be seen and felt in the affected areas. Fibrosis and scarring has also been demonstrated in the underlying muscle leading to further restrictive mobility of soft palate, tongue and jaw. Trismus is progressive, so much so that patient may not be able to put his finger in the mouth or brush his teeth. Oro-dental hygiene is affected badly and teeth become carious. Examination of oral cavity is difficult particularly to rule out other associated premalignant lesions or malignancy.

☒ **Treatment**

- ✓ **Not regress** by habit cessation
- ✓ In **mild type** may be treated by **intra-lesional corticosteroid injection**
- ✓ In **late stages** by **splitting or excision of fibrous bands** to improve opening
- ✓ Recent study show intralesional injection of **interferon gamma** for full improvement
- ✓ Frequent evaluation

❖ **Actinic cheilitis**

- ✓ It's a common Premalignant alteration of lower lip vermilion that results from continuous or over extended exposure to ultraviolet light of sun
- ✓ Most common in outdoor occupation farmer's lip or sailor's lip



☒ **Clinical features**

- ✓ Mostly above age of 45
- ✓ Male to female ratio is 10:1
- ✓ Very slow progression
- ✓ Atrophy ,Smooth and blotchy area then rough , dry and scaly area then chronic focal ulceration then **sq. cell carcinoma**

☒ **Treatment**

- ✓ Most of the changes are irreversible
- ✓ Use of sunscreen
- ✓ Biopsy should be submitted to rule out carcinoma
- ✓ If not carcinoma do shaving of lip vermilionectomy
- ✓ Alternatively CO2 laser ablation or electrodesiccation
- ✓ Long term follow up
- ✓ If sq cell carcinoma identified treat accordingly

❖ Erosive lichen planus

- ✓ its not as common as **reticular type** but is more significant for patient as its symptomatic
- ✓ Lesion is atrophic , erythematous with central necrosis and at the periphery there is border of fine white radiating striae or some time may beat the gingival margin produce a reaction called desquamative gingivitis
- ✓ Some data shown that there is an association between **erosive or atrophic lichen planus** and oral squamous cell carcinoma



☒ Treatment

- ✓ Biopsy is recommended
- ✓ Corticosteroid is recommended sp.Topical
- ✓ Be aware of candidacies development
- ✓ Topical retinoid and cyclosporine occasionally is advocated
- ✓ Re evaluation at 3 months intervals

❖ Sideropenic dysphagia (plummer-vinson syndrome)

- ✓ those patient suffer from iron deficiency anemia which cause atrophy of oral mucosa and difficulty in swallowing because iron is an essential requirement for growth of oral epithelium , mostly it leads to esophageal carcinoma also may leads to oral carcinoma



☒ Clinical features

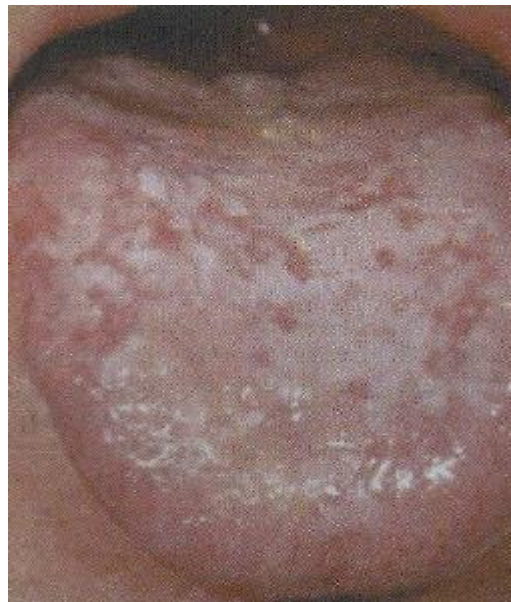
- ✓ Most of the pt. are north European background
- ✓ Mostly women 30-50 years
- ✓ They are typically complain from **burning mouth syndrome**
- ✓ **Angular cheilitis** is often present **Red , smooth tongue**
- ✓ Difficult swallowing (esophageal web)
- ✓ Spoon-shaped nail (koilonychia)
- ✓ Sign and symptom of anemia

☒ **Treatment**

- ✓ Treat anemia by dietary iron supplement
- ✓ Occasionally esophageal dilatation
- ✓ Regular follow up (5% to 50% risk of Regular carcinoma)

❖ **Syphilitic glossitis**

- ✓ In Tertiary syphilis the tongue may involve diffusely with Gumma
- ✓ appear large ,lobulated and irregular shape this lobulated form is termed diffuse **interstitial glossitis**
- ✓ loss of dorsal tongue papillae produce a condition called **Leukic Glossitis** which makes it more susceptible to environmental



carcinogens

☒ **Treatment**

- ✓ Penicillin
- ✓ Erythromycin or tetracycline for penicillin sensitive pt.
- ✓ For neurosyphilis pt. may not get total cure from such medication

☒ Factors for malignant transformation

- 1- habit like tobacco , betel nud
- 2- Clinical conditions of the patient ex.Immunity and nutrition
- 3- Female has high risk
- 4- Age of the patient
- 5- Size of the lesion
- 6- Period or duration of the disease

<i>Lesion/condition</i>	<i>Aetiology</i>	<i>Risk of malignant change^a</i>
★ Dysplastic leukoplakia	Unknown	High, but can regress
★ Erythroplasia	Idiopathic/smoking	Very high
★ Speckled leukoplakia	Idiopathic/smoking	High
★ Tertiary syphilis	<i>Treponema pallidum</i>	Very high
★ Oral submucous fibrosis	'Betel' chewing	High
★ Dyskeratosis congenita	Genetic	High
Pipe smokers' keratosis	Pipe smoking	Low and not in the keratotic area
Snuff-dippers' keratosis	Smokeless tobacco	Low
Chronic candidosis	<i>Candida albicans</i>	Low
Lichen planus	Idiopathic	Low
Discoid lupus erythematosus	Autoimmune	Unclear (mainly lip)

☒ Physical Examination & Investigation

- Extraoral
- Intraoral

☒ Work up and early detection

- Supravital staining
- Oral cytology
- Chemiluminescent light
- Tissue autofluorescence

☒ Toluidine blue (TB) is an acidophilic dye

- ✓ 1% solution is placed on the oral mucosa and removed after 1-2 minutes with 2% acetic acid.
- ✓ The clinician then examines the oral mucosa for areas of increased cellular staining
- ✓ In the evaluation of **potentially malignant oral lesions**, TB staining may provide better demarcation of lesion margins, may guide biopsy site selection, and may be valuable in the identification and visualization of lesions in high-risk patients

☒ **Oral cytology**

- ✓ A diagnostic technique used to sample oral tissue for histomorphological analysis.
- ✓ The clinician applies a stiff brush to the oral mucosa with enough pressure to induce pinpoint bleeding, which ensures a full-thickness or trans-epithelial tissue sample.
- ✓ These cellular samples can then be analyzed by a variety of unique diagnostic measures, including cytomorphometry, DNA cytometry, and immunocytochemical analysis

☒ **Chemiluminescent light based systems (ViziLite Plus, MicroLux DL)**

- ✓ use the application of a diffuse chemiluminescent light source to visualize abnormal oral mucosa not visible under normal incandescent light.
- ✓ A 1% acetic acid oral rinse is used to remove surface debris and slightly desiccate the oral mucosa before direct examination with the light source.
- ✓ Under illumination
 - normal epithelium absorbs the light (appearing light blue)
 - abnormal tissue reflects the light (appearing white, with sharper, distinct margins).
- ✓ The ViziLite system then uses a toluidine blue stain to aid in further lesion assessment.

☒ **Tissue autofluorescence**

- ✓ describes the exposure of epithelial tissue to specific wavelengths of light that results in the excitation of cellular fluorophores and emission of energy in the form of fluorescence.
- ✓ With the disruption of normal tissue morphology in dysplastic lesions, tissue fluorescence is scattered and absorbed, resulting in characteristic alterations in color that can be visually interpreted.

Oral Cavity Cancer

- 30% of all head and neck cancers (most common head and neck site).
- Oral cavity has the highest rate of second primaries (10–40%)
- >90% of occult metastatic disease in oral cancer involves nodal groups I–III
 - o Supraomohyoid dissection is oncologically sound especially in the N0 neck.
- Clinical picture:
 - o Non-healing ulcers
 - o Denture difficulties
 - o Dysphagia, odynophagia, trismus,
 - o Halitosis
 - o Numbness in the lower teeth (mandible involvement of inferior alveolar nerve).
- Risk Factors :
 - o Smoking, alcohol, and tobacco abuse
 - o Radiation and ultraviolet radiation exposure (lip cancer)
 - o Plummer-Vinson syndrome
 - o Human papilloma virus
 - o Poor oral hygiene
 - o Oral leukoplakia (5–20% malignant potential)
 - o Oral erythroplakia (25% malignant potential)

Lips Cancer:

- Most common location of oral cancer.
- Rules of 90's:
 - o 90% on lower lip.
 - o 90% squamous cell carcinoma.
 - o 90% 5-year survival if <2 cm.
- Basal cell carcinoma is more common on upper lip.
- 2–15% regional metastasis (for all stages).
- Lower lip has bilateral lymphatic drainage into level I–III nodal groups.
- Upper lip has ipsilateral lymphatic drainage into level I–III nodal groups (no contralateral drainage due to embryological fusion plates).
- Overall 5-year survival for all stages for squamous cell carcinoma is 70–90% for the lower lip and 40–60% for the upper lip.
- Poorer prognosis is associated with:
 - o Upper lip involvement
 - o Commissure involvement
- Typical patient:
 - o White
 - o Male
 - o Smoker
 - o Fair complexion
 - o 6th decade

- Prognostic factors for decreased survival:
 1. Primary > 3 cm
 2. Presence of neck metastasis
 3. Poorly or undifferentiated histology
 4. Involved surgical margins
- Causes:
 - Sunlight exposure
 - Pipe and cigarette smoking (thermal trauma?)
 - Poor dental hygiene
 - Chronic alcoholism
 - Chronic immunosuppression
- Clinical Evaluation
 - Complete history and physical examination
 - Incisional Biopsy
 - Imaging
 - Obliteration of fat in masticator space, along mandibular canal or in pterygopalatine fossa a bad sign.

TABLE 110.2 DIAGNOSIS OF LIP CANCER		
Diagnosis	Symptoms and Signs	Tests
squamous cell cancer	Nonhealing blister or recurrent crusting, lip induration, ulcer or mass for months to years, mental nerve numbness	Panorex CT/MRI Biopsy
basal cell carcinoma	Pearly nodule with central dimpling, upper lip or perioral skin	Biopsy
minor salivary gland tumor	Submucosal mass, usually upper lip	Biopsy
actinic keratoma	Rapid growth then spontaneous regression, may mimic squamous cell carcinoma in appearance	Incisional biopsy

CT, computed tomography; MRI, magnetic resonance imaging.
 From Baker SR, Krause CJ. Pedicled flaps in reconstruction of the lip. *Facial Plast Surg* 1983;1:68–69, with permission.

Oral Tongue:

- Movable portion, anterior to circumvallate papillae.
- Second most common site of oral cancer.
- 25–66% regional metastasis (for all stages).
- 60–80% 5-year survival for early disease (T1–T2)

Buccal Mucosa:

- Common site near mandibular third molar (site of chewing tobacco).
- Most common site for verrucous cancer.
- More common in India.
- 50% regional metastasis (for all stages).
- Occult neck metastasis is approximately 10%.

Alveolar Ridge:

- More common in edentulous and molar areas of the mandible.
- Must differentiate from invasive maxillary cancer.
- High rate of bony involvement.
- 50–65% overall 5year survival.

Retromolar Trigone:

- Triangle-shaped region with the base at the last mandibular molar and the apex at the maxillary tuberosity.
- Typically presents in an advanced stage.
- Bony invasion is common.
- 50% regional metastasis (for all stages).
- 25–55% 5year survival for all stages (due to advance initial staging and poor salvage potential).

Hard Palate:

- Incisive foramen allows tumor extension into anterior nose.
- Palatine foramen allows tumor extension to pterygopalatine fossa.
- Less aggressive (10–25% occult regional metastasis).
- Minor salivary gland tumors are common.

Floor of Mouth:

- Dependent site for alcohol and chewing tobacco.
- 30% present with regional metastasis.
- Overall 5-year survival is 30%–65%.

Staging and Pathological Classification Staging (based on the AJCC Staging, 2010):

- **T1:** Primary tumor <2 cm
- **T2:** Primary tumor 2–4 cm
- **T3:** Primary tumor >4 cm
- **T4:** Primary tumor invades adjacent structures,
- **T4a:** Moderate advanced local disease (invasion through cortical bone, **inferior alveolar nerve**, floor of mouth, skin, extrinsic muscles of tongue, maxillary sinus)
- **T4b:** Very advanced local disease (invasion through masticator space, pterygoid plates, skull base or encases internal carotid artery)

Pathology of Oral Cavity Cancer:

- Squamous Cell Carcinoma (SSC):
 - o >90% of oral cancers.
- Verrucous Carcinoma:
 - o Variant of SSC.
 - o Broad based, warty growth, lateral growth.
 - o Most common site is the buccal mucosa.
 - o Rare metastasis and deep invasion.
- Basal Cell Carcinoma:
 - o More common on the upper lip.
- Other types:
 - o Lymphoma, Salivary Gland Malignancies, Melanoma.

NOTE:

- Necrotizing Sialometaplasia and Granular Cell Tumors may be mistaken for squamous cell carcinoma in the oral cavity due to similar histology (pseudoepitheliomatous hyperplasia).

Management of Early Oral Cancer (T1–T2)

- Single Modality Therapy:
 - o Excision of primary tumor with primary reconstruction.
 - o May consider primary radiation (external beam versus brachytherapy).
- NO Neck:
 - o Elective ipsilateral or bilateral (if midline) selective neck dissection (supraomohyoid) versus external beam therapy for subsites with high risk of occult metastasis.
 - o Early stage hard palate or lower lip do not require elective neck dissections because of lower rate of occult metastasis.
- N+ Neck:
 - o Modified radical neck dissection for clinical nodes.
 - o Parotid nodes require a superficial parotidectomy.

Management of Advanced Oral Cancer (T3–T4)

- Double Modality Therapy:
 - o Excision of primary tumor with primary reconstruction followed by postoperative radiation therapy.
 - o Chemotherapy indicated for palliation or may be considered for adjuvant treatment for advanced disease (positive margin or extracapsular spread).
- NO Neck:
 - o Elective ipsilateral or bilateral (if midline) selective neck dissection (supraomohyoid) versus external beam therapy.
- N+ Neck:
 - o Modified radical neck dissection for clinical nodes.
 - o Parotid nodes require a superficial parotidectomy.

Surgical Management of Oral Cancer:

- Approach:
 - o Anterior and small tumors (<2 cm) may be approached intraorally.
 - o Larger and more posterior tumors require a trans-mandibular or trans-cervical approaches.
- Mandible:
 - o Invasion of the mandible (as diagnosed by imaging) requires a segmental mandibulectomy.
 - o Direct abutment of tumor against periosteum requires a mandible sparing procedure (marginal mandibulectomy).

- Reconstruction:
 - Primary closure and split-thickness skin grafts provides the best speech and swallowing outcomes.
 - Large defects require regional or distant flaps for bulk and adequate closure.

Lip Reconstruction

- Ideal reconstruction results in:
 - Sensate lip
 - Sphincter function creating water tight seal
 - Sufficient opening for food introduction, dentures and hygiene.
 - Aesthetically acceptable.
- Partial-thickness defects:
 - Usually from vermillionectomy or lip shave.
 - Reconstruction with labial buccal mucosa advancement.
 - Problems:
 - Lip thinning
 - Persistent deep red colour
 - Potential hair inversion at vermillion border
 - Alternative reconstruction:
 - Laser shave with secondary intention mucosalization
 - Ventral tongue flap possible but not often used.
- Full-thickness defects:
 - Uncommon to see vermillion only (mucosa plus orbicularis), but are often seen after outer surface recon with flap.
 - For small defect, can do a lateral vermillion advancement.
 - For larger defects, muscle/mucosa flap from ventral tongue; not a great result with bumpy lip surface
 - Better option: facial artery musculomucosal (FAMM) flap; axial flap with buccal mucosa and some buccinator.

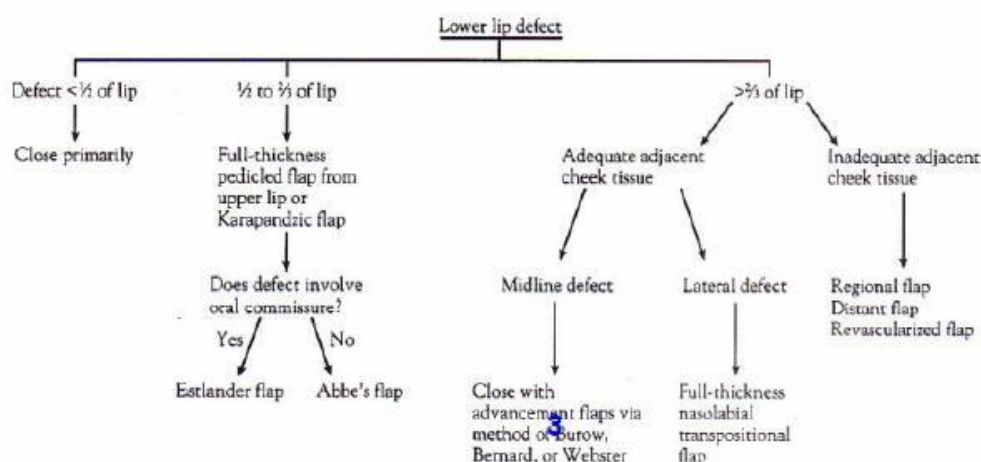


Figure 110.3 Lower lip reconstruction. (From Baker SR, Krause CJ. Pedicled flaps in reconstructive surgery. 2nd ed. Philadelphia: Elsevier; 2001.)

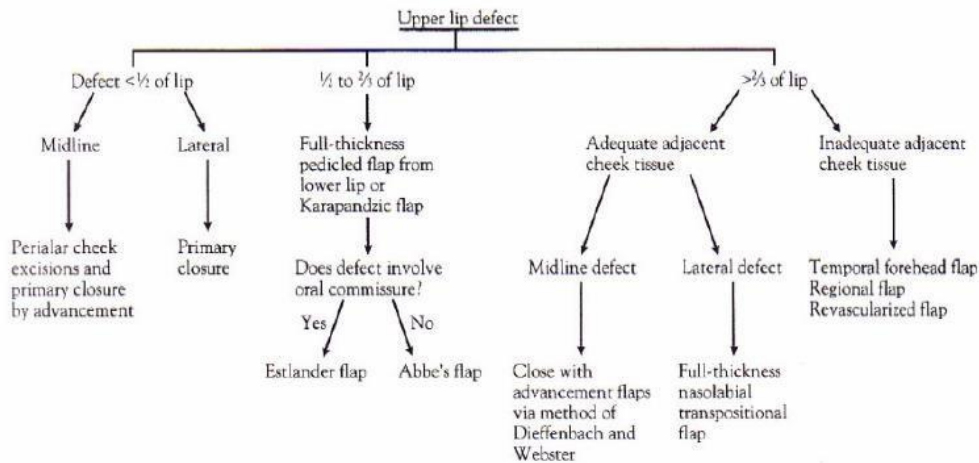


Figure 110.4 Upper lip reconstruction. (From Baker SR, Krause CJ. Pedicled flaps in reconstruction of the lip. *Facial Plast Surg* 1983;1:68–69, with permission.)

Defects of 1/2 to 2/3 of the lip:

- Majority can be closed with rotated or pedicled tissue from opposite lip.
- Three procedures are the workhorses:
 - 1. Karapandzic labioplasty**
 - 2. Abbe cross-lip flap**
 - 3. Estlander cross-lip flap**
- **Karapandzic:**
 - Circumoral advancement
 - Preserves vascularity, sensation and motor innervation.
 - Single-staged
 - Mostly lower lip, but also upper.
 - Advantages:
 - Easy to design
 - Preserves sphincter
 - Disadvantages:
 - Commissure blunting
 - Scars
 - Potential for microstomia which may preclude denture use
- **Abbe and Estlander:**
 - Full-thickness tissue transfer between opposing lips pedicled on labial artery.
 - Advantages:
 - Precise design; less microstomia; avoid circumoral incisions.
 - Disadvantages:
 - Denervation of tissue; pincushioning or trapdoor deformity; need second-stage.
 - Estlander causes ++ unilat commissure blunting.



Figure 110.6 The Karapandzic labioplasty. Circumoral skin incisions are made within the nasolabial and mental creases. The orbicularis oris muscle is bluntly dissected from supporting perioral muscles, taking care to preserve the neuromuscular pedicles, which enter from the periphery. The oral mucosa usually does not require transection.

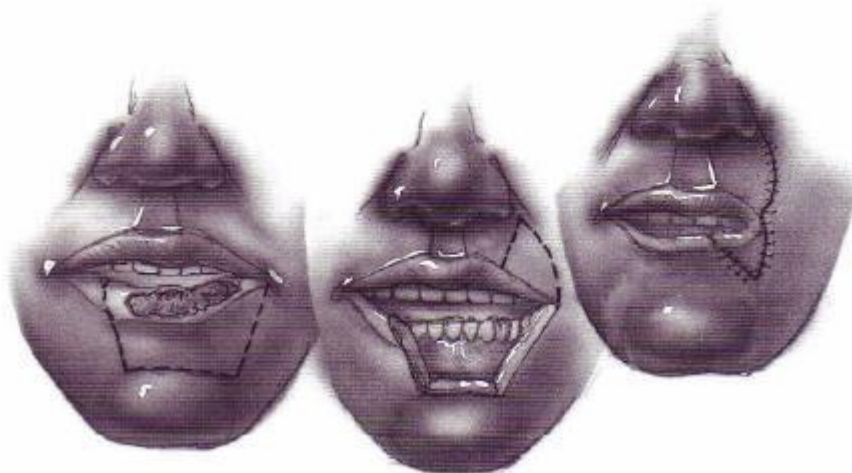


Figure 110.8 The superiorly based Estlander flap may be modified from its original description by designing the flap so that it lies within the nasolabial fold.

Defects greater than 2/3 of the lip:

- **Large central upper defects:**
 - Cross-lip plus lateral advancement.
 - Total upper:
 - Modified cheek flaps (large transverse scars across cheek).
 - Bilateral composite nasolabial flaps.
 - Free temporal scalp for hairbearing flap in men (not innervated)
- **Large lower defects:**
 - Karapandzic followed by cross lip.
 - Central lower:
 - Any modification of Bernard using full-thickness advancement of adjacent cheek tissue (need a mucosal flap to cover new tissue).
 - Total lower with adjacent defect:
 - Need distant free flap; RFFF can be folded around palmaris longus tendon for inner/outer lip lining.

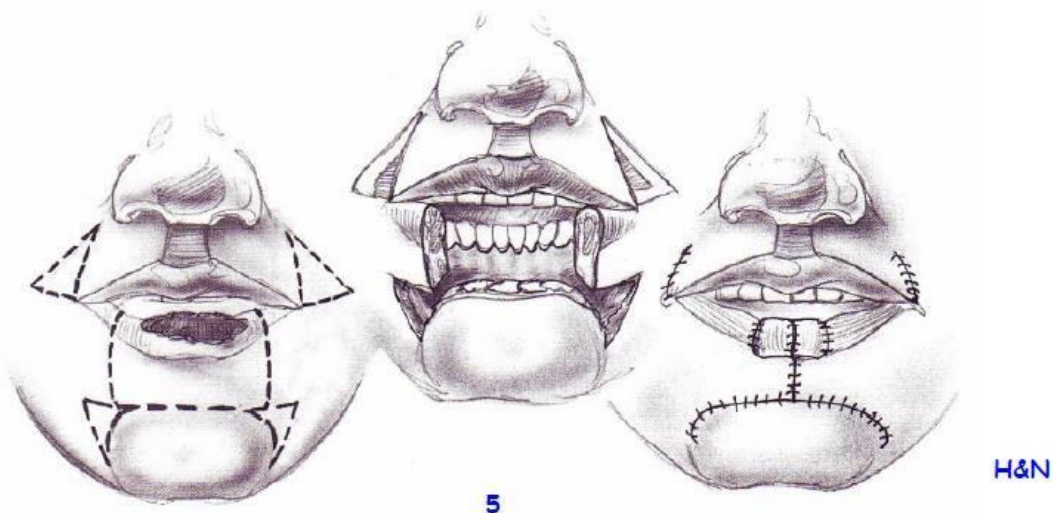


Figure 110.9 Webster modification of Bernard cheiloplasty. Cross-hatched area denotes mucosal flap used to create new vermilion (see text).

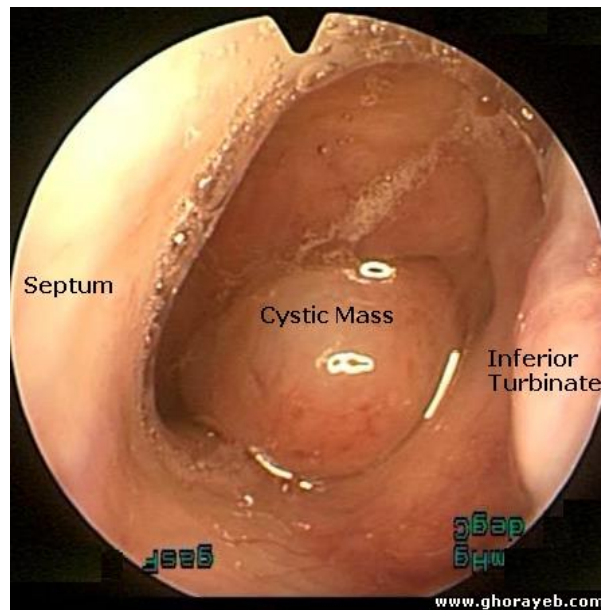
Flap	Use	Advantages	Disadvantages	Potential Complications
•Abbe	•1/2 - 2/3 defects	•sensory/motor innervation •Full-thickness lip •Restoration of orbicularis oris •No commissure violation	•Cross-lip flap •2 nd surgery •microstomia •Temporary denervation	•Vascular compromise •Vermillion notching •Lip asymmetry
•Estlander	•1/2 - 2/3 defects •oral commissure	•motor/sensory competence of lip •One stage •Scar hidden in skin crease •No mouth closure	Requires commissuroplasty	Commissure violation
•Karapandzic	•Central lower lip defects up to 80%	Preservation of neurovascular supply	•Microstomia •Inversion of vermillion •Flattened mentolabial junction	Dysesthesia/ anesthesia of lip
•Gillies fan	Defects up to 80%	Less microstomia	Full sensation may not return	Oral incompetence may result
•Bernard-Burow	Up to total lip defect	Aesthetic result	•Not for defects below labiomental crease •Adynamic reconstruction	Postoperative drooling

Nasopharyngeal Tumors:

Benign Tumors:

Thornwaldt Cyst:

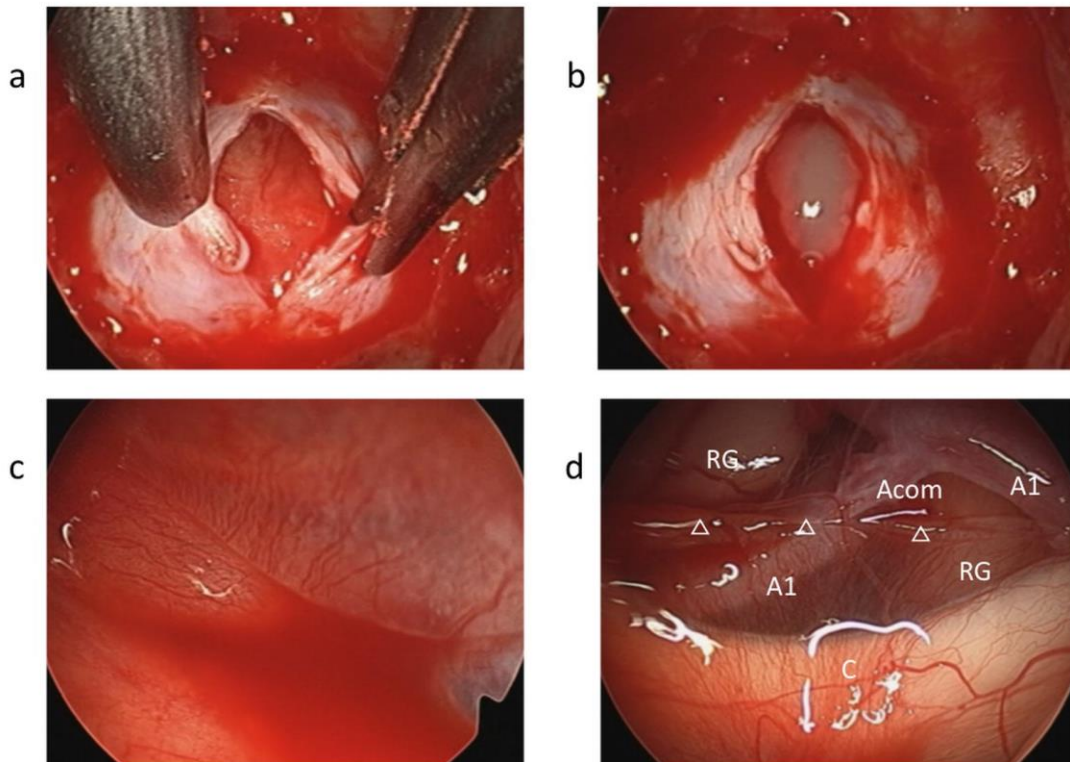
- Pathophysiology:
 - Arises from a pharyngeal **notochord remnant** (pharyngeal bursa or pouch of Luschka).
- Location:
 - **Nasopharynx** (midline, surrounded by adenoid tissue).
- Clinical picture:
 - Asymptomatic, smooth mass found in nasopharynx.
 - Rare infection.
 - Postnasal drip, referred otalgia, or otitis media with effusion
- Diagnosis:
 - Endoscopy
 - CT or MRI
 - Biopsy
- Treatment:
 - Observation
 - Antibiotics with marsupialization or excision for infected lesions.



Rathke's Pouch Cyst:

- Pathophysiology:
 - Persistent craniopharyngeal canal from failure of the obliteration of Rathke's pouch (a diverticulum of ectoderm that invaginates to form the anterior lobe of the hypophysis [pituitary] and pars intermedia).
- Location:
 - Nasopharynx
- Clinical picture:
 - Typically asymptomatic.
 - Smooth mass in nasopharynx
 - May enlarge and impinge on the pituitary
- Diagnosis:
 - Endoscopy
 - CT or MRI
 - Biopsy
- Treatment:
 - Endoscopic removal or marsupialization for symptomatic lesions,
 - add antibiotics if infected.

Supplemental Figure 1



Malignant Tumors:

Nasopharyngeal Cancer:

- Epidemiology and Geographic Distribution:
 - Multifactorial disease, Such as genetic susceptibility, environment, diet and personal habits.
 - Most common in China particularly in southern states and Taiwan.
- Etiology:
 - 1. Genetic:**
 - Chinese Even after migration, and regional distribution.
 - HLA-A2 & HLA-B17, BW46.
 - 2. Viral:**
 - Epstein-Barr virus is closely associated with nasopharyngeal cancer
 - 3. Environmental:**
 - Air pollution
 - Smoking
 - Nitrosamines from dry salted fish
 - Smoke from burning of incense and wood.
- Pathology:
 - Squamous cell carcinoma is the most common (85%).
 - Lymphomas constitute 10% .
 - The rest 5% are rhabdomyosarcoma, malignant mixed salivary tumour or malignant chordoma.
- WHO classification:
 - **Type I: Keratinizing SCC; intercellular bridges:**
 - 25% of North American NPC "nonendemic areas" and 3% of Chinese "endemic areas".
 - 5 year survival 10-35% "worst prognosis".
 - **Type II: Non-keratinizing carcinoma; +/- lymphoid stroma , little/no keratinization present; papillary morphology:**
 - 12% of North American NPC and 2% of Chinese
 - Associated with **EBV**.
 - 5 year survival 60%
 - **Type III: Undifferentiated carcinoma; +/- lymphoid stroma. clear, enlarged nuclei:**
 - 63% of North American NPC and 95% of Chinese
 - Associated with **EBV**; high local control; more mets
 - 5 year survival 60%.

	Present WHO terminology	Former terminology
Type I (25%)	Squamous cell carcinoma	Squamous cell carcinoma
Type II (12%)	Non-keratinising carcinoma	Transitional cell carcinoma
	- Without lymphoid stroma	Intermediate cell carcinoma
	- With lymphoid stroma	Lymphoepithelial carcinoma (Regaud)
Type III (63%)	Undifferentiated carcinoma	Anaplastic carcinoma
	- Without lymphoid stroma	Clear cell carcinoma
	- With lymphoid stroma	Lymphoepithelial carcinoma (Schminke)
		Spindle cell carcinoma

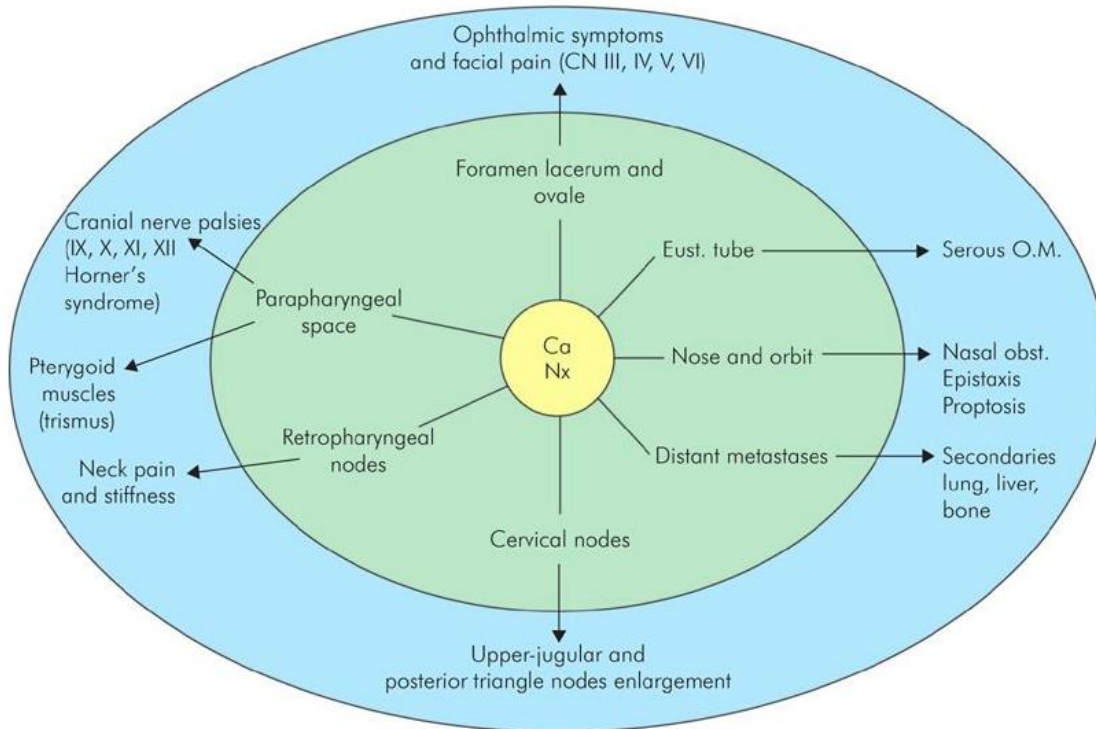
- EBV is associated with type II & III.
- The **commonest** site of origin is **fossa of Rosenmuller** in the lateral wall of nasopharynx.
- Appearance:
- Grossly, the tumor presents in three forms:
 - **Proliferative:**
 - When a polypoid tumour fills the nasopharynx, it causes obstructive nasal symptom.
 - **Ulcerative:**
 - Epistaxis is the common symptom.
 - **Infiltrative:**
 - Growths infiltrate submucosally.
- Clinical features:
 - Cervical lymphadenopathy (painless neck mass):
 - Most common presentation (60-90%).
 - Lymph node involvement is common because of rich lymphatic network in the nasopharynx.
 - Upper-most retropharyngeal nodes (Rouviere) or direct to level II LN.
 - Hearing loss (OME):
 - Second most common presentation (50%).
 - Epistaxis:
 - Third most common presentation (30%).
 - Tinnitus
 - Nasal obstruction
 - Cranial nerve palsies:
 - CN VI paralysis is the most common of CN involved.
 - Headache
 - Earache
 - Neck pain
 - Weight loss.

Trotter's Triad:

- Conductive deafness (eustachian tube blockage).
 - Ipsilateral temporoparietal neuralgia (involvement of CN V).
 - Palatal paralysis (CN X).
- At presentation, 76% have neck mass, 73% have nasal symptoms, 62% have aural symptoms and 20% have CN palsy.



- Squint and diplopia due to involvement of CN **VI**
 - Ophthalmoplegia (CN III, IV and VI) (cavernous sinus)
 - Facial pain and reduced corneal reflex may (invasion of CN **V** through foramen lacerum) occur.
 - Tumors may directly invade the orbit leading to exophthalmos and blindness (CN II at the apex of the orbit).
 - Involvement of IXth, Xth and XIth cranial nerves may occur, constituting *jugular foramen syndrome*. (Usually, this is due to pressure of enlarged lateral retropharyngeal lymph nodes on these nerves in the neck.)
 - CN XII may be involved due to extension of growth to hypoglossal canal.
 - Horner's syndrome may occur due to involvement of cervical sympathetic chain.
 - Intracranial, cavernous sinus/ infratemporal fossa extension.
- ***Distant metastases***
 - Involve bone, lung, liver and other sites. (<3%)



- **Diagnosis**

- Examination of postnasal space by a nasopharyngoscope is the most important.
- **CT** soft tissue extent; parapharyngeal extension; erosion; also shows post-treatment bone regeneration – tumour gone
- **MRI** with gadolinium enhancement reveals the intracranial extension
- PET: best for **recurrence or persistence**
- Biopsy is essential to show the exact histology of the malignancy.

- **EBV antigen:**

- Early intraellular antigen (EA) [early, 95% **specific**] (IgA)
- Viral capsule antigen (VCA) [late, 95% **sensitive**] (IgA)
- Antibody dependant cell-mediated cytotoxicity (ADCC) for other EBV antigens **TYPE 2&3**
 - Low antigens titer predict poor prognosis
 - High titer predict better prognosis
- Elevated IgA VCA &EA may be used for **screening** for nasopharyngeal Ca in high risk regions

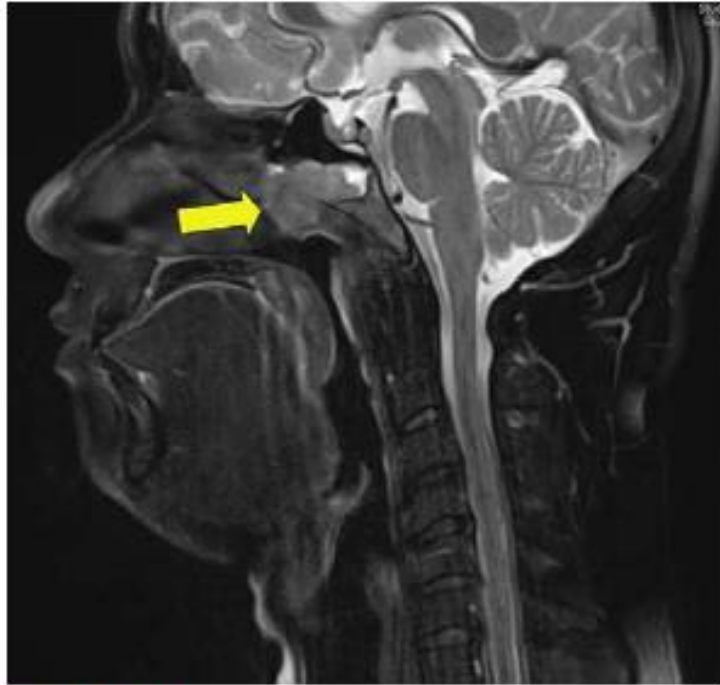


Figure 120.11 Sagittal T2-weighted MRI demonstrates NPC invading the sphenoid sinus (arrow).



Staging:

T1: Tumor confined to NP, or extend to Oropharynx and/or nasal cavity without parapharyngeal extension

T2: With parapharyngeal extension

T3: involves bony structures of skull base and/or paranasal sinuses

T4: intracranial extension and/or involvement of cranial nerves, hypopharynx, orbit, or with extension to infratemporal fossa/ masticator space

Rate of stage IV at presentation is 80%

**TABLE
120.1**

THE AMERICAN JOINT COMMITTEE ON CANCER STAGING

Primary tumor in nasopharynx (T)

T1—Tumor confined to the nasopharynx, or tumor extends to oropharynx and/or nasal cavity without parapharyngeal extension

T2—Tumor with parapharyngeal extension

T3—Tumor involves bony structures of skull base and/or paranasal sinuses

T4—Tumor with intracranial extension and/or involvement of cranial nerves, hypopharynx, orbit, or with extension to the infratemporal fossa/masticator space

Regional lymph nodes (N)

The distribution and the prognostic impact of regional lymph node spread from nasopharynx cancer, particularly of the undifferentiated type, are different from those of other head and neck mucosal cancers and justify the use of a different N classification scheme.

NX—Regional lymph nodes cannot be assessed

N0—No regional lymph node metastasis

N1—Unilateral metastasis in lymph node(s), 6 cm or less in greatest dimension, above the supraclavicular fossa, and/or unilateral or bilateral, retropharyngeal lymph nodes, 6 cm or less, in greatest dimension

N2—Bilateral metastasis in lymph node(s), 6 cm or less in greatest dimension, above the supraclavicular fossa

N3—Metastasis in a lymph node(s) >6 cm and/or to supraclavicular fossa

N3a—>6 cm in dimension

N3b—extension to the supraclavicular fossa

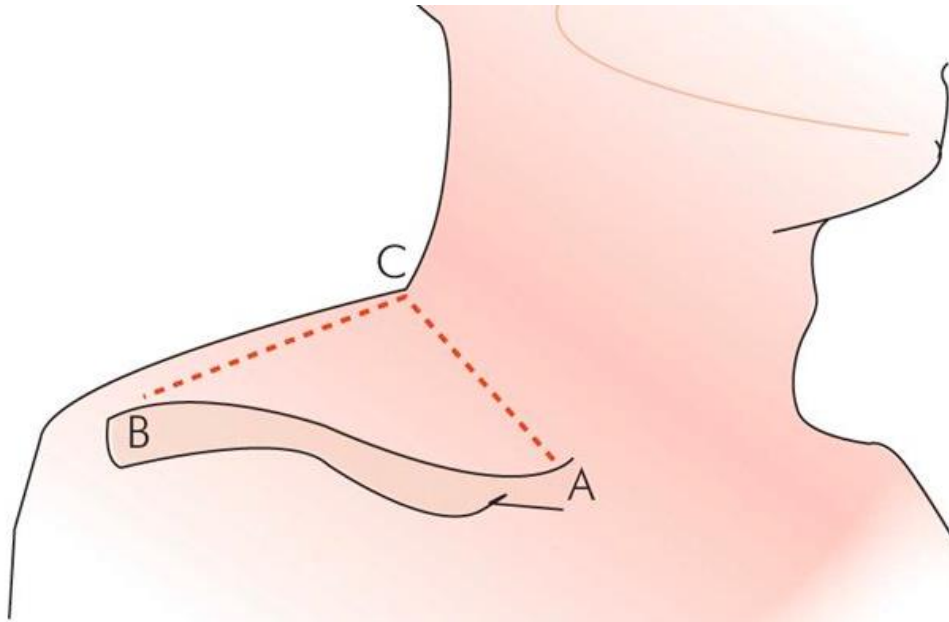
Distant metastasis (M)

M0—No distant metastasis

M1—Distant metastasis

Stage groups

Stage 0	T1s	N0	M0
Stage I	T1	N0	M0
Stage II	T1	N1	M0
	T2	N0	M0
	T2	N1	M0
Stage III	T1	N2	M0
	T2	N2	M0
	T3	N0	M0
	T3	N1	M0
	T3	N2	M0
Stage IVA	T4	N0	M0
	T4	N1	M0
	T4	N2	M0
Stage IVB	Any T	N3	M0
Stage IVC	Any T	Any N	M1



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Supraclavicular fossa (or Ho's triangle) is bounded by medial (A) and lateral (B) ends of clavicle and the point (C) where neck meets the shoulder. It includes caudal portions of levels IV and V.

Management:

- **Radiotherapy:**

- Primary modality; Radiosensitive.
- Radiation alone is effective for stage I & II.
- Multiple sensitive vital structures around NP limit total dose
- 65-75 Gy to primary; 65-70 Gy to nodes; node-negative necks get 50-60 Gy electively
- IMRT very useful to limit dose to surrounding structures

- **Chemotherapy:**

- Combined with radiation for treatment of stage III & IV
 - Cisplatin 100 mg/m² IV at weeks 1, 4 and 7
- NPC is the only entity also treated with chemo post radiation.
 - Cisplatin 80 mg/m² IV, and
 - 5-FU 1000 mg/m² IV daily for 4 days
 - Both drugs given on wks 10, 14 and 18.

- **Complications:**
 - Increased by a second course of radiation
 - Transverse myelitis “which leads to weakness and numbness of the limbs”.
 - Temporal lobe necrosis
 - Severe trismus,
 - Severe sensorineural hearing loss
 - Choanal stenosis
 - Palatal dysfunction,
 - Lower cranial nerve palsies neuroendocrine
 - Xerostomia
 - Poor oral/dental hygiene
 - Fibrosis
 - Carotid artery stenosis;
 - Memory & cognitive impairment

- **Management of Persistent or Recurrent Disease:**
- If persistent after 10/52, then worth going after.
- 5% to 10% of all newly diagnosed NPC patients will develop local recurrences.
- Persistence/recurrence in neck is harder to detect than in NP
- **Persistence or recurrence in neck nodes:**
 - The incidence of local failure was around 8.3% and they may present as residual or recurrent tumor.
 - And the incidence of isolated failure in the neck in recent years was around 1.6%.
 - Options of treatment:
 - **Additional radiation** = 20% 5-yr survival
 - **RND** = 66% 5-yr survival
 - If extending beyond node, consider **brachytherapy** to node bed.
- **Persistence or recurrence in NP:**
 - Options of Treatment:
 - **Second course of radiation:**
 - Higher dose = 32% salvage rate with 24% incidence of late sequelae and 2% incidence of death.
 - Other options (only good if focus is small and confined to NP) to minimize the effect on Quality Of Life from second course of radiation:
 - **Stereotactic radiotherapy:**
 - 86% local control rate at 3 yrs
 - **Bachytherapy:**
 - High dose to tumor; spares surroundings.
 - 87% local control for persistence
 - 63% local control for recurrence

- **Surgery (Nasopharyngectomy):**
 - Used if too big for brachy, or if extension into paranasopharyngeal space.
 - 5% to 10% of all newly diagnosed NPC patients will develop local recurrences.
 - Up to 50% of these patients will be amenable to salvage by surgery;
 - remaining 50% are usually too far advanced for surgical intervention.
 - Contraindications of surgery:
 1. internal carotid artery involvement
 2. skull base erosion,
 3. intracranial involvement
- Surgical approaches:
 - **Endoscopic and Robotic Approach**
 - **Open approaches** "One or a combination":
 - Transnasal
 - Transpalatal approach
 - Transmaxillary "Lateral Rhinotomy, Medial Maxillectomy Approach, Maxillary Swing
 - Midfacial degloving
- **Distant metastasis:**
 - Cisplatin-based combo chemotherapy (with 5-FU mostly)
 - Mostly palliative; some long-term disease-free survivors reported in literature
- **Prognosis:**
 - The poor prognostic factor in NPC:
 - Keratinizing histology
 - Length & symptomatology of disease (>2 months)
 - Extension beyond the nasopharynx
 - Low neck disease (Levels IV/V)
 - Distant metastasis
 - Stage I and II NPC patients treated by radiation alone have 5-year overall survival rates of 80% and higher.
 - Stage III or IV disease who have had concurrent CRT have a 5-year overall survival rate of about 70%.
 - The use of CRT for stage III and IV patients has seen improvements local and regional control, but distant metastasis remains the main failure leading to death in NPC patients

OTHER MALIGNANT TUMOURS OF NASOPHARYNX

They are rare and include

1. Lymphomas:

Non-Hodgkin's type is more common than Hodgkin's.
Almost all are B-cell type.

2. Rhabdomyosarcoma:

Commonly seen in Children.
Embryonal rhabdomyosarcoma presents as a polypoid mass in the nasopharynx.

3. Plasmacytoma:

It may be solitary or part of generalised multiple myelomatosis.

4. Chordoma:

From remnant of notochord.

5. Adenoid cystic carcinoma:

From minor salivary glands.

6. Melanoma (rare).

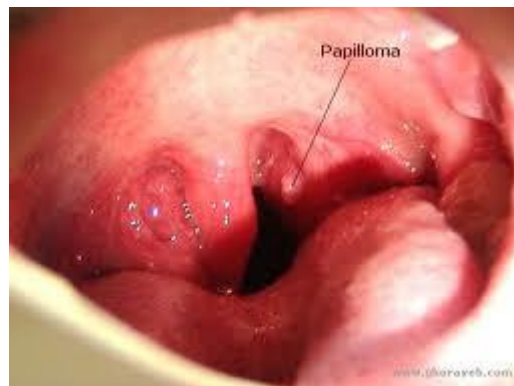
Oropharyngeal Tumors:

❖ BENIGN TUMOURS

- ✓ They are far less common compared to malignant tumors. The common ones are described here.

1. Papilloma

- ✓ It is usually pedunculated, arises from the **tonsil**, **soft palate** or **faucial pillars**.
- ✓ Often asymptomatic, it may be discovered accidentally by the patient or the physician.
- ✓ When large, it causes local irritation in the throat.
- ✓ Treatment is surgical excision.



2. Haemangioma

- ✓ It can occur on the **palate**, **tonsil**, **posterior** and **lateral pharyngeal wall**.
- ✓ It may be of capillary or cavernous type.
- ✓ Capillary haemangioma or asymptomatic cavernous haemangioma may be left alone.
- ✓ It is treated only if it is increasing in size or giving symptoms of bleeding and dysphagia.
- ✓ Treatment:
 - diathermy coagulation
 - injection of sclerosing agents.
 - Cryotherapy or laser coagulation is very effective.



3. Pleomorphic Adenoma

- ✓ common benign salivary gland neoplasm
- ✓ It is mostly seen submucosally on the **hard or soft palate**.
- ✓ It is potentially malignant and should be excised totally.



4. Mucous Cyst

- ✓ It is usually seen in the **vallecula**.
- ✓ It is yellow in appearance and may be pedunculated or sessile.
- ✓ When large, it causes foreign body sensation in the throat.
- ✓ Treatment
 - surgical excision, if pedunculated;
 - incision and drainage with removal of its cyst wall.



❖ **Lipoma, fibroma and neuroma** are other rare benign tumours.

❖ **MALIGNANT TUMOURS**

- ✓ The common sites of malignancy in the oropharynx are (Subsites in the oropharynx):

1. Posterior one-third (or base) of tongue.
2. Tonsil and tonsillar fossa.
3. Faucial palatine arch, i.e. soft palate and anterior pillar.
4. Posterior pharyngeal wall.

- ✓ Gross appearances of the tumor can be divided into 4 types:

1. Superficially spreading
2. Exophytic
3. Ulcerative
4. Infiltrative

- ✓ **Superficially & Exophytic types**

- seen in the **palatine arch**
- rarely associated with metastasis.

- ✓ **Ulcerative and infiltrative types**

- often involve the **base of tongue** and **tonsil**.
- They have poor prognosis and deeply invade the adjoining structures and have marked tendency for regional metastasis.

- ✓ Initially: grows along mucosa, submucosal and deep
- ✓ Late: vascular and thick fascial invasion; bone invasion rare (< 17%)
- ✓ Any time: perineural invasion; parapharyngeal space spread
- ✓ Lymphatic mets: common at presentation; usually start high; > 20% incidence of occult mets in N0 neck for all lesions > T1
- ✓ Distant mets: rare at presentation (2-5%); more common later

**TABLE
121.1**

THE INCIDENCE OF NODAL DISEASE AT PRESENTATION, SECOND PRIMARIES, AND DISTANT METASTASIS

Site	Palpable Nodes at Presentation (%)	Second Primary (%) ^a	Distant Metastasis ^b (%)
Base of tongue	72	19	18
Tonsillar region	58	25	15
Soft palate	45	26	7.5
Posterior wall	52	29	11

^aIncidence of synchronous and metachronous second primaries.

^bIncidence of distant metastasis with control above the clavicles.

- ✓ **Sign & symptom:** sore throat, odynophagia, dysphagia, voice quality changes, neck mass, referred otalgia, globus sensation, trismus, dysarthria, decreased tongue mobility, base of tongue mass (on palpation)

✓ **Risk Factors:**

- smoking, alcohol, and tobacco abuse
- Epstein-Barr virus (lymphoepitheliomas "arise from Waldeyer ring")
- human papilloma virus 16 (HPV) infection, especially in nonsmokers
- Other possible factors: poor oral hygiene; vitamin deficiency; poor diet; syphilis; occupational exposures; previous rads
- Immunosuppression accelerates development of Ca

**TABLE
121.2**



**DIAGNOSIS
OROPHARYNGEAL CANCER**

1. History
 - Alcohol and tobacco abuse
 - Pain and dysphagia
2. Physical
 - Nodal enlargement
 - Trismus
 - Cranial nerve deficits
3. Biopsy of the primary lesion and fine-needle aspiration of enlarged nodes
4. Imaging studies
 - PET/CT—for Stages III and IV
 - Chest radiograph—if not evaluated by CT or PET/CT
 - CT scan/MRI—to evaluate bone and deep invasion
5. Laboratory studies
 - Complete blood count and chemistry
 - Liver function tests
 - ECG
6. Examination under anesthesia
 - Look for second primaries
 - Assessment of submucosal spread, mandibular invasion, and tumor fixation

☒ **Histologically, the tumours may be:**

- **(a) Squamous cell carcinoma.**
- most common , >95% of oropharyngeal cancer
- various grades of differentiation (well, moderately or poorly differentiated)
- **(b) Lymphoepithelioma.**
- poorly differentiated variant of the SCC, with admixture of lymphocytes, which do not show any features of malignancy.
- arise from Waldeyer ring
- This is often seen in **tonsil, base of tongue** and **vallecula**.
- exophytic
- similar in behavior to the undifferentiated type of nasopharyngeal carcinoma
- **radiosensitive** (IMP)
- **(c) Adenocarcinoma.**
- It arises from minor salivary glands.
- It is mostly seen on the **palate** and **fauces**.
- **(d) Lymphomas.**
- Both Hodgkin and non-Hodgkin lymphomas arise from the **tonsil** and **base of tongue**.
- usually **non-Hodgkin** lymphoma
- 10–15% of base of tongue and tonsillar cancers
- They are seen in the young adults and sometimes in the children. Enlarged cervical nodes may co-exist.

☒ **AMERICAN JOINT COMMITTEE ON CANCER (AJCC) TUMOR STAGING BY SITE**

- **Oropharynx**
 - **T1** Tumor is 2 cm or less in greatest dimension.
 - **T2** Tumor is more than 2 cm but not more than 4 cm in greatest dimension.
 - **T3** Tumor is more than 4 cm in greatest dimension or extension to lingual surface of epiglottis
 - **T4a** Tumor invades the larynx, deep/extrinsic muscle of the tongue, **medial** pterygoid, hard palate, or mandible.
 - **T4b** Tumor invades the **lateral** pterygoid muscle, pterygoid plates, lateral nasopharynx, or skull base or encases the carotid artery.

H. TNM Staging for the Larynx, Oropharynx, Hypopharynx, Oral Cavity, Salivary Glands, and Paranasal Sinuses

Stage Grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
	T1	N1	M0
	T2	N1	M0
	T3	N1	M0
Stage IVA	T4a	N0	M0
	T4a	N1	M0
	T1	N2	M0
	T2	N2	M0
	T3	N2	M0
	T4a	N2	M0
Stage IVB	T4b	Any N	M0
	Any T	N3	M0
Stage IVC	Any T	Any N	M1

Clinical Stage Grouping by T and N Status

	<i>T1</i>	<i>T2</i>	<i>T3</i>	<i>T4a</i>	<i>T4b</i>
N0	I	II	III	IVa	IVb
N1	III	III	III	IVa	IVb
N2	IVa	IVa	IVa	IVa	IVb
N3	IVb	IVb	IVb	IVb	IVb

1. Carcinoma of Posterior One-third or Base of Tongue

- ✓ more aggressive tumor than oral tongue
- ✓ The lesion remains asymptomatic for a long time
- ✓ patient presents when metastases in cervical nodes make their appearance.
- ✓ **Earlier symptoms** "sore-throat, feeling of lump in the throat and slight discomfort on swallowing are often ignored or attributed to lingual tonsils"
- ✓ **Late features** "referred pain in the ear, dysphagia, bleeding from the mouth, and change in the quality of speech (hot potato voice).
- ✓ poorer prognosis (approximately 65% 5-year survival for all stages)



An exophytic growth at the base of tongue.

☒ Spread

- ✓ **Local**
 - deeply infiltrative and spread to the "rest of tongue musculature, epiglottis, pre-epiglottic space, tonsil and its pillars, hypopharynx"
 - **Lymphatic.** **70%** "high rate" of the cases show cervical metastases either unilateral or bilateral "20% "at the time of initial consultation. **Jugulodigastric nodes** are the first to be involved.
- ✓ **Distant metastases.** Bones, liver, lung may be involved.

☒ Diagnosis

- ✓ Lesions can be seen on indirect laryngoscopy
- ✓ palpation of the tumour should never be ignored "under anaesthesia when tissues are relaxed gives better idea of the degree of infiltration of tissues".
- ✓ Lesion is usually far more extensive than it appears on mirror examination.
- ✓ **CT scan** is recommended for tumour and nodal staging.
- ✓ **Biopsy** is essential to know its histology.

☒ Treatment

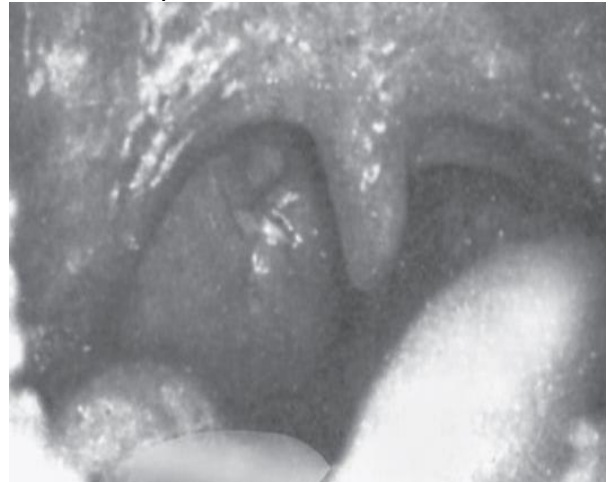
- ✓ SEE BELOW

2. Carcinoma Tonsil and Tonsillar Fossa

- ✓ most common site of oropharynx CA
- ✓ aggressive
- ✓ **Squamous cell carcinoma** is the most common and presents as an **ulcerated lesion with necrotic base**
- ✓ **Lymphomas** may present as unilateral tonsillar enlargement with or without ulceration and may simulate indolent peritonsillar abscess



Squamous cell carcinoma involving tonsil, pillar and soft palate.



☒ Spread

- ✓ **Local**
 - Tumour may spread locally to "soft palate and pillars, base of tongue, pharyngeal wall and hypopharynx".
 - may invade pterygoid muscles and mandible resulting in **pain and trismus**.
 - Parapharyngeal space may also get invaded.
 - **Lymphatic**. 50-60% of the patients have initial cervical node involvement at the time of presentation. Jugulodigastric nodes are the first to be involved "higher **lymphomas and lymphoepithelomas**"
- ✓ **Distant metastases**. They are seen in late cases.

☒ Clinical Features

- ✓ **presenting symptoms** "sore throat, difficulty in swallowing, pain in the ear or lump in the neck"
- ✓ **Later on**, bleeding from the mouth, fetor oris and trismus may occur.

☒ Diagnosis

- ✓ Palpation of tonsillar area should never be Ignored to find the extent of tumour.
- ✓ Biopsy is essential for histological typing.

☒ Treatment

- ✓ SEE below

3. Carcinoma of Faucial (Palatine) Arch

- ✓ Soft palate, uvula and anterior tonsillar pillar comprise the faucial arch.
- ✓ Rare
- ✓ since more visible than other sites typically found at early stages
- ✓ Carcinoma in these sites is often of squamous cell variety.
- ✓ Lesions are superficially spreading and well-differentiated with late tendency for nodal metastases "20–45% regional metastasis"
- ✓ Upper deep cervical and submandibular nodes may be involved.
- ✓ Behave more like carcinomas of the oral cavity.
- ✓ Spread may occur locally to the contiguous structures or lymph nodes.
- ✓ Patients with palatine arch cancer usually present with persistent sore throat, local pain or earache
- ✓ 70% 5-year survival
- ✓ Treatment is irradiation or surgical excision.



✓ An ulcerative lesion of palatine arch. It was well-differentiated squamous cell carcinoma.

4. Carcinoma of Posterior Pharyngeal Wall

- ✓ less common
- ✓ aggressive although less metastatic potential than base of tongue
- ✓ Lesions remain asymptomatic for a long time.
- ✓ They may spread submucosally to the adjoining areas such as tonsil, soft palate, tongue, nasopharynx or hypopharynx.
- ✓ They may also invade parapharyngeal space or anterior spinal ligaments.
- ✓ 60% of patients may have lymph node metastases. Bilateral nodal involvement is common
- ✓ Treatment
 - See below

☒ Management of Oropharyngeal Cancer

☒ **T 1/2 or N 0/1 stages**

- ✓ single modality "surgery alone = rads alone"
- ✓ pick one, then the other can be used for salvage
- ✓ except **base of tongue involvement; "squamous cell carcinoma "** surgical excision with block dissection is preferred and if neck dissection specimen reveals a stage **more than N₁**, post-operative radiation is added
- ✓ Patients treated nonsurgically should be evaluated using a posttreatment PET/CT to determine response at 8 to 12 weeks after completion of therapy

☒ **T 3/4 or N 2 b/c/3 staging**

- ✓ Multimodality : **excision** of primary tumor with primary reconstruction and postoperative **radiation** therapy versus primary radiation with salvage surgery

☒ **Neck**

- ✓ Neck needs treatment for all > T1
- ✓ N0/1- Neck: elective bilateral neck dissection versus external beam therapy
- ✓ N2–3 Neck: (combined modality surgery "radical neck dissection for clinically evident nodes" and postoperative chemoradiation)

☒ **Adjuvant Therapy**: postoperative radiation therapy may be considered for

- ✓ Close margin
- ✓ Advanced (T3,4)(N2,3) or aggressive disease (tongue base cancer)
- ✓ close or positive margins
- ✓ (N0,1) if multiple positive neck nodes or extracapsular extension;
- ✓ perineural or intravascular invasion
- ✓ bone, cartilage, or soft tissue invasion

☒ **Chemotherapy**

- ✓ not as much evidence for chemo as there is in **laryngeal Ca**
- ✓ induction protocols may be considered for select cases (surgical salvage for nonresponders and adjuvant radiation therapy for responders)

TABLE 121.4 TREATMENT OROPHARYNGEAL CANCER	TABLE 121.5 INDICATIONS FOR POSTOPERATIVE RADIATION (± CHEMO)
1. Team approach 2. Primary tumor treatment a. T1 and T2: single modality surgery or radiation b. T3 and T4: combined modality (chemoradiation or surgery and postoperative radiation) 3. Neck treatment a. N0 and N1: surgery or radiation b. N2 and N3: combined modality (surgery and postoperative chemoradiation or chemoradiation) c. Both sides of neck are treated with midline lesions d. Retropharyngeal nodes are always treated when cervical nodes are treated	Tumor factors 1. Close or involved resection margins 2. Perineural or vascular invasion 3. T3 4. T4 Neck factors 1. Clinically N0 or N1 neck a. Two or more histologically positive nodes b. Histologically positive nodes at multiple sites c. Perineural or vascular invasion d. Extracapsular nodal spread 2. N2 3. N3

1. Base tongue

- ✓ T₃ and T₄ SCC lesions require surgical excision with **mandibular resection**, neck dissection and post-operative radiation.
- ✓ T₄ lesions, which also extend into anterior 2/3 of tongue or vallecula, require extensive surgery with total glossectomy and laryngectomy in addition to the block dissection.
- ✓ Chemotherapy may be combined with radiotherapy and surgery T₃ and T₄.
- ✓ For advanced cancers, in patients with poor health, only palliation with radio- or chemotherapy may be required.
- ✓ They often end up into tracheostomy and gastrostomy in the terminal phase to restore their air and food passages and strong analgesics for relief of pain.

2. Tonsil and Tonsillar Fossa

- ✓ Excision of the tonsil can be done for early superficial lesions.
- ✓ Larger lesions and those which invade bone require wide surgical excision with hemimandibulectomy and neck dissection (**commando operation**).

3. Posterior Pharyngeal Wall

- ✓ irradiation or surgical excision of growth with **skin grafting**.
- ✓ Access to posterior pharyngeal wall is through lateral pharyngotomy with or without mandibular osteotomy.

• Other OP cancers

- Verrucous requires wide local excision
- Lymphoepitheliomas get rads for all stages " to the primary and neck nodes." – **very sensitive**
- Adenoid/adenosquamous and basaloid: surgery and rads
- Lymphoma: chemo and rads
- Malignant minor salivary gland tumours: wide local excision +/- post op rads
- Melanoma and sarcoma: wide local excision

☒ Surgical Management of Oropharyngeal Cancer

- ✓ typically requires an initial tracheotomy
- ✓ Unresectable if: in poststyloid space; prevertebral or carotid
- ✓ Need 1-2 cm margins; reresect positive margins if possible
- ✓ open procedures were developed during a time when surgery was the primary mode of therapy for most patients.
- ✓ These procedures have been largely obsoleted as primary therapy because of the success of CRT and minimally invasive transoral surgical approaches.
- ✓ Open approaches may still be required in in very advanced cancer with bone involvement & salvage of CRT failures

• **Approaches**

1. Transoral

- oral
- Mandibular Lingual Release/apron Flap

2. Transmandibular

- mandibular swing approach
- Mandibulectomy

3. transpharyngeal

- Lateral Pharyngotomy
- Transhyoid Pharyngotomy

○ **oral:**

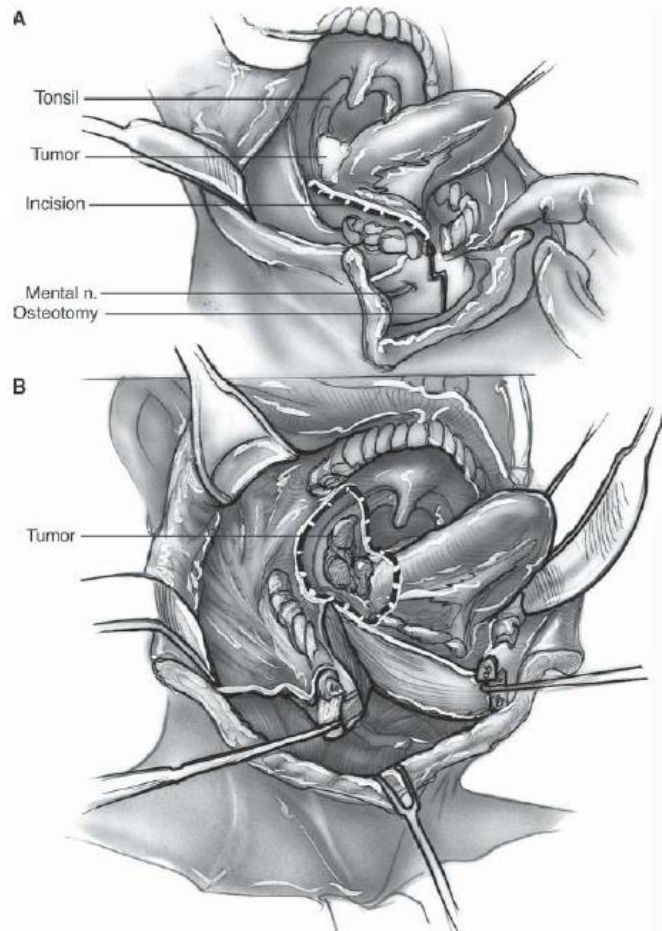
- ✓ may be used for limited tumors
- ✓ eg, posterior pharyngeal wall, anterior pillar, uvula, soft palate
- ✓ no external scar
- ✓ poor exposure

○ **Mandibular Lingual Release/Apron Flap:**

- ✓ may be considered for large tumors of the base of tongue or tonsil,
- ✓ access oropharynx from a transoral incision of the floor of the mouth, preserves mandibular integrity (not require mandibulotomy or a lower lip split)
- ✓ poor exposure lateral pharynx and parapharyngeal spaces
- ✓ The lingual arteries, lingual nerves, and hypoglossal nerve are also at risk for damage

- **Transmandibular (mandibular swing approach)**
 - ✓ spares mandible
 - ✓ may be approached laterally or midline with a lip-splitting incision, osteotomy is performed in a stepwise fashion to create a favorable repair followed by rigid fixation (recon plate)
 - ✓ provides excellent exposure to everything

Figure 121.6 Mandibular swing approach. **A:** Osteotomy is placed anterior to the mental foramen. **B:** The full thickness of the floor of the mouth is incised. The mandibular segments are then retracted laterally, allowing good access to the oropharyngeal structures and parapharyngeal space.



- ✓ less risk of malocclusion

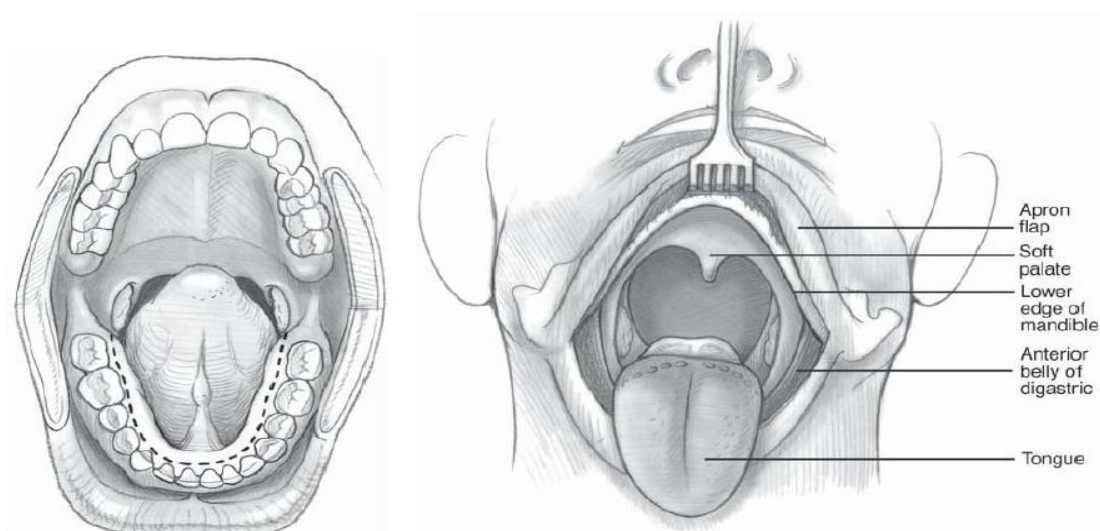


Figure 121.2 Mandibular lingual release. **A:** An incision is made through the lingual mucoperiosteum and the periosteum at the lower edge of the mandible. **B:** The anterior mandibular muscles are released with the periosteum from the inner mandibular table, delivering the tongue and floor of mouth into the neck.

- **Mandibulectomy:**
 - ✓ indicated for larger lesions, mandible extension,
 - ✓ approached laterally or medially with a lip-splitting incision
 - ✓ provides excellent exposure, easier soft tissue closure
 - ✓ risk of malocclusion and plate extrusion
 - ✓ Poor cosmetic and functional outcome, especially if not reconstructed properly

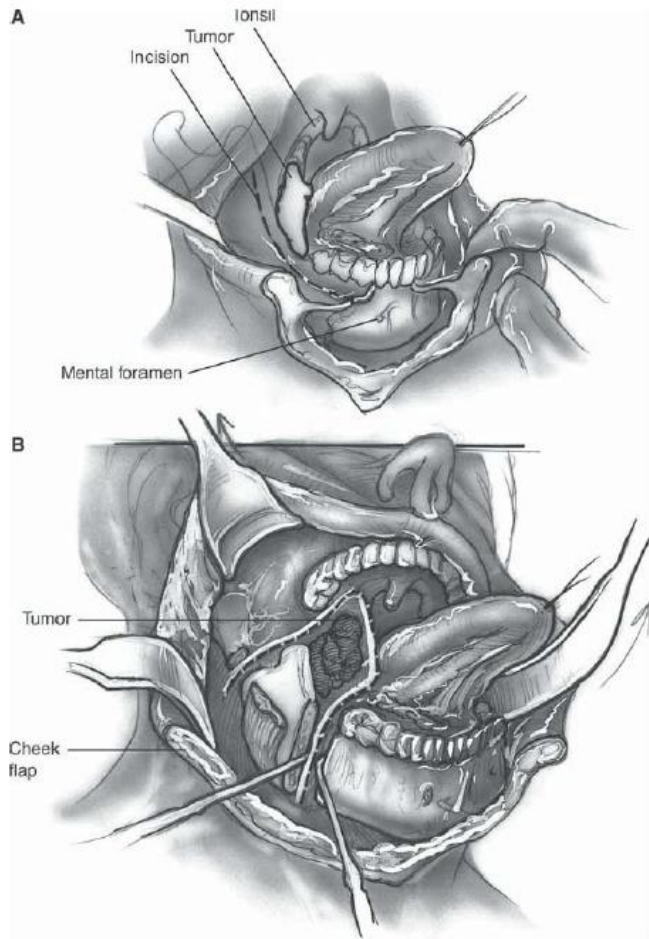
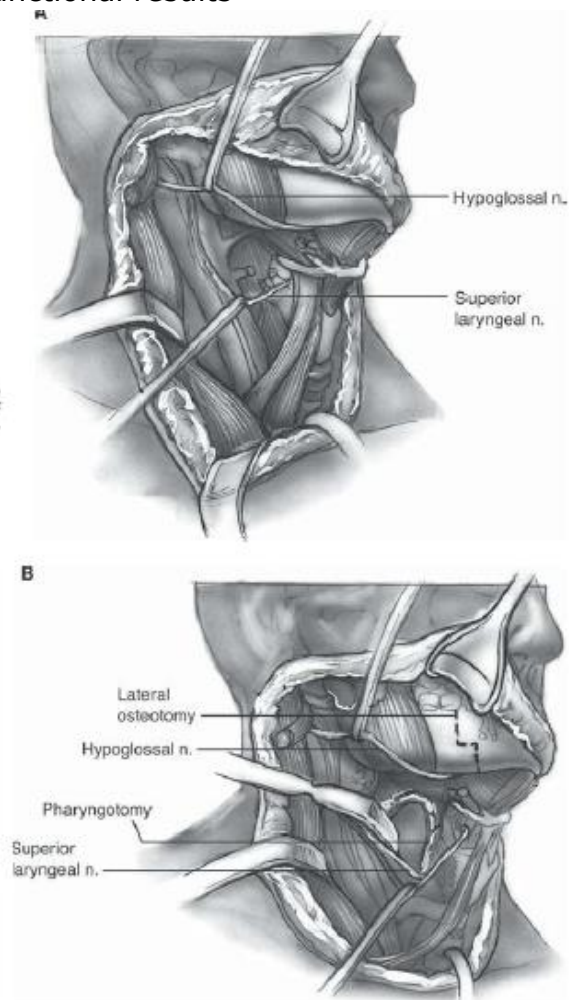


Figure 121.7 Mandibulectomy. **A:** A cheek flap is developed through a full-thickness incision of the gingivobuccal sulcus, exposing the mandible. **B:** The mandibular cuts are made well clear of the tumor and anterior to the mental foramen if the mandibular canal or inferior alveolar nerve is involved with tumor. The mandible is then retracted laterally and the soft tissue cuts are performed.

- **Lateral Pharyngotomy:**
 - ✓ may be considered for small base of tongue or posterior pharyngeal wall tumors
 - ✓ enters pharynx post to thyroid ala between hypoglossal and superior laryngeal nerves; on unaffected side
 - ✓ limited exposure
 - ✓ spares mandible
 - ✓ avoids lip-splitting incision
- **Transhyoid Pharyngotomy:**
 - ✓ may be considered for small base of tongue or posterior pharyngeal wall tumors without significant superior or tonsillar extension
 - ✓ enters pharynx above or through hyoid bone
 - ✓ spares mandible
 - ✓ avoids lip-splitting incision
 - ✓ vallecular must be free of tumor
 - ✓ poor exposure superiorly
 - ✓ Great cosmetic/functional results

Figure 121.4 Lateral pharyngotomy. **A:** Retraction of the hypoglossal and superior laryngeal nerves. **B:** Exposure of the lower oropharynx and the hypopharynx. Further superior exposure is obtained with lateral mandibulotomy.



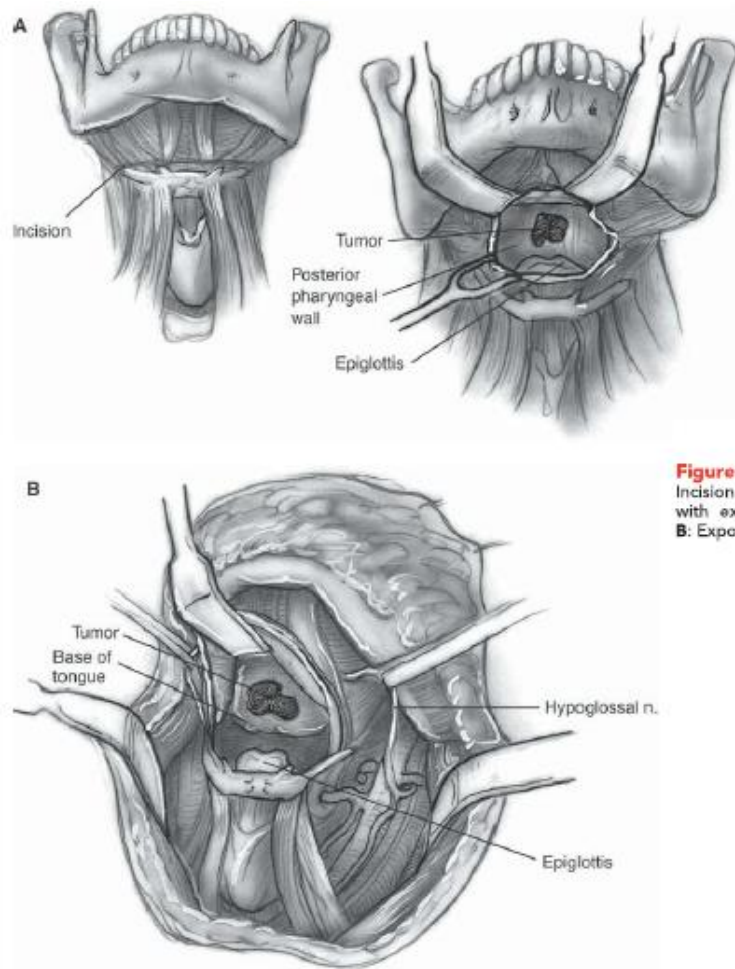
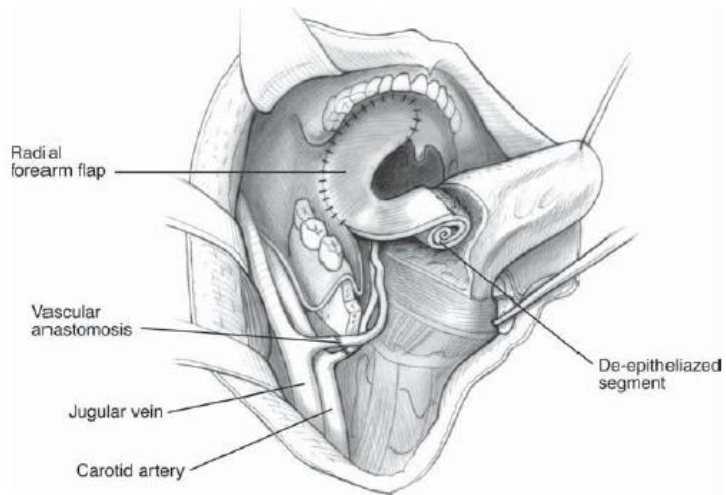


Figure 121.3 Suprahyoid pharyngotomy. **A:** Incision above the hyoid through the vallecula with exposure of the posterior pharyngeal wall. **B:** Exposure of the tongue base.

☒ Reconstruction

- ✓ **Primary Closure:**
 - simplest and best functional outcome (speech and swallowing)
 - ideal method of reconstruction if a tension-free closure
- ✓ **Split-thickness Skin Grafts:**
 - allows resurfacing with good functional outcome
 - does not provide tissue bulk
- ✓ **Pedicled Regional Flaps:**
 - provides soft tissue bulk with compromise of speech and swallowing function
 - easy; single stage; abundant tissue; limited bulk; significant rate of marginal skin necrosis; tough to tailor them to defects involving multiple sites
- ✓ **Free Tissue Transfers:**
 - provides soft tissue bulk with compromise of speech and swallowing function
 - **Fibular Free Flap** (or **Iliac Crest**) methods of choice for mandibular reconstruction of >5 cm of bone loss
 - can be reinnervated; prolonged OR time and special expertise limit their use

Figure 121.8 Reconstruction of the pharyngeal wall and tongue base with radial forearm fasciocutaneous flap. The distal flap is deepithelialized and rolled on itself to replace tongue base volume. The proximal flap is folded onto itself and sutured to the remaining nasal and oral aspects of the soft palate.



- Local flaps not used
- Lat wall defects < 3 cm, or tongue base defects < 1/3 of tongue base volume, can be closed primarily, skin grafted, or left to granulate
- Larger defects need free fasciocutaneous recon to prevent tether
- Primarily tongue base defects can be filled with myocutaneous flaps

☒ **Follow-up and Prognosis**

- Close follow-up needed for at least 5 years, and probably beyond
- 5-year actuarial survival for OP cancer:
 - Stage I: 67%
 - Stage II: 46%
 - Stage III: 30%
 - Stage IV: 30%

☒ **Complications and Emergencies**

TABLE 121.7	EMERGENCY CARE
1. Airway obstruction 2. Bleeding 3. Free tissue transfer vascular compromise	

**TABLE
121.6**



**COMPLICATIONS
OROPHARYNGEAL CANCER
TREATMENT**

1. Radiation
 - Mucositis
 - Xerostomia
 - Taste dysfunction
 - Dysphagia
 - Fibrosis
 - Ulceration and tissue necrosis
 - Osteoradionecrosis of the mandible
 - Hypoglossal palsy
2. Surgical
 - A. Approach related
 - Damage to teeth
 - Damage to nerves
 - Cerebral embolism and carotid artery thrombosis
 - B. Resection and reconstruction related
 - Hemorrhage
 - Wound infection and dehiscence
 - Positive resection margin
 - Pharyngocutaneous fistula
 - Aspiration
 - Dysphagia
 - Poor speech
 - Velopharyngeal incompetence
 - Eustachian tube dysfunction
 - Nonunion and osteomyelitis of the mandible
 - Malocclusion and TMJ dysfunction

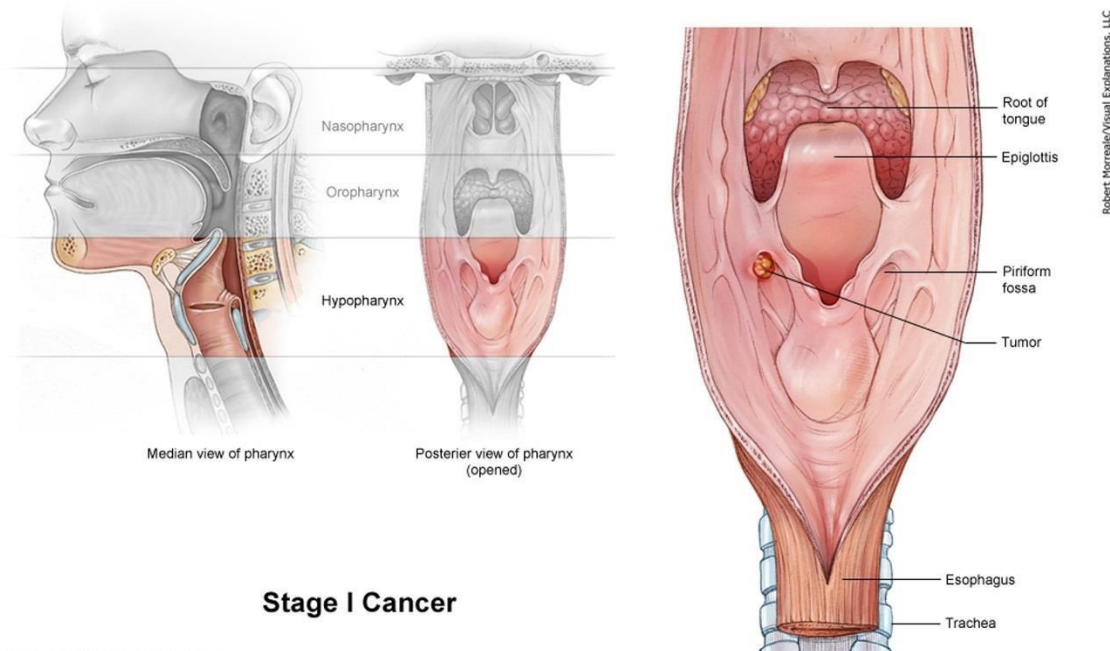


Hypopharyngeal Cancer:

- 5% to 10% of malignancies of the upper aerodigestive system
- **The pathologic features** of hypopharyngeal cancer are often unfavorable and are marked by:
 - Multicentricity o Submucosal spread
 - Early LN metastasis. unilateral or bilateral
- Patients often present with advanced disease and severe malnutrition □
The overall prognosis remains poor

Anatomy:

- Extends from hyoid to inferior border of cricoid; shaped like funnel; continuous with OP above and esophagus below
- Hypopharynx lies opposite the 3rd, 4th, 5th, 6th cervical vertebrae



OR © 2005 American Society of Clinical Oncology

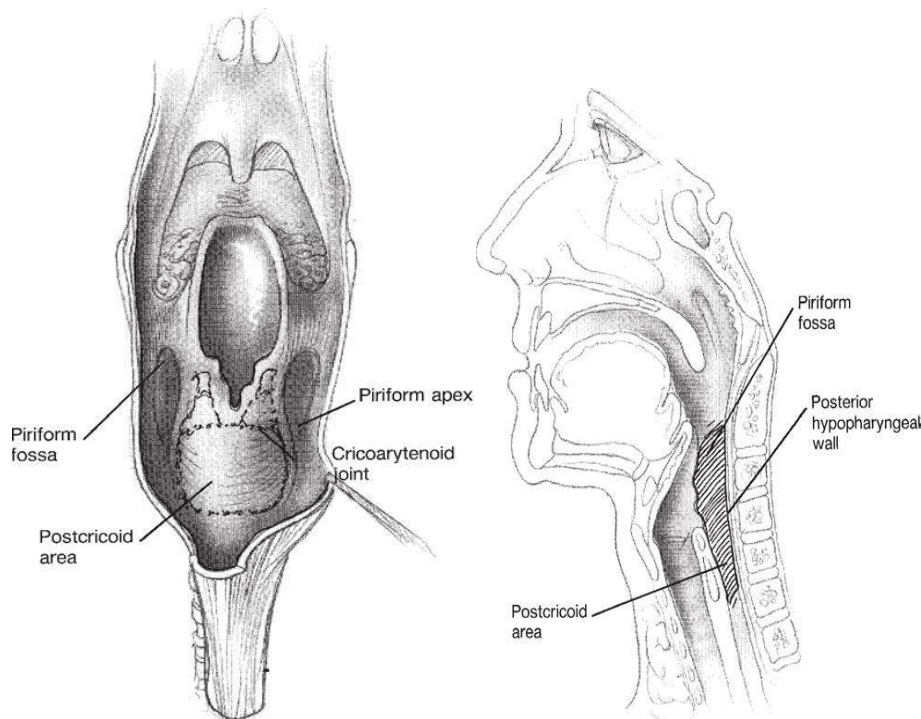
- Subdivisions:
 - **Piriform sinuses:**
 - Extend from the pharyngoepiglottic folds sup to apex at level of true cords inferiorly.
 - Bound laterally by thyroid cartilage and thyrohyoid membrane
 - Bound medially by larynx (the aryepiglottic fold, posterolateral surfaces of arytenoid and cricoid cartilages)
 - Internal laryngeal nerve runs submucosally in the lateral wall of the sinus

- **Post-cricoid region:**

- It is the part of the anterior wall of laryngopharynx between the upper and lower borders of cricoid lamina (From post surface of arytenoid mounds to inf border of cricoid)
- The proximity of the pyriform sinuses and the postcricoid area to the larynx can lead to direct invasion of tumors of these regions into the paraglottic space and laryngeal skeleton

- **Posterior pharyngeal wall:**

- The posterior pharyngeal wall is the portion of the hypopharynx overlying the vertebrae, extends from the level of hyoid bone to the level of cricoarytenoid joint.
- Tumors of this area can directly invade the potential retropharyngeal space. Paraspinous muscles, and prevertebral fascia, making complete resection extremely difficult



- Vascular supply:

- From the EXT Carotid system ○ branches of **sup thyroid**, **lingual** and **asc. Pharyngeal**
- Venous drainage mirrors the arterial system in addition to the prevertebral venous plexus.

- Sensation:

- **IX and X** via pharyngeal plexus and internal branch of SLN, the vagus nerve also gives rise to the **Arnold nerve**, which provides sensory innervation to the EAC and accounts for referred otalgia caused by hypopharyngeal lesions.

- Motor:
 - inf constrictor via pharyngeal plexus;
 - Post. CricoArytenoid via RLN
- Lymphatics:
 - **Ant path:** drains larynx and piriforms through thyrohyoid membrane to levels II-IV;
 - **Post path:** drains post wall to Retropharyngeal nodes and levels II-III through inf constrictor.
Inferior pyriform sinuses also drain through the cricothyroid membrane along the RLN into paratracheal nodes
 - **Bilateral lymphatic mets are common, especially for lesions on the medial wall of the pyriform sinuses and the posterior pharyngeal wall.**

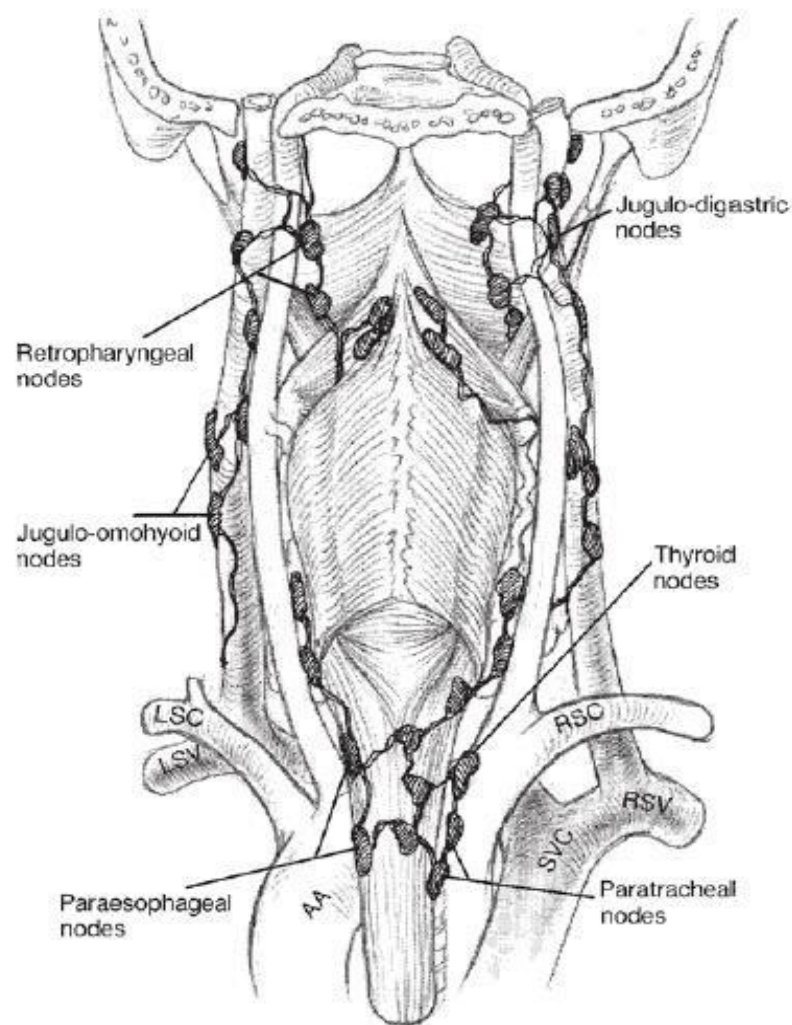


Figure 122.3 The primary lymph node drainage of the hypopharynx. Cancers of the superior hypopharynx metastasize to the jugulodigastric and retropharyngeal nodes, whereas those of the inferior hypopharynx, including the piriform apex, metastasize to the jugulo-omohyoid, paraesophageal, paratracheal, and thyroid lymph nodes. AA, aortic arch; LSC, left subclavian artery; LSV, left

History and Physical Exam

- Common presenting symptoms: odynophagia; dysphagia; otalgia
- Others: throat clearing; globus; dyspnea and hoarseness (late; direct invasion or RLN)
- **History of tobacco and ethanol abuse** □ Comorbidities: pulmonary or cardiac
- General status; wt; stridor; voice
- Thorough H&N exam:
 - OC: dentition; leukoplakia; 2nd primary
 - Laryngoscopy: look for edema and erythema
- Palpation of neck:
 - LN; loss of laryngeal crepitus may = postcricoid tumor
- Cranial nerve exam:
 - Vocal cord paralysis can be caused by the tumor invasion of the paraglottic space, cricoarytenoid joint, or the RLN
- Flexible direct laryngoscopy with Valsalva for piriforms
- **Most common symptoms in patients presenting with hypopharyngeal carcinoma were:**
 - Dysphagia (53%) Hoarseness (39%) Neck mass (37%) Weight loss (36%) Sore throat (34%) Otalgia (30%).

Radiology

- CT usually adequate for soft tissue; MR can help if CT not enough o The accuracy of CT for detection of laryngeal cartilage invasion varies from reported sensitivity of 46% to 91% and specificity of 80% to 91%
- MRI offers comparable rates of detection of laryngeal cartilage invasion with the sensitivity ranging from 89% to 94% and the specificity ranging from 74% to 88%.
- CXR for mets in early disease; CT chest if multiple neck mets o The risk of distant metastases, however, increases with higher nodal stage. **Patients with 3 or more LN mets have ~ 50% risk of developing distant mets.**
- The accuracy of FDG-PET in detecting mets in the lungs appears to be comparable to that of chest CT
- Ba swallow useful for second primary in esophagus if not doing endo

TABLE 122.1	DIAGNOSIS HYPOPHARYNGEAL CANCER
History	
Treated for pharyngitis	
Tobacco and ethanol abuse	
Weight loss	
Dehydration	
Odynophagia	
Dysphagia	
Hoarseness	
Dyspnea	
Otalgia	
Neck mass	
Airway obstruction	
Support group status	
Physical	
Weight loss or cachexia	
Decreased skin turgor	
Hyporesonant voice	
Hoarseness	
Stridor	
Status of dentition	
Indirect examination with mirror	
Flexible fiberoptic laryngoscopy	
Full neck examination	
Radiology	
Chest radiograph	
Barium swallow	
CT or MRI of the neck	
Laboratory	
Complete chemistries	
Staging endoscopy and biopsy	
Control of airway	
Complete paralysis	
Gross characteristics of tumor	
Three-dimensional relationships of tumor	
Biopsy	
Determination of inferior border	
Esophagoscopy	
Bronchoscopy, if warranted	

CT, computed tomography; MRI, magnetic resonance imaging.

Pathology

- SCC > 95% of lesions; smoking & alcohol strongly associated; **reflux**, other pathologies would be adenoma and salivary gland malignancies:
- **65-75%** are in piriforms; o 20-30% are on post wall
- 1%-5% are postcricoid area
- **M>F**; 6th-7th decade
- **Plummer-Vinson pts**:
 - (postcricoid dysphagia, upper esophageal webs, and iron deficiency anemia) are mostly **women** and can get alcohol/tobacco independent postcricoid lesions o The cause of PVS is unclear. Proposed etiopathogenic mechanisms include iron and nutritional deficiencies, genetic predisposition, and autoimmunity
 - chronic irritation of the webs is thought to be the causative factor in the progression to carcinoma

- Manifestations of iron deficiency (with or without anemia) may be evident, including the following:
 - Angular cheilitis
 - Glossitis
 - Koilonychia (spoon nails)
 - Pallor
- **Patterns of spread (distinguished features):**
 - **Submucosal spread** (due to rich sub mucus lymphatic network) up to 5-10 mm (**requires a wide surgical margin, especially inferiorly**; skip lesions common)
 - Its recommended to take wide margins **3 cm IN THE INFERIOR HYPOPHARYNX AND 2CM LATERALY and 1.5 cm superiorly.**
- **Lymph node metastasis:**
- In hypopharyngeal Ca in general is common 64-90% of patient presents with nodal metastasis
 - Bilateral LN in 8 -16% of cases o II to IV o Rarely I and V
 - Mediastinal nodal involvement in 80% of T4 pts

Pyriform sinuses tumors:

- Present late.
- Lateral spread **involves the thyroid cartilage and soft tissue of the neck** (can directly involve carotid)
- Medial extension **involves the larynx and the paraglottic space.**
- Posteriorly, the tumor can spread to and along the posterior pharyngeal wall into the contralateral pyriform sinus. o **High (50% to 80%) rates of regional LN mets**, most commonly to the **jugulodigastric nodes** (75% rate of LN mets in pasha).
 - In a review of 79 patients with pyriform sinus cancer who had radical ND for clinically N+, the incidence of LN metastasis :
 - level **II** and **III** → 72%
 - level **IV** → 47%
 - level I → 6.3%
 - level V → 7.6%
- Furthermore, bilateral metastasis is more common with lesions of the medial wall of the pyriform sinus than the lateral wall.

Posterior pharyngeal wall tumors:

- Tend to be exophytic without invasion of the prevertebral fascia, therefore, early lesions of the posterior pharyngeal wall are amenable to surgical resection.
- Tumors that have invaded prevertebral fascia considered unresectable.
- Because of the midline location, bilateral metastasis is common.
- First echelons are **levels II and III**, and (**RP** → 42% to 67%)
 - **Overall, incidence of cervical LN disease is between 35-45% (60% IN PASHA)**

Postcricoid tumors:

- Present late
- Tend to grow **circumferentially**.
- Involvement of the cervical esophagus is common.
- Because of the proximity of the cricoid cartilage anteriorly, **invasion of the cricoarytenoid joints and the cricoarytenoid muscles is common leading to cord immobility.**
- Often involve the thyroid gland, and paratracheal and paraesophageal nodes.
- **Up to 40% of patients will have regional metastatic disease (40% IN PASHA)**

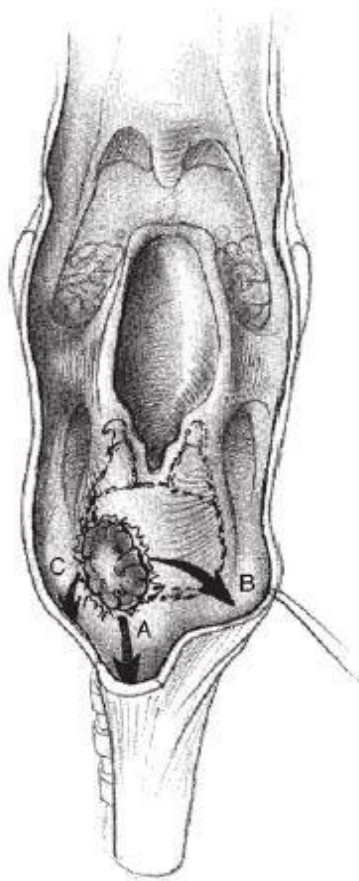


Figure 122.4 Lymph node and submucosal spread of piriform fossa cancer. Submucosal spread inferiorly (A) can involve the piriform apex and then metastasizes to the paratracheal, paraesophageal, thyroid, and jugulo-omohyoid nodes (B). Medial extension (C) involves the arytenoid and perilaryngeal compartments.

Molecular Pathology

The most common genetic abnormality found in head and neck cancer, including the hypopharynx, is **mutation in the p53 tumor suppressor gene**

Staging

T Staging for Hypopharyngeal Lesions

- **T1:** tumor 2 cm or less and limited to one subsite
- **T2:** 2-4 cm, or > 1 subsite of HP or adjacent site
- **T3:** > 4 cm in greatest dimension or hemilarynx fixation
- **T4a:** invades thyroid/cricoid cartilage, hyoid, thyroid gland, esophagus or central compartment soft tissue
- **T4b:** invades carotid, prevertebral fascia or mediastinum

T4 lesions have been divided into T4a (resectable) and T4b (unresectable), leading to the division of the stage IV into stage **IVA** and stage **IVB**. Patients with distant metastatic disease are classified as stage **IVC**.

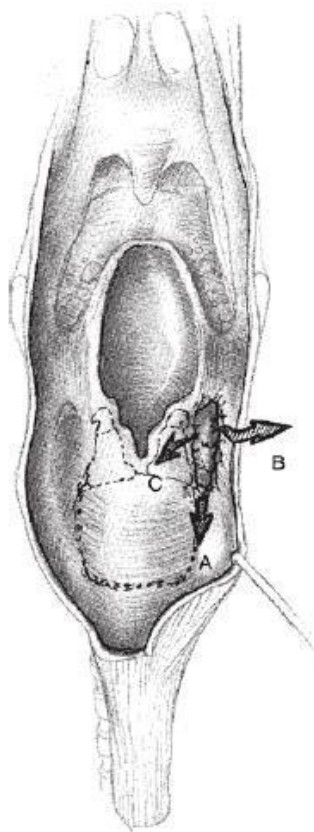


Figure 122.5 Lymph node and submucosal spread of postcricoid cancer. Submucosal spread inferiorly (A) can be extensive, and these lesions frequently metastasize to the paratracheal, thyroid, and paraesophageal nodes (B,C).

Management

Early stage (T1/T2) of all subsites:

- Radiation (w/ Elective B/L neck radiation ~pasha)
- Larynx-conserving surgeries can also be considered for highly selected patients (with ipsilateral elective neck dissection or B/L if medline lesion~pasha)

T3/T4 need either:

- Surgery with post-op rads or
- Organ-preservation chemorads which can achieve laryngeal preservation in **30% to 40%** of the patients without compromising survival is a valid therapeutic options

Posterior wall Cancer

- Often exophytic and do not invade the prevertebral fascia until advanced o Most often treated with **radiotherapy**
- May offer superior functional outcome due to preservation of the pharyngeal plexus (if resected may lead to aspiration because of the loss of propulsion and coordination of swallowing)
- Treat the RP lymph nodes in the same time
- **Surgery** is an option for small lesion.
- Approaches:

Lateral or suprahyoid pharyngotomy:

- Take resection down to prevertebral fascia; take RP nodes if suspicious; can recon with STSG and bolster; remove bolster transorally (Procedure's details in the book)
- If the resection site is on the posterolateral wall, lateral pharyngotomy alone or in combination with suprahyoid pharyngotomy may be used.
- In these cases, reconstruction can be accomplished with a **pec flap**, **rectus flap**, or **bipedicled prevertebral muscle flap** with STSG.
- Care should be taken, however, to ensure that the bulkiness of the muscle flap does not interfere with swallowing.
- For larger defects, reconstruction via **jejunal free flap** or **radial forearm free flap** can be considered.

Median labiomandibular glossotomy:

- Can be used for small lesions of upper pharyngeal wall
- Have significant morbidity

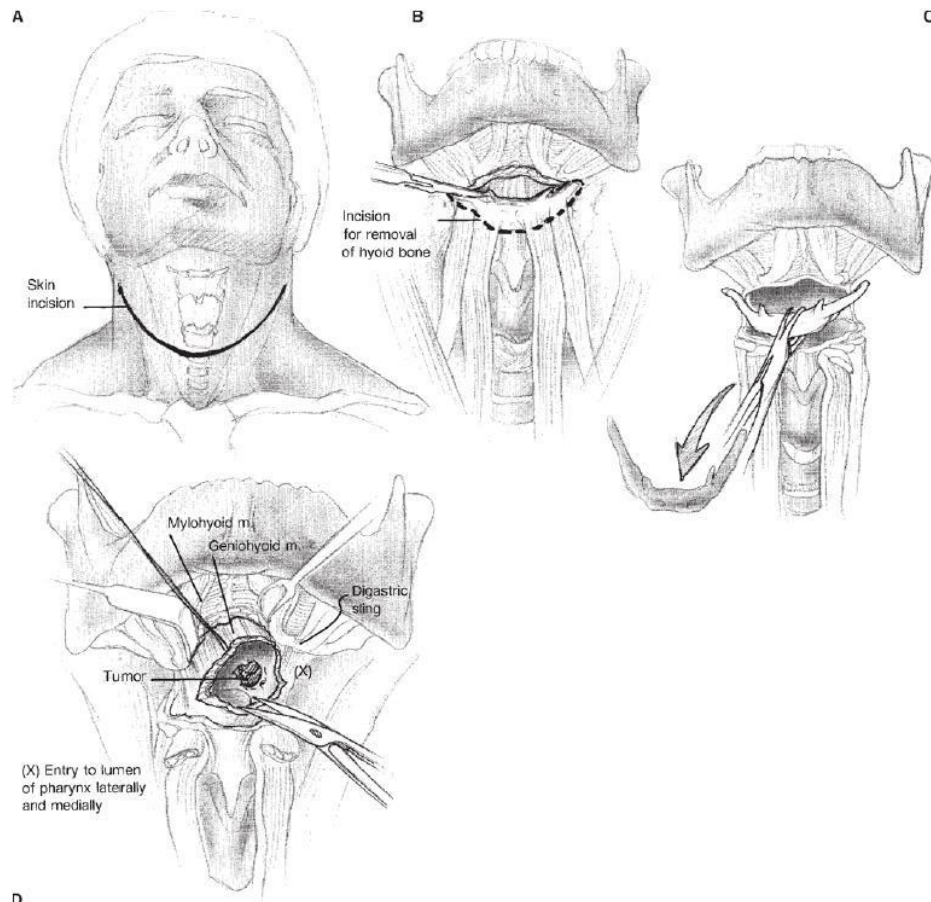


Figure 122.12 Suprahyoid pharyngotomy for posterior hypopharyngeal cancer. The initial incision (A and B) extends along the superior border of the hyoid for its entire length. The hyoid then is removed (C) to facilitate completion of the pharyngotomy (D).

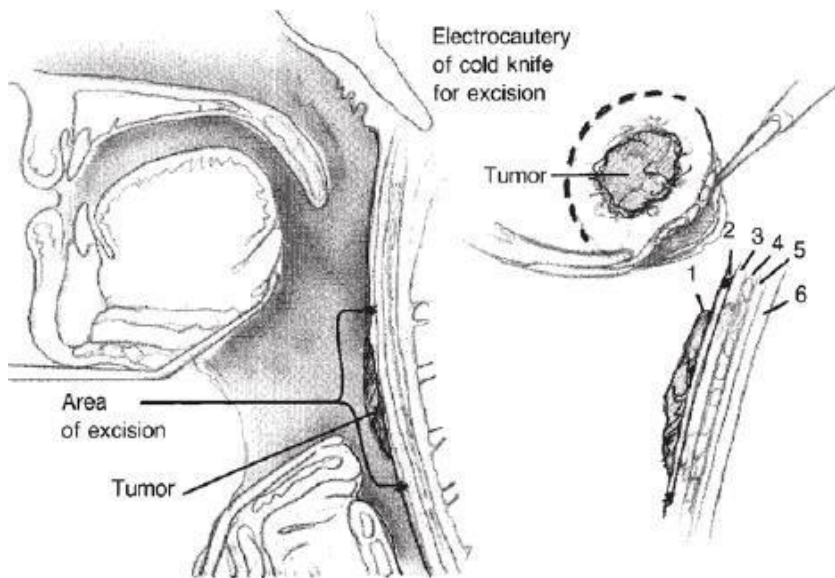


Figure 122.13 Suprahyoid pharyngotomy for posterior hypopharyngeal cancer. After the hyoid is removed and the pharyngotomy completed, superior and inferior retraction provides excellent exposure for wide excision of the cancer. 1, tumor; 2, mucosa; 3, constrictor; 4, longus colli; 5, retropharyngeal space; 6, prevertebral fascia.

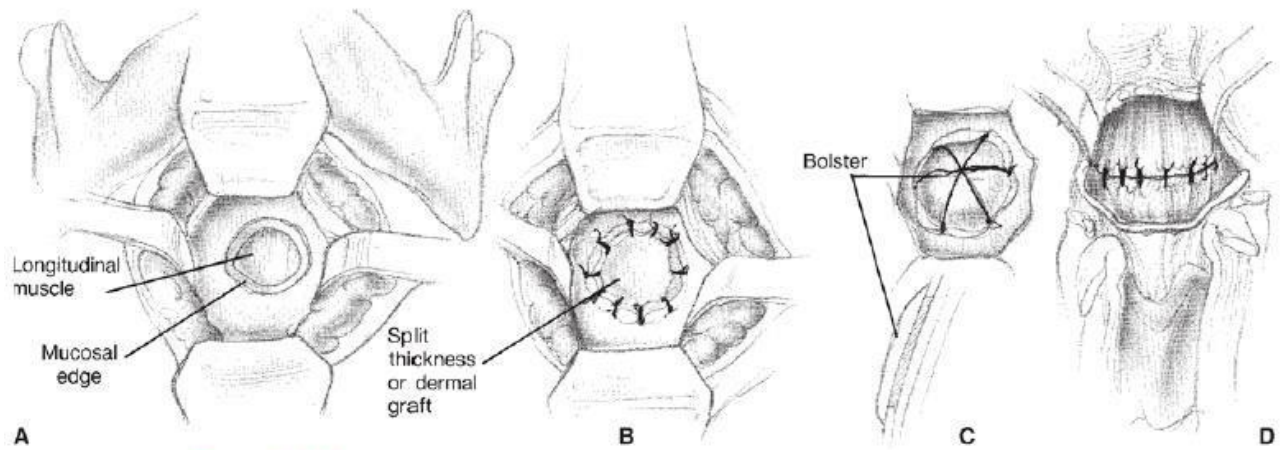


Figure 122.14 Suprahyoid pharyngotomy for posterior hypopharyngeal cancer. The incision is usually carried down to the prevertebral fascia (**A**), which acts as the surgical plane for excision. The defect then is covered with a split-thickness or dermal skin graft (**B**). This is held in place with a bolster (**C**) of nylon sheeting stuffed with cotton balls. The pharyngotomy is closed in layers, avoiding suture ligation of the hypoglossal and superior laryngeal nerves (**D**).

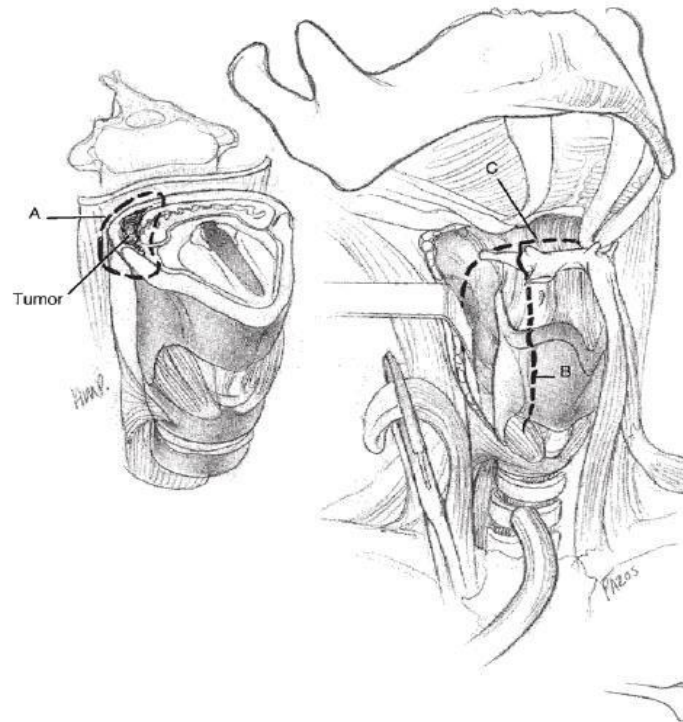


Figure 122.15 Combined suprahyoid and lateral pharyngotomy for posterolateral hypopharyngeal cancer. These cancers (A) are approached by excising the posterior third of the thyroid cartilage and combining the anterolateral pharyngotomy incision (B) with the suprahyoid incision (C).

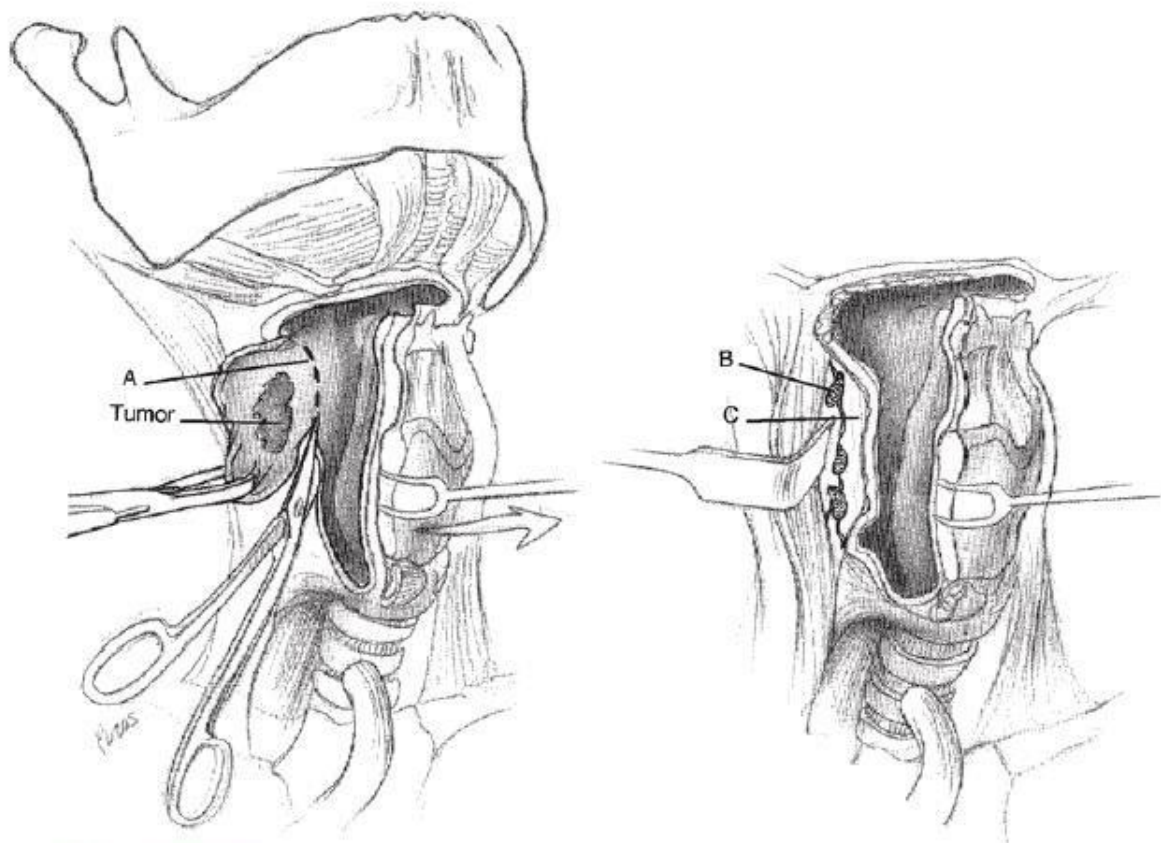


Figure 122.16 Combined suprahyoid and lateral pharyngotomy for posterolateral hypopharyngeal cancer. The final incision (A) is made under direct vision. The cervical sympathetic ganglion (B) should be preserved if it is not involved with the cancer. Reconstruction can be accomplished by using a portion of the prevertebral muscle (C) as a backing for a skin graft.

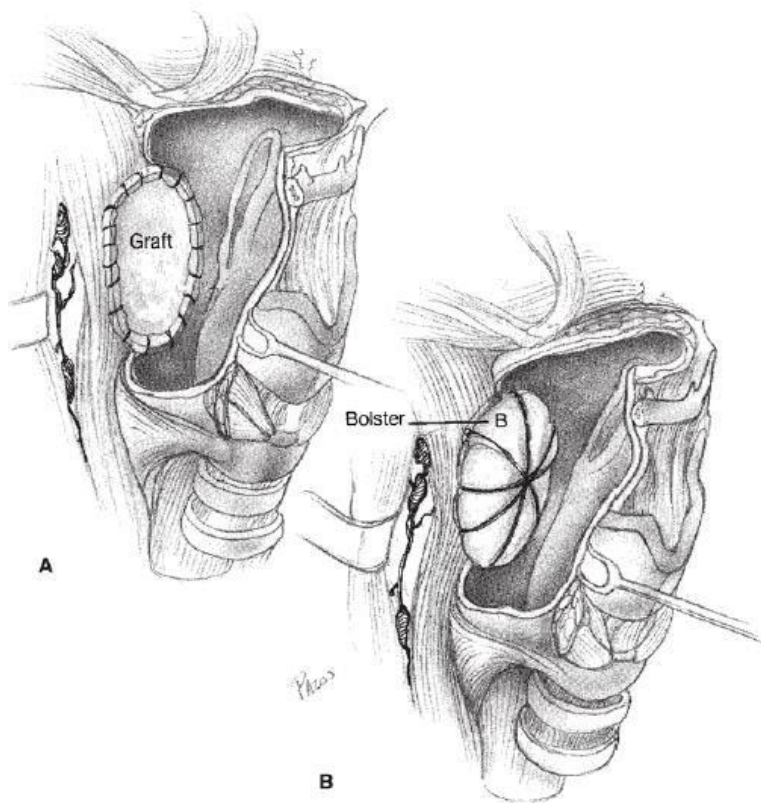


Figure 122.17 Combined suprahyoid and lateral pharyngotomy for posterolateral hypopharyngeal cancer. The split-thickness or dermal skin graft is sutured to the prevertebral muscle (A) and held in place with a nylon mesh and cotton-ball bolster (B).

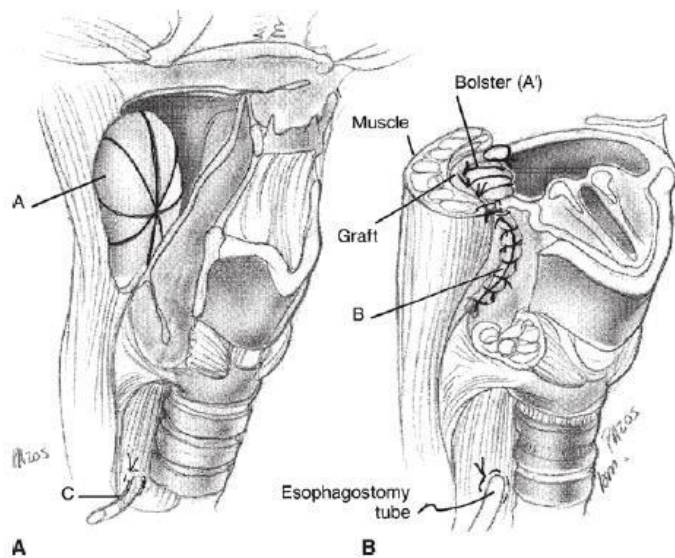


Figure 122.18 Combined suprahyoid and lateral pharyngotomy for posterolateral hypopharyngeal cancer. After the steps illustrated in Figures 122.15 and 122.16 the prevertebral muscle is mobilized as a bipedicle flap for medial rotation of the skin graft-bolster combination (A). The bolster is sutured in place with long sutures from the graft. A watertight closure of the pharyngotomy (B) is then accomplished, and a feeding esophagostomy is placed.

T1 and T2 piriform sinus lesions:

- High cure rates with **radiation** alone (preferred); include bilateral cervical and RP LN in the field
- Candidates for conservation surgery can be used for early lesions of the medial wall or pharyngoepiglottic fold (Only **2%** of patients with hypopharyngeal cancer will be candidates for conservation surgery)
 1. Does not involve apex of sinus or postcricoid area
 2. Normal ipsi VC movement
 3. Good pulmonary reserve
 4. No cartilage involvement

Approaches:

Partial pharyngolaryngectomy

- Combines supraglottic laryngectomy with the resection of the involved pyriform sinus.
- The resulting defect can be closed by suturing the tongue base to the thyroid perichondrium on the ipsilateral side. (Procedure's details in the book)

Supracricoid hemilaryngectomy

Can be used for lesions of the anterior part, the lateral wall, the medial wall, and the aryepiglottic fold

Complications:

1. Recurrence
2. Failed decanulation
3. Aspiration pneumonia (temporary gastrostomy may required)

T3 and T4 piriform sinus lesions:

- **Surgery with post-op radiation** is the gold standard:
 1. Postop rads results in significantly improved local control compared with surgery alone
 2. Surgery:
 - TL/PP(Total Laryngectomy and Partial or total Pharyngectomy)
 - If cervical esophagus is involved → cervical esophagectomy
 - If thoracic esophagus is involved → total esophagectomy and gastric pull-up
 - In all advanced hypopharyngeal cancer:
 - **Ipsi thyroidectomy**
 - **Ipsi dissection of paratracheal node**
 - **If it crosses midline or cervical esophagus is involved → need total thyroid and bilat dissection**
- Organ preservation **chemo/rads** a viable option

Postcricoid lesions:

- Usually advanced at presentation
- Often associated with invasion of the cricoid cartilage, cricoarytenoid muscle, and cervical esophagus
 1. Usually requires TL, PP and often cervical esophagectomy followed by postoperative radiation therapy
 2. Organ preservation chemo/rads again a valid option

Management of Regional Neck Mets

- LN mets in about **75%** of cases, with bilateral disease present in **10%**
- Rads or surgery, depending on modality chosen for primary Tx.
- Ipsilateral neck:
 1. T1/T2 lesions
 2. Lateral wall lesions
- Bilateral neck:
 1. Advanced lesions
 2. Lesion across midline
- Levels II-IV most commonly involved
- Paratracheal nodes need to be addressed as previously described
 - Paratracheal LN mets had a statistically significant negative impact on the survival
 - Elective paratracheal nodes dissection for patients with advanced lesions is recommended
 - Bilateral elective paratracheal nodes dissection for
 1. Lesion across midline
 2. Cervical esophagus is involved
- RP nodes very common for post wall lesions and need Tx (40-65%)

Role of Chemotherapy and Radiation Therapy:

- T1/T2 lesions:
 - 5-yr local control with rads alone → 70-100%
- T3/T4 lesions:
 - post-op rads takes 5-yr local control from **28% to 43%**
- Chemotherapy:
 - no role alone; no role for induction or adjuvant chemo.
 - concurrent chemo has survival advantage of **8%**
- **RTOG "Radiation Therapy Oncology Group":**
 - Locally advanced SCC of OC, OP, **HP (26%)** or SG larynx.
 - Randomized to pre-op rads; post-op rads or rads alone
 - Post-op did best
- **EORTC "European Organization for Research and Treatment of Cancer"**
 - T3/T4 HP lesions only
 - Randomized to induction chemo then rads or TL/PP with post-op rads
 - No difference in local control rates; survival better in chemorads group; 42% of chemorads group kept larynx
- **Intergroup R91-11 study:**
 - "Advanced laryngeal Ca"
 - Randomized to induction chemo followed by rads, concurrent chemorads, or rads alone
 - Concurrent did best (superior locoregional control)
 - No study yet done to prove that concurrent is best for HP lesions
- **RTOG 9501 and EORTC trial 22931**
 - "Role of concurrent chemorads post-op for pts with high risk of recurrence"
 - High-risk criteria in RTOG study:
 - Histologic evidence of involvement of 2 or more LN
 - Extracapsular invasion
 - Positive margins
 - In addition to these features, EORTC study extended the high-risk criteria to include:
 - Vascular or perineural invasion
 - PT3 or pT4 disease
 - Tumor stage of 1 or 2 with a nodal stage of 2 or 3 and no distant metastasis
 - Randomized post op (if high risk of recurrence) to postop rads or postop concurrent chemorads.
 - Both showed increased 2-yr **local/regional control** by 10% with the concurrent group; EORTC group **showed increased survival (53% vs. 40%)**
 - Both showed higher incidence of severe early side effects

Prognosis

- 35-40% overall survival; positive nodal status halves survival
- Post wall: all stages 5-yr survival = 35-45% with surgery + rads
 - Although radiation therapy is effective for the early lesions of the pharyngeal wall, surgery combined with rads is superior for advanced lesions
- Piriform: similar; see above for rads v. surgery and rads
 - The 5-year survival ranges from 40% to 60%.
 - Early lesions of the pyriform sinus can be treated with either radiotherapy or conservation laryngeal surgery.
 - Advanced lesions of the pyriform sinus, however, radiotherapy alone offers poor rates of local control and 5-year survival compared with surgery followed by adjuvant radiotherapy
- Postcricoid: limited data; low incidence
 - Radiotherapy alone may be adequate for early lesions but surgical resection with post-op rads offers superior outcomes
 - Vocal cord paralysis found to be a poor prognostic factor

**TABLE
122.4**

**HYPOPHARYNGEAL CANCER: 5-YEAR SURVIVAL BY LYMPH
NODE STATUS (EASTERN VIRGINIA MEDICAL SCHOOL
EXPERIENCE)^a**

Node Location (n)	Negative (n)	Positive (n)	5-Year Survival (%)
Piriform fossa (63)	14	49	8/14 (57) 11/49 (22)
Posterior wall (30)	7	23	5/7 (71) 12/23 (52)
Postcricoid (4)	1	3	1/1 (100) 0/3 (0)
Totals	22	75 ^b	14/22 (63) 23/75 (30)

^aEastern Virginia Medical School 1972 to 1985; 97 patients.

^bBilateral nodes in 10 of 97 (10%) of entire group or 10 of 75 (13%) of those with positive nodes. Only 1 of 10 (10%) survived 5 years.

Reconstruction

- Small lesions: primary closure or skin grafts
- Don't tube pharynx if < 2 cm of mucosa left (because of concerns of stricture); use pec, ALT or forearm
- Avoid pec if larynx preserved; bulk leads to aspiration
- For TP defect: tube forearm or ALT; free jejunum
 - The free jejunal graft, which allows early deglutition, is associated with a low incidence of stenosis and fistula
 - Supercharged (**pedicled and micro**): 2% failure in one study
 - "supercharging" of the arterial supply to the distal end of the enteric conduit can be performed where additional vascular supply to this area is established via microvascular anastomosis
 - All-comers have 85% rate of G-tube independence
 - Jejunal free flap may result in better swallowing ability compared with radial forearm free flap.
 - TEP better if HP not resected; still adequate for most
- For TP/esophagectomy: gastric pull-up is recon of choice
 - The need for major abdominal surgery and posterior mediastinal dissection, however, results in operative morbidity of 50% and mortality rates of 10%.
 - Another option in this situation is the use of pedicled jejunum or colonic interposition.

Complications

- Leakage:
 - The pre-operative factors:
 - Nutritional status
 - History of prior radiation therapy
 - The Intra-operative factors:
 - Type of reconstructive option used
 - Tight closure
- Infection, hemorrhage, and breakdown of skin wounds are also common in these high-risk patients
- Airway obstruction "with tracheostomy".
- A late complication is aspiration, which if severe, can lead to pneumonia.
- Stenosis after circumferential reconstructions may require repeated dilation

Early Laryngeal Cancer

- The term "early laryngeal cancer" refers to a mucosally derived neoplastic lesion that may invade deeply into soft tissue. but not into the underlying cartilage.
- The majority of literature reports include carcinoma in situ (CIS), T1 and T2 lesions in the spectrum of early laryngeal cancer.(T1N0M0, T2N0M0)

Epidemiology:

- Laryngeal cancer is the **2nd most common** type of head and neck cancer worldwide
- Male to female ratio 6:1
- Eastern Asia and central and eastern Europe
- Incidence declined in the states due to reduced smoking
- **60 %** of laryngeal CA cases are diagnosed as Early Laryngeal cancer (T1,T2)
- Most commonly from **the Glottis**

Risk factors:

- **Smoking & alcohol:**
 - multiplicative effect, tobacco and alcohol exposure was the highest in larynx of all head and neck cancer sites at 89%
 - 5% of laryngeal cancers occur in nonsmokers and non-drinkers
 - suggesting other factors such as diet, gastroesophageal reflux. previous radiation, and viral infection may also contribute
- **HPV 16, 18** has been analysed in 5-32% of laryngeal cancer specimens
- **Occupational exposures** to wood dust, polycyclic hydrocarbons, and asbestos

Genetics:

- Mutations in **p16**, a tumor suppressor gene located at chromosome 9p21, occur in over half of laryngeal squamous cell carcinomas
- Overexpression of **cyclin D1**, a key cell-cycle regulator protein, results in increased cellular proliferation and tumorigenesis (17). It has also been shown to correlate with poor prognosis, recurrence. and lymph node metastasis
- Alterations in **p53** a tumor suppressor gene result in destabilization of genomic repair processes leading to tumorigenesis

Pathology:

- Laryngeal cancer is thought to arise from precancerous lesions. characterized by atypical cellular changes with malignant features such as:
 - loss of cell maturation
 - nuclear atypia
 - increased mitotic activity
- precancerous lesions includes Dysplasia , graded from "mild" to "severe" depending on the extent of epithelial involvement and CIS (involvement of the whole thickness of the epithelium)
- progression to micro-invasive and invasive carcinoma characterized by infiltration of the basement membrane and the underlying tissue
- other histological types of primary laryngeal tumors includes :
 - verrucous SCC (highly diff. with low incidence of mets)
 - adeno ca
 - spindle cell ca
 - fibrosarcoma
 - chondrosarcoma
 - Neuroendocrine tumors (rare, but is the most common non epithelial)

Anatomical considerations:

- Supraglottis Extends from the Epiglottis to the ventricular apices, including :
 - Laryngeal surface of epiglottis
 - A-E Fold
 - Arytenoids
 - False Vc
- Glottis contains :
 - True VC
 - Anterior commissure
 - Inter Arytenoid region
 - Floor of the Ventricle
 - It extends 1cm below the plane of the apex of the ventricles
- Subglottis :
 - Extends from 1cm below the ventricle to the lower border of the cricoid

Laryngeal spaces (anatomy lecture)

Lymphatic drainage:

- **Supraglottis** rich lymphatic network , primary to Level II , then level III then extends to level IV
- **Glottis** , non-exiting lymphatic drainage
- **Subglottis**, paratracheal (level VI) is the primary echelon nodes , then may spread to Level VII or Level IV

Early supraglottic cancer

- **30-40% of laryngeal ca**
- **Often silent , Or non-specific sx, like otalgia and sore throat**
- **Dysphagia and aspiration with only significant bulk**
- **Cervical mets at presentation depends on T stage , site and differentiation**
- **Generally from 0-33% in T1 ,T2**

Diagnostic approach

- History and physical examination, office-based fiberoptic laryngoscopy
- Imaging: Office neck ultrasonography
- CT/MRI for assessment of primary tumor and neck
- chest CT or PET
- Pulmonary function test
- Direct laryngoscopy under anesthesia for biopsy assessment of
 - Site
 - Size
 - extent of primary tumor
 - Tumor extension to
 - ✓ Vallecular
 - ✓ tongue base
 - ✓ pharyngeal wall
 - ✓ medial wall of piriform sinus
 - ✓ arytenoid/ventricle
 - Cord mobility (hypomobile T2 , fixed T3)
 - Palpation of neck nodes
 - Adequacy of transoral exposure
 - UADT examination for second primaries

Surgical pathology:

- The **supraglottic** is a **different embryological** entity rising **from 3rd+ 4th** branchial arch (buccopharyngeal analge) and glottic and subglottic from the 6th (trachopulmonary) analge, but there's no anatomical barrier to separate it from the rest
- early tumors spreads within the supraglottis (easier resection)
- glottis invasion can happen with **epiglottic petiole** lesion extending inferiorly
- epiglottic perichondrium and thyroepiglottic ligament limit anterior extension
- tumor can spread into the fatty areolar tissue of PES via epiglottic cartilage fenestrations (**PES invasion implies T3** , usually happen with infra-hyoid lesions)
- a tumor free space usually presents between the hyoid and anterior front of the tumor (helps in saving the hyoid in SGL (Supraglottic laryngectomy) with oncological safety
- extra laryngeal spread occurs when extension anteriorly from the PES, breaching the thyro-hyoid membrane

- another extra laryngeal route is through the Hyo-epiglottic ligament toward the valcula and tongue base
- ossified cartilage prone to invasion
- Aryo-epiglottic fold lesions may spread laterally to invade medial wall of pyriform sinus

Staging and treatment planning

A complete workup to assess the patient, the subsite and stage primary tumor, and the presence of cervical lymphadenopathy is indicated

Cord hypomobility implies paraglottic space invasion. and cord fixation upstages the primary tumor to T3.

For T2 lesions, radiologic evaluation with computed tomography (Ct) or magnetic resonance imaging (MRI) is performed to determine the extent of the primary and to rule out involvement of the PES, the paraglottic space, or the inner thyroid cortex

Treatment

- **Single-modality therapy** with primary **surgery or definitive radiotherapy** (Rt) are the two alternative treatment approaches for early supraglottic tumors.
- **RT alone** however, **is less effective for local control** and high laryngectomy rates (14% to 31%) for rerurrent tumors, have made this option less acceptable than primary surgery. **(dr.khalid prefer Radiation)**
- T1 and T2 primaries are amenable to curative treatment by conservation surgery with an optimum oncologic control and minimal functional morbidity
- The modern paradigm of conservation surgery for supraglottic tumors, , is minimally invasive, endoscopic transoral laser microsurgery (TLM)

Surgery

Transoral laser microsurgery

- The basic technique of **trans-tumoral** cuts to assess the depth of disease and **multibloc** transoral laser resection .
- makes it possible to resect the primary tumor with optimally cleared histologic margins and, at the same time, preserve the anatomical and functional integrity of the noninvolved, surrounding tissue.
- The resection can be tailored according to tumor location and extension
- Contraindications is inadequate tranoral access to the entire tumor **8Ts** :
 - Teeth Prominent
 - Trismus
 - Transverse dimension (narrow mandibular arch)
 - Tori
 - tongue
 - Tilt (atlanto-ocipital extention)
 - Treatment (prior chemo or rad)
 - Tumor (site and size)
- Disease requiring Bilateral arytenoids resections is contraindicated

**TABLE
123.2****ELS CLASSIFICATION OF ENDOSCOPIC SURGERIES FOR EARLY SUPRAGLOTTIS
AND GLOTTIS CANCER**

Endoscopic SGL (42)	Endoscopic Cordectomy (43,44)
Type I, limited excision of small size, superficial lesions in any part of the supraglottis	Type I, subepithelial cordectomy
Type II, medial SGL without resection of the PES	Type II, subligamental cordectomy
Type IIa, superior hemiepiglotectomy	
Type IIb, total epiglotectomy	
Type III, medial SGL with resection of the PES	Type III, transmuscular cordectomy
Type IIIa, without extension to the ventricular fold	
Type IIIb, with extension to the ventricular fold	
Type IV, lateral SGL	Type IV, total cordectomy
Type IVa, includes ventricular fold	
Type IVb, includes arytenoid	
	Type V, extended cordectomy
	Type Va, encompassing the contralateral vocal fold and the anterior commissure
	Type Vb, encompassing the arytenoid
	Type Vc, encompassing the ventricle
	Type Vd, encompassing subglottis (1 cm)
	Type VI cordectomy for cancers of the anterior commissure
	Extension to one or both of the vocal folds, without infiltration of the thyroid cartilage

Surgical considerations:

- ET intubation with Laser tube (small)
- Laser safety and precaution (eyes)
- Mouth guard (Aquaplash) to protect teeth and gingiva
- Introducing laryngoscope (Stein, Weerda. Kleinsasser.)
- Transoral resection is performed with a co2 laser beam delivered either via micromanipulator or a handheld device
- rapid debulking may initially be performed with electrocautery
- For margin management Co2 laser is used for its precise and bloodless properties under microscopy

Surgical technique:**suprahyoid epiglottis:**

- Small and well lesions may be resected **en bloc**
- For Larger lesions epiglottis divided sagittally from sup – inf
Then lateral cuts across the vallecula , later P-E ligament and epiglottis bilaterally if necessary
- Wide resection margins can be obtained without swallowing morbidity

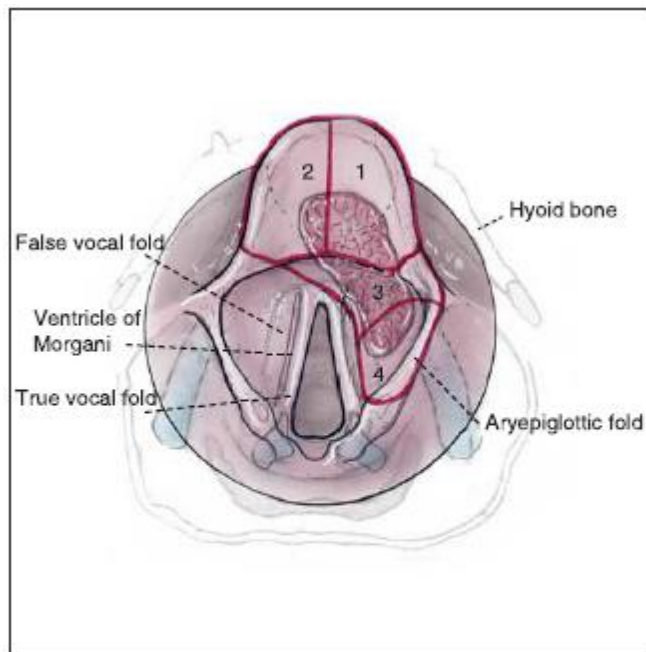


Figure 123.2 Incisions for multibloc TLM resection of supraglottic tumor. Blocs 1–4 are removed in numerical order.

Infrahyoid epiglottitis:

- Close to PES so caution
- Some tumors are key-holed out of the epiglottis by excising a disc of mucosa and epiglottic cartilage containing the disease, with perichondrium, using the preepiglottic fat as the deep margin.
- If extensive laryngeal surface of Epiglottis involved > Epiglottitis split sagittally and ipsilateral **Epiglottectomy** preformed
- Margins at least 5 mm

False vocal cords:

- Tumors at these sites can be completely resected using similar TLM techniques.
- The depth of excision is determined by a transverse cut through the tumor's epicenter.
- the surface extent of tumor determines the design of the mucosal incisions; inferiorly, the dissection is usually carried to the level of the ventricle or the superior vocal cord
- safe lateral margin 5mm
- safe inferior margin usually 1-2 mm

Aryepiglottic fold:

- Small and superficial lesions can be excised en bloc.
- The general principle is to split transversely through the tumor and the fold until sufficient deep tissue margin (2 to 5 mm) is established
- When necessary, the arytenoid may be shaved away serially from anterior to posterior, leaving the posterior body undisturbed

Hemostasis :

- Meticulous hemostasis should be ensured during and at completion of all TLM procedures for supraglottic tumors
- Bipolar cautery directly down the endoscope, is especially useful for mucosal bleeding, common along epiglottic and base of the tongue incisions
- Hemostasis is critical when the procedure involves transection of lateral pharyngoepiglottic fold or dissection of lateral supraglottic mucosa due to proximity to or exposure of the **superior laryngeal artery** or its branches
- Before extubation, the laryngoscope is desuspended to allow for reduction of any temporary hemostatic tamponade, and **Valsalva maneuvers** are used to stimulate dormant bleeders

Frozen section:

- Should be marked and labelled
- Shouldn't be crushed or burned (crush artefact make it diffc. to read)
- 2% discrepancy has been reported between final and frozen

Post-operative care:

- Antibiotic broad spectrum 3 weeks course
- AntiReflux 3 weeks
- Very rarely requires intubation or tracheostomy

Complications:

- Air way Fire ,prevention by:
 - decrease Fio2 to 30%
 - Laser tube
 - Laser proof coverage over Non laser tube
- Secondary haemorrhage (day 0 to day 21)
- Post resection Edema
- Aspiration and pulmonary complications infrequently resection of the following increase aspiration:
 - Artyenoids
 - Arye-Epiglottic folds
 - Tongue base
- Suprqlottic stenosis
- Laryngoscope pressure complications:
 - Dental injury
 - TMJ
 - Mandible
 - Paryngeal tears
 - Tongue swelling
 - Laceration
 - Numbness or loss of taste

Transoral Robotic surgery

- Transoral surgery using the da Vinci robotic surgical system was recently reported to be suitable for performing SGL
- 3 arms , suction device , surgical arms and endoscope for viewing
- PRONS :
 - The three-armed robotic system is said to confer improved
 - three-dimensional optics
 - tissue manipulating/cutting maneuverability
 - The "endowrist • design allows for flexibility in movement at the end of the robotic instruments
 -
- CONS:
 - Some optical resolution is lost over a high-quality operating microscope
 - the arms are straight are rigid, and cannot be maneuvered down a laryngoscope
 - necessitating spatulate retraction of the tongue.
 - significant instrument crowding
- Currently; the robot appears to offer **no real advantage** over endoscopic laser resection although its application in supraglottic tumors may increase with future refinements.

Open conservation procedures

Horizontal Supraglottic Laryngectomy:

- Open • SGL entails resection of :
 - the epiglottis
 - false cord
 - aryepiglottic folds
 - hyoid
 - upper half of the thyroid cartilage.
 - complete resection of PES.
- **Spare structures** that are vital to phonation, respiration, and deglutition which are :
 - the true cords
 - at least one arytenoid
 - tongue base.
 - avoids the need of a permanent tracheostomy
- In early-stage tumors, the basic SGL can be modified to preserve the hyoid bone
- **.Extended •SGL** may be performed for lesions extending :
 - vallecula,
 - tongue base
 - ipsilateral arytenoid
 - medial wall of pyriform sinus
 - A tracheostomy is always required at the time of resection.

▪ **Contraindications:**

- Paraglottic space involvement (T3)
- Cord fixation (T3)
- thyroid cartilage invasion (T4)
- bilateral arytenoid
- interarytenoid space involvement
- tumor extension to the pyriform sinus apex
- extensive tongue base involvement
- poor motivation
- respiratory insufficiency

Surgical technique (surgical steps):

- A preoperative laryngoscopy is performed to reassess the extent of primary tumor and a preliminary tracheostomy is secured
- An apron incision is made and subplatysmal flaps are raised
- The hyoid bone is skeletonized
- infrahyoid muscles are detached from the body and greater cornua of the hyoid
- in preparation for the superior pharyngotomy.
 - The greater cornu should be skeletonized with care because of proximity to the **hypoglossal nerve**
- **The superior thyroid neurovascular pedicle** is divided and ligated on the side of the tumor
- constrictors are separated from the posterior edge of the ipsilateral thyroid ala.
- An incision is made through the perichondrium along the superior border of the thyroid cartilage
- perichondrial flap is reflected inferiorly below the level of the true cord
- exposing the upper two-thirds of the thyroid cartilage.
- The thyroid cartilage is transected horizontally just above the midpoint between the thyroid notch and lower border including the superior cornu on the tumor side.
- The incision is extended horizontally across to the other side or obliquely to preserve the contralateral superior cornua of the thyroid cartilage
- The pharynx is entered superiorly infrahyoid, through the vallecula if the tumor is confined to the supraglottic larynx.
- For tumors with extension to the vallecula, a suprahyoid or lateral pharyngotomy approach through the contralateral pyriform sinus is utilized
- The epiglottis is delivered outward through the pharyngotomy
- retracted to visualize the laryngeal introitus.
- Connecting mucosal incisions are made through the aryepiglottic folds on both sides, anterior to the arytenoids
- The cuts are extended through the posterior part of the false cords into the ventricle and connected to the thyroid cartilage incisions to complete resection.
- An inferior margin of about 2 to 3 mm normal mucosa has been found to be adequate to prevent recurrence at the glottic level
- Resection of all supraglottic mucosa including uninvolved false cord is recommended to prevent postoperative edema.

- The role of cricopharyngeal myotomy in SGL for better swallowing outcomes lacks consensus.
- securing complete hemostasis,
- the defect is reconstructed by suturing the thyroid perichondrial flap to the deep musculature of the tongue base with 3-0 Vicryl.
- A second reinforcing layer is created by suturing the suprahyoid muscles to the cut edges of infrahyoid musculature.
- The neck incision is closed after placement of a dosed suction drain.
- The tracheostomy tube is reinserted through a separate incision in the lower skin flap and care is taken to isolate the stoma from the major resection wound in order to prevent contamination of the neck

Postoperative care:

- Routine care of wound, tracheostomy, and neck drain is instituted.
- Feeding is initiated through a nasogastric tube
- Deflate the cuff after the edema subsides
- Trial of decanulation, Depends on patients tolerance ,
- Intensive swallow rehabilitation
- Supraglottic diet initially followed by a normal diet

Complications:

- Severe aspiration
 - Recurrent aspiration pneumonia
 - Intractable life threatening aspiration (w/ Extended SGL Base of tongue or arytenoid)
 - May require functional Total Laryngectomy
- Bleeding
- Infection
- Accidental decanulation
- Subcutaneous emphysema

Supracricoid Laryngectomy

- It entails en bloc resection of:
 - the PES
 - paraglottic space
 - Thyroid cartilage
- **Indications :**
 - T2 supraglottic cancers extending to :
 - ❖ the ventricle,
 - ❖ the anterior commissure
 - ❖ the glottis with or without impaired cord mobility
- yielding outcomes comparable to total laryngectomy in **selected** cases
- preserving functions of phonation and deglutition without a permanent tracheostoma

- **contradictions:**
 - Invasion of the cricoid cartilage
 - Bilateral arytenoid involvement
 - extension into the base of the tongue
 - extension into vallecula
 - extension into hypopharynx.
 - fixation of arytenoids
- **surgical technique (steps):**
- A direct laryngoscopy is recommended at the onset for reevaluation of the tumor and its suitability for resection
- After adequately skeletonizing the larynx. the inferior constrictors are incised along the lateral border of the thyroid cartilage
- Followed by release of the pyriform sinuses from the inner thyroid cartilage.
- The external thyroid perichondrium is preserved bilaterally
- During division of the superior laryngeal vascular pedicle, care is taken to preserve the **internal branch of the superior laryngeal nerve**.
- The cricoarytenoid joints are disarticulated with care bilaterally to preserve both **recurrent laryngeal nerves**, which lie just medial and posterior to this joint
- A cricothyrotomy is performed just immediately above the cricoid following which ventilation continues through this new opening.
- The pharyngotomy is performed by entering the vallecular on the uninvolved side, grasping and retracting the epiglottis through the vallecular incision
- Sharp dissection is continued alongside the epiglottis to the aryepiglottic fold
- and in front of the arytenoid bilaterally, down to the cricoid cartilage and the cricothyrotomy incision
- thus releasing the specimen
- On the side of the tumor, incisions are made ensuring safe resection margins
- If one of the arytenoids needs to be included in the surgical specimen the incision down the aryepiglottic fold is **made posterior to**, instead of anterior to, arytenoid on the ipsilateral side
- Before closure, hemostasis is secured and the mucosa is approximated over the arytenoid in a manner that does not limit its movement
- To prevent posterior prolapse of arytenoid, a single 3-0 Vicryl suspension suture is passed through the arytenoid, anchoring it forward to the cricoid cartilage.
- Neck extension is removed to perform **laryngoplasty** by approximating the cricoid and hyoid using interrupted, nonabsorbable 1-0 Vicryl sutures passed submucosally
 - one in the midline
 - one 1 cm on either side of the midline
- Tightening of these cricohyoidopexy (CHP) sutures brings the arytenoids(s) closer to the tongue base.
- This facilitates vibration of the mucosa over arytenoids(s) against the tongue base, a mechanism that is responsible for phonation in these patients

- Once the trachea has been stretched up by the CHP, tracheostomy is performed at an appropriate site that allows the tube to exit skin through the neck incisions
- The inferior and superior margins of strap muscle flaps are approximated to reinforce the CHP area
- Neck incisions are closed after placement of suction drains

Complications:

- Aspiration:
 - Atelectasis
 - Pneumonia
 - Precautions and Counter measures to aspiration :
 - Preservation of the superior laryngeal nerves
 - anterosuperior suturing and positioning of the cricoarytenoid unit
 - early decannulation
 - early onset of swallowing rehabilitation
- Hemorrhage
- Infection
- wound dehiscence
- accidental decannulation
- subcutaneous emphysema
- pharyngocutaneous fistula

Management of the neck

- Clinically or radiologically positive neck disease requires the patient to undergo neck dissection.
- Ipsilateral N+ necks require ipsilateral dissection
- bilateral or contralateral N+ necks require bilateral neck dissection
- Neck dissection is performed at the same operative session as primary tumor resection
- Selective, lateral dissection of levels II, III, and IV is an effective and adequate procedure for management of the neck in early supraglottic cancer
- Presence of multinodal or multilevel neck disease with **extracapsular spread** is an indication for postoperative **RT or chemoradiotherapy**.
- The standard of care in early supraglottic tumors with an NO neck is controversial. An occult metastatic rate ranging from 4% to 35% combined with a high risk of contralateral metastasis, is cited as a rationale for performing a routine bilateral neck dissection.
- Although surgery is preferred in neck management RT alone to one or both necks can be used to control NO or N1 necks
- However, unwanted radiation to the larynx will occur, even with intensity-modulated radiation therapy, adding to the risk of prolonged airway edema and obstruction from arytenoid swelling.

Outcomes:

- Surgical therapy alone achieves oncologic outcomes equivalent to combined therapy in early supraglottic cancer
- The decision for administering adjuvant RT should be judicious and based on high-risk neck indication
- Lymph node metastasis is the most important prognostic factor in supraglottic cancer
- a decline in survival was observed among the early-stage supraglottic cancers, T1NOMO and T2NOMO
- At this stage. no definitive links can be identified between outcomes and patterns of surgical treatment.

- **Oncologic:**

- Excellent oncologic outcomes have been reported for TLM in early supraglottic cancer
- A 5-year recurrence-free survival of 83% for early tumors,
- a 5-year local control rate of 100% for T1 and 89% to 97% for T2 tumors,
- 5-year laryngeal preservation rate of 86% to 100%
- Few comparative studies found in the endoscopic laser group compared to open SGL group
 - higher 5-year disease-free survival (80% vs.72%)
 - 5-year laryngeal preservation rate (86% vs. 80%)
 - 5-year disease-specific survival (89% vs. 80%)

- **Functional**

- TLM superior to open conservative in
 - shorter hospital
 - stays, lower tracheostomy rate
 - better swallowing outcomes.
- a lower incidence of aspiration in TLM (11.5%) compared to open surgery (40%) for supraglottic cancer
- other advantages of **TLM** Avoidance of :
 - wound dehiscence
 - pharyngocutaneous fistula

Radiotherapy:

- Definitive RT is another treatment modality reported to be effective for management of early supraglottic cancers.
- The local control rates observed from review of large RT series (61,62) range from
 - 75% to 100% for T1 tumors
 - 71% to 83% for T2 tumors,
 - laryngeal preservation rate of about 80%
- The success of RT is often correlated with the tumor volume, better results are reported for
 - Superficial tumors
 - small-volume tumors
- Recurrences after primary RT for early supraglottic cancer have been reported to be as high as 29%
- Surgical salvage with conservation procedures or total laryngectomy is associated with a greater risk of complications:
 - pharyngocutaneous fistula
 - tissue necrosis, wound breakdown
 - carotid artery rupture.
 - The rate of complications has been related to the dose of radiation therapy

EARLY GLOTTIC CANCER

- Glottic cancer accounts for about 50% to 60% of all laryngeal cancer cases.
- A change in voice with hoarseness resulting in diagnosis at an early stage
- These lesions do not cause airway obstruction or swallowing difficulty unless they progress to higher T stages.
- Lymphatic spread of early glottic cancer is extremely rare.

Surgical pathology:

- Glottic cancer mostly arises on the mucosa of anterior true cords
- A region with paucity of lymphatics
- Tumor extension occurs along the cord in the submucosal space
- **The chief barriers** to spread of glottic cancer are:
 - the vocal ligament
 - conus elasticus
 - thyroid perichondrium
 - thyroid cartilage.
 - The Broyle ligament (barrier to superior and anterior spread)
 - site of insertion of the Broyle ligament at the anterior commissure is devoid of inner thyroid perichondrium, and once involved, it serves as a portal of entry of the tumor into the thyroid cartilage from where it can spread to involve the ossified portions of the cartilage or further to the extralaryngeal structure

spreading pattern:

- **Anterior :**
 - Spread may occur across the anterior commissure to the opposite cord
- **Posteriorly:**
 - the lesions can spread to involve the thyroarytenoid muscle
 - This may result in impaired mobility, which should be carefully assessed during preoperative planning to differentiate from cord fixation
 - Further posterior spread may occur to involve the arytenoid cartilage or the posterior commissure.
- **Inferior:**
 - Inferior extension can occur to the subglottis; spread of more than 1 cm below the free edge of the true cord can invade the cricoid cartilage
- **Superior:**
 - With superior extension, the tumor can spread along the floor of the ventricle and invade the glottic musculature.
- **Transglottic :**
 - Extension to the false cords, filling the ventricle, makes the lesion transglottic
- **Paraglottic space invasion:**
 - can also be involved following spread across the conus elasticus or lateral spread of the tumor along the superior surface of the true cord.

Staging and Pre-treatment Planning

- History and physical examination
- office-based fiberoptic laryngoscopy
- Videostroboscopy facilitate :
 - assessment of the superficial extent as well as the infiltrative depth of early lesions
- Imaging: Limited role, evaluation of invasion in bulky lesions (baily)
- CT H&N , CT CAP (khalid)
- Pulmonary function test
- Direct laryngoscopy under anesthesia for biopsy and assessment of
 - Site
 - Size
 - Surface
 - extent of primary tumor
 - Tumor extension to
 - subglottis,
 - Arytenoid
 - Supraglottis
 - anterior commissure
 - petiole of epiglottis
- Cord mobility
- Adequacy of transoral exposure, especially anterior commissure
- UADT examination for second primaries

Treatment

- Fundamental goals of treatment:
 - Disease cure
 - Voice preservation
 - Airway protection
 - maintenance of function-preserving options for recurrence (avoid salvage total laryngectomy after recurrence)
- treatment modalities :
 - Transoral laser microsurgical codectomy
 - Open surgeries (codectomy , hemi-laryngectomy)
 - Radiation
 - ❖ All options have been reported to have excellent survival outcome
- treatment decision is guided by other variables:
 - patient preference
 - age
 - expectations for quality of voice
 - tumor stage
 - site, for example, extension to anterior commissure
 - the physician's expertise and skills
 - treatment-related factors such as :
 - morbidity
 - cost-effectiveness
 - availability of function preserving salvage options(RT)

Carcinoma In situ management

- CIS can exist alone, involving limited or extensive areas of true cords
- or it can coexist with foci of invasive squamous cancer
- Management plans described in literature include:
 - watchful waiting (not practical 66% will progress to invasive)
 - cord stripping (Modest local control rates 56% -72% and worsening of voice have been reported)
 - excisional biopsy (EBx)
 - transoral laser excision (TLM) (high local control rate 75-100%)
- RT(Definitive treatment with RT is preferred at a few centers for the treatment of CIS) disadvantages with comparable control rate :
 - the long-term complications
 - increased treatment time
 - costs
 - exhausts the option of subsequent radiation for recurrent or second primary tumor.
- ❖ TLM when compared with Ebx it provide :
 - Better diagnostic yield
 - Offer complete removal of macroscopic disease in the same settings
- Approximately 29% of the laryngeal CIS lesions have a potential to progress to SCC
- Transformation rate:
 - higher in untreated (33-92%)
 - anterior commissure Risk of SCC is **92%**
 - In membranous cord only 17%

Invasive Squamous Cell Carcinoma management

Surgery

Endoscopic procedures:

Transoral laser microsurgery :

- Adequate laryngoscopic exposure of glottis is the basic technical requirement for TLM
- For procedures restricted to true cord:
 - the laser is kept at **continuous super pulse mode**
 - **low power** settings ranging from **1 to 3 W**

- **T1a (Unilateral) and T1b (Bilateral) Glottic tumor:**
- Small superficial lesions can be excised **en bloc**
- Resection of larger microinvasive tumors
 - Is formed in 2 more pieces
 - First incision pass through the middle of the tumor for depth assessm.
 - Prior resection partial or complete, of the ipsilateral false cord may be required.
 - Resection is commenced by this first incision through the center
 - followed by a further incision in normal tissue
 - lateral to the tumor
 - posterior to the tumor.
 - A deep margin of 1 to 3 mm recommended
 - mucosal margin of 1 to 2 mm recommended
- Bilateral tumors can be removed transorally in two sessions to prevent web formation

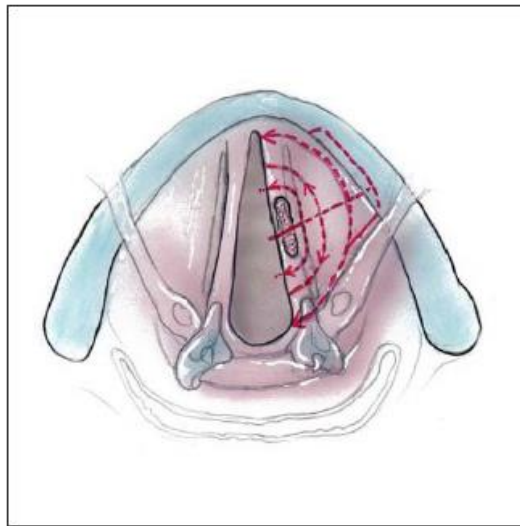


Figure 123.4 Transoral laser cordectomy for glottic tumors. Dotted lines indicate extended resections for lesions of increasing depth.

- ❖ **T2 Glottic tumor (Supraglottis and/or Subglottis Extension, and/or Impaired Vocal Cord Mobility):**
- T2 glottic cancers are amenable to complete excision with TLM irrespective of tumor extension into
 - Supraglottis
 - Subglottis
 - anterior commissure.
- The surgical principles applied are similar to laser surgery for other subsites
- For tumors extending to supraglottic larynx across the arytenoid cartilage or interarytenoid mucosa.
 - it is usually possible to preserve the arytenoid by peeling the tumor off the cartilage
 - if superficial. the exposed cartilage gets epithelialized after 3 to 4 weeks

❖ **Early Glottic Tumor with Anterior Commissure Involvement**

- **In T1a tumors** extending to the anterior commissure
 - a small tumor-free margin of the anterior part of contralateral cord should be included in the resection (sometimes causes Web formation)
- **In T1 b tumors**, the bilateral cord lesion is usually resected together with the anterior commissure
 - The dissection also continued anteriorly along the thyroid cartilage
 - with the help of frozen sections, it is ensured that the perichondrium toward the inside of the thyroid cartilage is free from the tumor.
 - Fibrin glue , sailastic sheet placed for 6 weeks are measures to prevent web formation
- **Postoperative care:**
 - **Follow up 2 weeks**
 - **Cessation of smoking and alcohol**
 - **Anti-reflux measures**
 - **Wound healing will take 4-6 weeks**
- **Complications:**
 - 2ndary bleeding (rare)
 - Post op laryngeal or tongue edema particularly T2 tumors or arytenoid involvement
 - IV steroid
 - Inhaled Vasoconstrictors
 - If airway obstruction > tracheostomy
 - Granulation tissue formation
 - Reflux makes it worse
 - If persisit more than 6-12 weeks or compromising voice Q
 - Surgical removal may be required
 - Laryngeal stenosis
 - rare, common with subgottis,arytenoid , intearytenoid)
 - common in radiated larynx
 - Poor voice quality always before Epithelisation
- **Phonosurgery can be offered after resections for poor voice improvement**
 - Fat injection etc,
 - VC medilization , Gore-tex etc,

Open conservative procedures:

- Laryngofissure and cordectomy
 - indicated for T1 lesions of vocal cord limited to the membranous cord when transoral resection is not possible

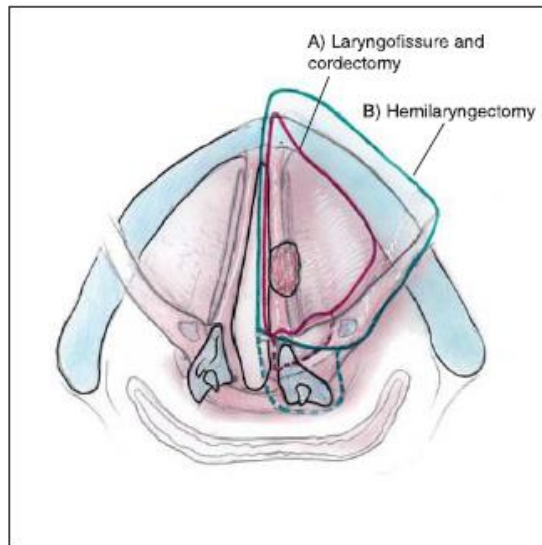


Figure 123.6 Extent of resection in laryngofissure and cordectomy (A) and hemilaryngectomy (B). Extensions (dotted line) in either procedures may include the vocal process of the arytenoids, if required.

- **Vertical Partial Laryngectomy. Hemilaryngectomy**
- involves resection of:
 - the ipsilateral thyroid cartilage
 - Arytenoid
 - true cord
 - false cord
 - underlying muscle
 - mucosa from the aryepiglottic fold to the upper border of the cricoid cartilage from the posterior to anterior midline.
- Indications include
 - T1 lesions with suspicion of deeply infiltrating tumor
 - 12 lesions of vocal cords with extension to arytenoids
- **Frontolateral vertical hemilaryngectomy**
- Indication
 - for T1 cord lesions approaching or extending to the anterior commissure and not more than 1 to 2 mm of the contralateral cord

- **Anterior commissure technique/ anterior frontal vertical partial laryngectomy**
- Indication
 - for T1 and T2 cord lesions with significant •hoiJeshoe• involvement of the opposite vocal cord
- Resection :
 - The vertical midline portion of the thyroid cartilage
 - inner perichondrium
 - true cord
 - false cord,
 - ventricle up to the vocal process bilaterally
- reconstruction:
 - performed by silastic or keel insertion to allow epithelialization on either side without web formation.
 - The keel is usually removed after 6 to 8 weeks

- **Reconstruction after partial laryngectomy**
- The glottis defect from open partial laryngectomy procedures may result in unsatisfactory voice outcomes.
- Resurfacing and reconstruction of the glottic defect enable better functional restoration
- Techniques:
 - strap muscle flaps.
 - Mucosal flaps
 - external thyroid perichondrium flap
 - a muscle-based cartilage flap
 - imbrication laryngoplasty are alternatives

- **post op care:**
 - nasogastric feeding
 - tracheostomy care
 - decanulation
 - wound healing complete
 - swallowing improves

- **Complications:**
- **Dysphagia**
- **aspiration**
- **Airway obstruction**
 - **Granulation tissue**
 - **Collapse of soft tissue into the airway**
 - **Managed by maintenance of tracheostomy**
- **Glottic stenosis**
 - **Specially in radiated cases**
- **Bleeding**
- **Fistula**

❖ **In general**

- **TLM has the lowest complication rate 1.2%**
 - **Vertical partial laryngectomy 9%**
 - **Frontolateral laryngectomy 13%**
 - **Open cordectomy 17%**
- **Supracricoid Laryngectomy**
 - SCL may be an effective procedure to conserve the larynx for T2 glottic cancers that involve
 - the anterior commissure
 - ventricle
 - bilateral vocal cords

OUTCOMES

- Early glottic tumors have a good prognosis and failure occurs mainly at the primary site
 - Poor outcome associated with:
 - T Stage
 - anterior commissure involvement
 - epidermal growth factor receptor
- multiple studies compared in T1 TLM and RT in
 - local control (89 vs 75%)
 - larynx preservation (100 vs 83%)
- TLM compared with open
 - Similar oncologic results
 - But TLM had :
 - Shorted clinical course
 - Lower complications
 - Best chance for salvage therapy
- Depending upon the stage and site of recurrence following TLM, options for treatment include
 - laser resection,
 - partial laryngectomy
 - RT
 - total laryngectomy
- poor prognostic factors for local recurrence:
 - the anterior commissure was the site of recurrent tumor in 54% of the cases
 - vocal muscle infiltration
 - subglottic involvement

- **lesions involving the anterior commissure**
- considered to be a poor prognosticator for local control in early glottis
- lower responses to RT, possibly due to underdosage.
 - The observed rates for local control in T1 carcinoma with anterior commissure extension vary from 57% to 89%
- Organ preservation rates
 - RT 80-85%
 - Open procedures 80-90%
- The oncologic outcomes for open surgery were found to be comparable with TLM but
- functionally less favorable in terms of
 - speech
 - swallowing
 - provisional tracheostomy

RADIOTHERAPY

- RT is employed as a treatment of choice for early glottic cancer at a number of centers
- The cure rates for T1 lesions range from 71% to 95%
- For T2 glottic cancer, the local control rates with RT vary from 50% to 80%
- Successful local control and laryngeal preservation with RT have been shown to be affected by several factors including:
 - high tumor volume/bulk
 - anterior commissure involvement
 - subglottic extension
- Avoidance of RT is recommended in the younger populations for prevention of morbidity from both acute and late RT-related complications.
 - patchy or diffuse mucosal coating
 - persistent atytenoid edema/polypoid lesion
 - xerostomia,
 - persistent laryngeal edema
 - cervical fibrosis
 - glottic stenosis
 - hypothyroidism,
 - chondronecrosis
 - radiation-induced malignancy.

Management of Radiation Recurrences

- Recurrences after failed radiation are often advanced at the time of diagnosis.
- A minority of radiation-recurrent glottis carcinomas can be salvaged by conservation surgery which is challenging due to:
 - persistent aspiration
 - pharyngocutaneous fistula
 - delayed decannulation,
 - laryngeal stenosis
- total laryngectomy may be commonly required for radiation failures

Advanced Laryngeal Cancer

- Over 40% of present with advanced-stage disease.
- Advanced-stage laryngeal cancer is defined as Stage III or IV disease which includes not only T3 and T4 tumors but also tumors with regional cervical metastasis (N1 to N3 disease)
- The ultimate goal is cure the patient of disease
- secondary goals:
 - preserve phonation
 - deglutition
 - maintain a safe airway
- The treatment of laryngeal cancer should be a multidisciplinary approach to provide the patient with every opportunity available and the best oncologic and functional outcome
- the 5-year survival for all stages of laryngeal cancer has worsened over the past 30 years.

Epidemiology

- The larynx is the second most common site of primary epithelial malignant tumors of the head and neck.
- Sub-sites for laryngeal malignancy:
 - Glottis (51%)
 - Supraglottis (32%)
 - Subglottis (2%)
- Male to female ratio of 3.6: 1.
- The ratio of male:female has decreased over the years likely secondary to the increased rate of tobacco use in females.

TABLE 124.1 OVERALL 5-YEAR SURVIVAL BY SUBSITE AND STAGE (8,9)			
	Supraglottis	Glottis	Subglottis
Stage III	50%–60%	50%–60%	43%
Stage IV	28%–53%	22%–52%	25%

Staging !!!!

TABLE
124.2 **STAGING LARYNGEAL CARCINOMA**

Primary tumor (T): TX: Primary tumor cannot be assessed T0: No evidence of primary tumor Tis: Carcinoma *in situ*

Supraglottis

- T1: Tumor limited to one subsite* of supraglottis with normal vocal cord mobility
- T2: Tumor invades mucosa of more than one adjacent subsite* of supraglottis or glottis or region outside the supraglottis (e.g., mucosa of base of tongue, vallecula, medial wall of pyriform sinus) without fixation of the larynx
- T3: Tumor limited to larynx with vocal cord fixation and/or invades any of the following: postcricoid area, preepiglottic tissues, paraglottic space, and/or minor thyroid cartilage erosion (e.g., inner cortex)
- T4a: Tumor invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of the neck including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus)
- T4b: Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Glottis

- T1: Tumor limited to the vocal cord(s) (may involve anterior or posterior commissure) with normal mobility
- T1a: Tumor limited to one vocal cord T1b: Tumor involves both vocal cords
- T2: Tumor extends to supraglottis and/or subglottis, and/or with impaired vocal cord mobility
- T3: Tumor limited to the larynx with vocal cord fixation and/or invades paraglottic space, and/or minor thyroid cartilage erosion (e.g., inner cortex)
- T4a: Tumor invades through the outer cortex of the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck, including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus)
- T4b: Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Subglottis

- T1: Tumor limited to the subglottis
- T2: Tumor extends to vocal cord(s) with normal or impaired mobility
- T3: Tumor limited to larynx with vocal cord fixation
- T4a: Tumor invades cricoid or thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck, including deep extrinsic muscles of the tongue, strap muscles, thyroid, or esophagus)
- T4b: Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Regional lymph nodes (N): NX: Regional lymph nodes cannot be assessed

- N0: No regional lymph node metastasis
- N1: Metastasis in a single ipsilateral lymph node, 3 cm or less in greatest dimension
- N2a: Metastasis in a single ipsilateral lymph node more than 3 cm but not more than 6 cm in greatest dimension
- N2b: Metastasis in multiple ipsilateral lymph nodes, not more than 6 cm in greatest dimension
- N2c: Metastasis in bilateral or contralateral lymph nodes, not more than 6 cm in greatest dimension
- N3: Metastasis in a lymph node more than 6 cm in greatest dimension

Distant metastasis (M): MX: Distant metastasis cannot be assessed

- M0: No distant metastasis
- M1: Distant metastasis

Stage

- Stage 0:** Tis, N0, M0
- Stage I:** T1, N0, M0
- Stage II:** T2, N0, M0
- Stage III:** T3, N0, M0 or T1, N1, M0 or T2, N1, M0 or T3, N1, M0
- Stage IVA:** T1, N2, M0 or T2, N2, M0 or T3, N2, M0 or T4a, N2, or less M0
- Stage IVB:** T4b, any N, M0 or any T, N3, M0
- Stage IVC:** Any T, any N, M1

*Subsites include the following: suprahypoid epiglottis, infrahypoid epiglottis, vestibular folds (false vocal cords), aryepiglottic folds, arytenoids (each fold is one subsite).

Adapted from AJCC. Larynx. In: Edge SB, Byrd DR, Compton CC, et al., eds. *AJCC Cancer Staging Manual*, 7th ed. New York: Springer, 2010:57–62.

Box 106-3. TUMOR/NODE/METASTASIS SYSTEM FOR THE LARYNX (EPITHELIAL MALIGNANCIES)**Primary Tumor (T)**

- T_x Primary tumor cannot be assessed
 T₀ No evidence of primary tumor
 T_{is} Carcinoma in situ

Supraglottis

- T1 Tumor limited to one subsite of supraglottis with normal vocal cord mobility
 T2 Tumor invades mucosa of more than one adjacent subsite of supraglottis or glottis or region outside the supraglottis (e.g., mucosa of base of tongue, vallecula, medial wall of piriform sinus) without fixation of the larynx
 T3 Tumor limited to larynx with vocal cord fixation and/or invasion of the postcricoid area, preepiglottic tissues, or paraglottic space with or without minor thyroid cartilage erosion (e.g., inner cortex)
 T4a Tumor invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea; soft tissues of neck, including deep extrinsic muscles of the tongue; strap muscles; thyroid; or esophagus)
 T4b Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Glottis

- T1 Tumor limited to the vocal cords; may involve anterior or posterior commissure; normal vocal cord mobility
 T1a Tumor limited to one vocal cord
 T1b Tumor involves both vocal cords
 T2 Tumor extends to supraglottis and/or subglottis or impairs vocal cord mobility
 T3 Tumor limited to the larynx with vocal cord fixation and/or invades paraglottic space and/or causes minor thyroid cartilage erosion (e.g., inner cortex)
 T4a Tumor invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea; soft tissues of neck, including deep extrinsic muscles of the tongue; strap muscles; thyroid; or esophagus)

- T4b Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Subglottis

- T1 Tumor limited to the subglottis
 T2 Tumor extends to vocal cords with normal or impaired mobility
 T3 Tumor limited to larynx with vocal cord fixation
 T4a Tumor invades cricoid or thyroid cartilage and/or invades tissues beyond larynx (e.g., trachea; soft tissues of neck, including deep extrinsic muscles of the tongue; strap muscles; thyroid; or esophagus)
 T4b Tumor invades prevertebral space, encases carotid artery, or invades mediastinal structures

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed
 N0 No regional lymph node metastasis
 N1 Metastasis in a single ipsilateral lymph node, <3 cm in greatest dimension
 N2 Metastasis in a single ipsilateral lymph node, >3 cm but not >6 cm in greatest dimension, or in multiple ipsilateral lymph nodes, none >6 cm in greatest dimension
 N2a Metastasis in a single ipsilateral lymph node, >3 cm but not >6 cm in greatest dimension
 N2b Metastasis in multiple ipsilateral lymph nodes, none >6 cm in greatest dimension
 N2c Metastasis in bilateral or contralateral lymph nodes, none >6 cm in greatest dimension
 N3 Metastasis in a lymph node >6 cm in greatest dimension

Distant Metastasis (M)

- MX Distant metastasis cannot be assessed
 M0 No distant metastasis
 M1 Distant metastasis

- While this system stages tumors based on size and extent of the tumor, other **bad important prognostic factors** are not included like:
 - Perineural invasion
 - angiolymphatic spread
 - extracapsular spread of nodal metastasis
 - histologic grade of the tumor.
 - Overexpression of mutant p53 protein (in some studies)

CERVICAL NODAL METASTASES

- Incidence of cervical metastases and levels involved depends of the Subsite of the laryngeal cancer

Supraglottic CA

- Rich lymphatic
- highest incidence of regional metastases clinical and occult
- incidence of occult mets in all T stages is 12-40%
- 10% of T1, 29% of T2, 38% of T3, and 57% of T4
- usually level II, III and IV and mostly bilateral if lesion is central
- Supraglottic N0 managed by Bilateral Selective neck dissection from level II to VI

- **Glottic ca**

- Low risk of cervical mets
- The nodes at risk of metastasis from glottic SCC are those in levels II, III, IV, and **VI**—the prelaryngeal, pretracheal, and paratracheal nodes
- Rarely bilateral or contralateral
- 0.1% of T1, 5% of T2, 18% of T3, and 32% of T4 tumors

- **Subglottic ca**

- Rare disease
- incidence of cervical metastasis is generally **low**, with a range of 4% to 27%
- nodes involved includes **Para-tracheal nodes (VI)**, and atypically III, IV and V

Distant Metastases

- Through hematogenous and lymphatics
- The most common site of metastases
 - Lungs
 - Liver
 - Skeletal system (less common)
- Mediastinum is the most common site of lymphatic spread
- Incidence depends on the subsite
 - Glottis 3-8%
 - Supraglottic 3-15%
- Clinical and pathologic factors associated with an increased risk of distant metastases include:
 - advanced-stage primary tumor (stage T4)
 - presence of cervical metastases, especially N2 and N3 disease
 - duration, level and extracapsular spread of cervical metastases
 - locoregional recurrence

Evaluation and diagnosis

History

symptoms depend on the location

- hoarseness (glottic carcinoma)
 - muffled/hot potato voice (supraglottic carcinoma)
 - dyspnea
 - dysphagia
 - otalgia (usually supraglottic)
 - cervical mass.
 - stridor (subglottic)
 - hemoptysis
 - sore throat
 - choking and recurrent pneumonia
-
- Constitutional symptoms include:
 - weight loss
 - fevers
 - chills
 - night sweats.
-
- Glottic tumors usually present earlier than supraglottic
 - Supraglottic tumors present with fewer symptoms in the early stage including pain, dysphagia and cervical mets therefore are usually diagnosed at a later stage.
 - Subglottic tumor typically presents as advanced stage with airway obstruction

Risk factors

- Smoking
 - +ve relation between quantity of cigarette and risk of developing laryngeal ca
 - more association with **glottic** cancer
- Alcohol (more association with **supraglottic**)
- Together they have synergistic effect (at least cessation for 20 years to return to baseline risk)
- HPV (subtypes 16 and 18, HPV 16 is the most common in laryngeal ca)
- GERD/laryngopharyngeal reflux
- Occupational exposures:
 - metal workers
 - construction workers (asbestos and textile processors)

- Delirium tremens and tobacco withdrawal can lead to a complicated and prolonged postoperative course and impaired healing

Examination

- Complete head and neck examination with skin and all subsite of erodigestive tract
- Cranial nerves exam
- Flexible endoscopy
 - check vocal cord mobility and position
 - SCC may appear as:
 - ulcerative lesion
 - exophytic lesion
 - sessile lesion
 - polypoid lesion
 - ventricular bulge/fullness
 - laryngocele (occult laryngeal ca causing the laryngocele)
 - use angled scope to view the ventricular system
- Systemic exam for co-morbidities
- Dentition should be assessed for caries and periodontal disease
- Video-stroboscopy (for subtle changes in VC movement, more helpful in early disease)
- Direct laryngobronchoscopy , esophagoscopy

Imaging

- CT, MRI, PET and chest X-ray
- Imaging can provide information regarding:
 - Cartilage invasion
 - extension into the pre-epiglottic and paraglottic spaces
 - Extralaryngeal spread of tumor
 - Cervical nodal disease
- CT scan
 - Standard imaging modality for laryngeal cancer
 - Sensitivity 67% to 81%
 - Specificity 47% to 81%
- MRI
 - For Cartilage invasion (diff T3 vs T4 , can change the management)
- Chest x-ray
 - Chest assessment is a must (for staging), if any abnormalities CT chest can be done (some centers do CT CAP from for staging)

- PET
 - For residual disease or recurrence mostly
 - Used sometimes for distant mets sometimes (sensitivity 86%, specificity 84%)
 - Can be used to assess for cervical mets (sensitivity 74-94%, specificity 82-100%)
- Ultrasound
 - Popular in Europe for nodal assessment

Consultations

- A multidisciplinary approach is used in the treatment of advanced laryngeal cancer. Prior to the start of treatment
- the multidisciplinary team usually include:
 - oncology
 - radio-oncology
 - Radiology
 - dental oncology
 - speech pathology
 - nutritional services
 - spiritual care

Differential diagnosis

- SSC
 - comprise 95-99% of laryngeal malignancy
 - There's multiple SCC variants
 - Most common is verrucous carcinoma (representing 1% to 3%)
 - pushing borders
 - high local recurrence
 - extremely low rate of distant metastasis.
 - classic appearance of exophytic white-gray fronds
- Neoplastic
 - Sarcomas (chondrosaroma)
 - Neuroendocrine tumors (mostly supraglottic)
 - Paragangliomas
 - Large Cell tumors and small cell tumors
 - Lymphoproliferative (mucosal-associated lymphoid tissue)
 - Minor salivary gland tumors
 - Minor salivary tumors (most commonly supraglottic)
 - adenoid cystic carcinoma,
 - mucoepidermoid carcinoma
 - acinic cell carcinoma,
 - adenocarcinoma
 - malignant mixed carcinoma

- Granulomatous disease
 - Tuberculosis
 - Wegener granulomatosis
 - Sarcoidosis
 - Histoplasmosis
 - Blastomycosis
 - may present with an exophytic or fungating mass resembling SCCA
- secondary spread from other sites or metastases
 - thyroid Ca invasion
 - metastases
 - breast
 - colon
 - renal (most common laryngeal mets)

Management

- Requires multimodality treatment with:
 - Surgery followed by radiation
 - Radiation and chemotherapy
- The goal of treatment for advanced laryngeal cancer is to cure
- secondary goals of preserving speech and swallowing function
- case should be discussed in tumor board

Radiation therapy

- The goal of radiation is tumor eradication while preserving normal tissue
- Intensity-modulated radiation therapy
 - allows the radiation beams to be more precisely focused to limit the effects of the radiation on surrounding tissue
- typical total treatment dose of radiation is 60 to 70 Gy given 5 per week over a period of 6 to 7 weeks
- Indications include T1, T2 and small T3
- adjuvant radiation indications:
 - advanced stage disease
 - positive margins,
 - extracapsular spread
 - perineural
 - angiolymphatic spread of tumor
 - multiple cervical lymph node involvement
 - subglottic extension of tumor

- **Complications of radiation**

Early (up to 6 weeks):

- mucositis
- odynophagia
- dysphagia
- skin breakdown
- loss of taste
- edema – might lead to the need of tracheostomy

Late:

- Fibrosis
- Xerostomia
- edema.
- loss of taste
- hypothyroidism
- laryngeal stenosis
- esophageal stenosis
- chondroradionecrosis (happens in 5%, arytenoid is most commonly involved)
- osteoradionecrosis (prevented by dental evaluation prior to radiation)

Chemotherapy

- The two most common methods of chemotherapy are induction and concomitant
- Can be used as Palliation
- Cisplatin and 5 fluorouracil are the most commonly used agents
- Concurrent Chemotherapy and Radiation Therapy offers better free survival and laryngectomy free survival rate in comparison to radiation and to induction chemo

Surgical management

Endoscopic Transoral Laser Microsurgery

- Designed for T1 and T2 tumors, now can be utilized for T3&4 supraglottic and glottic
- Less invasive
- Does not deconstruct the larynx
- Major complications very uncommon
- Frequent avoidance of tracheostomy
- Swallowing rehabilitation in shorter duration
- Need special surgical expertise and instruments

Open Conservation Laryngeal Surgery

- It is any procedure that maintains physiologic speech and swallowing without the need for permanent tracheostoma
- Goal is Maximum laryngeal function without compromising cure
- Proper patient selection is critical
- Needs at least on CA joint and one laryngeal valve (epiglottis, FVC, TVC)

Total laryngectomy

- Gold standard with best Oncologic outcome
- Indicated for
 - T3 and T4 advance laryngeal ca
 - Chemotherapy and radiation failures
 - Conservative surgery failures
- The procedure includes an en bloc resection of the larynx, including:
 - the hyoid bone, thyroid cartilage and cricoid cartilage
 - hemi-thyroidectomy, in the following conditions:
 - Palpable disease present
 - Subglottic carcinoma or Glottic with greater than 1cm subglottic extension
 - T4 glottic carcinoma (cartilage destruction)
 - pyriform sinus carcinoma
 - Positive Delphian node
 - Creation of permanent tracheostoma
 - Combined with this procedure may be resection of the pharyngeal wall and base of tongue
- The pharyngeal wall is closed upon itself to form a digestive path separate from respiration (should be assessed pre op (involvement of pyriform on CT one way to assess) , if involved pt will need a free flap to close the pharynx)
- key to success is maximizing QOL by maximizing alaryngeal function
- complete cricopharyngeal myotomy or pharyngeal plexus neurectomy (to improve **deglutition** and alaryngeal voicing)
- Voice restoration through:
 - tracheoesophageal speech (TEP)
 - primary (at time of resection)
 - secondary
 - esophageal speech (effort to produce with short duration)
 - artificial larynx (pneumatic vs electric)

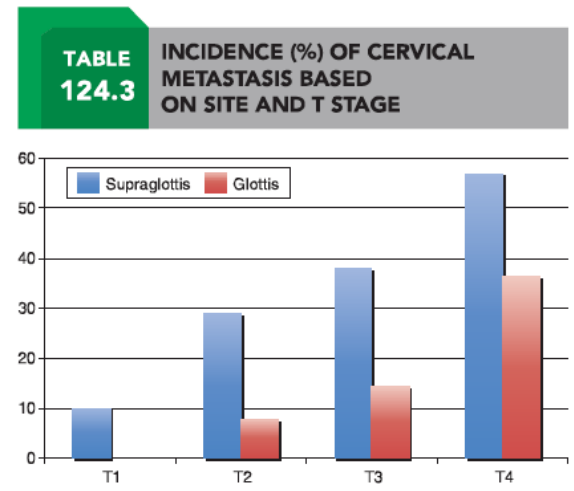
Salvage Total Laryngectomy

- Indicated for failure of organ preservation (chemorad)
- Higher surgical complication rate

Treatment of the Neck

- Neck treatment same as primary (surgery vs radiation)

- Generally accepted guidelines suggest that the neck should be treated if there is a 20-30% chance of regional metastasis
- Nodes at greatest risk Level II-IV + central compartment
- N0 and N1 = with single modality (Selective ND vs radiation depending on primary site treatment)
- N2-3 will require double modality (MRND and Rad)
- Supraglottic larynx has a rich lymphatic supply as compared to the glottis, and also tends to spread to bilateral cervical lymph nodes
- Glottic T3 and above to spread to cervical LNs
- Subglottic only T4 has chance of 20% or more to spread



Post laryngectomy Recurrence (stomal recurrence)

Regional, distant and local recurrence are common

Stoma recurrence

- Most common local recurrence location (2-25%)
- **Risk factors:**
 - Salvage laryngectomy
 - Advanced T and N stage
 - Subglottic involvement
 - Pre-operative tracheostomy (before laryngectomy by some time)
- **Sisson classification:**
 - Type I is localized to the superior aspect of the stoma without esophageal involvement.
 - Type II is localized to the superior aspect of the stoma with esophageal involvement.
 - Type III originates in the inferior aspect of the stoma with involvement of the superior mediastinum.
 - Type IV has extension laterally beneath the clavicle and into the superior mediastinum
- Type I and II best treated with salvage therapy
- Type III and IV are unresectable and for palliative measures (chemo-rad)

Surgical complications

- **Early General head and neck surgery complications:**
 - Hematoma
 - Infection
 - Skin flap necrosis
 - Aspiration pneumonia
 - Wound dehiscence
 - Hypocalcemia
 - Chyle leak/fistula (avoided by meticulous dissection in Level IV to avoid injuring the thoracic duct)
- **Late Complications:**
 - pharyngeal/esophageal stricture (dysphagia and failure of esophageal speech)
 - Tracheal stoma stenosis
 - Hypothyroidism
- **Pharyngocutaneous fistula** occurs as a result from failed surgical closure of the pharynx or larynx allowing contents to travel from the pharynx/larynx. through the neck to the skin.
- **The risk factors:**
 - poor nutritional status (albumin, hmg)
 - previous radiation therapy
 - preoperative chemotherapy radiation therapy
 - surgical technique
 - tracheostomy prior to primary management
- **Small fistula management:**
 - hold oral feeding
 - local wound care
- **Large fistula management:**
 - Debridement
 - closure of the fistula
 - regional tissue
 - free tissue transfer
 - role out residual or recurrence (in late occurring fistula and non responsive)

Post treatment function and quality of life

- Surgery, chemotherapy, radiotherapy, and combined modality treatments can have transient or permanent effects on
 - voice
 - swallowing
 - breathing
- such dysfunctions lead to:
 - depression
 - loss of social, cultural, and personal satisfaction with talking, eating, and drinking.
 - Loss of voice can also impact the patient's work and income
- Reasons for alaryngeal speech failure:
 - Inadequate air supply – Decreased respiratory support, improper stoma occlusion
 - Puncture site closure
 - Prosthesis failure – Position, patency, size, type, degradation, infection
 - Reflex pharyngeal constrictor spasm – Constrictors reflexively contract when dilated
 - Non vibrating pharyngoesophageal segment – Postradiation, reconstructed segment

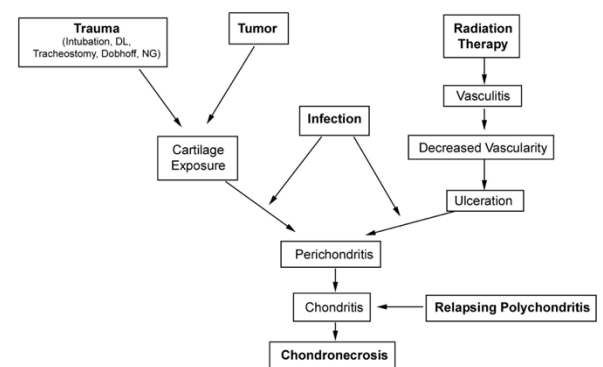
Emergencies

1. Airway obstruction

- Due to
 - As first presentation
 - After radiation therapy (edema)
- Best managed by avoidance of tracheostomy (increase risk of stomal recurrence)
- Flexible Endoscopic intubation better alternative then tumor deblinking
- If tracheostomy is a must should be made high up (stoma to be removed w/primary)

2. Chondroradionecrosis

- Radiation is the most common cause
- Incidence around 1-5%
- Risk factors:**
 - Smoking
 - tumor invasion
 - postoperative infection
 - trauma
 - radiation technique



- age, tumor grade and previous surgery are not risk factors
- they found 3 specific factors :
 - Radiation dose and duration
 - Presence of infection
 - General vascular condition of the patient

- **Chandler grading system for laryngeal radionecrosis**

- Grade I and II are common post-radiation changes and responds well to conservative management
- Grade III and IV more severe with less favorable outcome and considered radiation complications
- Larynx Framework becomes unstable altering airway size, mobility and structure

- **Presentation**

- dysphagia, odynophagia, hoarseness, stridor, dyspnea, respiratory obstruction, and recurrent aspiration.
- Odynophagia and neck pain and stiffness are late symptoms
- Cricoid joint fixation in either position can lead to airway obstruction and stridor or aspiration and breathy voice

- **Investigations (imaging)**

- No specific laboratory studies
- CT scan – evaluate cartilage and response to medical management
- MRI – better in differentiating fibrosis from inflammation or edema
- PET CT – 1995 proven to be most useful , in differentiating tumor form radionecrosis in > 80%
- MBS – to assess swallowing and ability to protect the airway

- **Laryngoscopy and biopsy**

- Post radiation therapy pt with persistent edema need multiple Biopsies to r/o recurrence or residual
- Mcguirt did biospies only for Grade VI or Grade III with +ve PET

- **Histology**

- The elastic fibers of the tunica media show degeneration, and hyalinization of smooth muscle fibers develops in postirradiated larynges, this is more marked in veins than in arteries
- Damage to the endothelium of the capillaries and lymphatics results in obliteration, atrophy, and fibrosis. In the presence of concomitant infection, overlying mucosal ulceration and purulent exudate is noted.

Conservative management:

- For grade I and II
- Humidification
- Voice restrains
- D/C of smoking
- Steroids
- Antibiotics (culture directed or Cipro)
- Hyper baric oxygen (HBO) in some studies

Surgical management

- Ranges from surgical debridement (failed GI & II after 10 days of medical management)
- Tracheostomy (securing airway in obstruction or aspirating larynx)
- Laryngectomy (dysfunctional larynx)

Significant morbidity and mortality, due to laryngeal instability and obstruction

- Recognition of perichondritis may prevent progression to chondritis then necrosis

3. Pharyngocutaneous fistula (puts the great vessels at risk)

Lymphoma

Introduction

Pathophysiology:

Lymphoproliferative disorder

Risks:

irradiation, Epstein-Barr virus (Burkitt's lymphoma), Congenital immunodeficiency disease, human immunodeficiency virus (HIV), immunosuppression, organic toxins (phenols, benzenes), human T-cell lymphotropic virus (HTLV-1, Tcell malignancies), immunological diseases (rheumatoid arthritis, celiac disease, Hashimoto's thyroiditis for thyroid lymphoma, Sjogren's for salivary gland lymphoma)

Clinical picture:

Nodal masses (may be painful), non-Hodgkin's lymphoma may present with extranodal masses as initial presentation (gastrointestinal tract most common extranodal site, also tonsils, paranasal cavity, base of tongue, salivary glands, thyroid, and orbit), fever, night sweats, weight loss, symptoms from mass effect from extranodal site (throat pain, nasal obstruction, exophthalmos, hoarseness)

Sites and presenting symptoms

- > 50% of extranodal in H&N is in Waldeyer ring
 - Tonsil swelling; dysphagia/odynophagia
 - Usually submucosal; SCC is ulcerative
- 33% occur in other extralymphatic sites
 - Presentation depends on site and mimics other Ca
- Waldeyer associated with GI in **3-11%** of cases; UGI or endoscopy
- 18% of extranodal H&N pts have bone marrow involvement; so a post iliac crest bone marrow biopsy is warranted initially
- If high grade, or if intermediate grade in paranasal sinuses, bone marrow, testes or paraspinal areas, do an LP

Evaluation

- 10% have H&N extranodal sites, including: Waldeyer ring; Nasal Cavity; paranasal sinuses; Oral cavity; larynx; salivary glands; thyroid; orbit
- Complete Physical Exam and History: including palpation of other nodal sites (supraclavicular, axillary, epitrochlear, inguinal, femoral) and spleen; inquire about fever, weight loss, and night sweats (Bsymptoms)
- Fine Needle Aspirate: may determine immunohistochemical subtype (B-cell or T-cell), DNA character (ploidy, phase), and RNA content (cannot differentiate follicular versus diffuse)
 - 10% have **discordant** lymphoma: two different types at different sites
 - 4% have **composite** lymphoma: two different types at one site
- Open Biopsy: usually required for definitive diagnosis prior to treatment, fresh sample required for immunochemistry
- Labs: complete blood count, blood urea nitrogen, lactate dehydrogenase levels, α_2 -microglobulin, liver transaminases

TABLE 115.1

HEAD AND NECK LYMPHOMAS—HISTOLOGIC CLASSIFICATION

Working Formulation (9)	WHO Equivalents (10)
Low grade	
Small lymphocytic	B-cell small lymphocytic or extranodal marginal zone
Follicular small cleaved cell	Follicular lymphoma, grade 1
Follicular mixed	Follicular lymphoma, grade 2
Intermediate grade	
Follicular large cell	Follicular lymphoma, grade 3
Diffuse small cleaved cell	Mantle cell, lymphoplasmacytic
Diffuse mixed	Diffuse large B cell, peripheral T cell
	Extranodal NK/T cell
Diffuse large cell	Diffuse large B cell, peripheral T cell
	Extranodal NK/T cell
High grade	
Immunoblastic	Diffuse large B cell, peripheral T cell
	Extranodal NK/T cell
Lymphoblastic	Lymphoblastic
Small non-cleaved cell	Burkitt lymphoma/Burkitt-like

- Head & neck lymphoma grades:
 - 12% low grade
 - 72% intermediate grade
 - 16% high grade
- Head & neck lymphoma types:
 - **Diffuse large cell most common**
 - Most H&N lymphoma have B-cell marker

Staging:

Bone scan (gallium); CT/ MRI of neck, chest, abdomen and pelvis; bone marrow biopsy

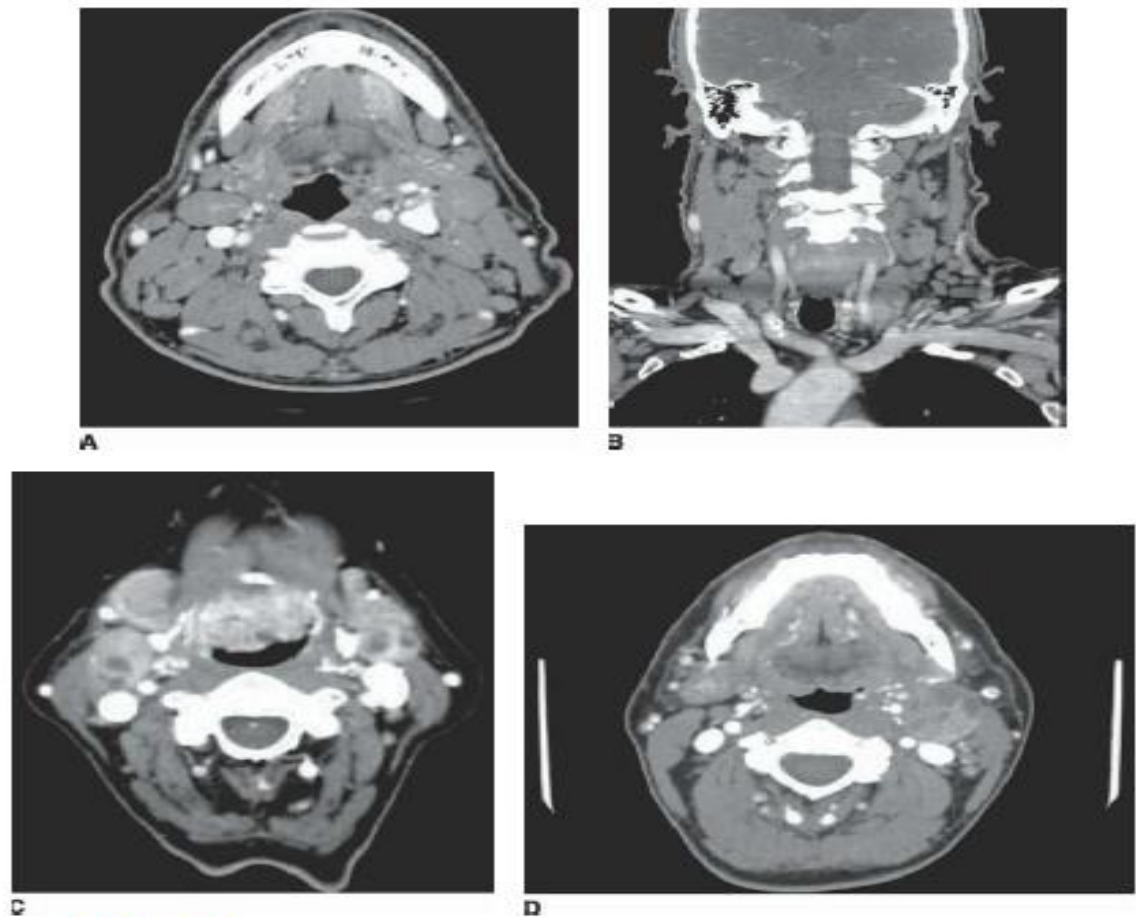


Figure 128.2 Contrast-enhanced (A) axial and (B) coronal CT images from patient with B-cell lymphoma, demonstrating multiple enlarged lymph nodes with poor contrast enhancement and uniform appearance. In contrast, lymph nodes with metastatic SCC demonstrate (C) heterogeneous enhancement as well as (D) central necrosis.

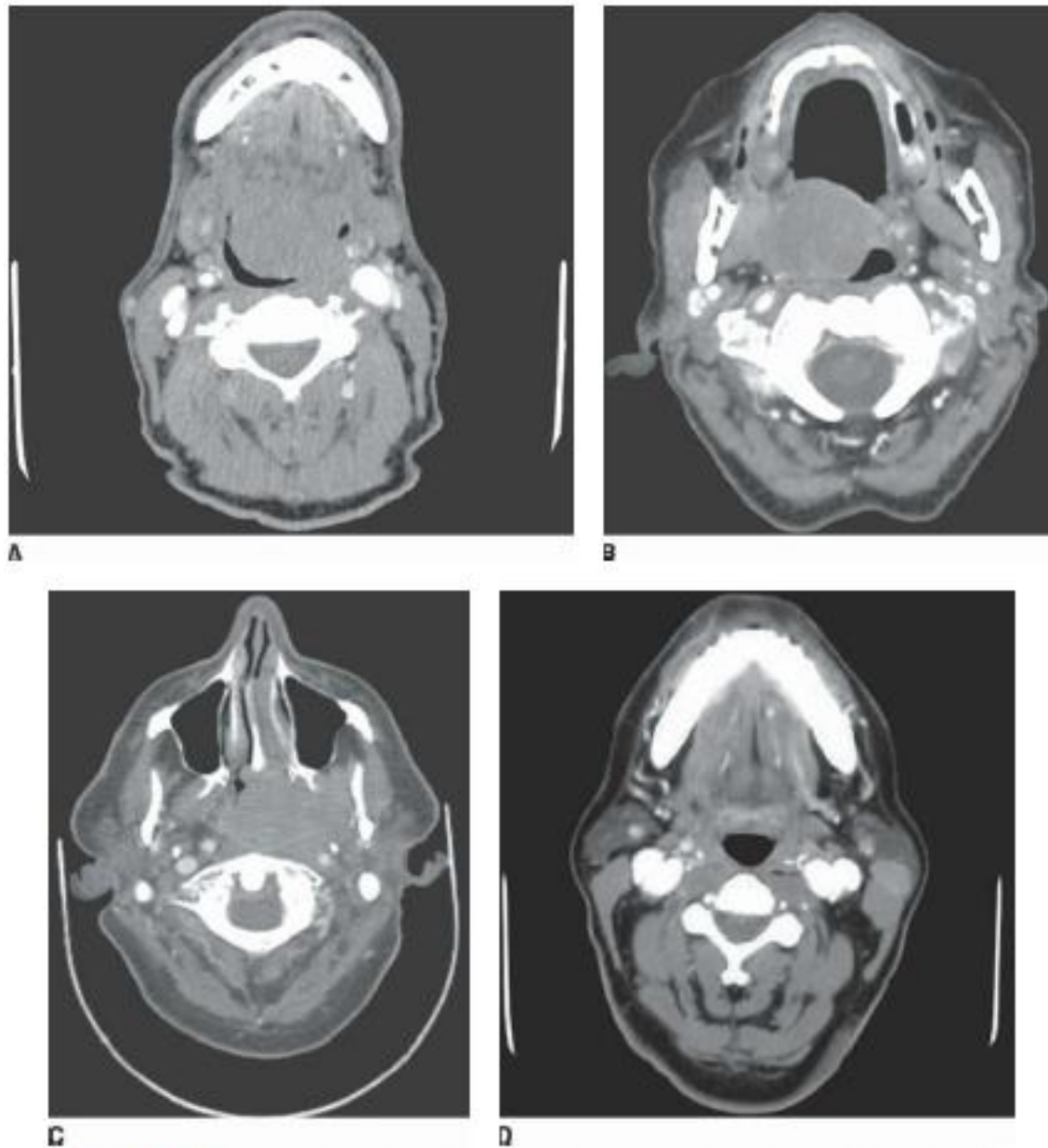


Figure 128.3 Contrast-enhanced axial CT images from patients with lymphoma of the (A) base of tongue, (B) left tonsil, (C) nasopharynx, (D) the left parotid gland.

Staging Laparotomy:

may be considered for aggressive Hodgkin's lymphoma (not NHL); includes inspection of abdominal cavity, liver biopsy, nodal biopsy, and oophoropexy

May also consider serum protein electrophoresis, upper gastrointestinal series

TABLE 115.2
HEAD AND NECK LYMPHOMAS—STAGING EVALUATION

- Pathologic review
- Complete physical examination, indirect laryngoscopy
- Complete blood count, liver function tests, LDH
- HIV test (positive risk factors)
- Chest radiograph
- CT or MRI of head and neck
- CT of chest, abdomen, and pelvis
- Upper gastrointestinal series with small bowel follow-through (Walden ring)
- PET scan
- Lumbar puncture (paranasal sinus)
- Bone marrow biopsy

CT, computed tomography; LDH, lactate dehydrogenase; MRI, magnetic resonance imaging; PET, positron emission tomography.

TABLE 31.2
ANN ARBOR CLASSIFICATION FOR LYMPHOMA

Ann Arbor Classification for Lymphoma

Stage I	Involvement of a single lymph node region or single extralymphatic site
Stage II	Involvement of two or more lymph node regions on the same side of diaphragm, localized contiguous involvement of only one extralymphatic site and lymph node region
Stage III	Lymph node involvement on both sides of the diaphragm
Stage IV	Disseminated involvement of one or more extralymphatic organs with or without lymph node involvement

Hodgkin's Lymphoma

- Bimodal incidence curve (teenagers and middle-aged adults)
- Spreads via contiguous lymph nodes
- **Reed-Sternberg Cells (R-S cells)**: binucleated, malignant giant cells, eosinophilic inclusions

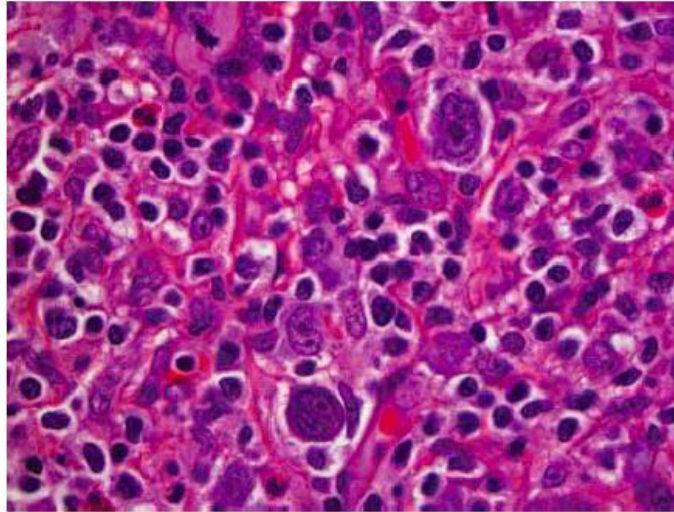


Figure 128.1 Reed-Sternberg cell in a lymph node involved by HL.

Treatment:

Early stages may consider single-modality radiation therapy; advanced disease requires multimodality therapy with radiation and chemotherapy: MOP (P) (**m**echlorethamine, **o**ncovin (vincristin), **p**rednisone, **p**rocarbazine)

ABVD (**a**driamycin, **b**elomycin, **v**inblastine, **D**TIC)

No surgical intervention except to obtain biopsy specimen

Prognosis:

Prognosis dependent on staging, overall >75% 5-year survival; better prognosis for lymphocytic predominance types; worse prognosis with presence of Reed-Sternberg cells, class B symptoms, and higher staging

Symptom Classes:

A class (asymptomatic),

B class (weight loss, fever, night sweats)

Rye Modification of the Lukes and Butler Classification

- 1. Lymphocytic Predominance:** predominantly lymphocytic cells with few R-S cells, typically presents at an early stage, better prognosis
- 2. Nodular Sclerosis:** more common in women, typically presents at an early stage
- 3. Mixed Cellularity:** most common type
- 4. Lymphocytic Depleted:** rare, few lymphocytes with many R-S cells, usually advanced, poor prognosis



Non-Hodgkin's Lymphoma (NHL)

Most are B-cells or null cells (except lymphoblastic lymphomas)

Treatment:

Typically treated with CHOP (**c**ytosan, **d**oxorubicin, **v**incristine, **p**rednisone) chemotherapy followed by radiation to involved sites, low-grade B-cell lymphomas and MALT syndrome may be observed.

General Management Principles FOR NHL

- Involves rads, chemo, immunotherapy or a combination of them
- Rads: 30-40 Gy for low grade; 50 Gy for intermediate grade; typical field includes Waldeyer ring and all H&N nodal regions (Waldeyer field); Mantle field includes H&N, mediastinal and axillary nodes
- Chemotherapy:
 - CHOP: cyclophosphamide 750 mg/m² IV day 1; doxorubicin 50 mg/m² IV day 1; vincristine 1.4 mg/m² IV day 1; prednisone 50 mg/m² PO days 1-5; repeat every 21 days
 - CVP: cyclophosphamide 400 mg/m² PO days 1-5; vincristine 1.4 mg/m² IV day 1; prednisone 100 mg/m² PO days 1-5; repeat every 21 days
- Immunotherapy:
 - Rituximab (R): 375 mg/m² IV per wk for 4 wks (monoclonal Ab against CD20 B-cell Ag)

Prognosis:

Grading important; better prognosis associated with smaller and nodular types; poor prognosis associated with larger or diffuse types, higher grading, and central nervous system or bone marrow involvement

International prognostic index (IPI) for NHL includes:

- Age
- Performance status
- LDH level
- Number of extranodal sites
- Stage

International prognostic indicator

	Better	Worse
• age	< 60	> 60
• stage	I-II	III-IV
• extranodal sites	<1	> 1
• LDH	<1x N	> 1x normal
• ECOG	0,1	> 2

ECOG PERFORMANCE STATUS*

Grade	ECOG
0	Fully active, able to carry on all pre-disease performance without restriction
1	Restricted in physically strenuous activity but ambulatory and able to carry out work of a light or sedentary nature, e.g., light house work, office work
2	Ambulatory and capable of all selfcare but unable to carry out any work activities. Up and about more than 50% of waking hours
3	Capable of only limited selfcare, confined to bed or chair more than 50% of waking hours
4	Completely disabled. Cannot carry on any selfcare. Totally confined to bed or chair
5	Dead

Mucosa-Associated Lymphoid Tissue (MALT):

Low-grade, B-cell tumors that may occur in the gastrointestinal tract, conjunctiva, lungs, and thyroid, associated with *Helicobacter pylori*, may respond to antibiotics and anti-ulcer medications



Rappaport Classification and Working Formulation

Low Grade:

Typically B-cell lymphomas, better survival

- Small Lymphocytic: may be found in the orbit, elderly, consistent with chronic lymphocytic leukemia
- Follicular (Small Cleaved Cells): common, indolent course
- Follicular (Mixed Cells): small and large cell components

Intermediate Grade:

Most common type in the head and neck, potentially curable with chemotherapy

- Follicular (Large Cell)
- Diffuse (Small Cleaved Cell)
- Diffuse (Mixed Cells): small and large cell components
- Diffuse (Large): may involve sanctuary sites (CNS, testes), rapidly fatal, most common NHL type in the head and neck

High Grade:

Usually presents in the head and neck as part of a more extensive disease, extremely aggressive, requires immediate chemotherapy

- Large Cell
- Lymphoblastic: usually of T-cell origin
- Burkitt's (Small Noncleaved Cell): maxilla or mandible
Involvement (EBV associated, common in Africa), abdominal organ involvement (HIV associated) its involve:

African Disease:

- Common in maxilla, may affect mandible
- Present with loss of dentation, facial distortion, proptosis, and trismus

North American disease:

- Present with abdominal
- 25% have H&N involvement [most commonly asymptomatic adenopathy]
[nasopharynx and Tonsils have been reported]

What is lethal midline granuloma?

- T-cell lymphoma

What is Richter's transformation?

- Low grade lymphoma transforms to aggressive high grade. Average survival < 1 yr

Generalized Tx Strategy for H&N Lymphoma

<u>Histology</u>	<u>Stage</u>	<u>Treatment Options</u>
Low Grade	I, II	Involved-field XRT
	III, IV	CVP, chlorambucil, R, observe

Tumour Lysis Syndrome

Findings: Hyperuricemia; Hyperkalemia; Hyperphosphatemia; Hypocalcemia; ARF Failure; Can occur within hours of starting chemo and can result in death from cardiac arrhythmias

Prophylactic Tx: IV hydration; Allopurinol; Alkalin-ization of urine for first 24-48 hrs of Tx

Hodgkin Disease v. Non-Hodgkin Lymphoma

HD

- Often single axial group of nodes
- Orderly spread
- Mesenteric and Waldeyers involvement rare
- Extranodal uncommon
- Bimodal peak in teens and in 60s
- Reed-Sternberg cells as classic histo finding

NHL

- Frequently multiple peripheral nodes
- No contiguous spread
- Mesenteric and Waldeyers involvement common
- Extranodal common
- ↑ risk with age

Treatment of Specific Subtypes

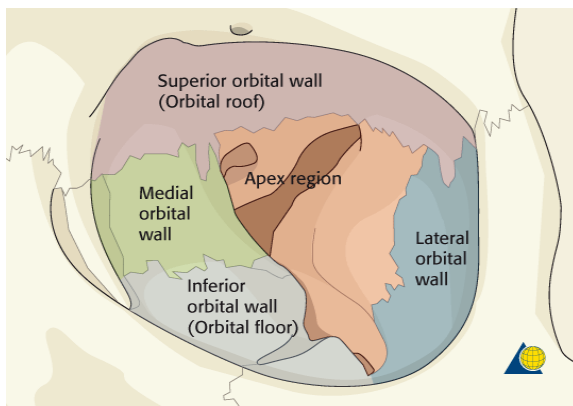
- Low-grade lymphomas, Stage I or II
 - 60% disease-free survival at 10 yrs
 - Involved or extended field rads; no evidence for chemo
- Low-grade lymphomas, Stage III or IV
 - 20-30% disease-free survival at 10 yrs
 - See table above
- Intermediate-grade lymphomas, Stage I or II
 - 80-100% disease-free survival at 5 yrs for stage I
- Intermediate-grade lymphomas, Stage III or IV
 - 30-60% disease-free survival at 5 yrs
 - RCHOP is the standard for diffuse large B-cell lymphoma
 - T and NK cell lymphomas have terrible prognosis
- High-grade lymphomas
 - Intensive combination chemotherapy required
 - At risk for tumour lysis syndrome (see text box)

What must you follow-up Hodgkin's patients for very closely?

- Watch out for post-treatment hypothyroidism – occurs in 2/3rds of patients

Orbital Tumors

- Anatomy of the Orbit
- a pyramid-shaped bony recess in the anterior part of the skull, lined by periosteum called the periorbital fascia
- The bony orbit is composed of
 - frontal
 - zygomatic
 - sphenoid
 - maxilla
 - lacrimal
 - palatine
 - ethmoid bones

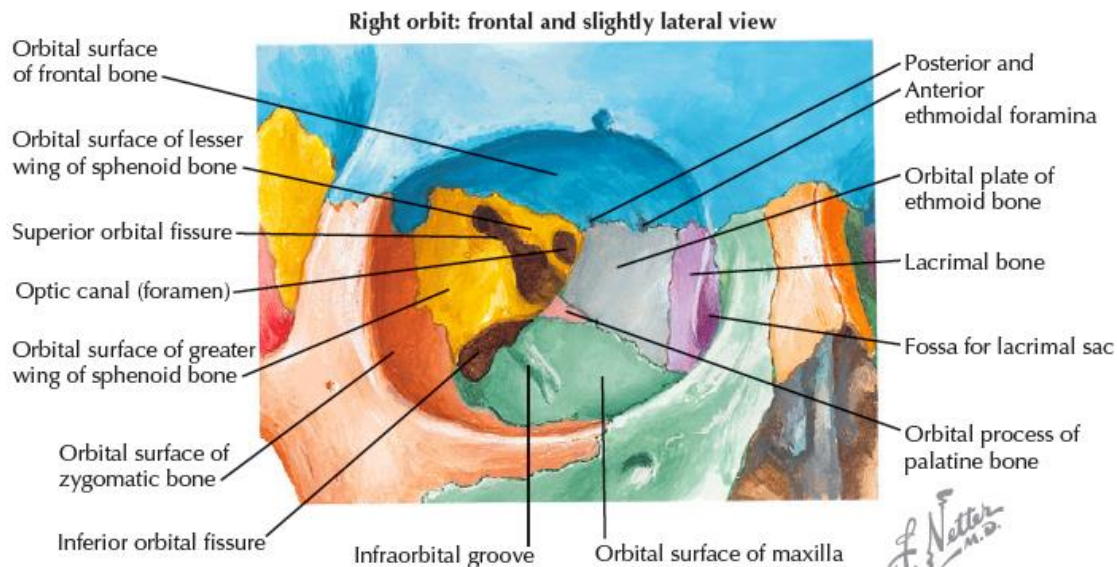


WALLS OF THE ORBIT

Superior	Frontal (orbital plate) Lesser wing of the sphenoid
Inferior	Maxilla Zygomatic Palatine (orbital process)
Medial	Ethmoid (lamina papyracea) Lacrimal Sphenoid Maxilla
Lateral	Zygomatic Greater wing of the sphenoid

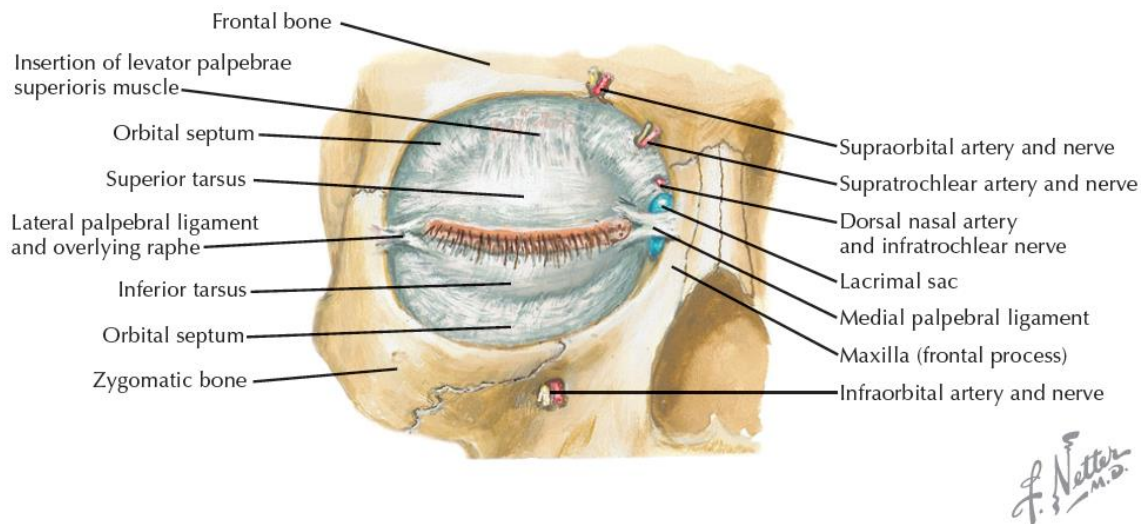
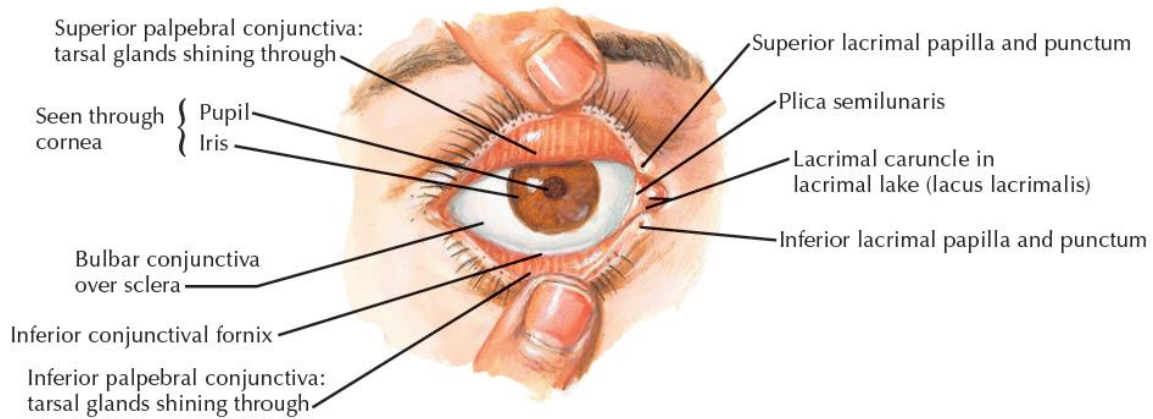
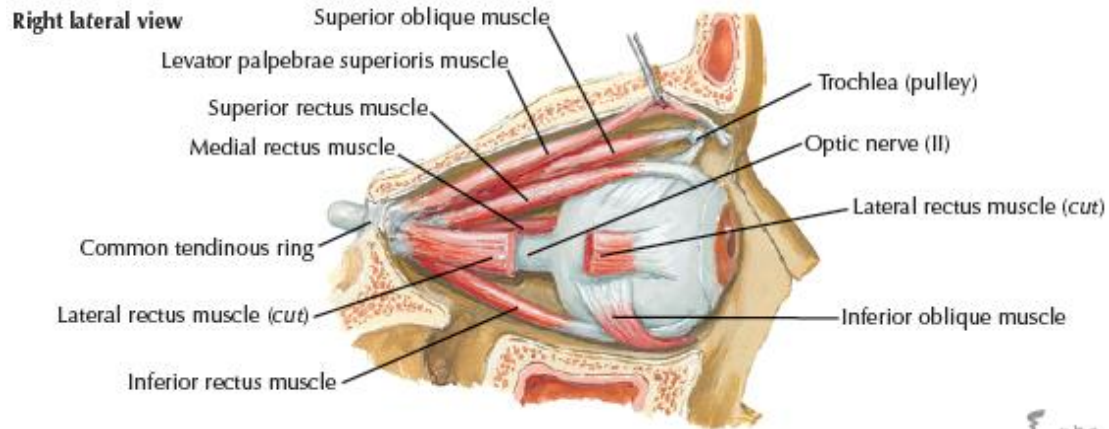
- The orbital walls lie adjacent to all four paranasal sinuses:
 - The orbital **apex** is located just anterolateral to the **sphenoid** sinus
 - The medial **lamina papyracea** borders the **ethmoid** sinuses
 - The orbital **floor** is adjacent to the **maxillary** sinus
 - The **roof** of the orbit often forms the floor of the **frontal** sinus
-
- The orbit is intimately associated with the skull base.
 - the orbital roof borders the anterior cranial fossa
 - the orbital apex borders the middle cranial fossa. (communicates via the superior orbital fissure and optic canal)
 - the orbit communicates with the pterygopalatine and infratemporal fossae via the inferior orbital fissure

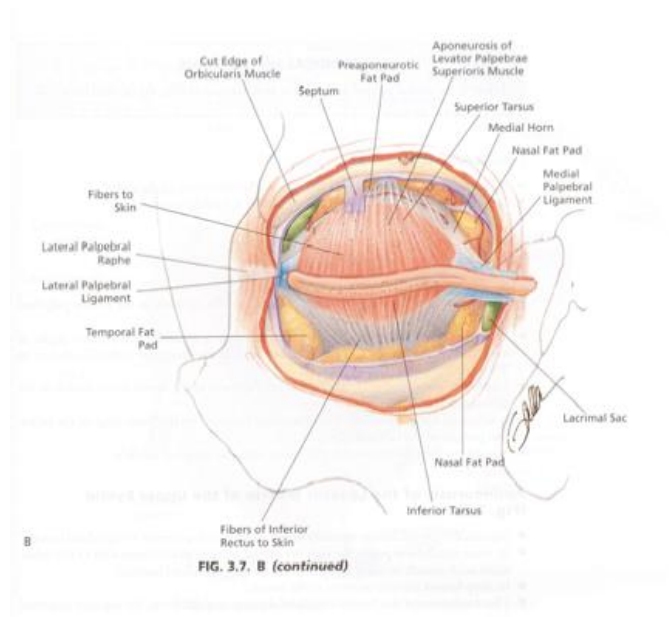
Opening	Bony Boundaries	Structures Passing through Opening
Optic foramen	Lesser wing of the sphenoid	Optic n. Ophthalmic a.
Superior orbital fissure	Greater wing of the sphenoid Lesser wing of the sphenoid	Lacrimal branch of the trigeminal n.'s ophthalmic division Frontal branch of the trigeminal n.'s ophthalmic division Nasociliary branch of the trigeminal n.'s ophthalmic division Oculomotor n. Trochlear n. Abducens n. Superior ophthalmic v. Inferior ophthalmic v.
Inferior orbital fissure	Greater wing of the sphenoid Maxilla	Infraorbital n. and vessels Zygomatic n. Branch of inferior ophthalmic v. that connects to the pterygoid plexus
Supraorbital foramen	Frontal	Supraorbital n. and vessels Supratrochlear n. and vessels
Infraorbital groove and canal	Maxilla	Infraorbital n. and vessels
Zygomatic foramen (1 or 2 openings)	Zygomatic	Branches of the zygomatic
Nasolacrimal canal	Lacrimal	Nasolacrimal duct
Anterior ethmoidal foramen	Ethmoid Frontal	Anterior ethmoidal n. and vessels
Posterior ethmoidal foramen	Ethmoid Frontal	Posterior ethmoidal n. and vessels



- The annulus of Zinn separates the superior orbital fissure into two compartments at the origin of the lateral rectus muscle.
- The **superolateral** portion transmits the lacrimal, frontal branches of V1 and trochlear nerves, as well as the superior ophthalmic vein and occasionally the orbital branch of the middle meningeal artery.
- The **medial** portion outside of the optic canal transmits the upper and lower division of the oculomotor nerve (CN III), the nasociliary branch of CN V1, and the abducens nerve (CN VI).

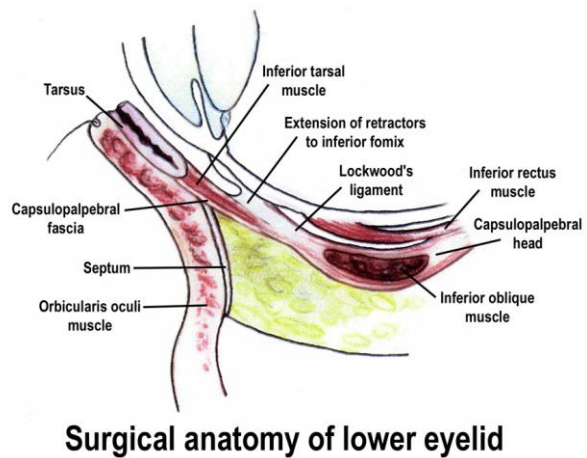
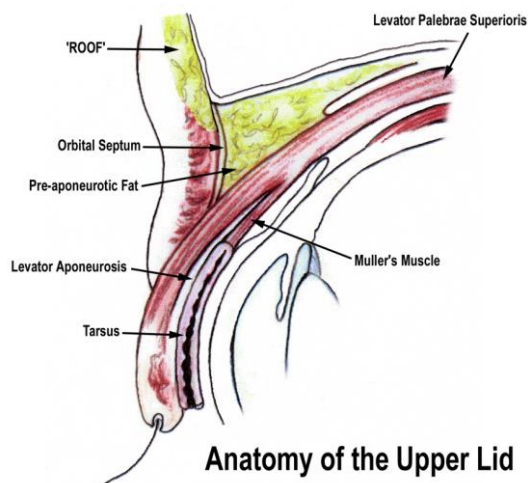
EXTRINSIC MUSCLES OF THE EYE					
Muscle	Origin	Insertion	Actions on Eye	Nerve	Comment
Superior rectus	Common tendinous ring on sphenoid (Annulus of Zinn)	Superior sclera	Elevation Adduction Intorsion	Superior division of the oculomotor	A check ligament attaches it to the levator palpebrae superioris m. to help elevate the upper eyelid
Inferior rectus		Inferior sclera	Depression Adduction Extorsion	Inferior division of the oculomotor	A check ligament attaches it to the inferior tarsal plate to help depress the lower eyelid
Medial rectus		Medial sclera	Adduction		The most medial of the extraocular muscles
Lateral rectus		Lateral sclera	Abduction	Abducens	Impaired in abducens n. palsy
Superior oblique	Body of the sphenoid	Superior portion of the posterolateral sclera	Depression Abduction Intorsion	Trochlear	Tendon passes through the trochlea, a fibrocartilaginous pulley
Inferior oblique	Maxilla (lateral to the lacrimal groove)	Inferior portion of the posterolateral sclera	Elevation Abduction Extorsion	Inferior division of the oculomotor	Only extraocular muscle that attaches to the maxilla
ASSOCIATED EXTRINSIC MUSCLE OF THE ORBIT					
Muscle	Origin	Insertion	Actions	Nerve	Comment
Levator palpebrae superioris	Roof of the orbit	Skin of the upper eyelid	Raises the upper eyelid	Superior division of the oculomotor Sympathetic fibers to the smooth muscle	Opposed by the palpebral part of the orbicularis oculi m. There are smooth muscle fibers which insert into the superior tarsal plate which are innervated by sympathetic fibers Lesions of the sympathetics will lead to a ptosis, or drooping of the upper eyelid



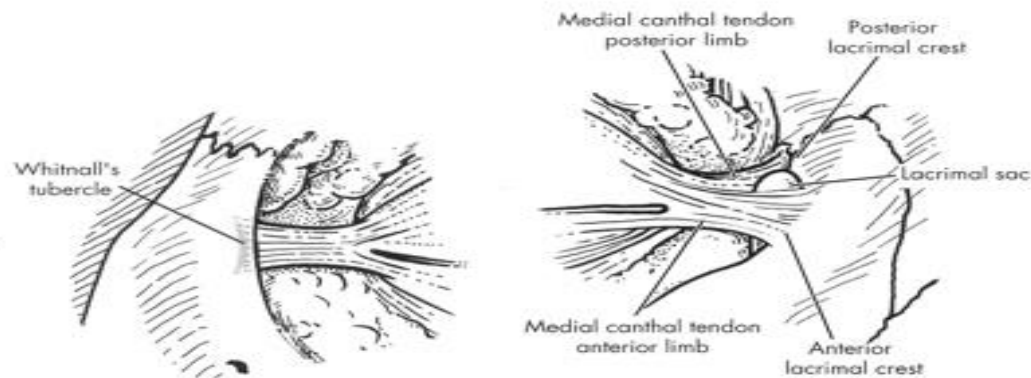
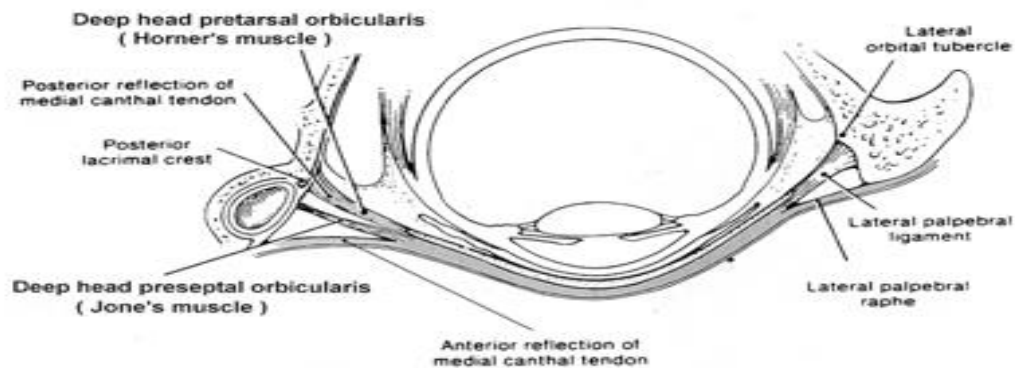


Orbital septum:

- Also known as palpebral fascia.
- Extends from margins of orbit where it is continuous with the periosteum of the orbit (periorbital) and periosteum of external surface of skull.
- The orbital septum separates the connective tissue spaces of the eyelids from the deeper structures, including the fascial sheaths of the ocular muscles and orbital fat.
- The superior orbital septum overlaps the anterior surface of the superior tarsus and attaches to it on both sides.
- The inferior orbital septum fuses with the connective tissue of the anterior part of the inferior tarsus

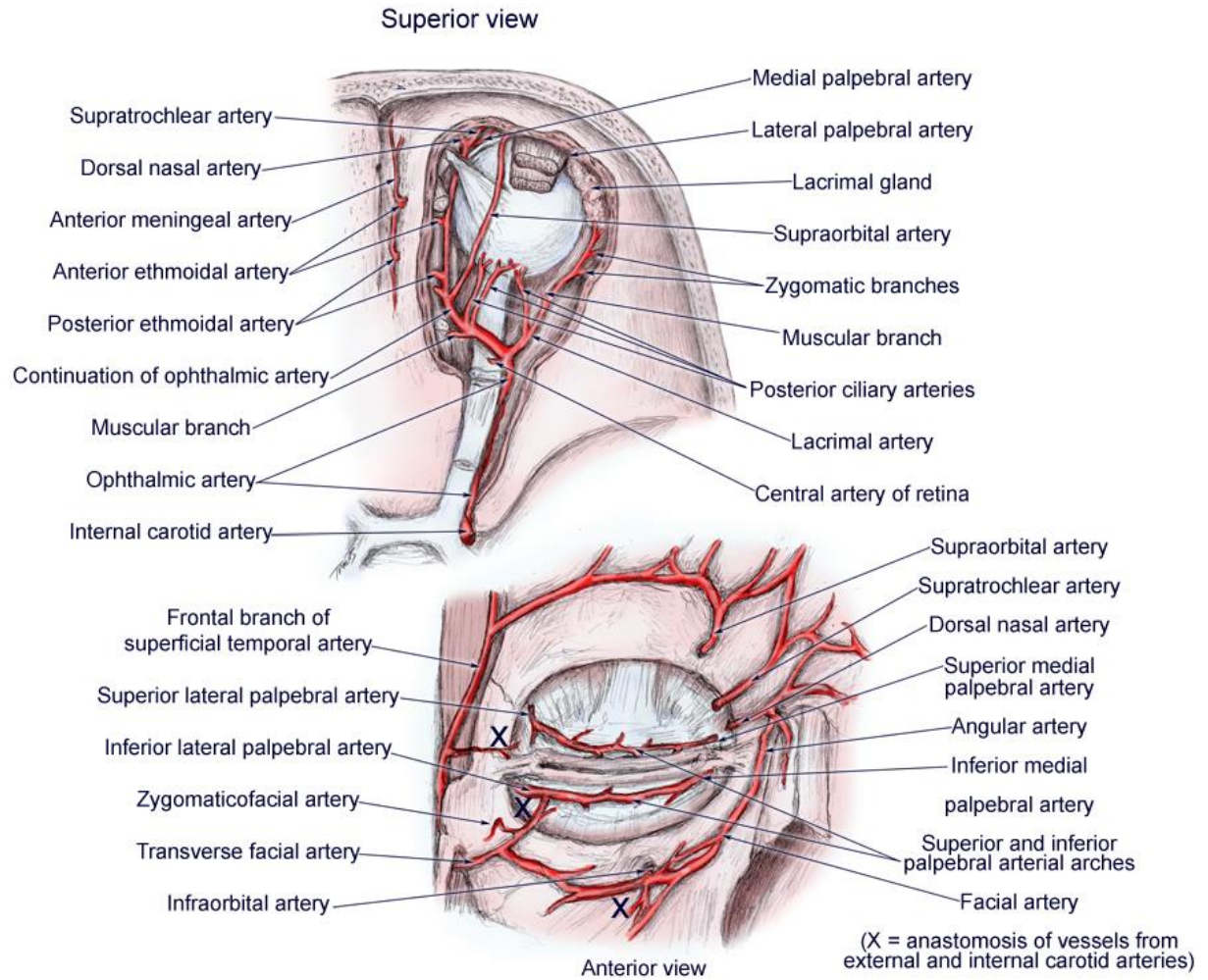


- Eyelid skin is the thinnest skin of the body and lacks subcutaneous fat.
- In the upper lid, there is a medial and central fat pad
- In the lower lid, there is a medial, central, and lateral fat pad.
- The medial canthal ligament attaches to the medial angle of both tarsi. The medial canthal tendon has two heads, superficial and deep.
- The lateral palpebral ligament lies posterior to both the orbital septum and the lateral horn of the levator palpebral aponeurosis
- The canthal tendons help maintain the horizontal stability of the eyelids



ORBITAL INNERVATION	
Orbital Innervation	Description
Sensory	2 Major Types Vision (special somatic afferent) via the optic n. General sensation (general somatic afferent) via the ophthalmic (and some maxillary) division of the trigeminal n.
Motor	2 Major Types Motor to the extraocular muscles (general somatic efferent) via the oculomotor, trochlear, and abducens nn. Autonomics to the intrinsic muscles of the eye (general visceral efferent) via: • Parasympathetics associated with the ciliary ganglion • Sympathetics associated with the superior cervical ganglion
Cranial nn.	5 cranial nerves provide innervation to the orbit: • Optic—vision • Oculomotor—extraocular motor and autonomics to the intrinsic muscles of the eye • Trochlear—extraocular motor • Trigeminal—general sensation • Abducens—extraocular motor

- **The arterial supply** of the orbit arises primarily from the internal carotid artery, which gives rise to the ophthalmic artery.
- The ophthalmic artery lies inferolateral to the optic nerve in the optic canal
- It gives off small branches including
- Branches to the optic nerve and retina (central retinal artery, posterior ciliary arteries), extraocular muscles,
- and lacrimal gland.
- Anteriorly it forms anastomoses with the external carotid arterial system to provide a rich blood
- supply to the eyelids and face.
- **Venous blood** is drained primarily by the superior ophthalmic vein, which originates in the superomedial orbit and travels posterolaterally to enter the cavernous sinus via the superior orbital fissure.



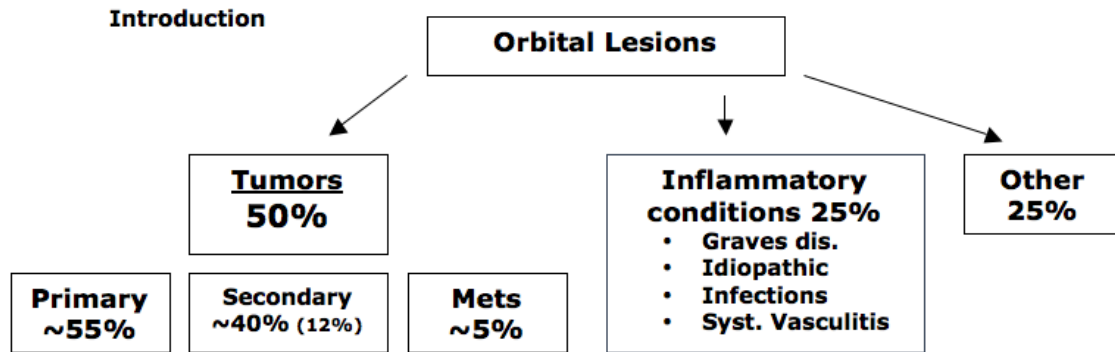
Differential diagnosis of orbital mass

Pediatric:

- Cystic
 - dermoid
 - epidermoid
 - teratoma
- Vascular
 - Capillary hemangioma (most common vascular in peds)
 - Lymphangioma
- Mesenchymal → Rhabdomyosarcoma (most common malignant in peds)
- Others
 - Optic nerve glioma
 - Fibrous dysplasia
- Primary globe (retinoblastoma)

Adult

- Primary
 - Hemangioma
 - Non Hodgkin's lymphoma
 - Inflammatory tumors
 - Meningioma
 - Optic nerve glioma
- secondary
 - Mucocele
 - SCC
 - Meningioma
 - Vascular malformation
 - BCC



- **Secondary** tumors (invasion from adjacent structures) account for 12% of orbital lesions and 33-45% of orbital tumours
- **Metastasia** to the orbit make up 2-8% of orbital tumours
Particularly **adenocarcinoma** of the lung and breast
- Pediatric orbital tumors
 - Most pediatric orbital tumours are **benign**; dermoids, vascular tumor (hemangioma and lymphangioma), optic nerve glioma & inflammatory tumors
 - Between 10% and 30% of childhood orbital tumors are malignant "**Rhabdomyosarcoma**" is the most common; other malignant are metastatic neuroblastoma, leukemia (granulocytic sarcoma), Ewing sarcoma, extension of retinoblastoma from the globe

Signs and symptoms

- **Proptosis** is the most common presentation for any lesion
 - **Axial** proptosis → **intraconal** space
 - **Nonaxial** proptosis → an **extraconal** position
- EOM disturbance and diplopia also common
- Other signs and symptoms include vision changes, conjunctival injection and chemosis, pain, and pupillary changes.

Vascular Tumors

- They represent 10% of orbital tumours; most are benign
- **Cavernous hemangioma**
 - **Most common adult primary intraconal neoplasm**
 - F>M; 3rd-5th decade; 88% intraconal
 - Presentation: **slowly** progressive, painless unilateral proptosis
 - CT and MR are diagnostic in 95% of cases
 - Discrete round/oval mass; no inflammation or infiltration of surrounding structures
 - Enhances with IV contrast
 - Path: multiple, large, blood-filled channels lined with flattened endothelial cells
 - Tx: complete excision of lesion with capsule; prognosis is excellent; recurrence rare
- **Capillary hemangioma**
 - **Most common vascular tumour of orbit and periocular tissues in infancy and childhood**; 1-2% of all newborns have one somewhere on their body (**strawberry mark on eyelid**)
 - Natural history: not present at birth; discovered within a month; grows for 12-18/12; stabilizes; involutes (50% gone at age 5; 70% at age 7; 90% at age 9)
 - Causes morbidity; present during critical developmental period
 - Sx: Deprivational amblyopia; strabismus; optic atrophy; corneal exposure
 - Amblyopia: reduced acuity in structurally normal eye due to incomplete visual system development which occurs during the 1st few years of life
 - Enlarge with valsalva
 - Dx usually clinical; CT/MR help assess orbital involvement
 - Diffusely infiltrating non-encapsulated mass
 - Conforms to surrounding structures; enhances
 - Expansion, but no erosion
 - Tx: let them regress unless symptomatic; otherwise, systemic steroids, intralesional steroid injection, or debulking surgery



Figure 130.2 Orbital cavernous hemangioma. T1 with contrast axial MRI showing a well-circumscribed enhancing orbital lesion.

- **Lymphangioma:**

- Benign; arise from pluripotent mesodermal tissue
- Usually identified in first decade
- **Can be superficial (conjunctiva and lids), deep or both**
- Visual prognosis better for superficial lesions
- Orbital presentation: **sudden** (spontaneous bleed – chocolate cyst) or fluctuate in size (cycle with URTIs) proptosis; no change with Valsalva
- Path: multiple, variably sized, thin-walled, endothelial lined vascular channels
- Dx usually clinical; CT/MR useful
 - Multiple, contiguous, dilated cystic spaces
 - No enhancement, but may have rim enhancement
- Tx: **infiltrative**, so complete resection impossible; may need debulking and cyst drainage to protect vision

Hematopoietic Tumors

- **Lymphoid tumours**

- Spectrum: benign reactive lymphoid hyperplasia v. atypical lymphoid hyperplasia v. malignant orbital lymphoma
- 4-13% of all orbital tumours despite **no lymphatics in orbit**
- 50% of orbital lymphoma is localized to orbit at time of Dx;
- 33% of malignant orbital lymphoma will develop systemic lymphoma over time (NHL)
- Almost exclusively in adults; F>M; 6th to 7th decade
- Presentation for most: slowly progressive, painless proptosis; lesions usually in anterior orbit and palpable along rim; visible **"salmon patch"** under conjunctiva
- Imaging
 - CT: **putty-like molding** around orbital structures; homogeneous with clear borders; mostly **extraconal**
 - 40% have lacrimal gland involvement
- Path: most are **monoclonal B-cell origin**
- Mx: biopsy for Dx; full-body survey by oncologist; for isolated lymphoid hyperplasia, low-dose rads curative; higher doses for atypical and lymphoma; systemic disease gets chemo/rads

- **Leukemia**

- 10% of pts with systemic leukemia have cells in orbit
- More in children (mean age is 7y); may be bilateral
- more with myelogenous than lymphocytic
- Presentation: **rapid onset proptosis** with inflammation
- **Granulocytic sarcoma**: collection of immature **myeloid cells**; high in **myeloperoxidase** giving it a green colour (chloroma); can be seen in AML or CML; can precede systemic disease (myelod sarcoma)
- CT scan shows a large irregular mass with possibly bony erosion.
- Tx: systemic chemo even if no evidence of systemic disease as all with ultimately get it; may use rads for orbital lesions
- Prognosis: most die within 18/12 of diagnosis of chloroma

- **Langerhans cell histiocytosis (histiocytosis X)**

- Rare; proliferation of Langerhans cells (epidermal Macrophage)
- The exact histogenesis is unclear, but it is thought to result from mononuclear phagocyte dysregulation.
- Three types
 - 1- eosinophilic granuloma (most common and benign)
 - 2- Hand-Schüller-Christian disease (intermediate)
 - 3- Letterer-Siwe disease (disseminated and most aggressive)
- One or more destructive (circumscribed lytic) bony lesions of skull, ribs, sternum, long bones, vertebrae or pelvis
soft tissue granulomas also seen if disseminated
- Only 11 cases reported in the orbit (hosam)
- CT: scan shows punched-out, osteolytic bone lesion with sharp borders.
- Tx: excision of isolated lesion can be curative; close follow-up

Neural Tumours

- **Schwannoma (neurilemoma)**

- Benign, non-invasive peripheral nerve tumour; unilat; solitary; painless
- 1% of all orbital tumours; mostly adults; slowly progressive
- More often intraconal; CT and MR useful
 - Well-circumscribed elongated ovoid mass; displaces
 - Often homogeneous; cystic degen or calcified if old
 - Small degree of enhancement with contrast
 - MR: hypo on T1 and hyper on T2
- Histo: encapsulated; Schwann cells
 - **Antoni A**: orderly palisading of **elongated spindle** cells
 - **Antoni B**: loose myxoid; **ovoid or stellate** cells
- Tx: complete surgical excision with capsule; great prognosis

- **Optic nerve glioma**

- **5th most common primary orbital tumour; 2nd in kids (15%)**
- **Fibrillary astrocyte** is the responsible cell; associated with **NF** in 18-50% of cases
- Presentation: axial proptosis and visual loss; optic nerve head edema present early; kids may not report visual loss
 - CT: lobular, isodense, homogeneous enlargement of optic nerve; nerve not distinguishable on coronal
 - MR (study of choice): assess optic canal and chiasm; hypo on T1 and hyper on T2;
- Tx: if confined to orbit with good vision and little progression, then simply observe; if growing or extending into optic canal, then resect whole nerve from globe to chiasm

- **Orbital meningioma**

- **Only 30% are primary; rest secondary from cranium**
- Primaries arise from optic nerve (dura); 3% of all orbital tumours
- F>M; 4th to 7th decade; rare in children
- Most common complain is altered vision; proptosis most common sign (85% have it)
- Also get disk pallor and nerve head edema; diplopia; HeadAche
 - CT: focal or exophytic mass anywhere along nerve, or fusiform enlargement of nerve; homogeneous; **infiltrative**; enhancing; normal nerve seen on coronal
 - MR: study of choice for intracranial involvement
- Tx: can't shell it off; follow for intracranial extension; rads may be helpful; can resect it if not worried about vision



Inflammatory Tumors

- **Idiopathic orbital inflammation (AKA orbital pseudotumour)**
 - **10% of all primary orbital tumours**
 - Diffuse or localized inflammation; **adults** mostly affected
 - Presentation: dull orbital pain; worse with eye movement; onset over a few days; proptosis in 67-85%
 - Vision loss in 20% (scleral, uvea or optic nerve inflammation)
 - CT: affected structures enlarged; shaggy margins; enhancing; often see multiple tissues affected
 - If EOM involved, tendon is too (tendon is not involved with thyroid orbitopathy)
 - MR: isointense to muscle on T1; iso-hyperintense to fat on T2; enhancing
 - Bx: vasculitis not seen; diffuse cellular infiltrate common
 - Tx: most respond to oral steroids; some need cyclosporine or cyclophosphamide or rads; try steroids before Bx if suspected
- **Orbital vasculitis**
 - Rare; associated with systemic vasculitis syndromes (Wegener; polyarteritis nodosa, giant cell arteritis, SLE)
 - Painful proptosis in addition to systemic symptoms
 - Bx shows vasculitic changes

Thyroid-Associated Orbitopathy

Although not considered an orbital lesion

The most common cause of unilateral and bilateral proptosis in adults.

The thyroid status:

- 90% are hyperthyroid,
- 6% are euthyroid
- 3% have Hashimoto thyroiditis
- 1% is hypothyroid.

Smoking exacerbates the disease

Pathogenesis: involves circulating autoantibodies to thyroid-stimulating hormone receptor (TSH-R) and insulin-like growth factor receptor (IGF-IR), which cross-react with orbital fibroblasts and stimulate them to differentiate into adipocytes and produce glycosaminoglycans, with resulting increase in orbital soft tissue volume.

Sx: The most common presenting sign is eyelid retraction

Proptosis, lagophthalmos, conjunctival chemosis and hyperemia over the rectus muscle insertions, extraocular muscle restriction, and optic neuropathy may also be observed

CT scan demonstrates the classic finding of fusiform extraocular muscle enlargement spares the tendon

Tx: establish euthyroid (anti thyroid Rx, radio ablation, total thyroidectomy)

A transient worsening of TAO may be noted after radioactive iodine ablation and total thyroidectomy, likely due to the release of TSH-R antigens.

Corticosteroids and/or orbital radiation therapy

Definitive treatment most often requires orbital decompression

Cysts

- **Dermoid and simple epithelial cyst most common primary orbital cysts** (near suture line trapped ectoderm)
- **Mucocele is most common secondary cyst**
- Dermoids
 - Superficial: **frontozygomatic**; **frontoethmoid**; **frontolacrimal**; diagnosed by age 5
 - Deep: **sphenozygomatic** or **sphenoethmoid**; diagnosed as adult
 - CT: round; well-defined; thin-walled; non-enhancing lumen; calcium deposits in wall for deep ones
 - Path: stratified squamous epithelium lined; appendages
 - Tx: small ones followed; large ones excised
- Simple epidermal cysts
 - Much less common; present in adults
 - Preferred Tx is surgical excision

Lacrimal Gland Tumours

- Most masses are due to idiopathic orbital inflammatory disease
- Staph and strep acute infections occur, as do viral (herpes, mumps)
- Of non-inflammatory/infectious lacrimal masses, 50% are lymphoproliferative and 50% are epithelial neoplasms
- On imaging, lymphoproliferative are smooth elongation and epithelial neoplasm are globular
- Lymphoproliferative (benign reactive lymphoid hyperplasia, atypical lymphoid hyperplasia, malignant lymphoma and leukemias)
- 50% of epithelial lesions are pleiomorphic adenoma; the other half are carcinomas (Adenoid cystic, malignant mixed and Adenocarcinoma)
- If Pleiomorphic Adenoma suspected, cut it out without doing an excisional Bx; high recurrence rate with incomplete removal
- Adenoid cystic carcinoma is most common epithelial malignancy of the lacrimal gland; mortality > 50%; Tx involves complete excision; RadTx

Mesenchymal Tumours

- **Rhabdomyosarcoma**
 - **Most common soft tissue malignancy and primary malignant orbital tumour in children**; 66% of orbital ones are **embryonal**
 - **Types (embryonal, pleomorphic, alveolar)**
 - Mean age 6-7 yrs; 90% < 13 yrs; M mildly > F
 - Presentation: progressive, painless, unilat proptosis; days-wks
 - 33% also have ptosis and palpable mass
 - May have redness and swelling; not warm like cellulitis
 - CT: poorly defined homogeneous orbital mass
 - Tx: biopsy (take as much of tumour as is safe); assess for disseminated disease → BM Bx, CXR, LFTs, CBC and LP; **chemo/rads**
 - 5-yr survival ~ 90% (< 35% if disseminated)



- **Fibrous dysplasia**
 - Uncommon; hamartomatous; monostotic (1 site of skeleton) Vs. polyostotic (multiple)
 - Replacement of cortical bone with cellular fibrous stroma containing multiple nests of immature woven bone
 - Monostotic most common for orbit; orbital roof most common
 - Polyostotic more likely to affect sternum, ribs and vertebrae
 - Presentation: adolescent with proptosis and/or asymmetry
 - CT: **bone thickened and deformed**; multiple lucent lytic spaces and sclerotic "ground-glass" spaces
 - Dx: clinical usually adequate; may need Bx
 - Tx: Observe; resection and recon if significant cosmetic deformity or vision loss (compression)
- **Osteoma**
 - Benign bony tumour; can be orbital or in sinuses
 - Most orbital ones are from the frontal bone; unilat; solitary
 - **Gardner syndrome**: multiple lesions with colon cancer
 - Presentation: sinusitis; dull orbital pain; facial asymmetry; globe displacement; proptosis
 - CT or plain film: smooth, round lobulated mass with density similar to that of bone; sessile or pedunculated
 - Tx: smaller ones are followed; larger ones causing deformity or loss of vision are resected; excellent prognosis
- **Osteosarcoma**
 - Highly malignant; usually affects long bones; rarely in face
 - Gene for osteosarcoma and retinoblastoma recently found
 - Presentation: **rapid progression** of proptosis and globe displacement; orbital pain; numbness; vision loss
 - CT: bony mass; enhances; lytic and sclerotic spaces
 - Tx: resection and chemo; extremely poor prognosis

Solitary Fibrous Tumor

It is a relatively new pathologic entity/classification

It is an uncommon spindle cell tumor that rarely occur in the orbit

The mean age of onset is 40 years, and patients present with proptosis and diplopia

A palpable mass with or without pain has also been observed

They are hypervascular lesions

CT scan findings of a well-circumscribed mass with moderate contrast enhancement and possible calcification and bony remodeling

MRI, these lesions appear as an intermediate intensity mass on T1-weighted images with moderate contrast enhancement

They share histologic similarities to fibrous histiocytoma and hemangiopericytoma lesions (immunohistochemical studies can differentiate)

Treatment includes en bloc tumor resection, although residual tumor has been found to remain stable for some time.

Secondary Tumours

- **44% of all orbital tumours**
- Mucoceles
 - 8% of orbital tumours; > 80% arise from frontal or ethmoid
- Sinus neoplasms
 - 40% can have orbital involvement; **SCC** most common
 - Maxillary sinus is site of origin in 80% of cases
- Cranial vault
 - Meningioma most common; most arise from sphenoid ridge
 - Odd findings: temporalis fossa fullness; boggy ipsi lower lid
- Eyelid
 - **BCC** (90%), sebaceous cell and SCC most common
 - Look for skin fixation to orbital rim
- Conjunctiva
 - SCC and melanoma most common; rare to see orbital invasion
- Globe
 - Choroidal melanoma most likely to spread to orbit (8-10% have extrascleral spread at time of enucleation)
 - Retinoblastoma most common intraocular tumour of kids; exenteration in < 10% of cases

Metastatic Tumours

- **8% of all orbital tumours;**
- In 25%, the orbital tumor is the presenting manifestation of an occult cancer
- **Breast**, by far most common; 50% of mets in orbit
- Others: **lung; prostate; GI; renal cell; thyroid; melanoma**
- Most common metastatic tumour in children is **neuroblastoma**
- Interesting finding: enophthalmos in 25% of cases

**TABLE
130.2****COMMON METASTATIC
ORBITAL LESIONS**

- Breast (adenocarcinoma): 29%–59%
- Lung (bronchogenic carcinoma): 8%–15%
- Prostate: 3%–10%
- Cutaneous melanoma: 5%–8%
- Renal cell carcinoma: 5%–7%
- Carcinoid tumors: 4–5%
- Gastrointestinal: 2%–5%
- Sarcomas: 1%–2%
- Thyroid carcinoma: <1%

**TABLE
130.1****OVERALL PREVALENCE OF ORBITAL LESIONS**

Type of Lesion	% of Orbital Tumors	Type of Lesion	% of Orbital Tumors
Metastatic:	5%–8%	Cysts:	6%–12%
Breast	2%–4%	Dermoid	2%–7%
Lung	<1%	Epithelial inclusion cysts	1%
Prostate	<1%		
Neural:	9%–17%	Mesenchymal:	6%–11%
Glioma	1%–4%	Rhabdomyosarcoma	2%–3%
Meningioma	4%–10%	Fibrous dysplasia	<1%
Schwannoma	1%	Solitary fibrous tumor	<1%–2%
		Osteomas	<1%
Vascular:	3%–10%	Sarcomas	<1%–2.6%
Capillary hemangioma	1%–3%		
Cavernous hemangioma	3%–6%	Lacrimal:	6%–9%
Lymphangioma	1%–4%	Lymphoid	2%–4%
		Epithelial	4%
Inflammatory:	7%–11%	Secondary:	11%–44%
Pseudotumor	3%–8%	Mucocele	3%–8%
Orbital vasculitis	1%–2%	Skin cancers	4%–11%
Thyroid-associated orbitopathy	1%–6%	Meningiomas	1%–7%
Hematopoietic:	9%–10%	Pediatric:	n/a*
Leukemia	<1%	Rhabdomyosarcoma	
Lymphoid	2%–9%	Neuroblastoma	
Langerhans cell histiocytosis	<1%	Granulocytic sarcoma	

*Percent estimates of all orbital tumors in adults.

Types of surgical approaches:

1. Transorbital approach
2. Extraorbital approach
 - Depend on: location, size, extent, goal of surgery And to lesser extent the pathology of the lesion

Transorbital:

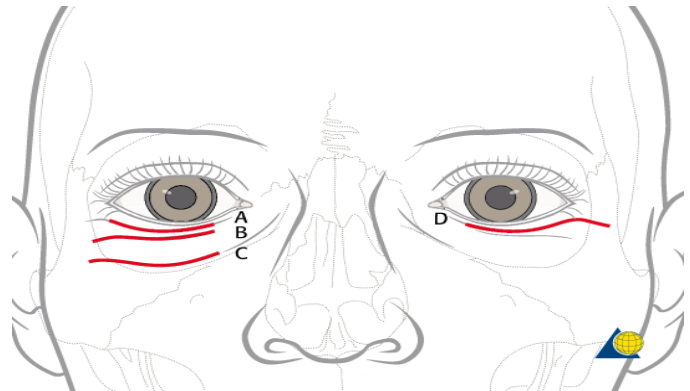
1. the anterior orbitotomy without osteotomy (superior [eyelid, supraorbital, or subbrow incision] or inferior [transconjunctival, subciliary, or lower-eyelid incision])
Or with osteotomy of the superior orbital rim (for large lesions)
2. lateral orbitotomy
3. medial orbitotomy
4. combination of the lateral and medial orbitotomies.

A subciliary

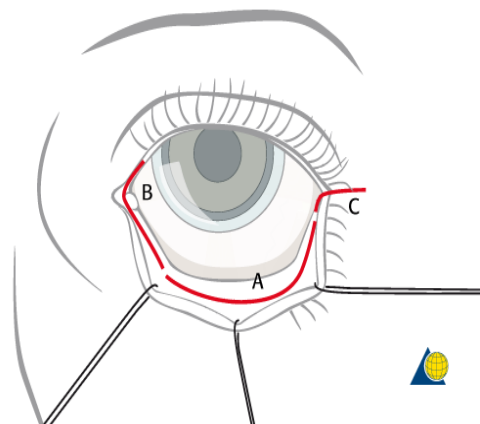
D sub ciliary with lateral extension

B sub tarsal

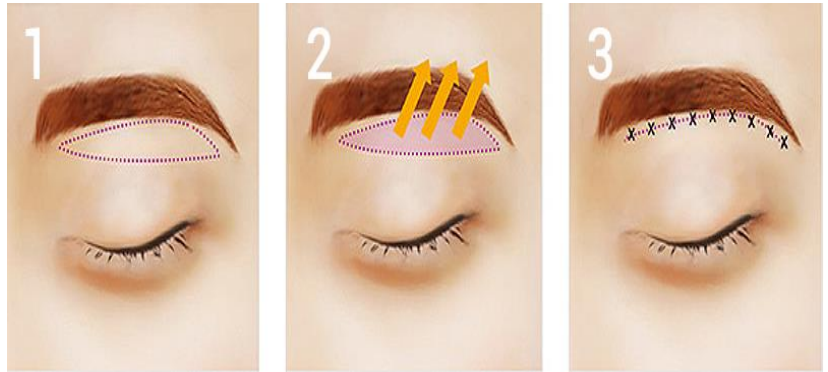
C infra orbital



Transconjunctival

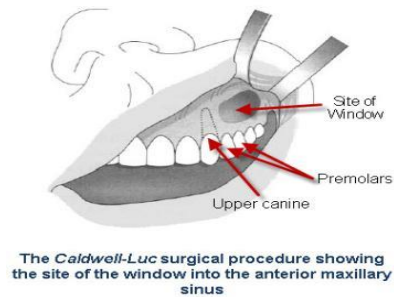
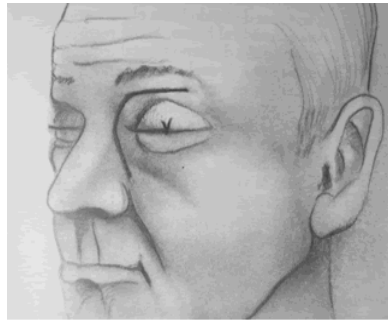


Sub-brow



Extraorbital:

1. The frontotemporal approach with or without orbital osteotomy (neuro) Orbital apex, superior orbital fissure or intracranial extension
2. The inferior orbital approach (Caldwell Luc)
3. lateral rhinotomy (Lynch)
4. Nasal endoscopic approach



Types of eye removal

Evisceration – removal of the iris, cornea, and internal eye contents, but with the **sclera and attached extraocular muscles left behind**

Enucleation – removal of eyeball, but the **eyelids** and adjacent structures of the eye socket remaining

Exenteration – **removal of the contents of the eye socket**, including the eyeball, fat, muscles, and other adjacent structures of the eye. The eyelids may also be removed in cases of cutaneous cancers and unrelenting infection

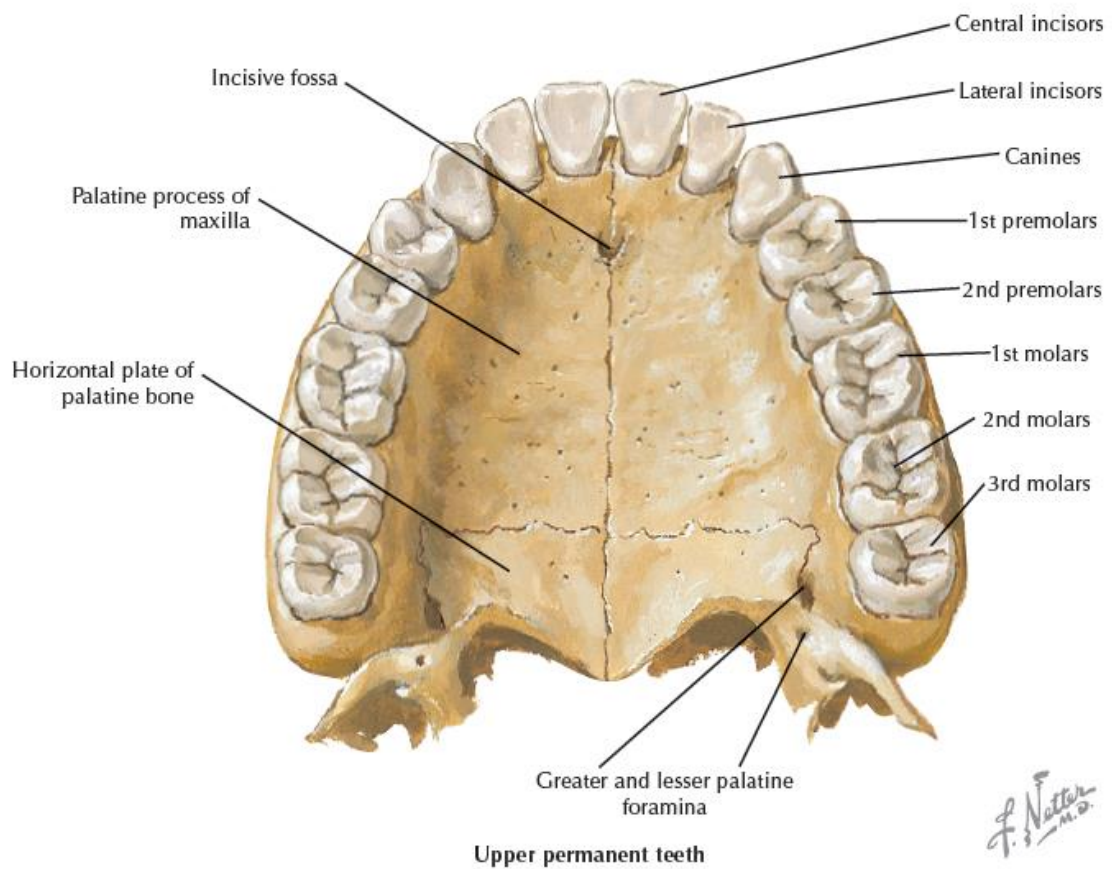
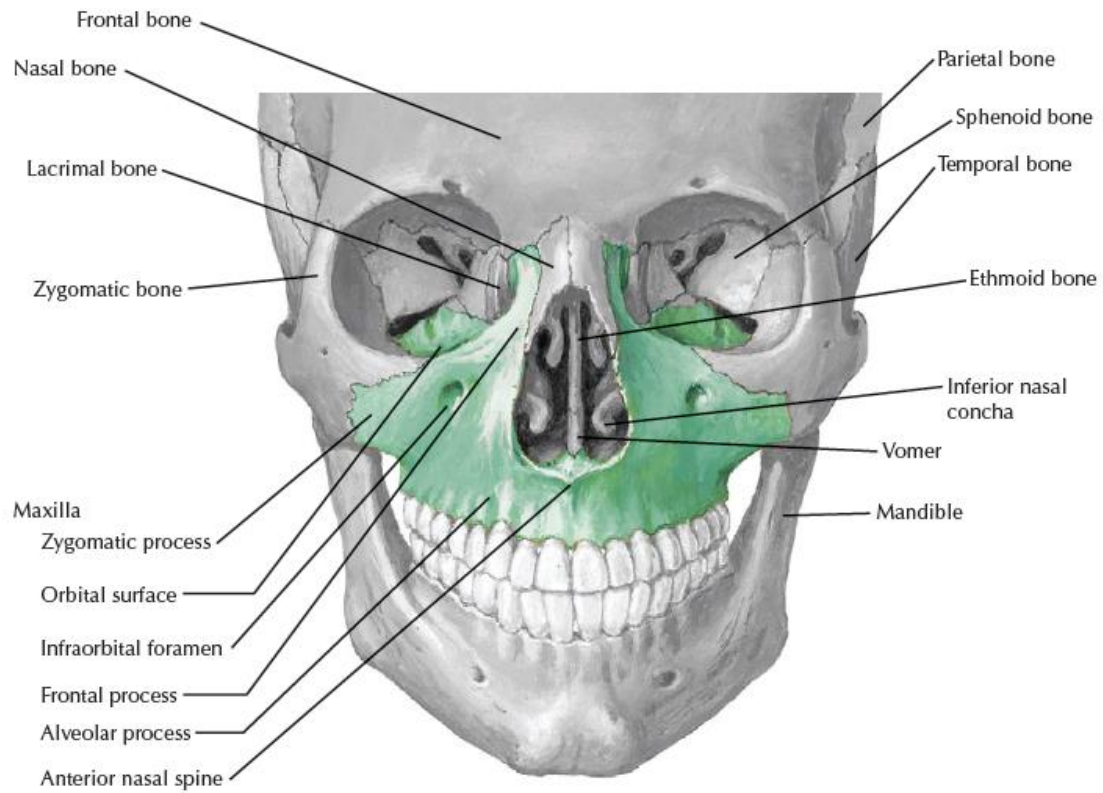
HIGHLIGHTS

- The most common benign tumors of the orbit are hemangiomas and meningiomas.
- The most common malignant tumor of the orbit is non-Hodgkin lymphoma.
- RMS is the most common primary malignancy of the orbit in the pediatric population.
- Proptosis is the initial and cardinal sign of an orbital lesion.
- Visual acuity loss is a late finding of advanced disease or a finding of a space-occupying lesion within the optic canal.
- The orbital vault is made of seven bones: frontal, zygomatic, sphenoid, maxilla, lacrimal, palatine, and ethmoid bones.
- Consideration for orbital metastases should be given for infiltrative and rapidly progressing orbital tumors. The most common primary site for metastasis in women is breast and for men is lung or prostate.
- Surgical approach needs to be dictated by the anatomic location of the lesion, the pathology of the lesion, and the primary goal for surgery (e.g., biopsy vs. resection).
- That extreme post-op pain, especially in association with chemosis, decreased acuity, and elevated intraocular pressure, indicates a possible retroorbital hemorrhage, which can lead to permanent ischemic injury to the optic nerve.
- There are several options for surgical care of orbital tumors; however, for any of these approaches team planning and surgery usually provides the best outcome.

Odontogenic Cysts and Tumors:

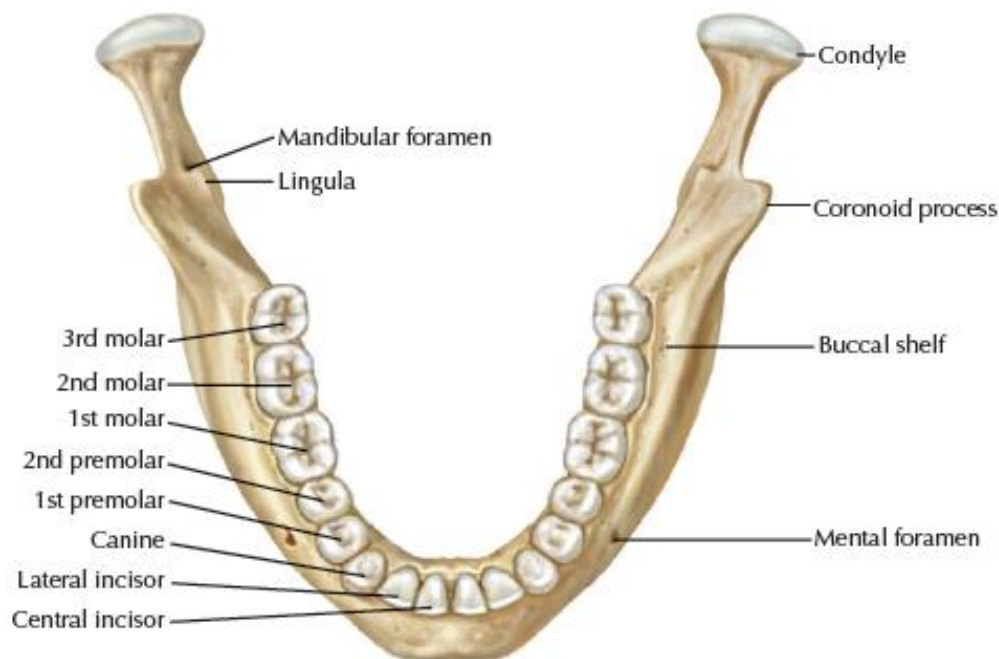
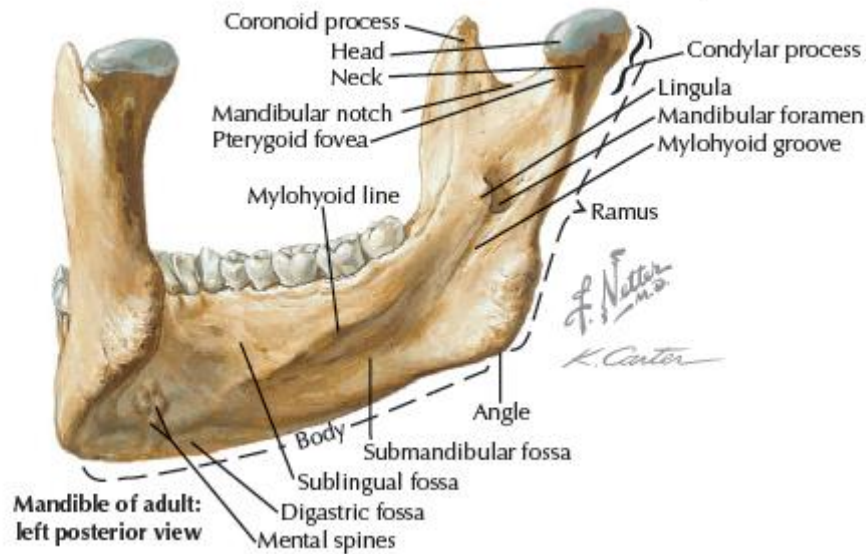
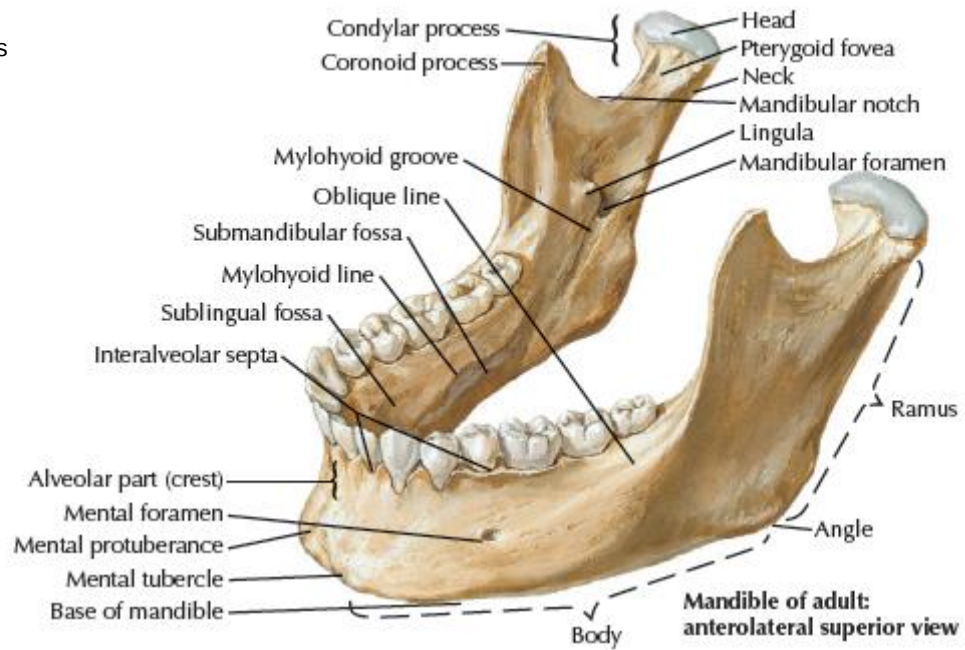
Maxilla

Characteristics	Part	Ossification	Comments
Forms the majority of the skeleton of the face and the upper jaw Contains the maxillary paranasal sinus Articulates with the opposite maxilla and the frontal, sphenoid, nasal, vomer, and ethmoid bones; inferior nasal concha; palatine, lacrimal, and zygomatic bones; and the septal and nasal cartilages There are 2 maxilla bones (maxillae)	Body	Intramembranous	Major part of the bone Shaped like a pyramid Contains the maxillary paranasal sinus <i>Gives rise to 4 different regions:</i> • Orbit • Nasal cavity • Infratemporal fossa • Face Infraorbital canal and foramen pass from the orbit region to the face region
	Frontal process		Extends superiorly to articulate with the nasal, frontal, ethmoid, and lacrimal bones Forms the posterior boundary of the lacrimal fossa
	Zygomatic process		Extends laterally to articulate with the maxillary process of the zygomatic bone
	Palatine process		Extends medially to form the majority of the hard palate Articulates with the palatine process of the opposite side and the horizontal plate of the palatine bone Incisive foramen is located in the anterior portion
	Alveolar process		The part of the maxilla that supports all of the maxillary teeth Extends inferiorly from the maxilla Each maxilla contains 5 primary and 8 permanent teeth Alveolar bone is resorbed when a tooth is lost



Mandible

Characteristics	Part	Ossification	Comments
Forms the lower jaw Described as horseshoe shaped All muscles of mastication attach to the mandible There is 1 mandible	Body	Intramembranous (ossifies around Meckel's cartilage)	Mental foramen lies on the anterior part of the lateral surface of the body External oblique line is observed on the lateral side of the mandible On the medial side of the body lies the mylohyoid line Mylohyoid line helps divide a sublingual from a submandibular fossa Posterior border of the mylohyoid line provides for attachment of the pterygomandibular raphe At the midline on the medial side are the superior and inferior genial tubercles, as well as the digastric fossa
	Ramus		Meets the body of the mandible at the angle of the mandible on each side Masseter m. attaches to the lateral side Medial pterygoid m. and sphenomandibular lig. attach to the medial side Mandibular foramen is located on the medial side of the ramus Superior part divides into a coronoid process anteriorly and a condylar process posteriorly, separated by a mandibular notch
	Coronoid process		The anteriormost superior extension of each ramus Temporalis m. attaches to the coronoid process
	Condylar process		Articulates with the temporal bone in the temporomandibular joint Has a neck that forms a condyle superiorly Lateral pterygoid muscle attaches to pterygoid fovea on the neck
	Alveolar process		Extends superiorly from the body Created by a thick buccal and a thin lingual plate of bone The part of the mandible that supports the mandibular teeth Each side of the mandible contains 5 primary and 8 permanent teeth Alveolar bone is resorbed when a tooth is lost



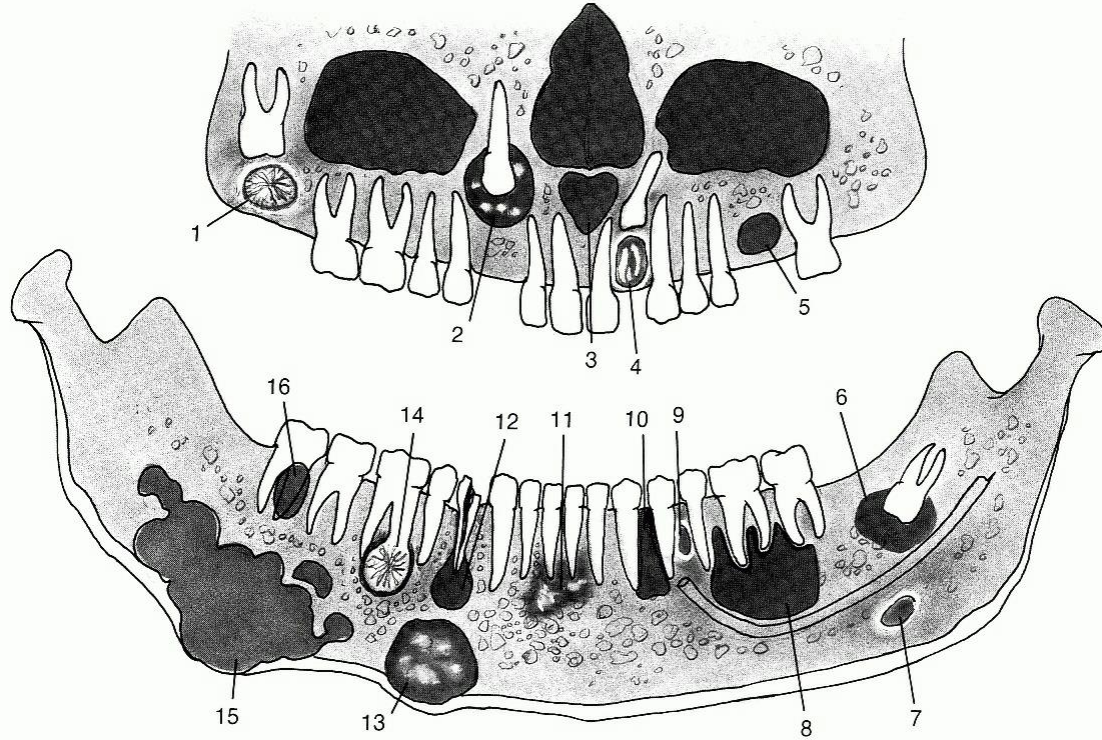
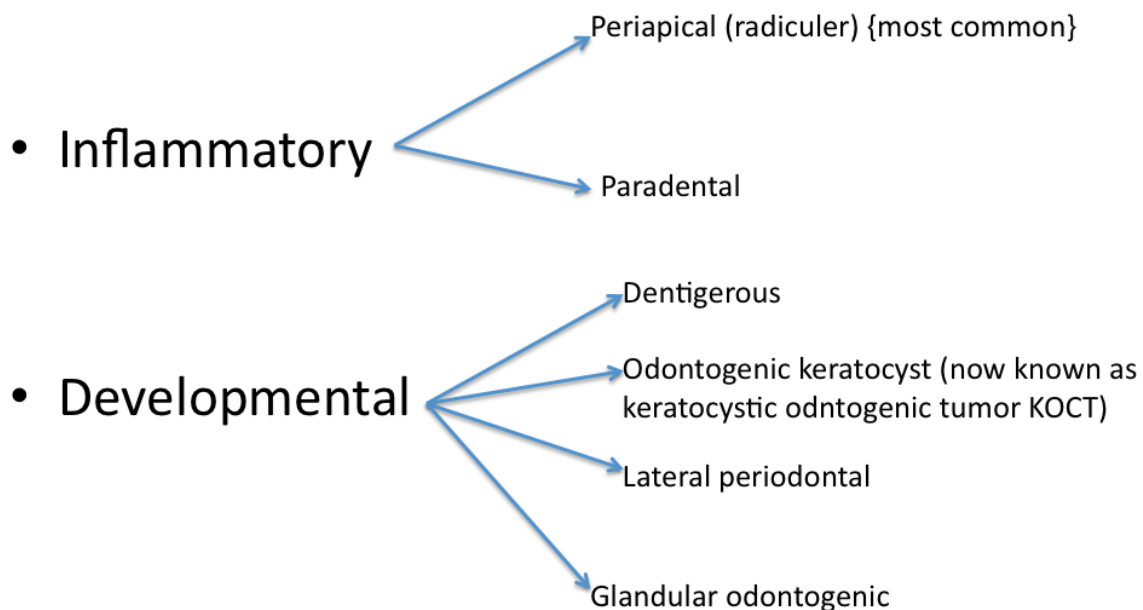


FIGURE 110.16. Prototypical radiographic appearance and location of selected odontogenic cysts and tumors. 1, Compound odontoma. 2, If opacities present: adenomatoid odontogenic tumor; if no opacities: dentigerous cyst, odontogenic keratocyst, ameloblastic fibroma, odontogenic myxoma. 3, Incisive canal cyst. 4, Complex odontoma. 5, Residual cyst, odontogenic keratocyst, surgical ciliated cyst of maxilla. 6, Dentigerous cyst, odontogenic keratocyst, unicystic ameloblastoma, ameloblastic fibroma, odontogenic myxoma. 7, Stafne bone cyst. 8, Traumatic bone cyst, odontogenic keratocyst. 9, Lateral periodontal cyst, odontogenic keratocyst. 10, Periodontal inflammatory disease, central giant cell granuloma, odontogenic keratocyst, histiocytosis X, malignant tumor. 11, Cementoma. 12, Periapical cyst, periapical granuloma. 13, Ossifying fibroma, calcifying epithelial odontogenic tumor, calcifying odontogenic cyst. 14, Cementoblastoma. 15, Ameloblastoma, odontogenic keratocyst, odontogenic myxoma, central giant cell.

Oodontogenic cysts:

- Odontogenic tumors are relatively uncommon lesions
- Odontogenic cysts can become inflamed or infected, causing significant signs or symptoms
- Most cysts of the oral cavity are **true cysts** because they contain an epithelial lining.
- The lining of these cysts are derived from one of three epithelial structures:
 - **Reduced enamel epithelium**: residual epithelium that surround the crown after enamel formation is complete
 - **Epithelial Rests of Malassez**: remnants of Hertwig root sheath; persist in periodontal ligament after root formation complete
 - **Rests of Serres** (aka remnants of dental lamina): islands of epithelium that originate from the oral epithelium and remain in tissues after inducing tooth development
- Two categories: inflammatory and developmental



- Radicular cysts, dentigerous cysts, OKC: make up 80% of dental cysts

Inflammatory Cysts:

- Non-keratinising stratified squamous epithelium lining an inflamed collagenous cyst wall
- **Periapical (radicular) cyst:**
 - Most common odontogenic cysts (65%)
 - Develops at the apex of an erupted tooth in response to pulpal necrosis secondary to dental **caries** or **trauma**
 - Arises from inflammatory stimulation and proliferation of the **Rests of Malassez**
 - Cellular debris in cyst → osmotic gradient → cyst expands
 - Clinical and radiologic features
 - Asymptomatic; incidental x-ray discovery (radiolucent)
 - Rarely > 1 cm; round-ovoid; contiguous with apex



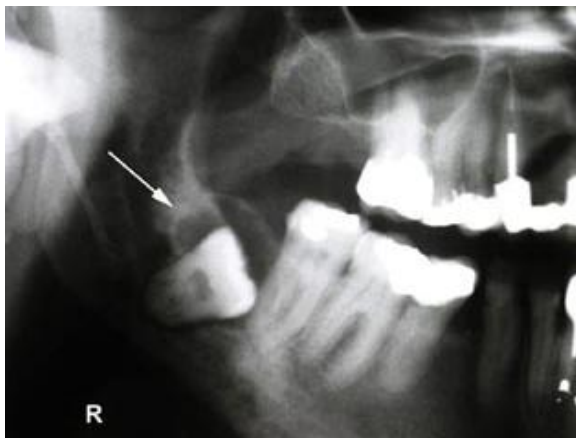
Figure 132.1 Periapical cyst. Radiograph demonstrating periapical lesion associated with grossly carious tooth # 20.

- Histopathology
 - Variably thick stratified squamous epithelium
 - Lymphocytes and neutrophils; necrotic debris
 - Some have crescent-shaped hyaline **(Rushton) bodies** within the lining
- Treatment and prognosis
 - Extract tooth and enucleate cyst; can do root canal
 - Complete healing within 6/12 if thoroughly removed

Developmental Cysts:

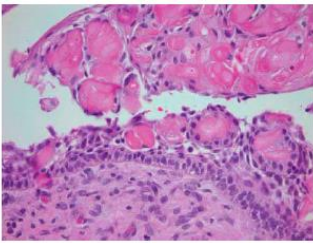
I.Dentigerous (follicular) cyst:

- Second most common cyst (recurrence rare)
- Must be associated with crown of unerupted tooth (dental follicle)
- Attached to cervical region of tooth; usually mandible. 3rd molar
- Maxillary 3rd molar, canain and mandible 2nd bicuspid
- Clinical and radiologic features
 - 2nd/3rd decade; M>F; affected tooth not erupted
 - Usually asymptomatic; can be big; **bony expansion**
 - Circumscribed & unilocular lucency; *surrounds crown*
 - Can displace tooth into ramus or inf border of mand.
- Histopathology
 - Non-keratinized stratified squamous epithelial lining
 - Lining can be 2-10 cells thick; often inflamed
 - Like radicular cysts, can have **Rushton bodies**, cholesterol clefts and hemosiderin deposits
- Treatment and prognosis
 - Removal of tooth and cyst enucleation
 - If really big, can first marsupialize to allow for decompression with compensatory bone fill before definitive removal of the lesion
 - **Lining can differentiate into ameloblastoma**



II. Calcifying odontogenic cyst (Gorlin cyst)

- Arise from remnants of **dental lamina** "**Rests of Serres**" within the gingival or jaws
- ¼ of them occur extraosseous (between bone and gingiva) anterior to the 1st molar
- Clinical and radiologic features
 - Little potential for recurrence; has a wide age range with the peak in the 2nd decade
 - Most are located in the anterior portion of either jaw; **F>M**
 - **Painless** gingival expansion; incidental findings
 - Initially lucent, then calcifies causing a well-circumscribed mixed radiolucent-radiopaque appearance
- Histopathology
 - Unilocular cyst; lined by odontogenic epithelium
 - The basal layer has columnar to cuboidal cells; polarized hyper-chromatic nuclei away from basement membrane
 - Ghost cell: eosinophilic altered epithelial cell with loss of nucleus that represent abnormal keratinization; later become calcified and may cause a foreign body reaction (from keratin release)
- Treatment and prognosis
 - Surgical enucleation with complete resolution



III. Glandular odontogenic cyst (Sialo-Odontogenic Cyst):

- Locally aggressive; likely to recur; but it's exceedingly rare
- Numerous **mucus secreting cells** within the epithelial lining
- Clinical and radiologic features
 - Middle-aged adults; anterior mandible most common
 - **Multilocular** radiolucencies; frequently cross midline
- Histopathology
 1. Epithelial lining of variably thickness and a flat epithelial-connective tissue junction
 2. Small microcysts/glandlike structure within the lining
 3. Single layer of cuboidal or columnar cells lining glandlike structure
- Treatment and prognosis
 - Enucleation and curettage
 - Because of its recurrent potential some advocate marginal resection

NON-ODONTOGENIC CYSTS

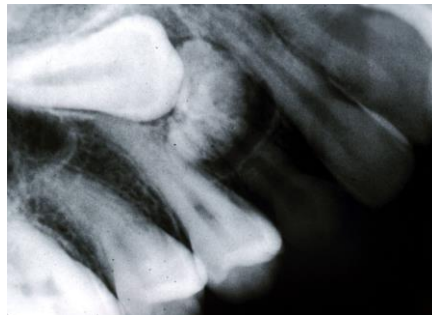
- Nasopalatine duct cyst (Incisive canal cyst)
 - From epithelial remnant of the two nasopalatine ducts
 - Clinical and radiologic features
 - Anterior maxilla near incisive foramen; **M>F**; 4th–6th decades
 - Asymptomatic or present as soft tissue swelling in the midline of the anterior hard palate
 - Well-circumscribed; heart shaped; **unilocular**
 - Can resorb roots of adjacent teeth
 - Can be confused with enlarged incisive canal;
cyst if > 6-10 mm
 - Histopathology
 - The lining can be stratified squamous, ciliated columnar or cuboidal, or a combination of both
 - The fibrous tissue has elements of nerve and vessel (incisive canal content)
 - Mucous glands occasionally seen
 - Treatment and prognosis
 - Enucleation curative; may damage nasopalatine nerve
 - Recurrence is rare
- Stafne bone cyst (static bone cyst; lingual salivary gland depression)
 - It is not a true cyst
 - It is a depression on lingual aspect of posterior body of the mandible
 - Typically filled with accessory lobe of SMG (secondary salivary tissue)
 - Classic X-ray: small ovoid, well-circumscribed lucency below inferior alveolar canal in region of 2nd or 3rd molar
 - No treatment needed
- Idiopathic bone cavity (traumatic bone cyst/cavity)
 - Also not a true cyst; does not have epithelial lining
 - Usually found in the posterior mandible; less well-defined radiolucency
 - Straw coloured fluid on exploration
 - Bleeding into lesion at surgery allows for granulation and healing
- *Surgical ciliated cyst of maxilla*
Surgical implantation usually result of Caldwell-luc antrostomy

ODONTOGENIC TUMOURS

- Rare but diverse group; arise from epithelium, mesenchyme or both
- Most odontogenic tumors are true neoplasms, but some behave as hamartomatous growths
- Common presentation: asymptomatic swelling; leads to bone loss, tooth displacement and jaw expansion; rare to have sensory loss

Odontoma (mixed origin)

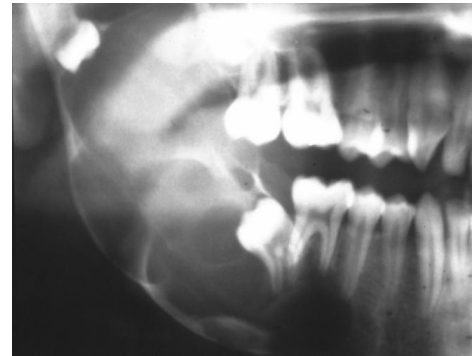
- Not true neoplasm, but rather **hamartomatous growth** because it forms during normal tooth development then reaches fixed size; has elements of enamel, dentin, cementum and pulp tissue
- Can be classified as either:
 - **Compound** if the lesion resembles tooth-like structure;
 - **Complex** if the lesion appears as an amorphous mass
- Clinical and radiologic features
 - Most common tumour
 - Usually asymptomatic; 1st & 2nd decades; M=F
 - Can block eruption of permanent tooth
 - Radiopaque mass; clear border; compound looks like multiple tiny tooth structures; complex appears as a dense irregular mass



- Histopathology
 - It is composed of enamel, dentin, cementum, pulp tissue; sparse fibrous tissue
- Treatment and prognosis
 - Removal for diagnosis or for tooth eruption
 - Enucleation and curettage is curative

Keratocystic Odontogenic tumor (KCOT): “used to be considered cyst, OKC”

- 5-8% of all odontogenic cysts
- Develop from **dental lamina remnants “Rests of Serres”**; in any jaw location
- The mandibular ramus and posterior body are the most common (Angle also)
- In the maxilla favors the posterior or canine regions
- Aggressive growth causing bony expansion and destruction; frequent recurrence (5-60%)
- Clinical and radiologic features
 - Peak incidence in 2nd/3rd decades; can present anytime
 - Nevoid basal cell carcinoma syndrome if multiple
 - Well-circumscribed radiolucency; **uni/multilocular**; bone expansion or erosion of either cortex common
- Histopathology
 - Its lining is parakeratinized stratified squamous; 6-8 cells layers thick
 - Luminal surface has corrugated surface of **parakeratin**
 - The **basal layer is palisaded** with prominent, polarized and hyperchromatic nuclei
 - Daughter (satellite) cells may be seen in the connective tissue and may be indicative of (NBCCS)
- Treatment and prognosis
 - En-bloc enucleation and bony curettage
 - **Modified Carnoy solution** or **liquid nitrogen** to kill remnants, but carcinogenic; some advocate marginal resection to avoid recurrence
 - Can marsupialize large lesions for decompression (risk of secondary infection; needs good compliance)
 - Most recur within 5 yrs; occasionally as late as 10 yrs; close F/U is important (up to 60% due to fragile cyst wall)
 - Malignant transformation rare (?)
- Orthokeratotic variety
 - ~ 15% of OKC
 - Much less recurrence rate
 - Has a prominent granular layer below a noncorrugated surface, and the basal layer is less prominent.



Nevoid BCC syndrome (Basal cell nevus syndrome, Gorlin syndrome)

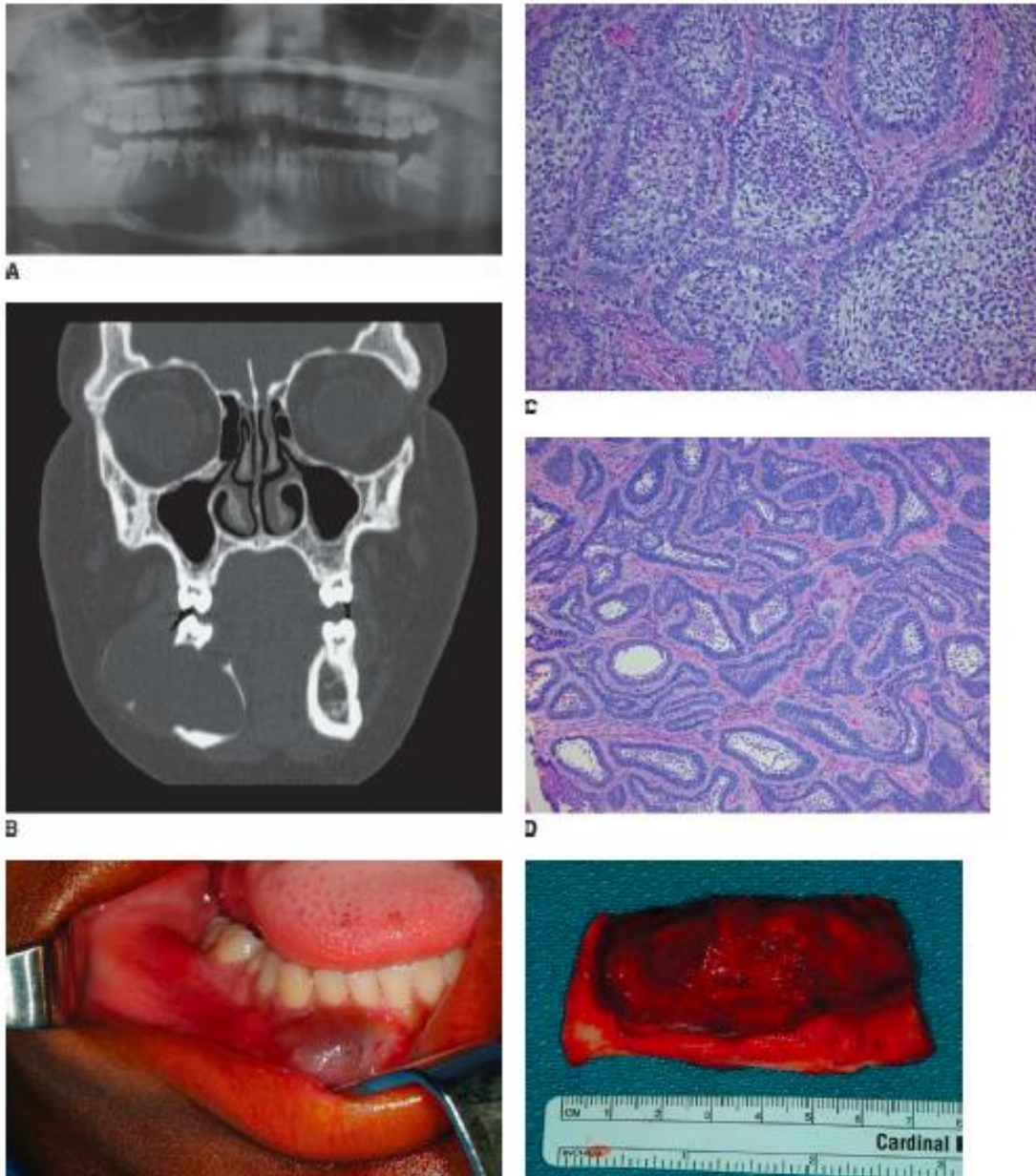
- AD condition that exhibits high penetrance and variable expressivity
- Result of a mutation of the **PTCH** tumor suppressor gene
- Clinically:
 - Multiple OKC of the jaws
 - Early development of multiple basal cell carcinomas
 - Bifid ribs, frontal bossing, depressed midface, wide nasal bridge, hypertelorism, mandibular prognathism, calcification of falx cerebrii
- Genetic counseling should be recommended

Ameloblastoma (epithelial)

- Arises from epithelial elements (rests of Serres/Malassez; reduced enamel epithelium; basal layer of oral mucosa) or from within dental follicle or dentigerous cyst
- Classification: Unicystic / Multicystic or solid/ Peripheral ameloblastoma
- Clinical and radiologic features
 - 2nd most common (3rd with OKT included); benign; locally aggressive; 3rd to 4th decades as peak; M=F
 - Slow growth; can get huge and may cause gross facial deformities; usually asymptomatic; **posterior mandible** is the commonest location
 - Uni/multilocular radiolucency; ill-defined borders; cortical expansion is common; root resorption occurs infrequently (Soap bubble appearance)



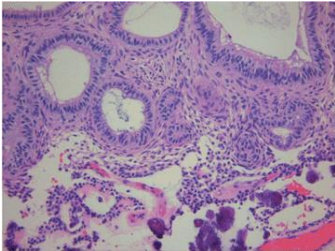
- Histopathology
 - Diverse; they can exhibit areas of focal variation, so adequate sampling is required
 - Unencapsulated / infiltrative into surrounding tissues
 - Columnar, hyperchromatic basal cells; reverse polarity (the nuclei move from the basement membrane pole of the cell to the opposite pole)
 - Numerous histologic patterns; the two most frequent patterns are the **follicular** and **plexiform** patterns; (no significance)
- Treatment and prognosis
 - Known to extend beyond radiographic margin
 - If limited to epithelial lining or proliferate into the lumen, enucleation and curettage is sufficient
 - When through connective tissue layer, more aggressive approach; 1-1.5 cm bony margin and one uninvolved overlying anatomic barrier (Mandible 1 cm, maxilla 1.5 cm)
 - Cure rate of 98% with negative margins (E&C)
 - Recurrence of 90% if inadequately treated
- **Malignant ameloblastoma** can metastasize; lesions still benign (both primary and mets)
- **Ameloblastic carcinoma** shows features of malignancy
- Ameloblastic fibrosarcoma is most common malignancy



- Peripheral ameloplastoma (soft tissue mass involving the gingival, often cause separation of teeth)

Adenomatoid odontogenic Cyst (AOC) (epithelial)

- Benign hamartoma of odontogenic epithelium (Hertwig root sheath)
- Clinical and radiologic features
 - Slow, progressive growth; most in **maxilla**
 - **"Two-thirds tumour"**: 2/3 occur with unerupted tooth, of which 2/3 are a canine, 2/3 in the maxilla, 2/3 in young **F**
 - Most < 3 cm, but they can be too big and can cause pain and displace roots
 - Well-circumscribed, unilocular lucency with an impacted tooth; some have small foci of **calcification**
- Histopathology
 - Highly cellular; **spindle-shaped cells** in whorls/rosettes
 - Enameloid material throughout lesion, duct like structures
 - Calcifications, a common feature of this lesion
- Treatment and prognosis
 - Curettage is curative; recurrence is not seen; bony regeneration within 1 yr



Calcifying epithelial odontogenic tumour (Pindborg tumour)

- Rare; < 1% of odontogenic tumours
- Originates from epithelial rests of the dental lamina or reduced enamel epithelium
- Clinical and radiologic features
 - Peak incidence in the 4th decade
 - Slow; firm; painless; highly infiltrative destructive, usually in the posterior mandible, the molar area is the most frequent region in either jaw
 - A peripheral variant exists and typically presents as soft tissue swelling in the anterior aspect of the mouth
 - Diffuse radiolucency; areas of calcification if large
- Histopathology
 - Sheets/strands of polyhedral epithelial cells
 - Prominent nucleoli; can appear pleomorphic
 - **Liesegang rings**: scattered rings of **calcification**
 - Large areas of amorphous eosinophilic tissue; **stains positive for amyloid with Congo red "unique characteristic feature"**
- Treatment and prognosis
 - Same as ameloblastoma
 - Peripheral variant: 5 mm margin with periosteum
 - Long-term F-U necessary for possible recurrence

Odontogenic myxoma

- Rare; derived from odontogenic ectomesenchyme
- Clinical and radiologic features
 - Locally aggressive; 3rd decade; most frequently in the posterior mandible
 - Usually asymptomatic and slow growing; can displace teeth
 - Uni/multilocular radiolucency; **honeycomb** appearance
- Histopathology
 - Infiltrative and gelatinous tumour – like snot
 - Scant, randomly arranged, spindle-shaped mesenchymal cells within mucoid ground substance
 - Fibromyxoma if more collagenous
- Treatment and prognosis
 - Very aggressive, same as ameloblastoma and KOT

Ameloblastic fibroma (fibro-odontoma)

- It is a true neoplasm composed of odontogenic epithelium and ectomesenchyme
- Clinical and radiologic features
 - Tumor of young patients, rarely > 40 yrs; painless swelling in **post mandible**
 - Uni/multilocular radiolucency; often over an unerupted tooth or displacing the tooth
- Histopathology
 - Islands of odontogenic epithelium; columnar; palisaded; reverse polarity identical to ameloblastoma
 - Stroma is myxoid, resembling dental papilla
- Treatment and prognosis
 - Encapsulated, therefore enucleation/curettage
 - If recurs, think of **ameloblastic fibrosarcoma**

Related Jaw Lesions

- **Torus**

- Developmental overgrowth; thought to arise because of bone stress
- Clinical and radiologic features
 - **Torus palatinus:** midline palate; 20% of adults; develops after puberty; slow growing; can either have a smooth ovoid appearance or multiple pedunculated loculations and normal overlying mucosa; can inhibit speech/swallowing
 - **Torus mandibularis** (lingual tori): bilat; bicuspid region; can get big and impair speech; both impair dentures
 - **Exostosis:** if on buccal aspect of either jaw
- Histopathology
 - Consist of nodular masses of dense cortical, lamellar bone with central trabecular bone containing few areas of fatty marrow
- Treatment and prognosis
 - Only excise if impairing function
 - Elevate mucosa; take bone down to the surrounding bone



Figure 132.10 Palatal torus.

- **Osteoma**

- Hamartomas or reactive proliferations of bone, composed of dense compact bone
- Periosteal (arises on the surface of bone) or endosteal (within bone)
- Clinical and radiologic features
 - Consider **Gardner syndrome** if multiple lesions
 - AD; intestinal polyps; skin fibromas; osteomas; impacted normal and supernumerary teeth, and odontomas
 - Malignant transformation is essentially 100% of the polyps
 - Slow-growing exophytic bony masses
 - Either jaw in areas not typically affected by tori or exostoses; mandibular angle common site
 - Incidental finding; circumscribed radiopaque mass
- Histopathology
 - Similar to tori, except periosteum more active
- Treatment and prognosis
 - Follow; excise for Bx if needed; unlikely to recur

- **Osteochondroma**

- Benign hamartomas of long bones; **mand condyle/coronoid**
- Theory: proliferation of epiphyseal cartilage into surroundings tissues
- Clinical and radiologic features
 - 2nd/3rd decades; M>F; cause swelling and pain
 - Teeth and chin deviate/point to opposite side
 - Irregular "**popcorn-like**" radiopaque mass on medial side of condyle; may replace the coronoid
- Histopathology
 - Bony masses with cartilaginous cap; endochondral ossification between cartilage and bone
- Treatment and prognosis
 - Coronoidectomy with minimal removal of temporalis
 - Condylectomy with immediate costo-chondral recon
 - Recurrence is rare

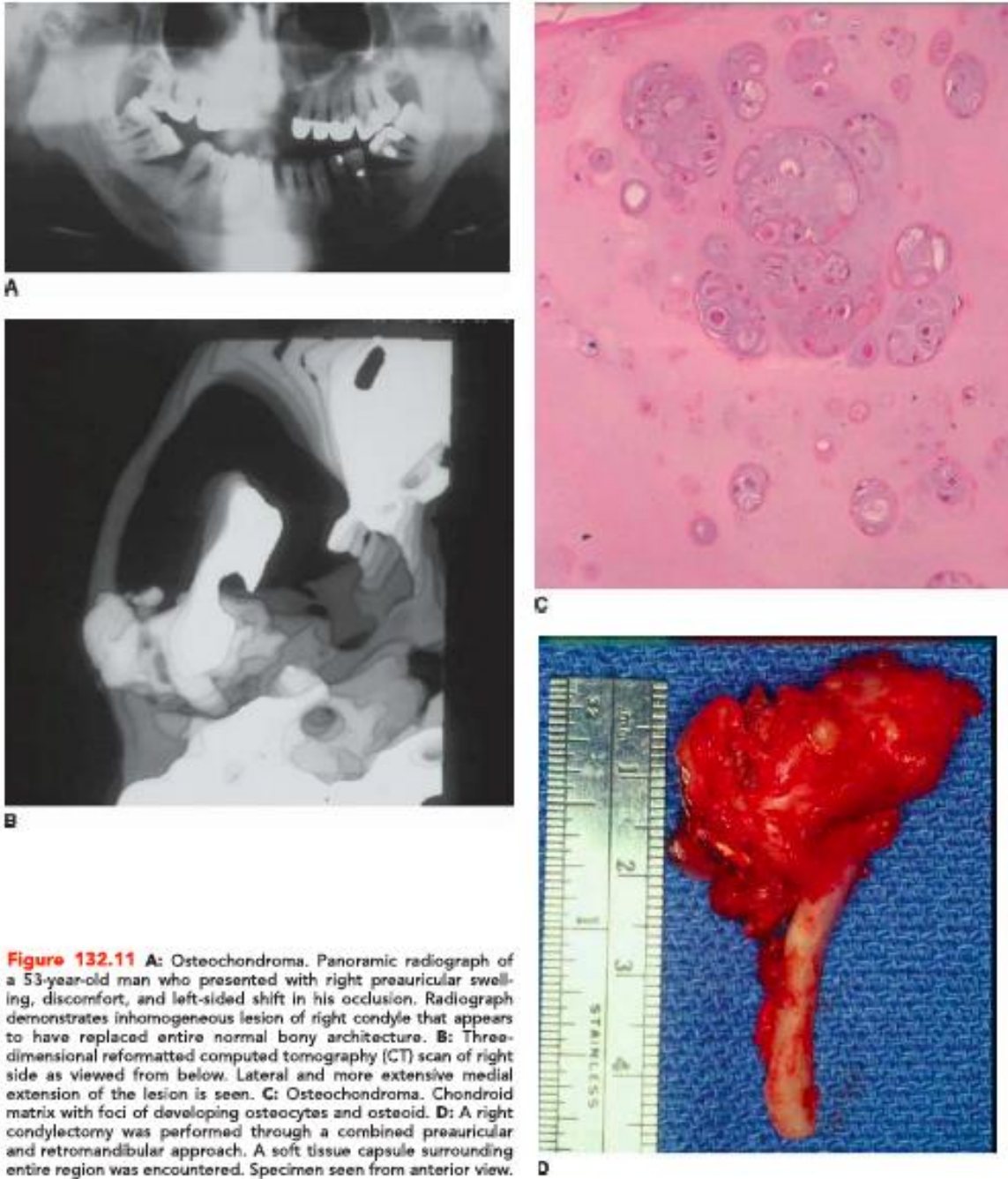


Figure 132.11 A: Osteochondroma. Panoramic radiograph of a 53-year-old man who presented with right preauricular swelling, discomfort, and left-sided shift in his occlusion. Radiograph demonstrates inhomogeneous lesion of right condyle that appears to have replaced entire normal bony architecture. B: Three-dimensional reformatted computed tomography (CT) scan of right side as viewed from below. Lateral and more extensive medial extension of the lesion is seen. C: Osteochondroma. Chondroid matrix with foci of developing osteocytes and osteoid. D: A right condylectomy was performed through a combined preauricular and retromandibular approach. A soft tissue capsule surrounding entire region was encountered. Specimen seen from anterior view.

- **Ossifying fibroma (cemento-ossifying fibroma)**

- Benign tumour; mesenchymal origin; juvenile form is a rare and aggressive variant
- Predilection for tooth-bearing portions of jaws
- Clinical and radiologic features
 - Slow growing; expansile; can reach enormous size
 - F>M; 3rd/4th decade
 - Initially radiolucent; mixed as it matures; ends opaque
 - Aggressive ones expand cortices and displace roots
 - In the mandible, appear as a midbody growth at the inferior border, enlarging outward and downward as if “hanging off the lower lateral border.”
- Histopathology
 - Well demarcated from the surrounding bone; composed of a dense cellular fibrous layer; varying amounts of calcified trabeculae of osteoid, bone or spherical cementum-like structures
 - Does not exhibit diffuse infiltration of adjacent tissues
- Treatment and prognosis
 - Enucleation/curettage if early; resect if big; 5 mm margin is safe; recurrence unlikely

- **Fibrous dysplasia**

- Normal medullary bone replaced with fibrous connective tissue mixed with irregular bony trabeculae
- Monostotic or polyostotic (latter part of McCune-Albright – café-au-lait spots and multiple endocrinopathies)
- Genetics: usually from spontaneous mutation
- Clinical and radiologic features
 - Slow growing; asymptomatic; bony hard swelling
 - Can be self-limited; growth stops with bone growth
 - May cause tooth displacement and malocclusion
 - Radiolucent early; become more opaque as the lesion matures; “the normal medullary bone is replaced with a fine trabecular bone, giving the lesion its **“ground-glass”** appearance
 - Expands cortices; does not displace inferior alveolar canal
 - Can cause cranial neuropathy or facial distortion
- Histopathology
 - Irregularly shaped trabeculae of woven bone in background of loose and cellular fibrous connective tissue
 - Not encapsulated; abnormal bone coalesces with surrounding tissue, in contrast to ossifying fibroma

- Treatment and prognosis
 - Only treat if disfiguring or cranial neuropathy
 - Recontour rather than resect; wait until mature
 - Sarcomatous transformation has been reported
 - Recurrence more likely if treated during active growth
- **Central giant cell lesion**
 - Locally aggressive; benign; long bones and jaws
 - Clinical and radiologic features
 - Asymptomatic; painless swelling; pain or **paresthesia** if aggressive
 - Bluish mass as a result of the thinning of the overlying bone and mucosa and its highly vascular nature
 - 2nd/3rd decades; F>M; prefers anterior region of jaws, particularly the mandible
 - Well-circumscribed or multilocular lucency
 - Can expand cortices; can move teeth; roots not eroded
 - Histopathology
 - Multinucleated giant cells vary in size and their number of nuclei in background of spindled mesenchymal cells; behave like osteoclasts
 - Unencapsulated; fibrous tissue present
 - Identical to cherubism, Brown tumour of hyperPTHism, and focal areas of fibrous dysplasia
 - Treatment and prognosis
 - Curettage usually curative; but recurrent or larger lesions occasionally require resection
 - Recurrence an issue if resection difficult/incomplete
 - Intralesional triamcinolone (10 mg/ml) weekly X 6 is advocated by some as a first line of therapy
 - Injectable calcitonin may also work
- **Aneurysmal bone cyst (ABC)**
 - Rare, expansile, osteolytic bone lesion; blood filled spaces that do not have an endothelial lining and, thus, is not a true cyst
 - Clinical and radiologic features
 - First 3 decades; posterior jaws; firm swelling
 - Expansile radiolucencies; can displace teeth
 - Histopathology
 - Hemosiderin, bone, osteoid within septae; giant cells
 - Treatment and prognosis
 - Curettage; intraoperative bleeding a challenge

- **Vascular malformations**

- Clinical and radiologic features
 - Slow growing, asymptomatic, expansile lesions; teeth mobile/elevated
 - Thrills or bruits; dilated veins if tongue involved
 - Some are well-circumscribed lucencies; others mixed
 - Tooth roots can be resorbed
 - CT, MR and angio helpful
- Treatment and prognosis
 - If suspicious, needle aspirate before biopsy to confirm
 - Arterial lesions can be embolized, followed by resection surgery
 - Venous ones can be sclerosed
 - If blood flow controlled, can consider curettage

ODONTOGENIC CYSTS	
▪ Inflammatory Cysts	
Periapical (radicular) cyst	Commonest (65%); at the apex of an erupted tooth; secondary to dental caries or trauma X-R: Rarely > 1 cm; round-ovoid; contiguous with apex
▪ Developmental Cysts	
1. Dentigerous (follicular) cyst	Second most common cyst; associated with crown of unerupted tooth X-R: Circumscribed & unilocular lucency; surrounds crown of unerupted tooth
2. Odontogenic keratocyst (OKC)	Develop from dental lamina remnants "Rests of Serres" ; most common at mandibular ramus and posterior body X-R: Well-circumscribed radiolucency; uni/multilocular ; bone expansion or erosion of either cortex common
3. Calcifying odontogenic cyst (Gorlin cyst)	Develop from remnants of dental lamina "Rests of Serres" ; ¼ of them occur <u>extraosseous</u> ; anterior portion of either jaw; X-R: Initially lucent, then calcifies causing a well-circumscribed mixed radiolucent-radiopaque appearance
4. Glandular odontogenic cyst (Sialo-Odontogenic Cyst)	Rare; locally aggressive; likely to recur; most common at anterior mandible mucus secreting cells within the epithelial lining X-R: Multilocular radiolucencies; frequently cross midline
NON-ODONTOGENIC CYSTS	
1. Nasopalatine duct cyst (Incisive canal cyst)	From epithelial remnant of the two nasopalatine ducts 4 th –6 th decades; Asymptomatic or anterior soft tissue swelling X-R: Well-circumscribed; heart shaped; unilocular
2. Stafne bone cyst (static bone cyst; lingual salivary gland depression)	not a true cyst; a depression on lingual aspect of posterior body of the mandible; filled with accessory lobe of SMG X-R: small ovoid, well-circumscribed lucency below inferior alveolar canal in region of 2 nd or 3 rd molar
3. Idiopathic bone cavity (traumatic bone cyst/cavity)	not a true cyst; found in the posterior mandible; less well-defined radiolucency; Straw coloured fluid on exploration

Cutaneous Malignancy:

Risk Factors (for basal and squamous cell carcinomas are strikingly similar)

- Most common malignancy **BCC** is 90% of them in H&N
- 2nd most common SCC then melanoma
- Most common in pts > 60 yrs
- **Sunlight**
 - UVB is most carcinogenic (280-320 nm); causes common sunburn; 1% reduction in ozone = 2% increase in UVB
 - Progressive over yrs from normal to actinic to cancerous
 - Tanning booths (UVA – 320-400 nm) synergistically augment UVB responses and can independently cause skin cancer
- **Traits at risk**
 - Fair complexion; light hair; blue/green eyes
 - Inability to tan/propensity to burn
 - History or multiple or severe sunburns
- **Other factors (indirect causes of sun exposure)**
 - Age; occupation (exposure); habits (tanning); where you live
- **Other factors (not related to sunlight)**
 - Chronic chemical exposure (arsenic in Fowler solution)
 - Chronic radiodermatitis
 - Trauma (burns; ulcers; scars); Marjolin ulcer (regions of previous burns)
 - Immunosuppression
 - HPV may be related
 - Genetic syndromes (xeroderma pigmentosum "autosomal recessive"; nevoid basal cell carcinoma syndrome "autosomal dominant") related to BCC
 - Albinism
 - Nevoid BCC syndrome (Gorlin's)
- The most common precancerous skin lesions are
 1. actinic keratosis
 2. Bowen's disease
 3. keratoacanthoma

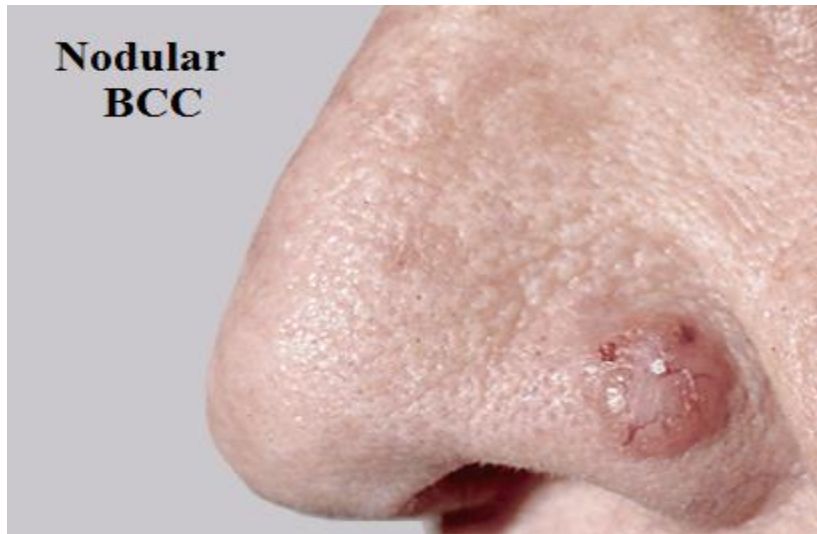
☒ Basal Cell Carcinoma

- indolent course, extends **peripherally** without vertical invasion
- rare metastasis
- cutaneous lesions located in the embryonic fusion plates of the face (nasolabial folds, floor of nose, columella, preauricular regions, inner and outer canthus of the eye) are **more aggressive** and have a higher
- risk of recurrence, therefore require close follow-up
- 90% cure rate in the head and neck regions
- **Dx**: excisional biopsy of suspicious lesions

FIVE CLINICAL SUBTYPES

1. Nodular or Noduloulcerative (**most common**)
2. Morphealike or Sclerosing (**most dangerous**)
3. Superficial Multicentric
4. Pigmented
5. Fibroepithelioma

• Nodular	○ Discrete; raised; circular; pink/waxy; capillary network ○ May have area of central ulceration with rolled border
• Morphealike (sclerosing; fibrosing)	○ Macular whitish/yellow plaques; indistinct margins ○ May look like a scar; may ulcerate
• Fibroepithelioma	○ Initially firm & pedunculated lesions; look like fibromas ○ Common location is the back
• Superficial multicentric	○ Scarring/atrophy; waxy border; red/scaling patches ○ Crusted; irregular borders; peripherally extend ○ Uncommon in the H&N
• Pigmented	○ Less common; brown; looks like nevus or melanoma ○ Similar to nodular aside from pigment



- **Nevoid BCC syndrome (Gorlin's)**
 - autosomal dominant; hundreds of small cutaneous nodules during childhood
 - Neoplastic change as patient ages; invasive; destructive
 - Other findings: jaw cysts; bifid ribs; scoliosis; MR; frontal bossing'
 - Rx: excise malignant lesions, close follow-up every 3–6 months
- **Histopathology**
 - Characteristics of typical BCC:
 - principal cells resemble basal cells (small, dark-blue cells with minimal cytoplasm) of the epidermis
 - Four basic histologic patterns
 - Solid – no differentiation
 - Keratotic – differentiated to hair-like structures
 - Cystic – differentiated to sebaceous gland-like structures
 - Adenoid – differentiated to tubular structures, lace-like histological

☒ Squamous Cell Carcinoma

FIVE HISTOLOGIC SUBTYPES

1. Generic
2. Adenoid
3. Bowenoid
4. Verrucous
5. Spindle-Pleomorphic

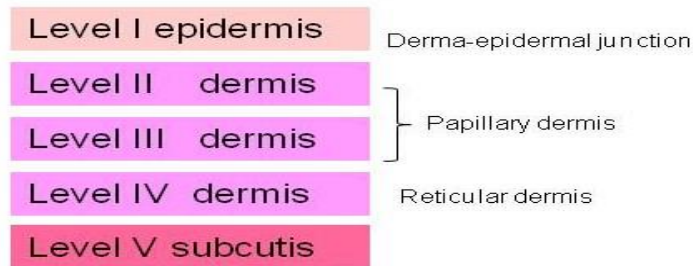
- Initially red, ulcerated hyperkeratotic, opaque nodule ,crusting; granular base; bleeds easily
- Elevated indurated area at margin
- 1–4% metastatic potential
- tendency for **vertical** growth
- more aggressive lesions and higher recurrence rates arise from
 - lesions from previous scars and wounds,
 - lesions located on embryonic fusion Plates
 - lesions arising de novo (non-sunexposed skin), and lesions deep (>6 mm) and large in size (>2 cm)



- **Various patterns for SCC**
 - Can have thickened hyperkeratotic patch or area of crusting; underneath is ulcerated base with rolled margin
 - Persistent ulceration
 - Site of previous trauma, burn or old scar (Marjolin's ulcer)
- **Histopathology**

- Irregular masses of epidermal cells **invading dermis**
- Keratin pearls in those that are well differentiated
- **Broder classification based on differentiation:**
 - **Grade 1:** < 25% undifferentiated cells
 - **Grade 2:** < 50% undifferentiated cells
 - **Grade 3:** < 75% undifferentiated cells
 - **Grade 4:** > 75% undifferentiated cells
- If arising from sun exposure, **less** metastatic potential
- Clark's level IV or V have 20% regional mets rate

Clark Levels



- **Generic:** actinic changes
- **Adenoid:** pseudo-glandular arrangement more aggressive form
- **Bowenoid:** invasion coexistent with findings of Bowen's disease
- **Verrucous:** rare as a H&N skin lesion; OC and larynx more common; (low-grade malignancy) white cauliflower-like lesion; hyperkeratosis, parakeratosis and acanthosis
- **Spindle-pleomorphic:** little differentiation; occurs more commonly at site of a scar, trauma, or burn, **more aggressive form**

☒ **Keratotic Basal Cell Carcinoma**

- AKA **basosquamous cell carcinoma**
- Histologically has features of both BCC and SCC
- Felt to be more aggressive than most BCC lesions

Prognosis

- Worse prognosis based on path:
 - morphealike BCC
 - spindle cell variants of SCC
 - basosquamous BCC
- **Nose and ear** lesions more likely to recur; embryonic fusion planes used by tumour as avenues for spread
- Swanson's H-zone (includes septum) (**high-risk areas of aggressive cutaneous malignancies**)
- Aggressive BCC lesions tend to be in those predisposed to them (arsenic ingestion; nevoid BCC syndrome; burns; radiodermatitis)
- Larger ones also more difficult to treat

TABLE 113.1

AJCC CLASSIFICATION FOR CUTANEOUS SQUAMOUS CELL CARCINOMA AND OTHER CUTANEOUS CARCINOMAS (EXCLUDING THE EYELID)

Primary tumor (T)

TX: Primary tumor cannot be assessed.

T0: No evidence of primary tumor

Tis: Carcinoma *in situ*

T1: Tumor 2 cm or less in greatest dimension with less than two risk features

T2: Tumor >2 cm or tumor of any size with two or more high-risk features

T3: Tumor with invasion of the maxilla, mandible, orbit, or temporal bone

T4: Tumor with invasion of the skeleton (axial or appendicular) or perineural invasion of the skull base

Regional lymph nodes (N)

NX: Regional lymph nodes cannot be assessed.

N0: No regional lymph node metastasis

N1: Metastasis in a single ipsilateral lymph node, 3 cm or less

N2a: Metastasis in a single ipsilateral lymph node, more than 3 cm but not more than 6 cm

N2b: Metastasis in multiple ipsilateral lymph nodes with none more than 6 cm

N2c: Metastasis in bilateral or contralateral lymph nodes with none more than 6 cm

N3: Metastasis in a lymph node more than 6 cm

Distant metastasis (M)

M0: No distant metastasis

M1: Distant metastasis

**TABLE
113.2**

**HIGH-RISK FEATURES OF CUTANEOUS SQUAMOUS CELL
CARCINOMA AND OTHER CUTANEOUS MALIGNANCIES**

Depth/invasion	More than 2 mm of thickness Clark level IV or greater Perineural invasion
Location	Ear Non-hair-bearing lip
Differentiation	Poorly differentiated or undifferentiated

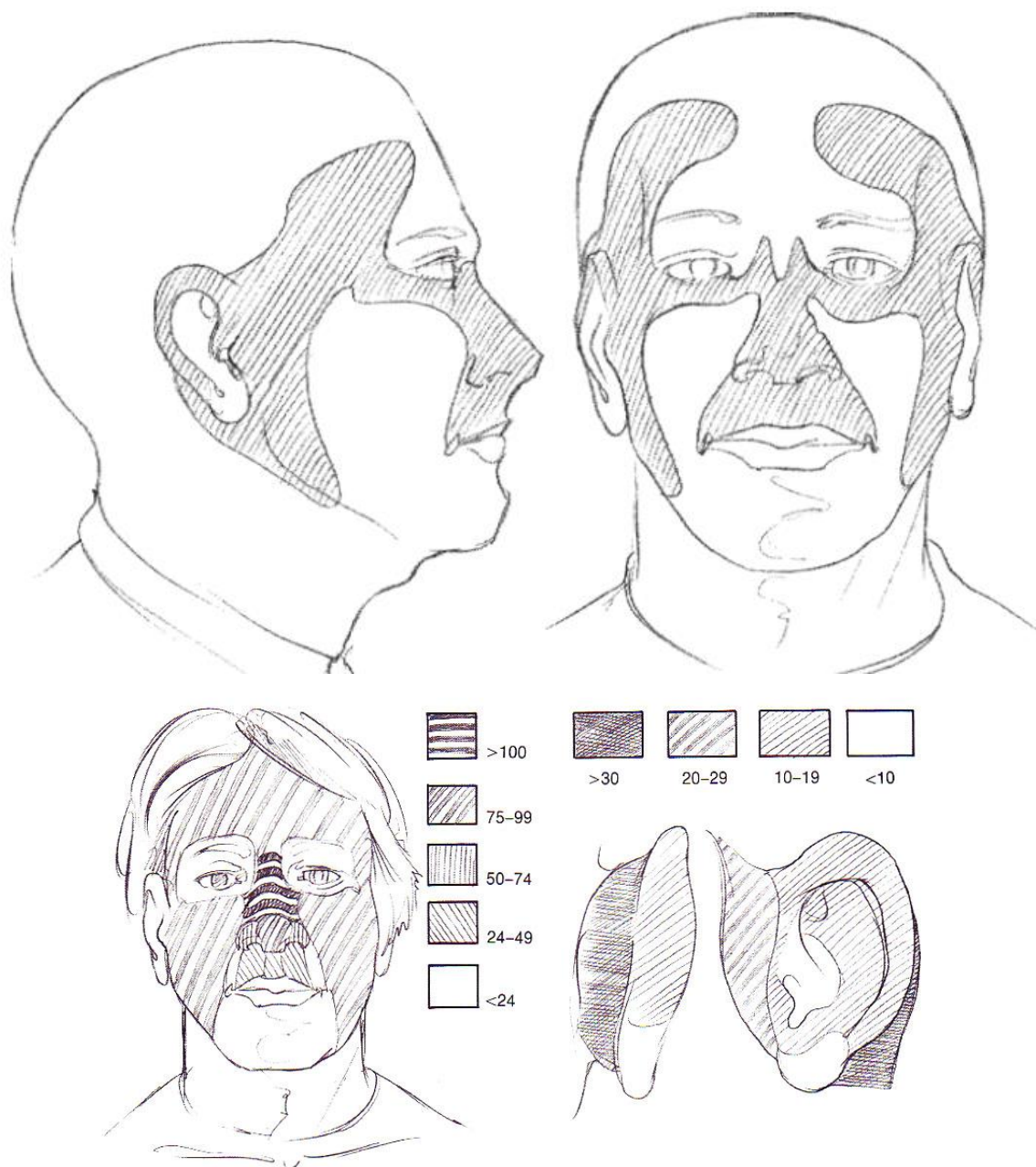
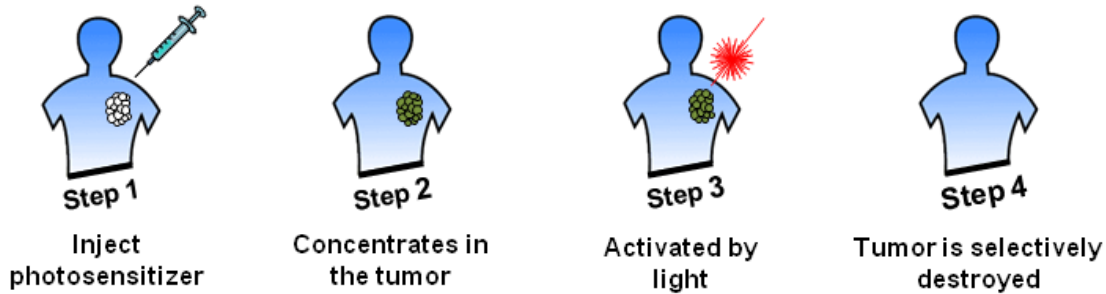


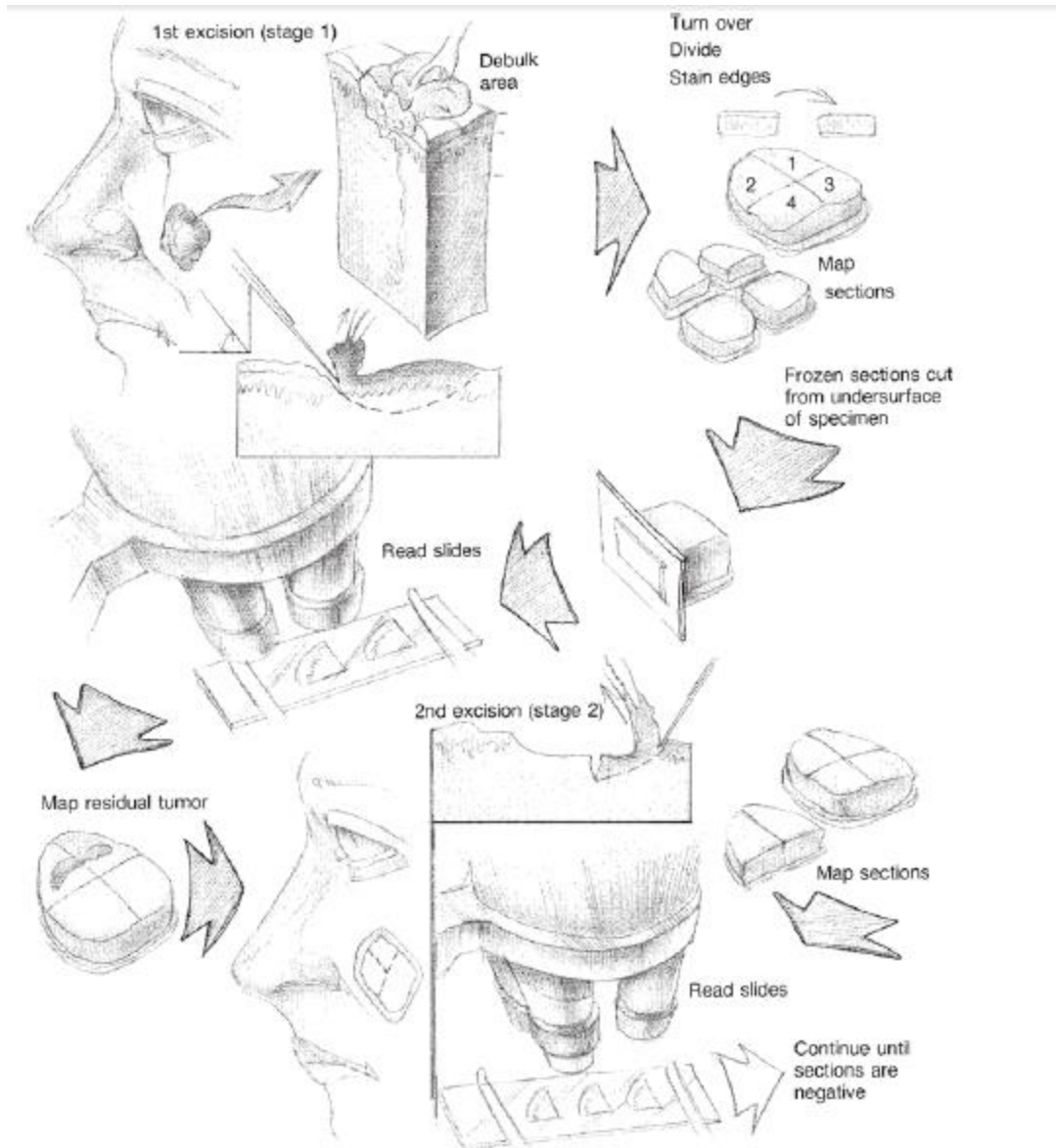
Figure 105.7 Distribution of recurrent cutaneous malignancies.

Management

- avoid excess sun exposure (sunblock)
- careful follow-up for recurrence and second primaries every 4–6 months
- **Curettage with electrodesiccation**
 - most Common for **BCC**; cure rates of 92-98%; dermatologists use it
 - Rational: tumours have soft feel that is evident when curetting
 - Once normal tissue felt over entire base, burn it; repeat 2-6 times; treat topically then allow secondary healing
 - Advantages: expedient; easy; sparing of normal tissue
 - Disadvantages: open wound; scarring; delayed bleeding; only **good for lesions < 2 cm D**; **SCC shouldn't be done this way**
 - **Contraindications: deep invasion; morphea BCC; recurrence**
- **Cryosurgery**
 - Appropriate for some BCC lesions; **not for SCC**
 - Most common is **liquid nitrogen** (–30°C lethal to malignant cell)
 - Freeze lesion and margin of surrounding tissue; allow to thaw; repeat freeze-thaw cycle
 - requires a 5 mm margin, may be considered for small (<1 cm) lesions
 - Advantages: high cure rate (for appropriately selected lesions); tissue sparing; expedient; useful over cartilage or in those with multiple lesions
 - Disadvantages: prolonged healing and wound care; **hypopigmentation** and **scarring**; **not for morphea,, SCC or recurrence**
- **Radiation therapy**
 - Can cure most; extensive use in past
 - Advantages: considered where cosmetic outcome is important (eyelid, nose, lip) or avoid surgery; wide field treatable
 - Disadvantages: protracted course; expense; adjacent tissue effects; **not** effective if cartilage/bone involved; radiodermatitis and delayed carcinogenesis possible
 - Currently used as advanced stages post surgical adjunct or palliation
- **Photodynamic therapy (PDT)**
 - Photosensitizing drug; light; preferential tumour necrosis
 - Investigational modality only at this point
 - **Porphyrin** most widely used drug; laser for light
 - Advantages: multiple lesions treatable at once; good cosmetic results; no anesthesia required
 - Disadvantages: severe pain and skin necrosis common; lack of predictable response; occasional photosensitivity



- **Interferon- α**
 - Investigational modality; excellent results for nodular BCC
 - Low-dose intralesional IFN- α 3x/wk
 - Mechanism: immunomodulating & antiproliferative properties; focal increase in host response to neoplastic tissues
 - Disadvantages: local pain and persistent erythema common; flu-like illness; leuko/thrombocytopenia
- **Excisional surgery**
 - 93-95% success rates reported; can also use CO₂ laser
 - should have 4 mm margin margins for early disease and 1-2 cm margins for advanced disease
 - Advantages: tissue for Dx; assess margins; excellent cosmesis
 - Disadvantages: time consuming, expensive and inconvenient for patients
- **CO₂ laser**
 - Laser indicated in those who can't tolerate epi in local (cardiac status or other medical condition)
 - Good for ablation of multiple small lesions not needing recon (7-8 mm largest size; heals well in 10 days)
 - Good for palliation of multiple large lesions
- **Mohs micrographic surgery**
 - Developed it as a student in 1930s; serial excision & mapping
 - Cure rates from 96-99%;
 - Indications: recurrence, morphea type, high risk of recurrence regions ("H-zone"), cosmetic regions in which reconstruction is difficult (eyelids, lips, nose, ears, nasolabial fold)
 - Advantages: examine margins entirely as you go; re-excise areas of microscopic foci; maximal preservation of surrounding tissue; can immediately reconstruct rather than waiting for path; highest cure-rate for advanced or recurrent lesions
 - Disadvantages: special expertise; time-consuming; expensive; limited availability



- **Neck Dissection and Superficial Parotidectomy:** indicated for clinically positive nodes only
- **Postoperative Radiation Therapy:** may be considered for
 - close or positive margins
 - multiple positive neck nodes or extracapsular extension
 - perineural or intravascular invasion
 - recurrent lesions
 - bone or cartilage invasion
- **Complications**
 - Inadequate excision – too worried about cosmesis/function
 - Nodal mets – 0.5% of BCC and up to 12.5% of SCC

Malignant Melanoma:

Epidemiology

- **15-30%** of melanomas present in head and neck (most common)
- site in the head and neck is on the skin of the **cheek** and the **occipital scalp**
- Number of new cases increases by 5% annually
- 2% of all US cancer deaths;
- M:F = 2:1; median age 55
- Increased relative risk in:
 - Fair skinned people who burn easily
 - Geographic location (intense **sunlight** – Australia)
 - Light pigmentation; 10x higher risk in whites than blacks
 - **UV-A** and **UV-B** both implicated in lab setting
 - B-ras oncogene positive in >65% of cutaneous melanoma
 - **skin lesions** "50-70% of all melanoma arise from preexisting benign nevi"
 - Large congenital nevi (> 20 cm)
 - Multiple benign nevi (>20 increases relative risk to 12:1)
 - Junctional nevi
 - dysplastic nevi " dysplastic nevus syndrome, aka Familial atypical multiple mole-melanoma (FAMMM) syndrome"
 - Xeroderma pigmentosa "rare autosomal recessive genetic disorder of DNA repair in which the ability to repair damage caused by ultraviolet (UV) light is deficient"



Clinical Presentation and Diagnosis

- Classic finding: **pigmented lesion** that changes over **weeks to month**
- ABCD's of Melanoma:
 - **A**symmetric" **A**pperance (irregular shaped)
 - **B**order irregularity
 - **C**olor variation (color changes within the lesion)
 - **D**iameter (**>1 cm**, increasing size), also bleeding and ulceration
- **Dx**: excisional biopsy of suspicious lesions (shave biopsy, curettage, and laser excision is **contraindicated** if melanoma is suspected)
- **Common Immunohistochemical Markers**
 - **HMB-45** (Sensitive and specific, does not stain spindle cell type).
 - **S-100** (High sensitivity, Low specificity, also found in neural and cartilaginous tumors).
 - **Mitf and Tyrosinase** (Sensitive and specific).
 - **MART-1 and melan-A** (Newer, sensitive and specific for melanocytes).
- Not all are pigmented; 10% can have no melanin
- Relevant history: family history of skin cancer; Hx of severe sunburn
- Exam: look at any/all pigmented lesions

CLINICOPATHOLOGIC SUBTYPES

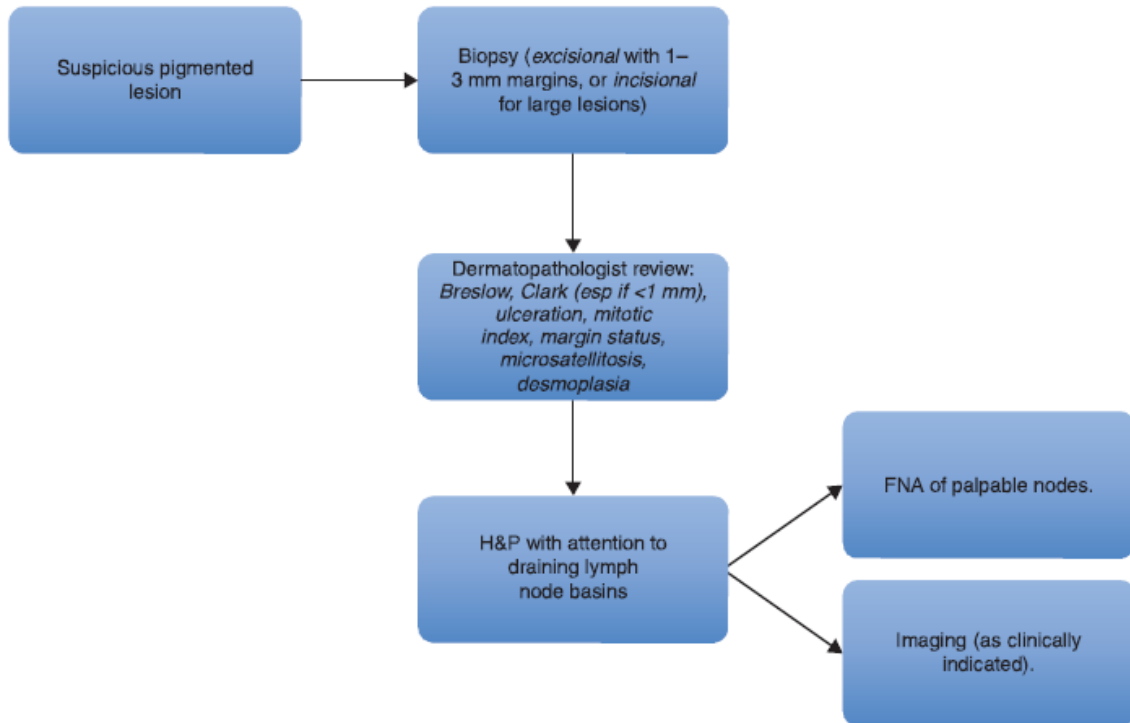
1. Superficial Spreading Melanoma
2. Nodular Melanoma
3. Lentigo Maligna Melanoma (LMM)
4. Acral Lentiginous Melanoma
5. Desmoplastic (very rare histo variant)
6. Mucosal
7. Metastatic with unknown primary

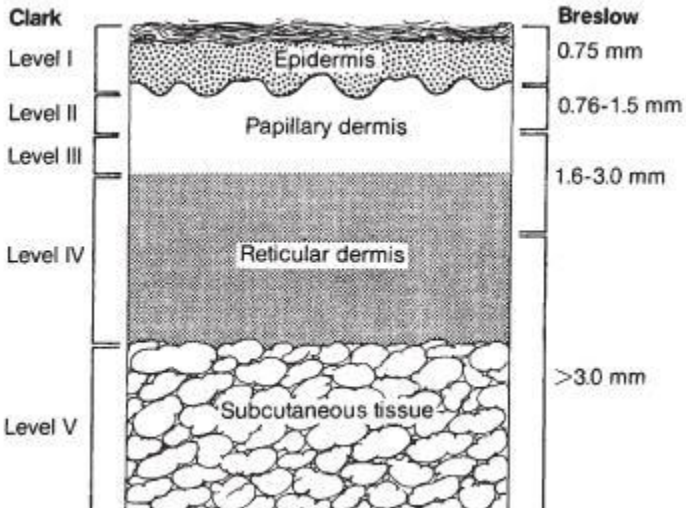
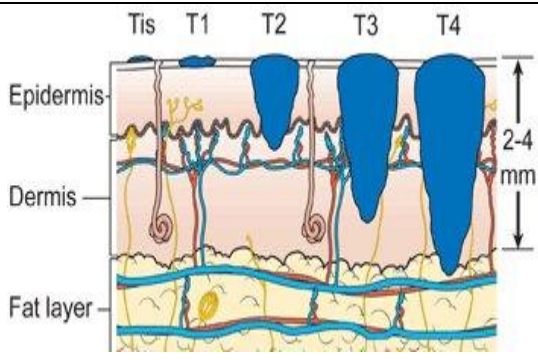
- **Radial growth**: intraepithelial; confined to junction
- **Vertical growth**: intradermal; invasion through junction
- **DDx**: seborrheic keratosis; benign nevi; hemangioma; blue nevi; pyogenic granuloma; pigmented BCC

Sub type		
Lentigo maligna (not LMM) Hutchinson's freckle	- AKA melanoma in situ; common in elderly - Atypical melanocytes; spread radially along junction; focal nesting; can extend along appendages - Up to 5% progress to invasive LMM	
Lentigo maligna melanoma (LMM)	- Progression from above; 6-10% of cases least common Best prognosis - only 30% ever metastasize - Slow radial growth phase; up to 10 yrs to progress 0.5 cm margins on resection	
Superficial spreading melanoma	Most common; 75% of cases - Wide color variety: pink; blue-gray; brown; tan; black Late invasion "Radial growth for 5-7 yrs; then invasion (ulcer and bleed)" Intermediate prognosis "High cure rates if detected before invasive phase"	
Acral lentiginous	- Most common in blacks - Soles of feet, palms of hands and oral/anogenital mucosa - not from sun Rarely seen in H&N	

• Nodular melanoma	• 10-15% of all melanoma may affect any part of body • Worst prognosis due to vertical growth, early invasion • Usually in pts > 50 yrs;	
• Mucosal Melanoma	• Only 2% of all H&N melanomas; > 50% are in nasal cavity , hard palate • Poor prognosis; advanced stage once detected • Characteristic melanosis may be absent • Depth of invasion has no prognostic value in these lesions • 5-yr survival 10-45%; NC better than OC	
• Desmoplastic Melanoma	• < 1% of overall cases; 75% of them in H&N • Amelanotic; neurotropism (perineural invasion/spread); • lower rates of regional mets, but higher recurrence rates from neural spread • Wider margins and rads recommended • 10% can have no melanin	

- Before biopsy, ulceration must clearly be documented
- Excisional biopsies preferred if possible; include fat for depth analysis



Primary Tumor Staging		
Breslow classification	Clark classification	T-staging for TMN
thickness in millimeters (from granular layer) - Stage I :- less or equal to 0.75mm - Stage II : 0.75 mm - 1.5mm - Stage III :1.51 mm - 2.25mm - Stage IV : 2.25 mm - 3.0mm - Stage V : greater than 3.0 mm Not used anymore	depth of invasion based on tissues - Level I – Only epidermis (in situ melanoma); no invasion - Level II – Invades papillary dermis but not papillary-reticular dermal interface - Level III – Invades and expands papillary dermis up to the interface with, but not into, reticular dermis - Level IV – Invades reticular dermis; not into subQ tissue - Level V - Invades into subcutaneous tissue	(any Ta has no ulcer; any Tb has ulcer) - Tis – Melanoma in situ; involves only epidermis (Clark I) - T1 – Tumour 1 mm or less in thickness - T2 – Tumour 1.01-2 mm in thickness - T3 – Tumour 2.01-4 mm in thickness - T4 - Tumor greater than 4 mm in thickness +/- invades subQ +/- satellite(s) within 2 cm of the primary tumor
		

Regional Node Staging

- Breslow: 1 no risk of nodes; 2 has 25%; 3 has 50%; 4 has > 75%
- Number of nodes more predictive than size of nodes
- N-staging for TNM

**TABLE
114.1**

CUTANEOUS MELANOMA TNM STAGING

Melanoma TNM Classification

T Classification	Thickness	Ulceration Status
T _{is}	Not applicable	Not applicable
T1	≤1.0 mm	A: w/o ulceration and mitosis <1/mm ² B: with ulceration or mitosis ≥1/mm ²
T2	1.01–2.0 mm	A: w/o ulceration B: with ulceration
T3	2.01–4.0 mm	A: w/o ulceration B: with ulceration
T4	>4.0 mm	A: w/o ulceration B: with ulceration
N Classification	Number of Metastatic Nodes	Nodal Metastatic Mass
N1	One node	A: micrometastasis ^a B: macrometastasis ^b
N2	—Two to three nodes	A: micrometastasis ^a B: macrometastasis ^b C: in-transit met(s)/satellite(s) without metastatic nodes
N3	Four or more metastatic nodes, or matted nodes, or in-transit met(s)/ satellite(s) with metastatic node(s)	
M Classification	Site	Serum LDH
M1a	Distant skin, subcutaneous, or nodal metastases	Normal
M1b	Lung metastases	Normal
M1c	All other visceral metastases	Normal
	Any distant metastasis	Elevated

^aMicrometastases are diagnosed after sentinel or elective lymphadenectomy.

^bMacrometastases are defined as clinically detectable nodal metastases confirmed by therapeutic lymphadenectomy or when nodal metastasis exhibits gross extracapsular extension.

- Detection of occult nodal disease
 - Controversial; PET may be useful; not gold standard yet
- poorer prognosis also associated with the [presence of ulceration, scalp lesions, microscopic satellites, and nodal disease](#)
- distant metastasis has a grave prognosis

Primary Resection and Neck Dissections

- Current margin recommendations
 - Tis , T1 < 1 mm depth: 0.5 cm down to fascia
 - T2 1-2 mm depth: 1.0 cm down to fascia
 - T3 2-4 mm depth: 2.0 cm down to fascia
 - T4 > 4 mm depth: > 2.0 cm down to fascia
- Should delay recon until permanent margins are reported
- Neck dissections for suspected nodal mets
 - **For face, ear and ant scalp:** **anterolateral neck and parotid**
 - **For postauricular and post scalp:** **posterolateral neck**
- N0 Neck: elective neck dissections not indicated
- N1–3 Neck: neck dissection (may require a superficial parotidectomy), may consider chemotherapy
- Elective lymph node dissection
 - Large trial showed survival benefit only in those < 60 yrs or those with lesions 1-2 mm in depth
 - **Mucosal Melanoma (T):**
 - **T3:** Mucosal disease.
 - **T4a:** Tumor involving deep soft tissue, cartilage, bone, or overlying skin.
 - **T4b:** Tumor involving brain, dura, skull base, lower cranial nerves (IX, X, XI, XII), masticator space, carotid artery, prevertebral space, or mediastinal structures.
 - **Mucosal Melanoma Regional Lymph Node Metastasis (N0-1):**
 - **N0:** No regional LN metastasis.
 - **N1:** Regional LN metastasis present

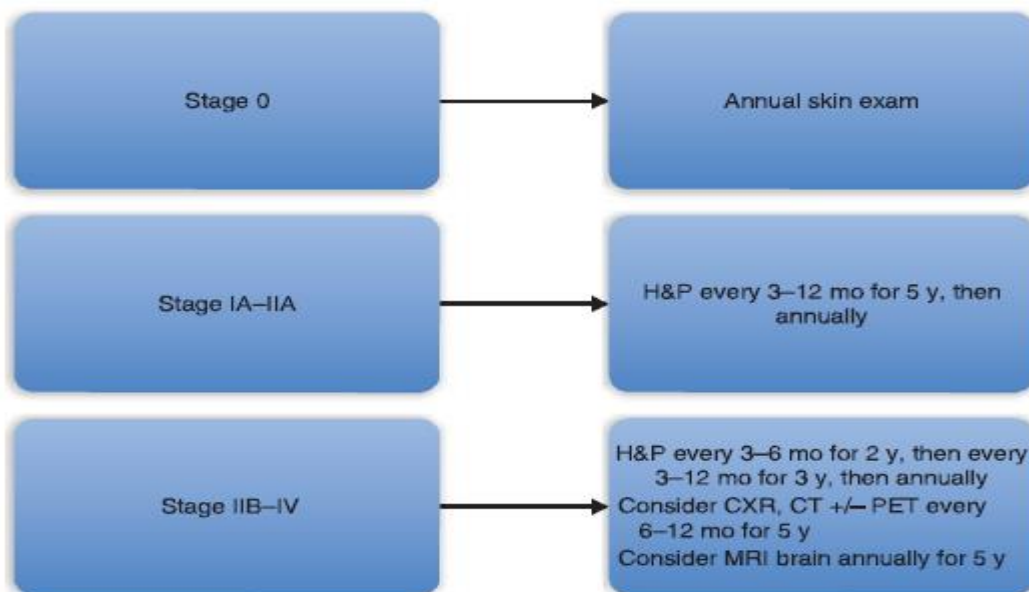
Radiotherapy

- Indications for rads as primary therapy for melanoma
 - Poor surgical candidates
 - Extensive facial "Lentigo maligna melanoma" that precludes adequate surgical resection based on cosmetic considerations
- Postop adjuvant for
 - High risk lesions, defined as >1.5 mm depth or ulcerated lesion
 - Positive Regional Metastasis
 - Desmoplastic histology

Systemic Therapy

- Agents: interferon, ipilimumab, dacarbazine, imatinib
- Two major indications
 - Adjuvant therapy for those who have completed primary treatment and are at risk of systemic relapse ((Primary Ulcerated lesions, >4 mm depth or Clark level IV, Satellitosis, in-Transit Metastases, Nodal metastase)
 - Presence of systemic mets
- **Interferon- α**
 - Well-studied; yet to show improved survival (M.Davis says 3% survival benefit)
 - increase NK cell activity; increase free radical production in M-phages
 - Toxicity: fever; chills; myalgias; fatigue; anorexia
- Systemic therapy for mets
 - Very poor prognosis
 - Liver, brain or bone mets have median survival of 3-4/12
 - Chemo option & combined with IL-2 or IFN- α (biochemotherapy)

Surveillance



Prognosis and Metastasis Potential

- overall 5–20% recurrence rate

Table 5–2. Melanoma 5-Year Survival and Metastasis Potential Based on Pathological Staging

Stage	5-Year Survival	Nodal Metastasis	Distant Metastasis
pT ₁	>95%	2–3%	
pT ₂	80–94%	20–25%	8%
pT ₃	40–84%	57%	15%
pT ₄	10–30%	62%	72%

Head and Neck Reconstruction Pearls:

Reconstruction Ladder:

- 1- 2ndary intension
- 2- Primary intension
- 3- Skin graft
- 4- Tissue Expander
- 5- Local flap
- 6- Regional pedicled flaps
- 7- Free flap

Types of Grafts:

1- AUTOGRAFT

- From patient own tissue
- Bone (Calvarium, Iliac crest, Septal, Rib, Fibula)
- Cartilage (Septal, Costal, Conchal)
- Fascia (Temporalis , Fascia lata)

2- ALLOGRAFT/HOMOGRAFT:

- Same species
- Irradiated cadaveric

3- XENOGRAFT:

- Different species
- Animal
- Not used anymore

4- ALLOPLAST:

- Synthetic material (Silastic, Gortex, Medpore, Calcium hydroxiapatite)

SKIN GRAFT

- Should be done over vasclurized bed
- NOT over -> Bone or Tendon
- Mechanism of healing:
 - 1- ADHERENCE
 - o Fibrin bonds
 - 2- IMBIBITION
 - o 2nd-4th day
 - 3- REVASCULARIZATION
 - o Starting from 4th day

TYPES OF SKIN GRAFTS

1- STSG:

- o Full epidermis + part of dermis
- o LESS Primary contracture
- o MORE Secondary contracture
- o Advantages:
 - 1- Better survival
 - 2- Easy to mesh -> Increase surface area
 - 3- No closure of donor site

- Disadvantages:
 - 1- Poor color match
- Indications:
 - 1- Oral/Nasal mucosal loss
 - 2- Orbital cavity
 - 3- Donor site defect

2- FTSG:

- Entire epidermis and dermis
- MORE primary contracture
- LESS secondary contracture
- Advantages:
 - 1- Better color match
- Disadvantages:
 - 1- High graft failure
 - 2- Slow Revascularization
- Indications:
 - 1- Nasal defects

Causes of Skin graft failure:

- 1- HEMATOMA
 - Most common cause
 - Avoided by
 - A- Hemostasis
 - B- Meshing
 - C- Pressure dressing
- 2- INFECTION
 - Bacteria count > 10(5)
- 3- SEROMA
- 4- GRAFT MOVEMENT:
 - Disrupt neovascularization between graft and its bed
- 4- INAPPROPRIATE BED:
 - NOT Grow on Tendon, Cartilage or Bone
- 5- IRRADIATED FIELD

LOCAL FLAPS:

- Adjacent tissue transfer
- TYPES:
 - 1- ADVANCEMENT
 - 2- ROTATIONAL
 - 3- TRANSPOSITION

REGIONAL PEDICLED FLAPS:

- Advantages:

- 1- Quick and easy to harvest
- 2- Minimal donor site morbidity

- Disadvantages:

- 1- Bulky
- 2- Limited range of mobility
- 3- Distal necrosis

- TYPES:

1- PEC MAJOR:

- ThoracoAcromial artery
- Most common in H&N
- Reach up to lateral canthus

3- TRAPEZIUS:

- A- Superiorly based: Occipital artery
- B- Lateral island: Transverse cervical artery
- C- Inferiorly based: Transverse cervical + Dorsal scapular artery

4- LATISSMUS DORSI:

- ThoracoDorsal artery
- Reach up to skull vertex

5- SCM:

- A. Superior 1/3: Occipital artery
- B. Middle 1/3: Superior thyroid artery
- C. Inferior 1/3: SupraScapular artery

6- DELTOPECTORAL:

- Internal mammary artery

6- TEMPEROPARITAL FASCIA:

- Superficial temporal artery

7- PARAMEDIAN FOREHEAD:

- SupraTrochlear + SupraOrbital artery

8- NASOLABIAL:

- Angular artery

9- PLATYSMA:

- Submental artery

10- POSTAURICULAR:

- Postauricular artery

FREE FLAP:

- Advantages:
 - 1- Potential for sensate, motor & secretory function.
 - 2- Improved vascularity and healing
 - 3- Unrestricted positioning and reach
 - 4- Large amount of composite tissue
- Disadvantages:
 - 1- Longer time
 - 2- Requires experience
 - 3- Risk of failure
- Pre-op:
 - 1- NO IV lines at site of flap
 - 2- Allen test for RFFF
 - 3- Ateriograph for Fibula flap
- Post-op:
 - 1- Avoid hypotension and hypovolemia
 - 2- No pressor
 - 3- Aspirin and Heparin
- 4- Flap monitoring:
 - A. Color
 - B. Temp
 - C. Pulse
 - D. Prick
 - E. Capillary refill

FLAP FAILURE:

- 5-15%
- Within 1st 3 days
- Neovascularization complete on 8th day
- 50% salvage rate
- Types:
 - 1- Venous anastomosis failure
 - Most common
 - Dark flap
 - Dark blood on prick
 - 2- Arterial anastomosis failure:
 - Pale flap
 - No blood on prick

COMMON FREE FLAPS:

1- RFFF

- Radial artery
- Allen test or Doppler pre-op to evaluate ulnar collateral
- Fascio/osseocutaneous
- 10 cm of bone
- Risk of pathological fracture
- Not fit for dental implants

2- ALT:

- Descending branch of Lateral circumflex femoral artery
- Fascioutaneous

3- FIBULA:

- Peroneal artery
- Osseocutaneous
- Best for large bony recon
- 25cm of bone
- Allows dental implant
- Needs pre-op Arteriogram to r/o peripheral artery disease

4- SCAPULA:

- Circumflex scapular artery
- Osseocutaneous
- 15cm of bone
- Not fit for dental implants

5- ILIAC CREST:

- Deep circumflex iliac artery
- Osseomyocutaneous
- 15cm of bone
- Allows dental implants

6- GRACILIS

- Adductor artery
- Muscle flap
- For facial re-animation

7- JEJUNUM:

- Superior mesenteric artery
- Best for CERVICAL esophagus
- Contraindicated for THORACIC esophagus -> Use Gastric pull up instead

Mandibulectomy:

1. MARGINAL Mandibulectomy:

- Resection of partial thickness (alveolar process or the lingual cortex)
- while leaving the mandibular arch intact (>1cm in height should be retained to avoid a stress fracture).
- Indications:
 1. Tumors of oral cavity adjacent to or involving the periosteum of the mandible without gross cortical invasion.
- Contraindications:
 1. Irradiated mandible
 2. Edentulous patients
- Types:
 1. Horizontal
 2. Vertical
 3. Oblique

2. SEGMENTAL Mandibulectomy:

- Resection of full-thickness segment of the mandible.
- Indications:
 1. 1-Gross bone erosion clinically or radiologically
 2. 2-Significant paramandibular disease
 3. Irradiated mandible
 4. Edentulous
- Types:
 1. Mid segmental
 2. Posterior segmental (posterior to mental foramen)
 3. Hemi-mandibulectomy

Reconstruction of Mandible:

- Avoid ANDY GUMP deformity
- **Reconstruction of MARGINAL Mandibulectomy:**
 - o Options:
 1. No plating
 2. AO plate to support remaining mandible
- **Reconstruction SEGMENTAL Mandibulectomy:**

1. AO PLATES ONLY:

- ➔ Risk of Extrusion
- ➔ Indications:
 1. Lateral defects
 2. Old patient
 3. Poor prognosis

2. AO PLATES + TISSUE TRANSFER:

- Reduce extrusion rate
- Free Flap (RFFF/Best) or Regional flap (PEC Major/Elderly)

3- OSSEOCUTANEOUS FLAP:

- Gold standard
- Indications:
 1. Large bony defect > 5cm
 2. Irradiated patient
- OPTIONS:
 1. **FIBULA (BEST)**
 - Peroneal artery
 - 25cm of bone
 - Allows dental implant
 2. **ILIAC CREST:**
 - Deep circumflex iliac artery
 - 15cm of bone
 - Allows dental implants
 3. **SCAPULA:**
 - Circumflex scapular artery
 - 15cm of bone
 - Not fit for dental implants
 4. **RFFF:**
 - Radial artery
 - 10 cm of bone
 - Risk of pathological fracture
 - Not fit for dental implants

Glossectomy:

- Types of Glossectomy
 1. **Partial Glossectomy:**
 - Excision < 1/3 of tongue
 - Closed by primary intension
 2. **Hemiglossectomy:**
 - Excision of 1/3-1/2 of tongue
 - Close by primary intension vs STSG vs RFFF
 3. **Near Total Glossectomy:**
 - Excision of > 1/2 of tongue
 - Closed with Free flap
 - RFFF
 4. **Total Glossectomy:**
 - Total excision of tongue including tongue base
 - Closed with Free flap (ALT)

Maxillectomy:

- Types of Maxillectomy:

1- MEDIAL MAXILLECTOMY:

- Resection of medial wall of maxilla
- Lateral nasal wall
- Endoscopic vs Open
- Indicated in Inverted Papilloma

2- INFERIOR MAXILLECTOMY:

- Resection of maxilla inferior to Infra-orbital nerve
- Indicated for:
 1. Hard palate Ca
 2. Maxillary alveolar Ca

3- TOTAL MAXILLECTOMY:

- Enblock resection of entire maxilla including Medial wall, Orbital floor, Hard palate, Medial orbital wall.
- Indicated for Maxillary Ca

4- RADICAL MAXILLECTOMY:

- Total Maxillectomy + Orbital exsentrination

APPROACHES of Maxillectomy:

1- LATERAL RHINOTOMY:

- For Medial Maxillectomy

2- WEBER-FERGUSON:

- Lateral Rhinotomy + subcilliary
- For Total Maxillectomy

3- TRANS-PALATAL:

- For Inferior Maxillectomy

4- MIDFACIAL DEGLOVING:

- For Medial or Inferior Maxillectomy

RECONSTRUCTION of Maxillectomy:

1. OBTURATOR:

- Quick
- Allows Naso-oral separation
- Allow for recurrence surveillance
- Risk of nasal regurgitations
- Needs repeated size adjustment

2. TITANIUM MESH:

- Reconstruct floor of orbit

3. BONE GRAFT:

- Reconstruct floor of orbit

4. OSSEOCUTANEOUS FLAP:

- Fibula (Best)

HYPOPHARYNGEAL Reconstruction:

1. Small Partial Defects:

- Remaining mucosa (>3cm)
- Closed by PRIMARY intension

2. Large Partial/Circumferential:

- No remaining mucosa or (<3cm)
- Closed by:
 1. Tubed ALT or RFFF
 2. Jejunum
 3. Gastric pull up
 4. PEC Major

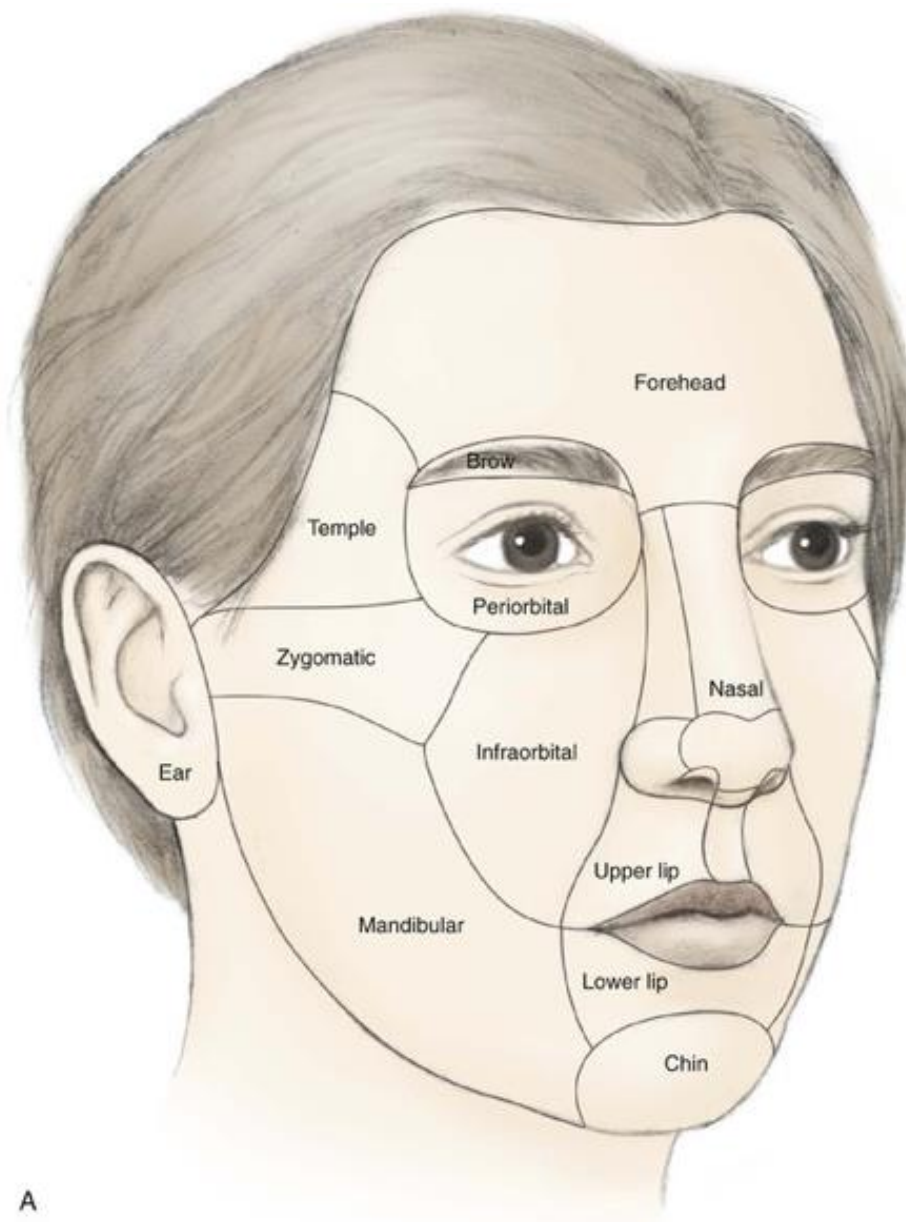
ESOPHAGEAL Reconstruction:

- CERVICAL Esophageal:
 - Tubed ALT
 - Jejunum
- THORACIC Esophageal:
 - Gastric pull up

Facioplasty:

- 1- Facial Analysis**
- 2- Anatomy of External Nose**
- 3- Rhinoplasty**
- 4- Otoplasty and Auricular Reconstruction**
- 5- Scar Revision**

Facial Analysis



Preoperative Assessment

- Establishing a Relationship:
 - Establish a good relationship.
 - Assess psychological stability and motivation for surgery.
- Patient Selection: Concept of Deformity
 - Body dysmorphic disorder:
 - Preoccupation with a minor or imagined defect.
 - Ensure no dramatic difference is present between pt's perceived self-image and the surgeon's observation.
 - S.I.M.O.N: (single, immature, male, overly expectant, narcissist).
- Preoperative Interviews:
 - Establish good communication.
 - Patient's expectations; are they realistic and reasonable; can they be fulfilled; can the patient be satisfied.
 - Having a family member or friend present can be helpful.
- The Sensitization, Aesthetic self-assessment. Peer Group comparison and Avoidance behaviors (SAGA).

**TABLE
170.1**

**RED FLAG SYMPTOMS. PATIENTS
EXHIBITING "RED FLAGS" MAY
WARRANT FURTHER INVESTIGATION**

Trying to please someone else
Unrealistic expectations
Pervasively unhappy
Poor self-image
Overly flattering
Perfectionists
Rude or demanding
Know-it-all
History of doctor-shopping
History of litigation
Multiple revision surgeries

Psychopathology in the Aesthetic Patient

- Depression:
 - Half of aesthetic surgery pts display post-op depression, even with good outcome.
 - Depression is not a contraindication but there is a risk of worsening depression.
 - They should undergo pre-op psych assessment.
- Personality Disorder
 - Most common psychopathology among patients seeking cosmetic surgery.
 - Poor candidates often exhibiting body dysmorphism.
 - Most commonly, borderline personality.
 - Need to handle them firmly; establish that no manipulation can occur.
 - Narcissist and histrionic personality

Body dysmorphic disorder:

- Only contraindication among psychiatric disorders.

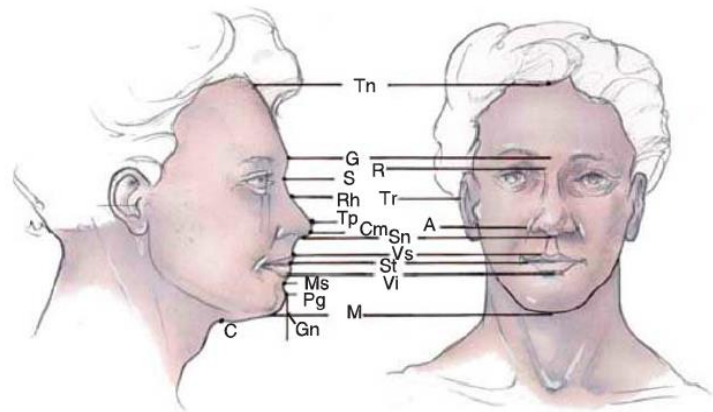


Figure 170.2 Basic anatomic landmarks for facial analysis. See Tables 170.2 and 170.3 for explanation.

**TABLE
170.2**

**BASIC ANATOMIC LANDMARKS
AS SEEN ON FRONTAL VIEW**

Trichion (Tn)	Superior margin of the forehead at the midline of the frontal hairline
Radix (R)	The region of the root of the nose
Subnasale (Sn)	Soft tissue point at the junction of the columella and upper lip in the midline
Superior and inferior vermillion border (Vs and Vi)	Mucocutaneous junction of the upper and lower lips
Stomion (St)	Midline point at the embrasure of the lips when closed
Menton (M)	Soft tissue point at the inferior-most border of the chin in the midline

**TABLE
170.3**

BASIC ANATOMIC LANDMARKS ON LATERAL VIEW

Nasion (N)	Posteriormost bony point at the root of the nose
Sellion (S)	Posteriormost soft tissue point at the root of the nose
Glabella (G)	Anteriormost point of the forehead in the midline
Rhinion (Rh)	Region at the junction of the bony and cartilaginous nasal dorsum
Tip-defining-point (Tp)	Anteriormost projection of the nasal tip, corresponding to the dome of the lower lateral cartilages
Columellar point (Cm)	Anteriormost point of the columella
Alar crease (A)	Sulcus defining the posterior border of the alar subunit of the nose
Mentolabial sulcus (Ms)	Crease between the lower lip and chin
Pogonion (Pg)	Anteriormost point of the chin in the midline
Gnathion (Gn)	Virtual point defined by the intersection of a vertical line lying tangent to the pogonion and a horizontal line that is tangent to the menton.
Cervical point (C)	Virtual point defined by the line tangent to the anterior margin of the neck and a line tangent to the menton
Tragion (Tr)	Point at the supratragal notch of the ear

General Facial Assessment

1- Evaluation of symmetry:

- A line drawn through the **midsagittal** plane on frontal view.
- The midline points of the forehead, nose, lips, and chin should lie on this axis.

2- Dividing the face into vertical fifths:

- Equal to the width of one eye = nose.
- Width to length ratio 3:4.

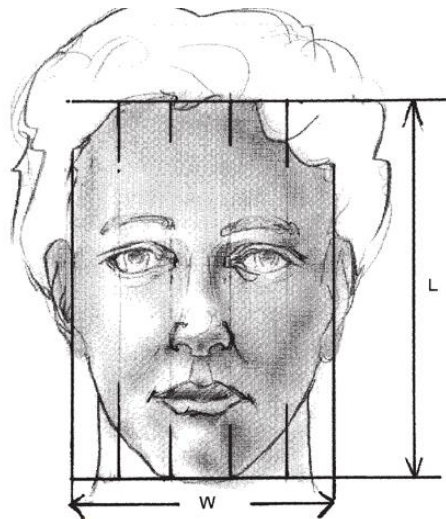


Figure 170.3 Ideal frontal symmetry and proportion is approximated by dividing the face into vertical fifths, with each fifth approximating the width of the eye. The overall width-to-length ratio should approximate 3:4.

3- Dividing the face into horizontal thirds:

- Evaluates facial **height**.
- Examined by on the frontal and lateral views.
- Lower face is divided in 1/3 and 2/3.

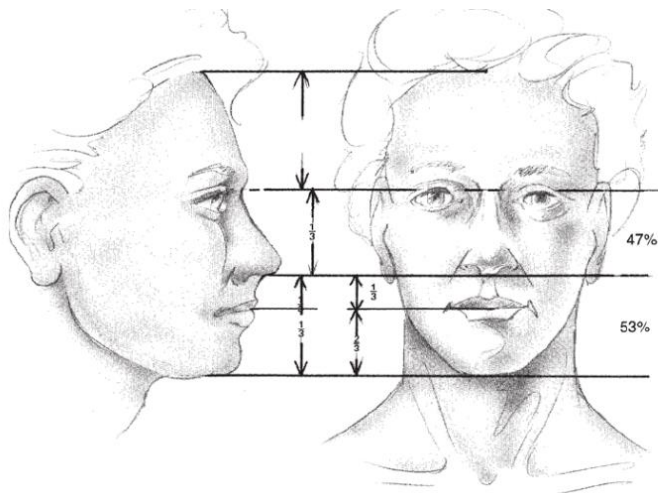


Figure 170.4 The facial thirds: trichion to glabella, glabella to subnasale, subnasale to menton. The lower face also is divided into thirds: The upper lip from subnasale to stomion is one-third, and the lower lip and chin is two-thirds. The face from nasion to subnasale is 47% and from subnasale to menton is 53% of the total height from nasion to menton. (From Powell N, Humphries B. *Proportions of the aesthetic face*. New York: Thieme-Stratton, 1984, with permission.)

4- Frankfort line and zero meridian (Gonzalez-Ulloa):

- Evaluated on facial profile (lateral).

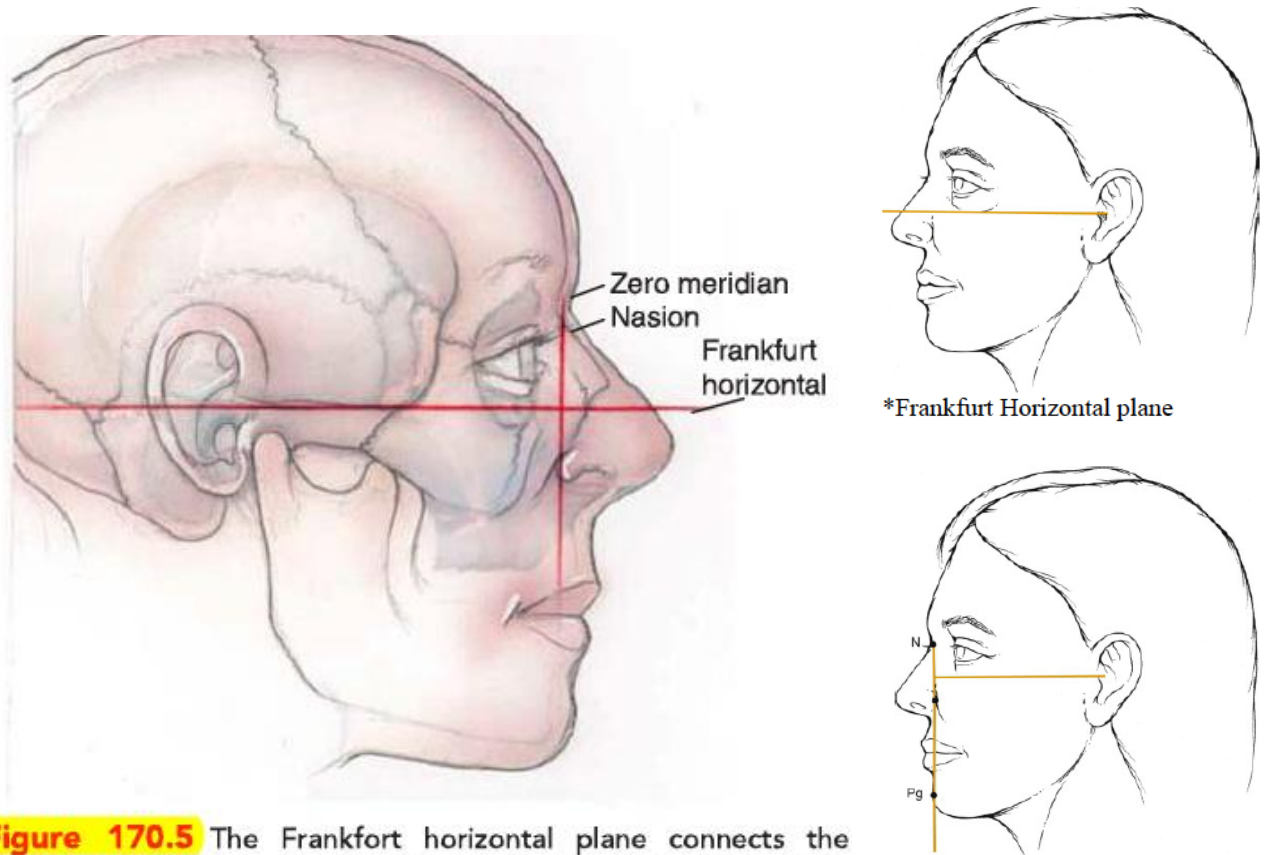


Figure 170.5 The Frankfort horizontal plane connects the inferior orbital rim to the tragus and is parallel to the horizon. Gonzalez-Ulloa defined the aesthetic profile in relation to the Zero Meridian, a line perpendicular to the Frankfort horizontal plane through the nasion. In the aesthetically pleasing profile, the outline of the midforehead, subnasale, upper and lower lips, and pogonion rest on this line (From Gonzalez-Ulloa M. Quantitative principles in cosmetic surgery of the face (profi leplasty). *Plast Reconstr Surg Transplant Bull* 1962;29:186–198.).

- Convex-sloping forehead and retrusive chin
- Concave-retrusive maxilla (dish-face deformity)

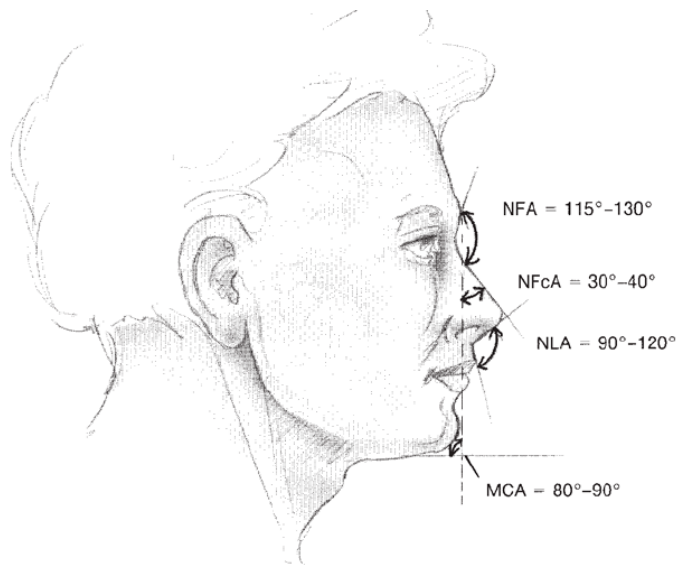


Figure 170.6 Profile angles for facial analysis. See Table 170.4 for definitions.

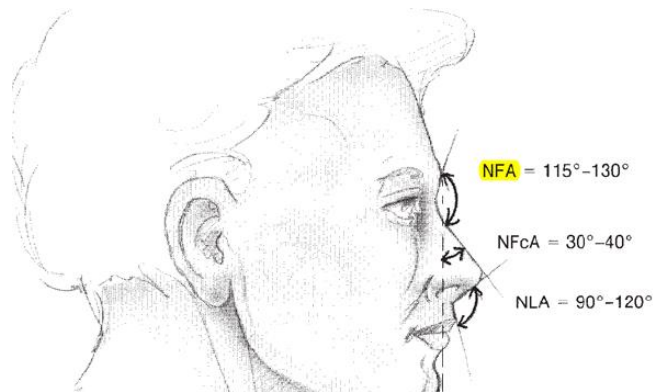
**TABLE
170.4**

PROFILE PLANES AND ANGLES USED IN FACIAL ANALYSIS

Facial Plane	Reference plane created that is tangent to the glabella and pogonion
Frankfort Horizontal	Reference line from the superior margin of the external auditory canal to the inferior border of the infraorbital rim
Zero-Meridian	Reference line perpendicular to the Frankfort Horizontal intersecting at the nasion
Nasofrontal angle (NFA)	Obtuse angle formed by lines tangent to the glabella and nasal dorsum with the vertex at the sellion
Nasofacial angle (NFcA)	Angle of inclination of the nasal dorsum from the facial plane
Nasolabial angle (NLA)	Angle of inclination between the columella and the upper lip
Lower Face-Throat Angle	Angle formed at the gnathion by the intersection of a line from the cervical point to menton with a line from the subnasale to the pogonion.
Mentocervical angle (MCA)	Angle formed by the intersection of a line from the cervical point to menton with a line from the glabella to pogonion.

Forehead:

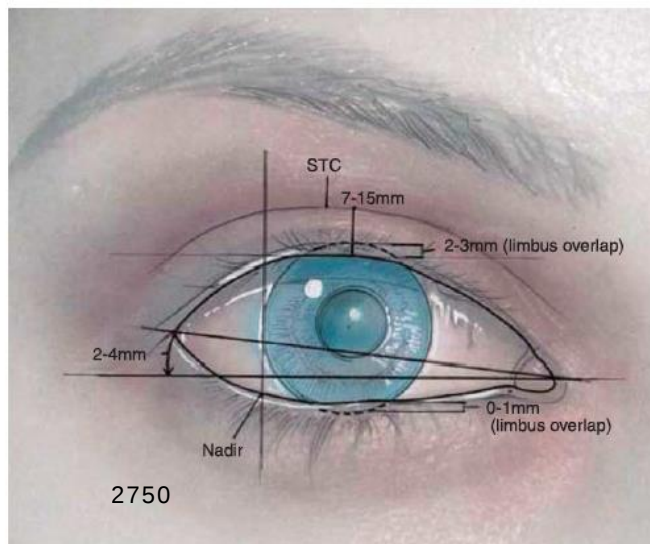
- Attractive female forehead contour has a gentle convexity on profile with its most anterior point at the supraorbital ridge lying tangent to the Zero Meridian.
- Men tend to have a more protruding forehead than women with a degree of supraorbital bossing.
- Men tend to have a more acute NFA when compared to females,
- In men the NFA ideally measures 115 to 120 degrees.
- Women ideally measure 120 to 135 degrees.
- **Position of NFA vertex determines the nasal length**
 - Superior NFA = lengthening of nose
 - Deepening of the angle = increased nasal projection



Eyes:

- Inter-canthal distance = width of 1 eye.
- Average intercanthal distance is 30-35 mm.
- Average interpupillary distance is 60-70 mm.
- Female eyebrows are above the supraorbital rim, with a higher arch; male eyebrow lie is directly over supraorbital rim with less arch.
- Upper lid covers 2-3 mm of superior limbus; lower lid lies below 1-2 mm inferior limbus.
- Distance from lash line to lid crease on the upper lid is 7-15 mm.

Figure 170.8 Characteristic relationships of the aesthetically pleasing eye. The upper eyelid overlaps the superior limbus of the iris by 2 to 3 mm, while the lower eyelid approximates or overlaps the inferior limbus by 1 mm. The supratarsal plate is tallest at the medial limbus. The nadir of the lower eyelid lash line is aligned with the lateral limbus. The distance from the upper eyelid lash line to the supratarsal crease (STC) varies from 7 to 15 mm. The intercanthal axis is tilted upward laterally, with the lateral limbus lying 2 to 4 mm above the medial canthus.



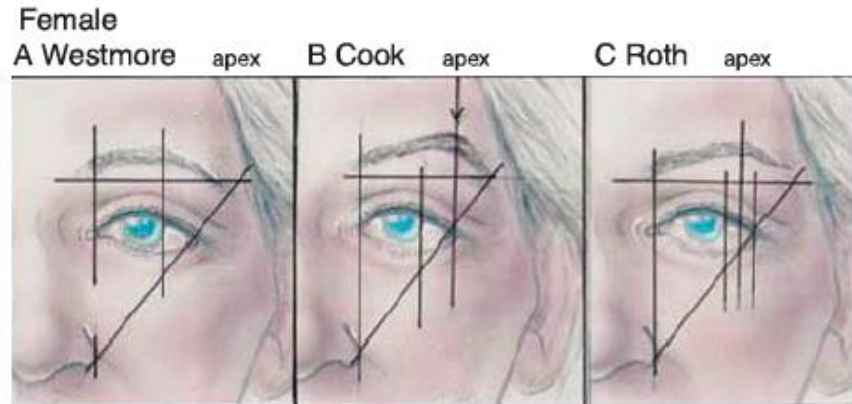


Figure 170.7 **A:** The ideal brow begins at the medial canthus and ends laterally along an oblique line through the alar–facial junction and the lateral canthus. Westmore proposed that the apex of the eyebrow should reach its maximal arch along a line tangent to the lateral limbus. (Adapted from Brennan GH. Correction of the ptotic brow. *Otolaryngol Clin North Am* 1980;13:265–273, with permission.) **B:** Cook Modification: With shifting societal aesthetic ideals, the aesthetic position of the female brow has also shifted. Cook proposed that the peak of the eyebrow lies over the lateral canthus rather than the lateral limbus. **C:** Roth Modification: Most recently, Roth et al. found the ideal eyebrow arch position lies between the lateral limbus and lateral canthus and that the lateral brow lies slightly superior to the medial brow in the horizontal plane.

lower eye lid

- The upper 5 mm has 4 layers: skin, orbicularis, tarsus, and conjunctiva.
- The lower 5 mm has 7 layers: skin, orbicularis, septum, pre-CPF (capsulopalpebral fascia) fat, inferior sympathetic muscle (equivalent to Müller's in the upper lid), CPF (equivalent to levator aponeurosis in upper lid), and conjunctiva.
- Upper eyelid (Fig. 2–6) (mnemonic: 4-5-7 rule):
 - The lower 5 mm has 4 layers: skin, orbicularis, tarsus, and conjunctiva.
 - The middle 5 mm has 5 layers: skin, orbicularis, levator aponeurosis, tarsus, and conjunctiva.
 - Above 10 mm, the upper eyelid has 7 layers: skin, orbicularis, septum, preaponeurotic fat, levator aponeurosis, Müller's muscle, and conjunctiva.

Ears:

- The auricle reaches 80% of the adult size by age 5, permitting early surgical intervention.
- Prominauris: excessively protruding ears.
- Top of the helix should align with lateral eyebrow.
- Inferior ear lobe should be at the level of alar-facial junction.
- Width to length ratio is 0.6:1.
- Longitudinal axis is inclined about 20 degrees off the vertical.
- Angle of protrusion from posterior scalp is about **20-30** degrees.
- In assessing ear protrusion, the distance from the mastoid to the helix (**S**), at the level of the external auditory canal (**M**), and at the lobule (**L**).
- The ideal values for these in the adult should be
 - 1- (S) **16 to 18** mm
 - 2- (M) **14 to 16** mm
 - 3- (L) **16 to 18** mm



* Auricular unit. 1, Helical subunit; 2, antihelical subunit; 3, triangular fossa subunit; 4, conchal subunit; 5, lobe subunit.

Nose:

- Nasal width, length, symmetry, presence of dorsal nasal curvature or deviation.
- Ideal NASAL WIDTH (alar groove to alar groove) = **70% of the NASAL length from nasion to tip-defining point.**
- Nasion at level of upper eyelid.
- Wider interalar distance in Asians and blacks.
- **Nasal length:**
 - Nasal analysis: lower face divided into 2 parts:
 - Nasion-subnasale-menton.
 - Nasal length (nasion-subnasale) should be $3/4^{\text{th}}$ the distance from subnasale-menton.

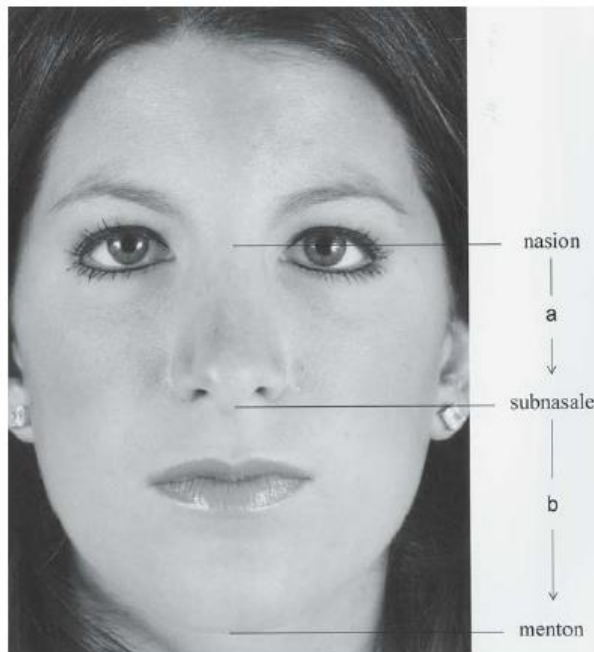


Figure 181.5 Determination of nasal length using the nasion, subnasale, and menton: ideally $a = \frac{3}{4} b$.

- **Nasal Width and Radix Contours:**
 - Nasal width should increase along its length.
 - Maximal width occurs at the alae ($1/5^{\text{th}}$ the width of face).
 - Upper nasal contour should follow a gentle curve from medial eyebrow to ipsilateral tip-defining point ([radix contours or lines](#) can be highlighted by nasal light reflex).

Four profile measures (contour, tip projection, rotation and length)

- **Contour:** should be relatively straight

- Nasal Profile & rotation:

- Extent of protrusion of the rhinion from anterior facial plane determines the degree of nasal hump or sloop deformity.
- Dorsal nasal profile is ideally straight or slight prominence at rhinion.
- Fullness in the supratip region causes a pollybeak deformity.
- Tip projection: extent of protrusion of nasal tip from anterior facial plane.
- Rotation: degree of inclination of the NLA:
 - NLA in males: 90-95 degrees
 - NLA in females: 95-105 degrees
- More rotation is acceptable in short pts.
- **Nasal length:** distance from nasion to tip-defining point.
- Presence or absence of a **supratip breakpoint** depends on the dorsal height of the cartilaginous septum and the projection of the domal region of the LLCs.

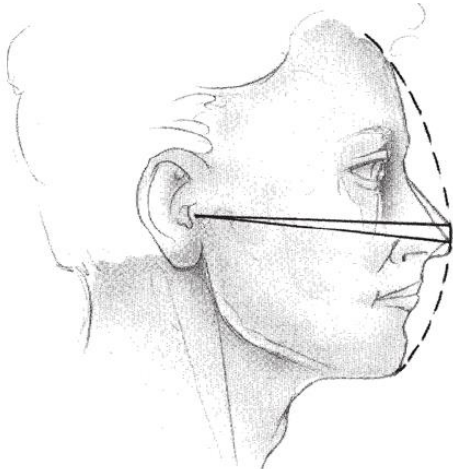
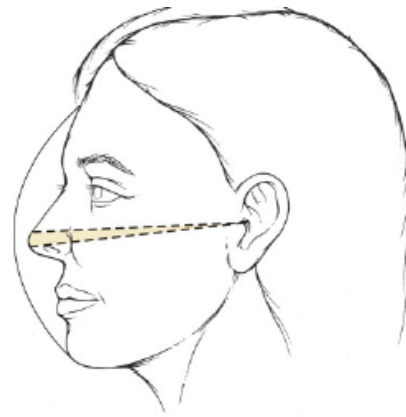


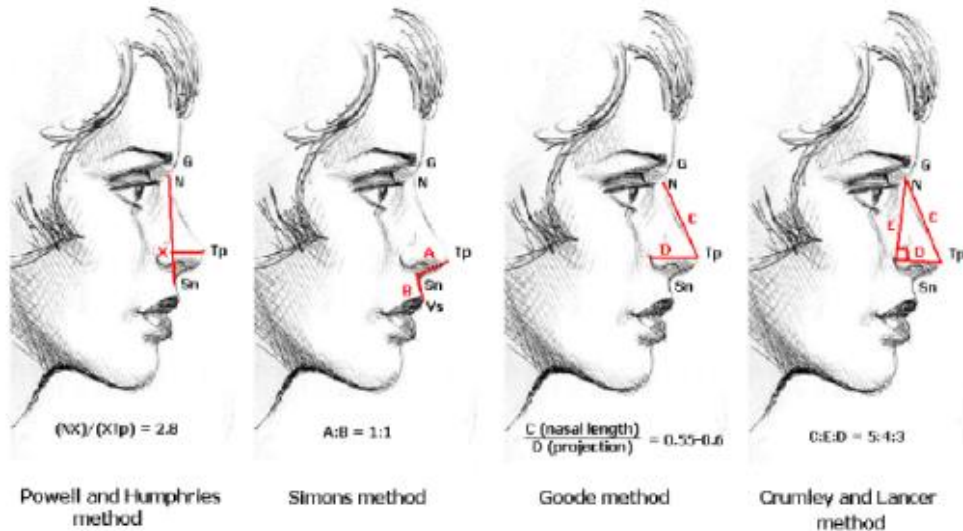
Figure 170.9 Nasal tip rotation is defined by the movement of the nasal tip on an arc around the trignon.



***Nasal Tip rotation**

- Tip Projection:

- Various methods.
- Nasofacial angle: line drawn from glabella to pogonion and line tangent to nasal dorsum; optimal NFA is 36 degrees.
- Powell and Humphries: tip projection-to-nasal height ratio 2.8:1.
- Simons: tip projection (Sn-Tp) equal upper lip length.
- Goode: projection-to-length ratio should equal 0.55 to 0.6:1.
- Crumley and Lancer: described a right triangle with projection to height (from N-A) to length equaling 3:4:5.
 - If dorsal projection exceeds this limit, dorsal hump may be present.
 - Slight concavity can be attractive (excess → saddle-nose).



*Tip projection methods

Nasal Base

- Nostrils should be slightly pear-shaped.
- The width of the nasal lobule should be 75% of the width of the nasal base.
- The lobule-to-columella ratio should be 1:2.
- On lateral view, the ala-to-lobule ratio should be 1:1.
- There should be 2 to 4 mm of columellar show .

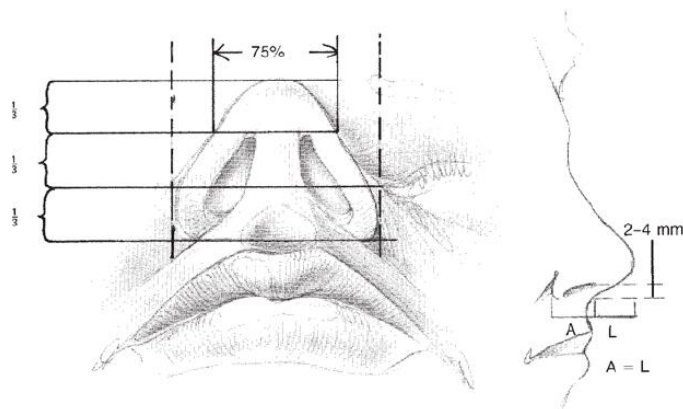


Figure 170.11 The width of the nasal lobule should be 75% of the width of the nasal base. The lobule-to-columella ratio should be 1:2. On lateral view, the ala-to-lobule ratio should be 1:1, and there should be 2 to 4 mm of columellar show.

<u>Profile</u>	<u>Frontal</u>	<u>Basal</u>
- Radix - Dorsal height - Tip projection - Alar columellar relation - Nasolabial angle - Caudal septum - Chin projection	- Straightness - Bony-cartilagenous Transition - Tip bulbousness - Tip definition - Alar columellar relation	- Dorsal line - Tip definition - Nostril symmetry - Alar base width

Glabellonasal angle > upper 1/3 projection

Nasofrontal angle (radix depth) = 115-135

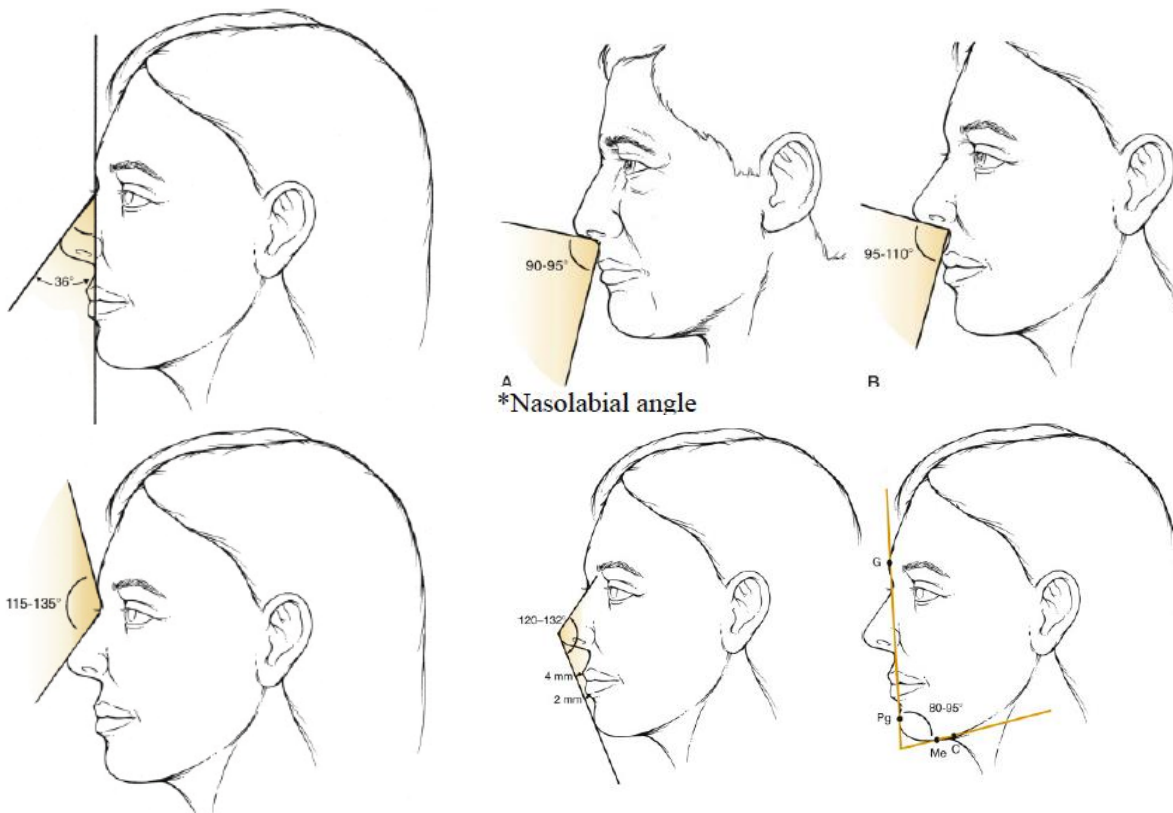
Nasofacial angle (tip projection) = 30-40

Nasolabial angle (tip rotation) = M 90-95, F 95-105

Nasomental angle (tip projection) = 120-132

Mentocervical angle = 80-95

Nasal tip angle & ala slope angle > tip projection



*Nasofacial and nasofrontal angles

* Nasomental (note the lip position) and Mentocervical angles

Malar Region (Midface):

- Face widest at zygomatic arches.
- Frontal view:
 - The prominence of the malar eminence should lie within the space formed between lines drawn from the nasal ala to the tragus and from the oral commissure to the lateral canthus

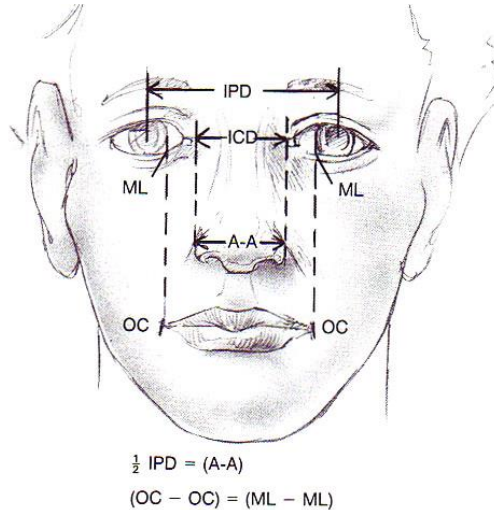


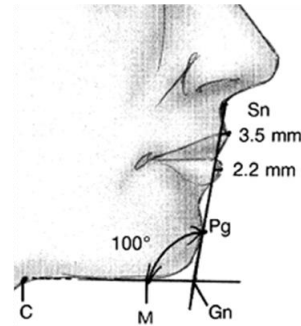
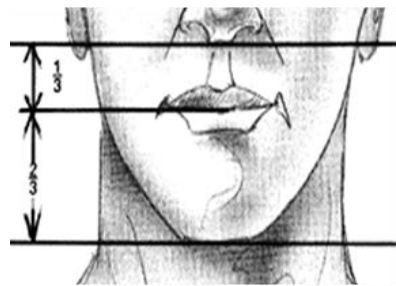
Figure 168.8 The alar-alar distance (A-A) should equal the intercanthal distance (ICD) or half the interpupillary distance (IPD). The lateral oral commissure (OC) should lie on a vertical line tangent to the medial limbus (ML) of the eye. An oblique line parallel to the lateral brow should parallel the vermillion border of the upper lip.

Chin and Neck

- Ratio of chin and lower lip to upper lip is 2:1.
- Projection of chin:
 - Chin extends from the mentolabial sulcus to the menton.
 - Vertical line from lower vermillion border.
 - Males: tangent line
 - Females: 2-3 mm posterior
 - Mentolabial sulcus lies 4 mm post to the line.
- Neck length from menton to sternal notch should be half of head height (vertex to menton).
- This angle is at the gnathion and is formed by the intersection of a line from subnasale to pogonion (Sn-Gn), with a line drawn through the cervical point and menton.
- Mentocervical angle takes into account the upper face.
 - Junction between glabella to pogonion line and cervical point and menton line

Lips:

- Line from subnasale to pogonion:
 - Upper lip is 3.5 mm anterior
 - Lower lip is 2.2 mm anterior
- Commissure should lie along a vertical line tangent to **medial limbus** of the iris.
- At rest, there should be no more than 3 mm interlabial gap.
- No more than 2 mm of maxillary incisal edge should show with lips in repose.
- No more than 2/3rd of maxillary incisor should show on smile; gingival show on smiling is undesirable.



- The vertical length of the upper lip from subnasale to stomion = 1/3 of the lower facial third
- The lower lip and chin from stomion to menton = 2/3 of the lower facial third

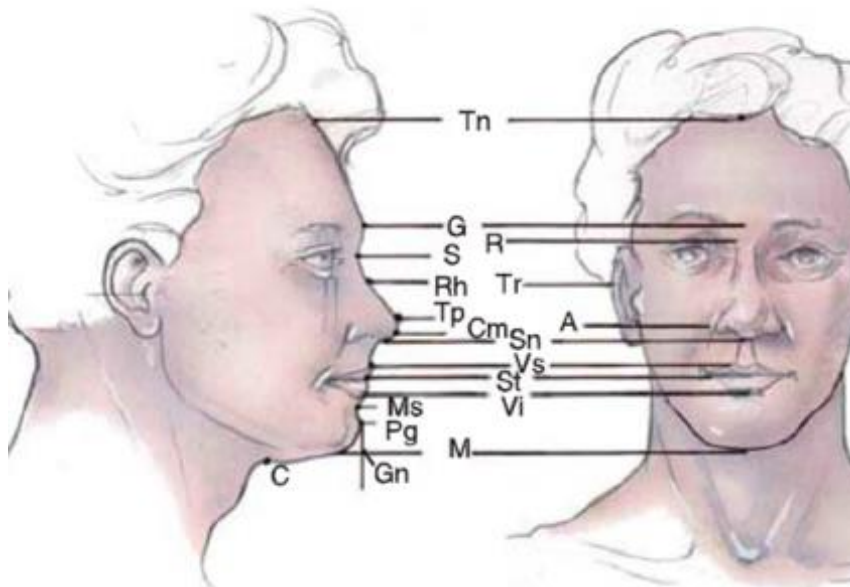


TABLE 168.1
NORMAL VALUES FOR FACIAL ANALYSIS

Vertical facial height [Powell and Humphries (11)]

Nasal height ratio (N-Sn): 47%

Lower facial height ratio (Sn-Gn): 53%

Facial convexity angle (G-Sn, Sn-Pg): 8–16 degrees (12 degrees)

Aesthetic triangle [Powell and Humphries (11)]

Nasofrontal angle (G-N-Tp): 115–130 degrees

Nasofacial angle (G-Pg, N-Tp): 30–40 degrees (36 degrees)

Nasomental angle (N-Tp-Pg): 120–132 degrees

Mentocervical angle (G-Pg, M-C): 80–95 degrees

Facial proportions [Peck and Peck (22)]

Nasal angle (N-Tr-Tp): 20–27 degrees

Maxillary angle (Tp-Tr-Vs): 12–17 degrees

Mandibular angle (Vs-Tr-Pg): 14–20 degrees

Nasal projection (Goode ratio, A-Tp/N-Tp): 0.55–0.6:1

Nasal projection (Powell ratio, Sn-Tp/N-Sn): 2.8:1

Nasolabial angle (Cm-Sn-Vs): 90–120 degrees

Alar-lobule ratio: 1:1

Nasal width = interocular distance = $\frac{1}{2}$ interpupillary distance:
30–35 mm

Lower facial third ratio (Sn-St/St-M): 0.5:1

Horizontal projection upper lip (from Sn-Pg): 3.5 mm

Horizontal projection lower lip (from Sn-Pg): 2.2 mm

Mentolabial sulcus (from Sn-Pg): 4.0 mm

Interlabial gap: 0–3 mm

Lower face–throat ratio: 1.2:1

N, nasion; Sn, subnasale; Gn, gnathion; G, glabella; Pg, pogonion; Tp, tip-defining point; M, menton; C, cervical point; Tr, tragion; Vs, superior vermillion border; A, alar crease; Cm, columellar point; St, stomion.

Methods of Facial Analysis:

- **Cephalometrics**
- Lateral radiographs; bony and soft tissue landmarks
- **Photometrics**
- Direct consideration of soft-tissue consideration on facial photographs is preferred by surgeons.
- In Rhinoplasty:
 - A: Frontal
 - B: Oblique
 - C: Lateral (Profile)
 - D: Basal
- Frontal and lateral = Frankfort line.
- Oblique = tip of the nose on contra lateral cheek.
- Basal (classic): tip between medial canthi.
- Basal (high): tip between eye brows.
- Chin down.
- In Otoplasty:
 - Frontal view
 - lateral view
 - Oblique view
 - Rear view

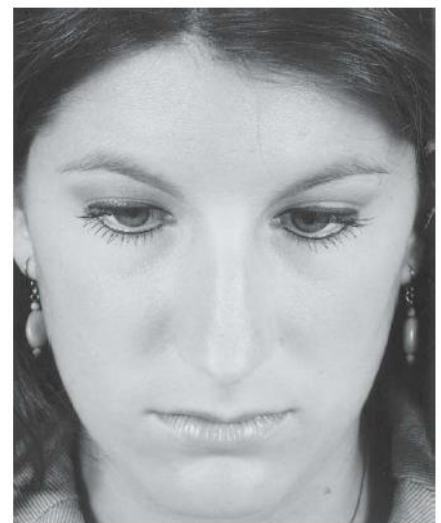
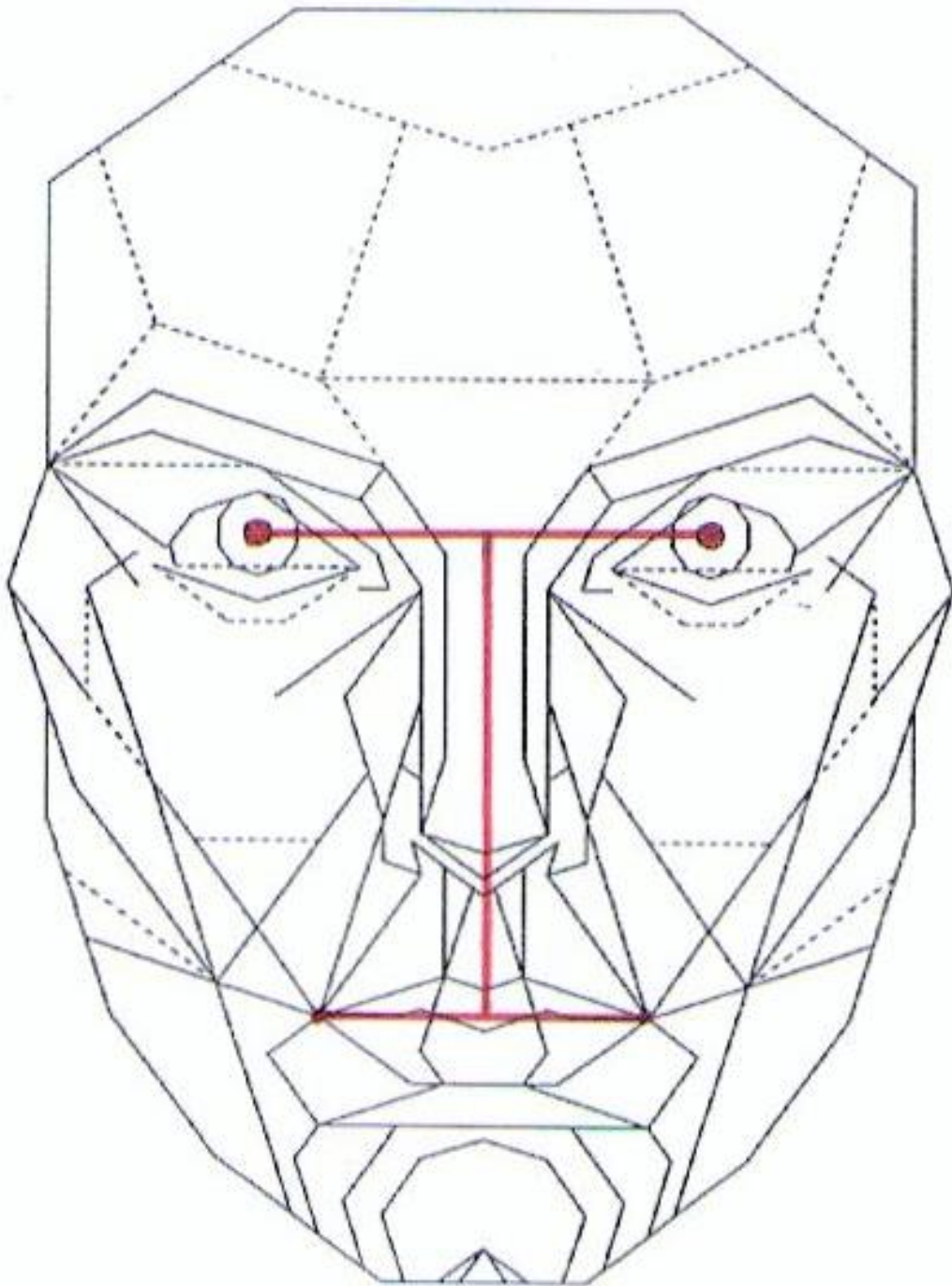


Figure 181.4 The chin-down view of nasal dorsum is useful to detect the presence of subtle radix contour irregularities or asymmetry.

**TABLE
171.1****RECOMMENDED, STANDARDIZED PHOTOGRAPHIC VIEWS
FOR SPECIFIC FACIAL PROCEDURES**

Procedure	Standardized Views
Botulinum toxin (Botox)	Frontal; frontal smiling; frontal frowning; frontal brow elevation (All photographs are taken before the first session regardless of what areas will be treated in order to enhance patient dialogue should the patient complain as well as to reduce the need for further photography if the patient desires other areas treated during future sessions.)
Rhinoplasty	Frontal; basal; L/R oblique; L/R lateral; +/- dorsal (head tilted down for a crooked nose) +/- smiling laterally (If the patient has an active depressor septi muscle that alters the nasal tip position during animation.)
Blepharoplasty/browlift	Frontal; L/R oblique; L/R lateral; closeup (May also take with eyes closed and upward gaze.)
Rhytidectomy	Frontal; L/R oblique; L/R lateral
Otoplasty	Frontal; L/R oblique; L/R lateral; posterior; R/L lateral closeup (Remember to tie long hair back in a bun and/or wear a headband to lift away obstructing hair.)
Malar augmentation	Frontal; L/R oblique; L/R lateral; basal
Hair transplant	Frontal; L/R oblique; L/R lateral; posterior; posterior with head tilted back; frontal with head tilted down +/- closeup of anterior/lateral hairline (When photographing the hair, the hair should be styled in a standardized fashion and "combovers" eliminated.)
Lip augmentation	Frontal; L/R oblique; L/R lateral; closeup mouth (Remember to remove lipstick, lip liners, and other makeup that can interfere with reproducible photography.)
Scar revision	Frontal; L/R oblique; L/R lateral; closeup (Consider reducing the exposure value to highlight the scar.)

Repose Frontal Mask



Fitzpatrick skin type

SKIN TYPE		DETAILS
I	Pale, porcelain, or ivory skin 	Skin burns very easily and doesn't tan. Likely to have light blonde or red hair.
II	Fair, beige, or cream-colored 	Skin will usually burn in the sun, and has difficulty tanning.
III	Light brown, golden, or olive 	Skin will sometime burn and will tan gradually.
IV	Caramel or medium brown 	Skin will tan easily and rarely burn.
V	Bronze or rich brown skin 	Skin will tan without burning.
VI	Mahogany or dark brown 	Skin never burns and will tan very quickly.

Anatomy of External Nose

Surface Anatomy

- Nose occupies central horizontal 1/3 (glabella to the subnasale).
- Also it occupies vertical 1/5 of face (between the medial canthi).

Figure 179.1 Topographic key landmarks and accepted designations for (A) frontal and (B) oblique views of the nose. 1, Glabella; 2, nasion, nasofrontal angle; 3, rhinion (osseocartilaginous junction); 4, tip-defining point; 5, infratip lobule; 6, columella; 7, columella-labial junction; 8, facet; 9, alar sidewall; 10, alar-facial junction; 11, medial crural footplate; 12, supra-alar crease; 13, alar margin; 14, philtrum; 15, philtral crest; 16, supratip dorsum. (From Tardy ME. *Surgical anatomy of the nose*. New York: Raven Press, 1990, with permission.)

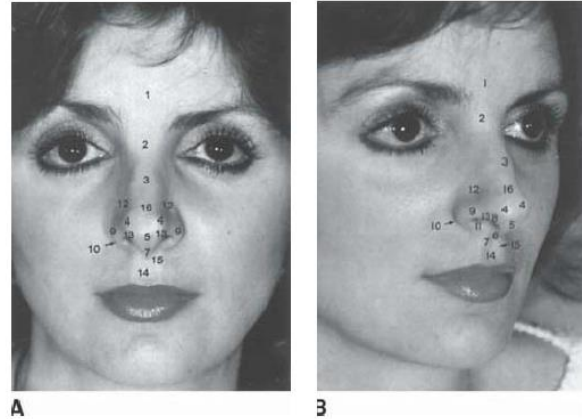


Figure 179.2 A: Topographic key landmarks and accepted designations for lateral view of the nose. 1, Glabella; 2, nasion (nasofrontal junction); 3, rhinion (osseocartilaginous junction); 4, tip-defining point; 5, infratip lobule; 6, columella; 7, columella-labial junction; 8, facet; 9, alar lobule; 10, alar-facial junction; 11, medial crural footplate; 12, supra-alar crease. B: Topographic key landmarks and accepted designations for basal view of the nose. 1, Tip-defining point; 2, interdomal area, alar lobule; 3, infratip lobule; 4, columella; 5, medial crural footplate; 6, columella-labial junction; 7, philtrum; 8, nostril aperture; 9, facet; 10, alar sidewall; 11, alar-facial junction; 12, nostril sill. (From Tardy ME. *Surgical anatomy of the nose*. New York: Raven Press, 1990, with permission.)

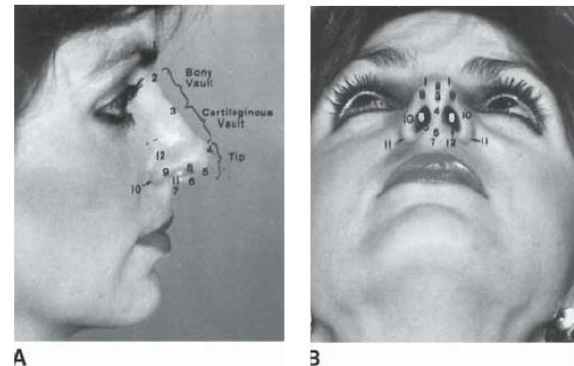
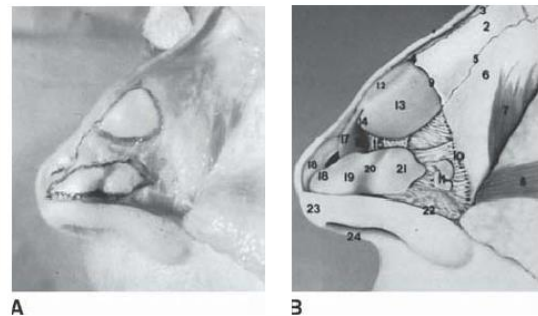


Figure 179.3 Key anatomic landmarks (A) and standard terminology (B) for lateral view of fresh cadaver dissection of nose. 1, Nasofrontal suture line; 2, nasal bone; 3, intranasal suture line; 4, osseocartilaginous junction (rhinion); 5, nasomaxillary suture line; 6, ascending process of maxilla; 7, levator labii superioris muscle; 8, transverse nasalis muscle; 9, cephalic portion of ULC (articulates to undersurface of nasal bone); 10, piriform margin; 11, sesamoid cartilages; 12, cartilaginous dorsum; 13, upper lateral cartilage; 14, caudal free margin of ULC; 15, intercartilaginous ligament; 16, quadrangular cartilage; 17, anterior septal angle; 18, tip-defining point alar cartilage; 19, lateral crus of alar cartilage; 20, concavity ("hinge") of lateral crus; 21, lateral aspect of lateral crus; 22, alar lobule; 23, infratip lobule; 24, columella. (From Tardy ME. *Surgical anatomy of the nose*. New York: Raven Press, 1990, with permission.)



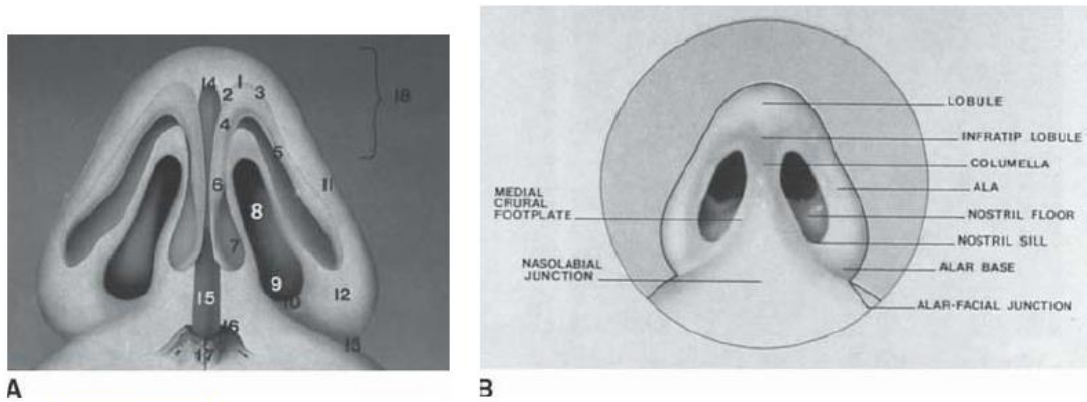
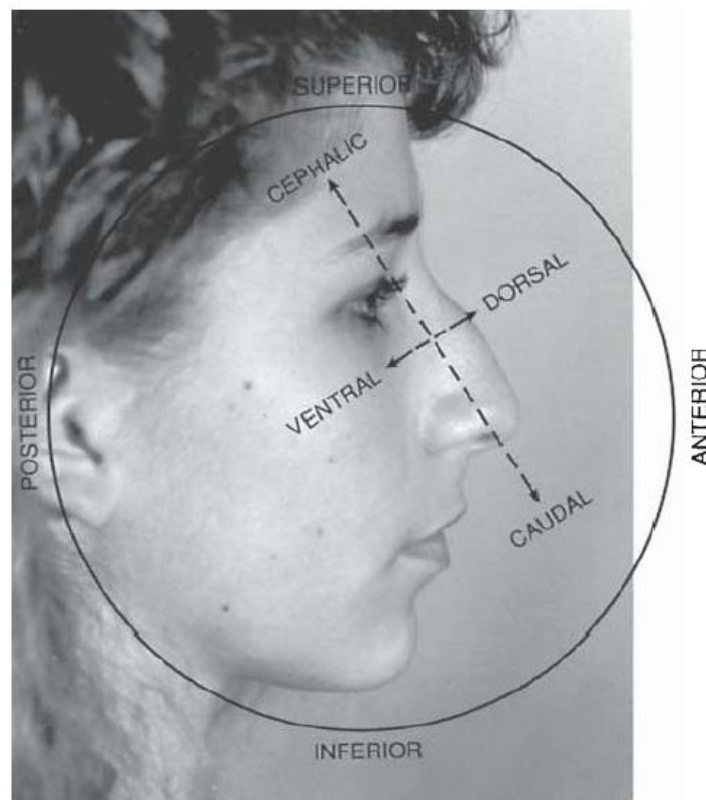
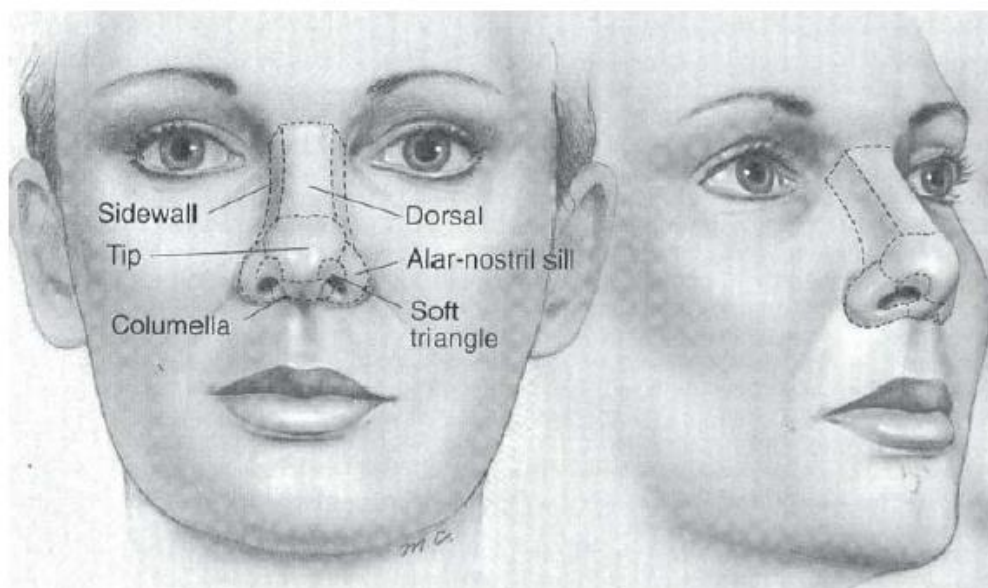
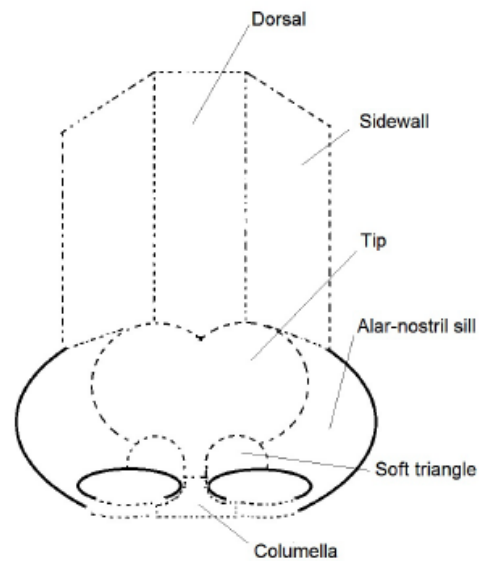
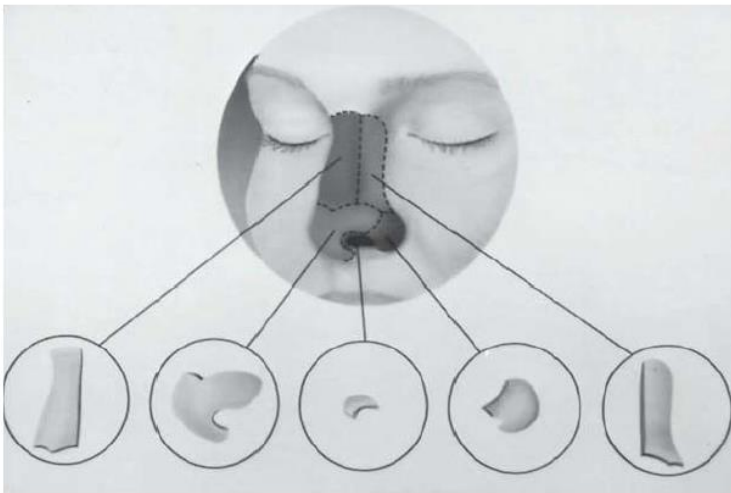


Figure 179.4 Additional anatomic landmarks (A) and standard nasal terminology of base of nose (B). 1, Apex of alar cartilage; 2, medial genu; 3, lateral genu; 4, transitional segment; 5, lateral crus; 6, medial crus; 7, medial crural footplate; 8, nostril aperture; 9, nostril floor; 10, nostril sill; 11, lateral alar sidewall; 12, alar lobule; 13, alar-facial junction; 14, anterior septal angle; 15, caudal septum; 16, maxillary crest; 17, nasal spine; 18, infratip lobule. (From Tardy ME. *Surgical anatomy of the nose*. New York: Raven Press, 1990, with permission.)



Subunits of the Nose:

- **Six subunits:** (3 pairs & 3 singles)
 1. Nasal dorsum (Single)
 2. Sidewalls (Pair)
 3. Tip (Single)
 4. Columella (Single)
 5. Ala/sill (Pair)
 6. Soft triangles (Pair)
- Effort should be made to place scars within subunit borders whenever possible



Variant Anatomy

- Change one thing, and others appear to change.
- **Dorsal Height:**
 - Depend mainly on cartilaginous and osseous **nasal septum**.
 - Increased projection of the dorsum on the "lateral view" corresponds to a narrower appearance on the frontal view.
 - Low nasal bridge creates a wider-appearing nose from the front.
 - Dorsal convexity creates an illusion of an underrotated nasal tip.
 - Dorsal concavity creates a perception of increased tip rotation and a shorter nose.
- **Radix and Nasofrontal Angle:**
 - Affect nasal starting point, nasofrontal angle, overall dorsal profile, and apparent intercanthal distance.
 - Low radix → illusion of increased intercanthal distance – illusion of dorsal hump when associated with tip under projection).
 - High radix → illusion of decreased intercanthal distance + blunts nasofrontal angle and makes nose look long.
 - Acute nasofrontal angle → illusion of a shorter nose.

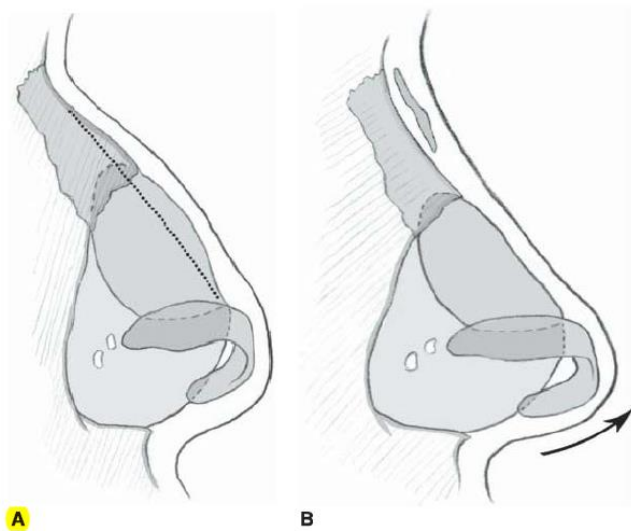


Figure 179.7 **A:** Profile view of a nose with a true dorsal hump in the region of the rhinion. Correction may involve reduction of the hump. **B:** Nose with a relative dorsal convexity due to a low radix and an underprojected nasal tip. Correction might involve elevating of the radix with a graft and increasing tip projection. Final external dorsal contour is similar in both examples.

- **Nasolabial Angle:**

- Formed by the upper lip and columella, represents degree of tip rotation.
- Increased nasolabial angle creates an illusion of increased tip rotation.

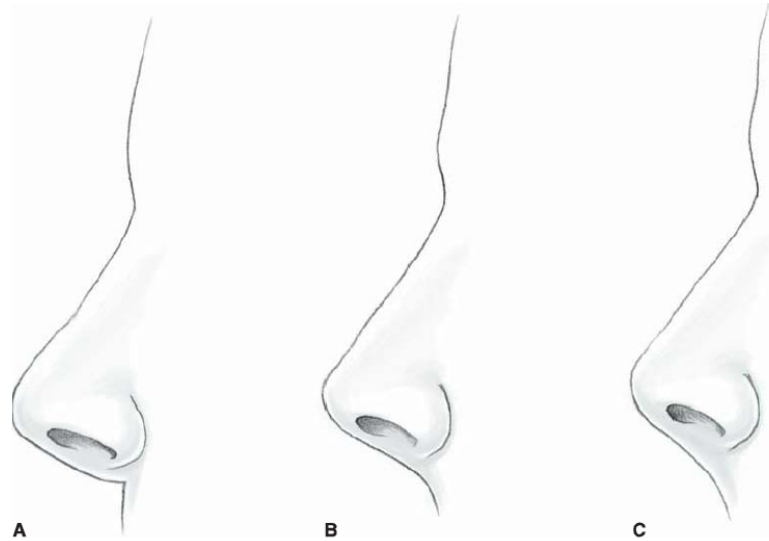
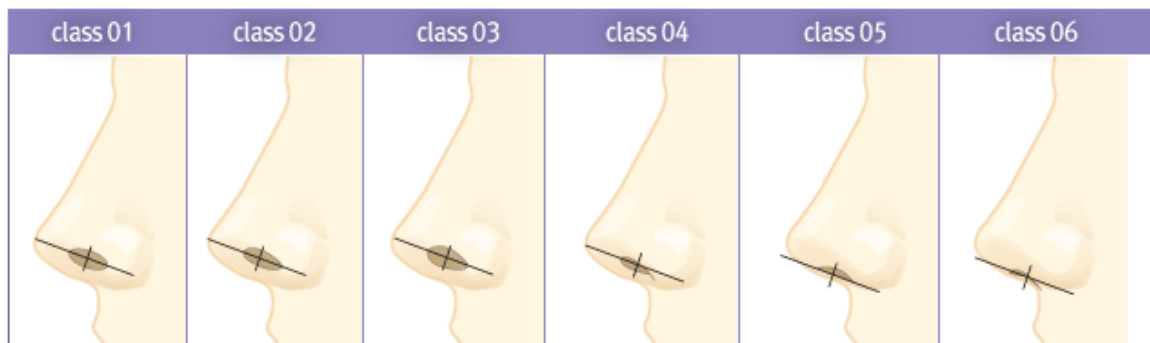


Figure 179.8 Lateral view of three noses with identical tip position. **A:** Relative deficiency of tissue at the nasolabial area and an acute nasolabial angle creates an illusion of underrotation of the tip. **B:** Nose with a moderate nasolabial angle. **C:** Fullness at the nasolabial area with an obtuse nasolabial angle creates an illusion of increased tip rotation.

- **Alar–Columellar Relation:**

- Alar retraction or columellar hang → excessive columellar height is visible on the lateral view.
- Hanging columella may create an illusion of a tip ptosis.
- Retracted caudal septum or a low alar margin → relative lack of columellar → illusion of increased tip rotation.



Skin-Soft Tissue Envelope (SSTE)

Skin:

- Skin: thick at nasion and tip; thin at rhinion.
- Thicker again at tip with sebaceous glands; very thin again at caudal-most aspect of alar margins and columella.
- Thin epithelial sleeve around alar rim and nasal facets – notching.
- Thick at nasion (2-5 mm)
- Thin/mobile over dorsum (3.2 mm)
- Thinnest at rhinion (2 mm)
- Thick/sebaceous at tip (5 mm)
- Nasal SMAS is immediately superficial to the periosteum/perichondrium.
- Dissection is done just below SMAS (preserves neurovascular structures and lymphatics).
- Because the skin is thinnest at the rhinion, a straight external profile requires a small relative convexity to remain in this region
- If the dorsum (bone) is reduced so that a straight skeletal profile results, a slight concavity at the mid-dorsum is likely to result after skin re-draping.
- This may create a cartilaginous **Pollybeak**, in which the supratip dorsum projects above the plane of the dorsum.
 - Other causes: Under resection of cartilaginous dorsum, **Ptotic tip**, Thick soft tissue envelope, Supratip dead space.

Subcutaneous tissue:

- Four soft tissue layers between skin and skeleton:
 1. Superficial fatty panniculus (directly connected to the dermis):
 - Adipose plus vertical fibrous bands.
 2. Fibromuscular layer (SMAS):
 - Musculature and aponeurotic system.
 3. Deep fatty layer: major superficial vessels/nerves.
 4. Periosteum/chondrium: fuses with that of piriform process; nice plane immediately above for dissection.

Muscles:

- Elevators shorten nose and dilate nostrils.
- Depressors lengthen nose and dilate nostrils.
- Compressors lengthen nose and narrow nostrils.
- Minor dilators widen nostrils.
- Dissection in the sub-SMAS layer of the soft-tissue envelope allows the surgeon to avoid the nervous supply to the nasal muscles.
- Division of the depressor septi nasi may correct drooping of the nasal tip and shortening of the upper lip during facial animation (may reduce gingival show and nasal-tip descent during smiling).
- All muscles are innervated by zygomaticotemporal division of CN VII.

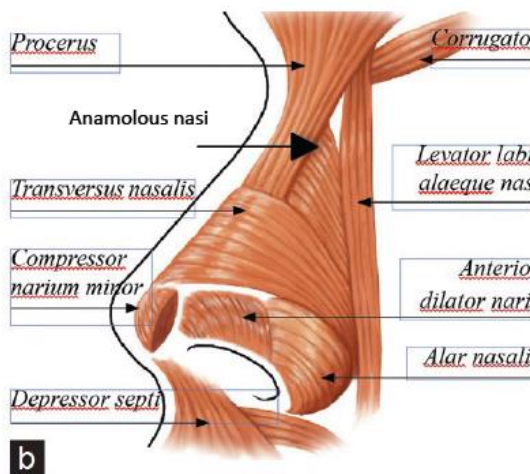


TABLE
179.1

INVESTING NASAL MUSCULATURE

Elevator muscles
Procerus
Levator labii superioris alaeque nasi
Anomalous nasi
Depressor muscles
Alar nasalis
Depressor septi nasi
Compressor muscles
Transverse nasalis
Compressor narium minor
Minor dilator muscles
Dilator naris anterior

Variant anatomy:

- **Thickness** of the SSTE varies with ethnicity, age, and gender
 - Underlying nasal architecture does not accurately transmit through the thick SSTE.
 - **Modifications of framework may not result in significant changes in external appearance.**
 - Thick SSTE cannot conform onto the underlying structure after reduction, scar tissue may fill the resulting void → **soft-tissue/scar pollybeak** that results after aggressive supratip hump reduction.
- In **Thin** SSTE → graft must be placed in a precise manner, and any edges that may transmit through the skin should be beveled or crushed to blend into surrounding structures; soft-tissue or crushed cartilage onlay grafts may aid in camouflaging irregularities.

Skeletal Framework:

- Divided into thirds:
 - Upper = osseus vault.
 - Mid = upper cartilaginous vault.
 - Lower = lower cartilaginous vault.
- Septum supports all three and divides it in 2 lateral halves.
- Septal angles:
 - **Anterior septal angle** → transition from dorsal to caudal aspect.
 - **Posterior septal angle** → quadrangular cartilage articulation with the anterior nasal spine.
 - **Intermediate septal angle** → in between the anterior and posterior septal angles.
- Septal cartilage adherent to upper and lower laterals; can have significant effect on nasal shape.

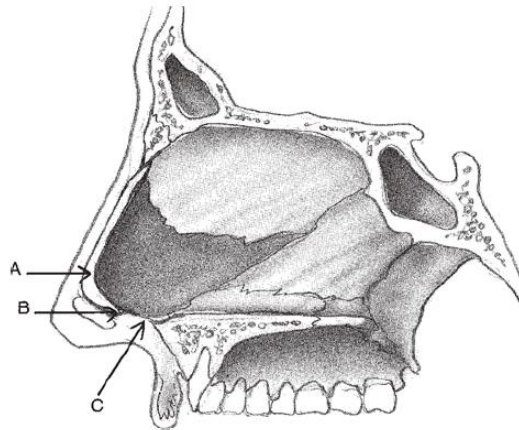


Figure 179.10 Arrows, The three anatomic angles composing the caudal aspect of the nasal quadrangular cartilage. A, Anterior septal angle; B, midseptal angle; C, posterior septal angle.

- Septal overgrowth → may lead to excessive projection of LLCs and ULCs → septum pulls the cartilaginous elements of the nose under tension —the underpinnings of the so-called **tension-nose deformity**.
- Tip-defining point is determined by a projecting anterior septal angle (rather than the domes).
- Low-hanging columella that is created by a prominent caudal septal border.
- Short nasal bones and long ULCs tend to have a quadrangular cartilage and less vomer and ethmoid plate (may be useful when looking for graft materials).
- The septum rarely exists in a true midsagittal plane.

Anatomy

- Upper 2/3rd : from nasion to alar groove, includes dorsum and sidewalls
- Lower 1/3rd : tip, columella, soft tissue triangles, alar-nostril sill
- Radix: region of nasal dorsum that defines the contour of the paired nasal bones from the glabella toward the nasal tip
- **Osseous vault:**
 - With bony septum, provides principal nasal structural support.
 - Includes: frontal process of maxilla and nasal bones.
 - Articulate at nasofrontal suture cephalic margin.
- Nasal bones:
 - Average length is 2.5 cm.
 - Fuse with frontal bone (11 mm superior to intercanthal line) (rests on the nasal spine of the frontal bone).
 - Free caudal edge is sup portion of piriform aperture; same joins upper cartilaginous vault at **keystone area (nasal bone with ULL)**.
 - Bones articulate with each other and frontal process of maxilla
 - Wider and thinner caudally (most # occurs in the caudal aspect).
 - Medial osteotomies disconnect the two halves of the osseous vault so each may be moved independently.
 - Lateral osteotomy is lateral to nasomaxillary suture.
 - Nasal bones are subject to age-related osteopenia and may become thinner and more fragile over time.

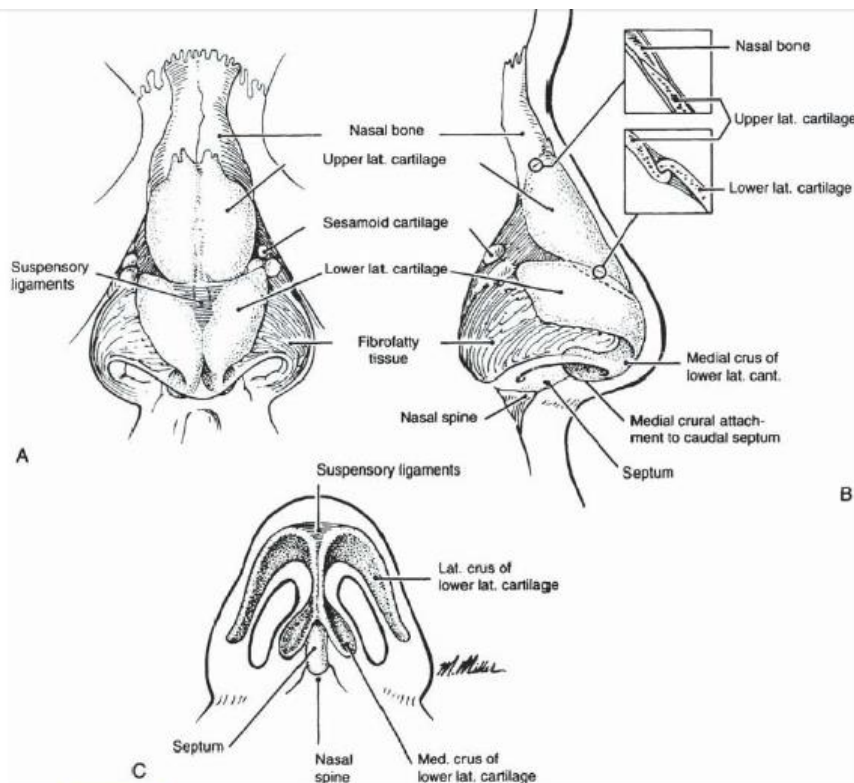


Figure 181.3 Anatomic relationships of the nasal bones, upper lateral cartilages, and lower lateral cartilages. (Modified from Papel ID. Management of the middle vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:408, with permission.)

- **Upper cartilaginous vault:**

- Paired upper laterals fused in midline to cartilaginous septum
- **Nasal bones provide most of their structural support.**
- **Keystone area:** bony overlap over cartilage, up to 11 mm; dislocation causes collapse of upper cartilaginous vault.
- Wider cephalically than caudally.
- Caudal free edge is site of internal nasal valve.
- Lateral margin of ULC fuse with dense Connective Tissue and medially is fused to septum (loose inferiorly).
- **Lengths of the osseous vault and upper cartilaginous vault have an inverse relation; shorter nasal bones have longer cartilage → floppier caudal end so internal valve more susceptible to collapse (may require a spreader graft).**
- **Scroll region:** junction of ULC and LLC.

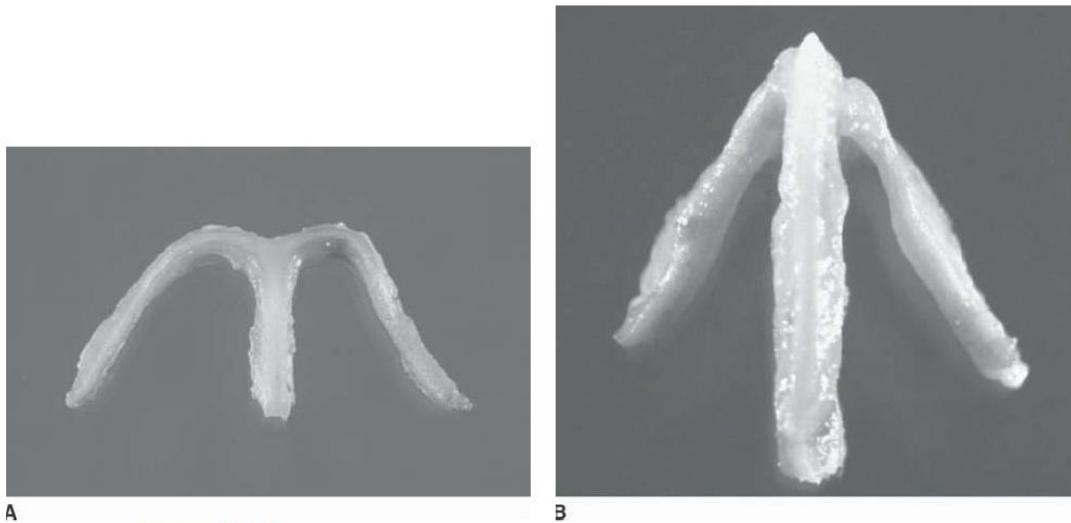


Figure 179.11 A: Cross section of the upper cartilaginous vault just below the rhinion. The broad arch of the ULCs closely follows the caudal margin of the nasal bones in this region. B: Further caudal in the area of the internal nasal valve, the structures are more flexible and a much narrower relation exists between the ULCs and nasal septum. (Photograph courtesy of Dean Toriumi.)

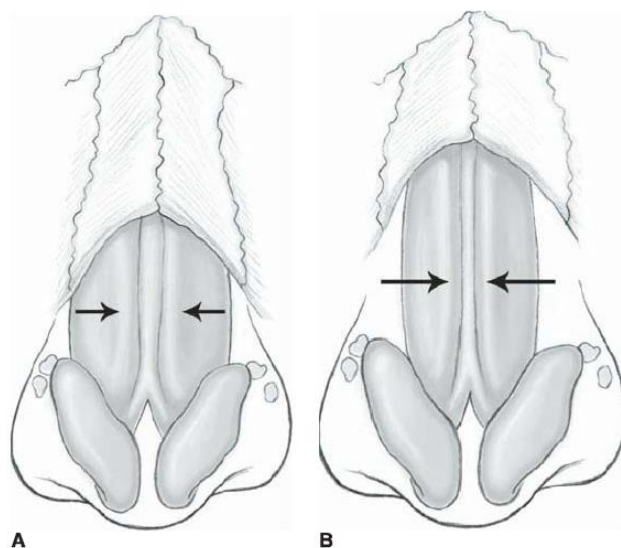


Figure 179.12 A: Long nasal bones create more stabilization of the caudal cartilaginous elements of the nose with less tendency for inferomedial collapse of the ULCs with inspiration or with scarring after hump reduction. B: Short nasal bones and a long upper cartilaginous vault may cause narrowing of the ULCs after they are separated from the nasal septum following hump reduction. Restabilization with spreader grafts may prevent this complication in such patients.

- **Medial crus:** connected to caudal septum, provides support to columella; narrowest in columella (as little as 4-5 mm); each medial crus divided into an anterior columellar segment and a posterior footplate segment; footplate segment flares causing normal widening at base; the anterior limit of the medial crus corresponds to the columellar–lobular junction at the apex of the nostril.
 - Variant Anatomy
 - Short medial crus → short columella and small AP dimension of nostril → deficient nasal tip projection and a small nostril–lobular ratio.
 - Long medial crura (beyond nostril apex) creates flat projecting nasal tip (may lead to a bifid appearance of the columella in thin SSTE) (minimal bifid is beauty).
 - Exaggerated flare of the posterior medial crura (footplates) may compromise airflow.
 - Low medial crura will lead to excessive columellar show → trimming caudal end of medial crura is one way to reduce columellar show.
- **Intermediate crus** “domal segments”:
 - Bridge medial to lateral crura; extending from medial to lateral genur.
 - Lateral genu is often most ant point → correlating to tip-defining point.
 - **Angle of divergence:** angle formed between the two intermediate crura as they flare away from each other laterally (approximately 60 degrees); provides a natural-appearing width to the infratip lobule as it transitions from the columella to the nasal tip.
 - Intermediate crura also diverge from the medial crura in a cephalic direction by approximately 50 degrees → this bend corresponds to the columellar–lobular angle, dividing the nasal base into the infratip lobule anteriorly and columella posteriorly—the basis of the double break.
 - Variations in angle affect tip and infratip shape.
 - Narrow angle makes triangular basal appearance.
 - Wide angle makes boxy tip (or bifid).
 - If intermediate crura are concave, a “double dome” segment is created.

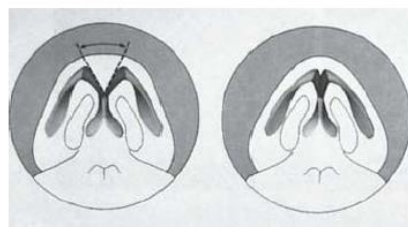


Figure 179.13 The intermediate crura bridge the lateral and medial crura of the LLCs. The angle of divergence is shown on the left. If the intermediate crura are widely bifurcated, a broad trapezoid appearance of the tip results, as shown on the left. If the intermediate crura are closely opposed, a triangular appearance results, as shown on the right.

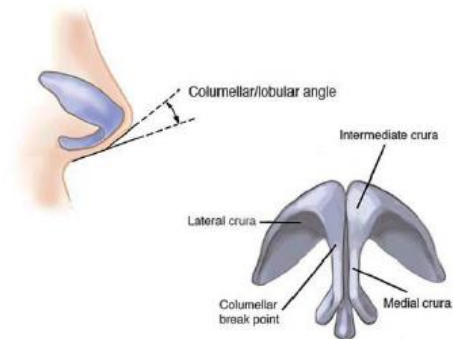


Figure 182.11 The double break is formed by the natural divergence of the intermediate crura. The columellar lobular angle is formed by the intermediate crura diverging into the lateral crura.

Lateral crus: lateral genu posteriorly; wider in midsection; **scroll area** where upper and lower laterals are connected.

Interlocking scroll 52%
Overlapping 20%
End-to-end 17%
Opposed scroll 11%

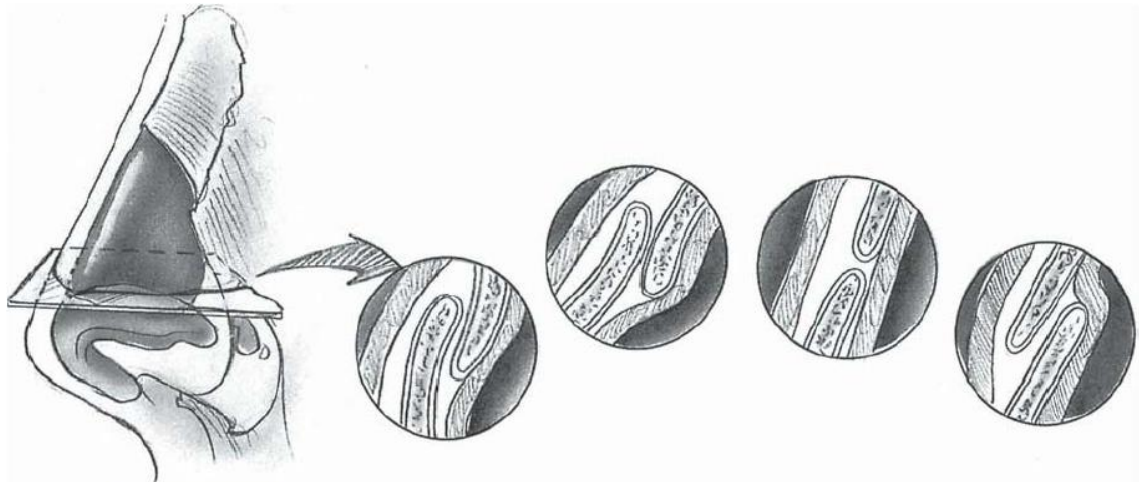
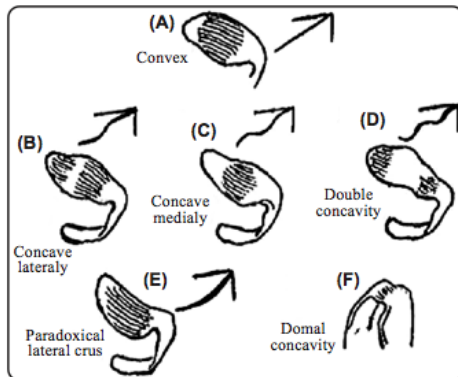


Figure 179.14 A variety of anatomic relations between the cephalic margin of the LLC and the caudal margin of the ULC is found in dissection of cadaver specimens. As individuals age, the intimate relation between the cartilages may be lost. (From Tardy ME. *Surgical anatomy of the nose*. New York: Raven Press, 1990, with permission.)



- Normally, it assumes a gentle convex shape for its medial half then flattens and turns posterosuperiorly for its lateral portion.
- If lateral crus curvature (convex) too great, gives illusion of fat tip (bulbous).
- LLCs may be concave as they diverge away from the domes; this may manifest as a hollow, sunken appearance at the alar lobule with an exaggerated supra-alar crease; **If severe, the concavity may narrow the nasal vestibule, impairing nasal airflow.**
- Risks of **bossae formation (pinched)** after excisional techniques to narrow the tip in patients who possess the triad of tip bifidity, thin skin, and strong LLCs (avoid with suture techniques).



Bossae tip is an irregular knoblike protuberance of the alar cartilages that creates a visible asymmetry of the nasal tip.

Parenthesis deformity, the tip cartilage (cephalad projection of the upper border of the lateral cartilage) is directed towards the inside corner of the eye instead of the more natural position towards the outer corner of the eye → cause buckling or pinching of the ala which may lead to nasal obstruction.

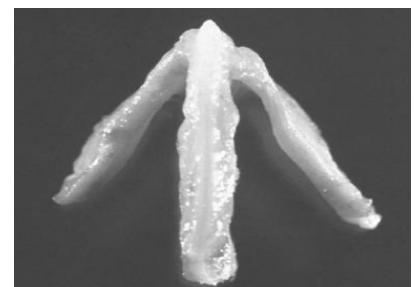


Nasal Base:

- Main structures: paired alae; nasal sills; soft triangles; columella
- Ala: most lateral; only soft tissue; changes to framing cartilage will alter shape; sesamoid cartilages in soft tissue over piriform aperture (0-5 cartilages) connect free edge of lateral crus to aperture rim; vertical insertion position affects columellar show.
- **Sill: connection between ala and columellar at the nasal spine (floor of the nostril); thin skin easily injured.**
- Soft triangle: borders columella medially, ala laterally and apex of nostril anteriorly; minimal subcutaneous tissue here; easily damaged
- Columella: paired medial crura bound by fibrous attachments; membranous septum; cartilaginous septal attachments only things limiting movement.
- Variant anatomy:
- Normal nostril-to-infratip lobule ratio of 2:1.
- Asian and African noses likely to have:
 - Greater alar base width, alar flare, a shorter columella, and more horizontally oriented nostrils.
 - Weaker, flatter LLCs; thick SSTE; and deficiency of the premaxilla and nasal spine.

Mechanics and Stability:

- Structural elements and relations:
 - Strength and resiliency of components and their various interconnections largely responsible for structural support.
 - Three echelons: bony vault; quadrangular; ULCs and LLCs.
- Cantilever concept:
 - The fusion of the osseous vault and the upper cartilaginous vault with the dorsal edge of the septum forms this structural 1-beam that extends from the nasion toward the nasal tip.
 - Hump reduction can detach 1-beam greatly reducing support; needs to be fixed.
- Role of the nasal septum:
 - Caudal end of quadrangular supports much of tip.
 - Cantilevering dorsal element and buttressing caudal element make up L-shaped strut.
 - Remove dorsal element and get saddle-nose deformity.
 - Remove caudal element and get tip ptosis.
- Ligamentous attachments:
 - **Scroll area:** (junction of upper borders of lateral crura and ULCs) :
 - Bound by intercartilaginous ligament (weakens with aging); structurally important.
 - Nasal tip support provided from scroll area.
 - Intercrural ligament (ligamentous sling):
 - Binds the medial aspect of the lateral crura, the intermediate crura, and the medial crura to each other.
 - Sesamoid complex (lateral crura complex):
 - Connection between lateral crura and piriform aperture.
- **Nasal-tip support: Tripod concept:**
 - Apex of tripod is nasal tip.
 - Individual lateral crura and paired medial ones make tripod.
 - Not a representation of tip structure/support, only a model to depict inter-relations among the structures.
 - Tip recoil tested through digital palpation.
 - Define tip rotation.



**TABLE
179.2**

TIP-SUPPORT MECHANISMS

Major

- Size, shape, and resilience of the medial and lateral crura
- Medial crural footplate attachment to the caudal border of the quadrangular cartilage
- Attachment of the upper lateral cartilages (caudal border) to the alar cartilages (cephalic border)

Minor^a

- Ligamentous sling spanning the paired domes of the alar cartilages
- Cartilaginous septal dorsum
- Sesamoid complex extending the support of the lateral crura to the pyriform aperture
- Attachment of the alar cartilages to the overlying skin and musculature
- Nasal spine
- Membranous septum

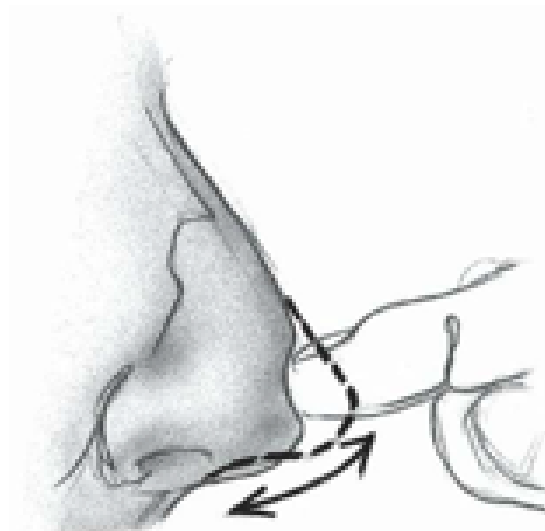


Figure 112.4 Demonstration of finger palpation of the recoil mechanism of the nasal tip. The relative resistance to deformity demonstrated by the nasal tip provides a useful indication of the integrity of nasal tip support mechanisms.

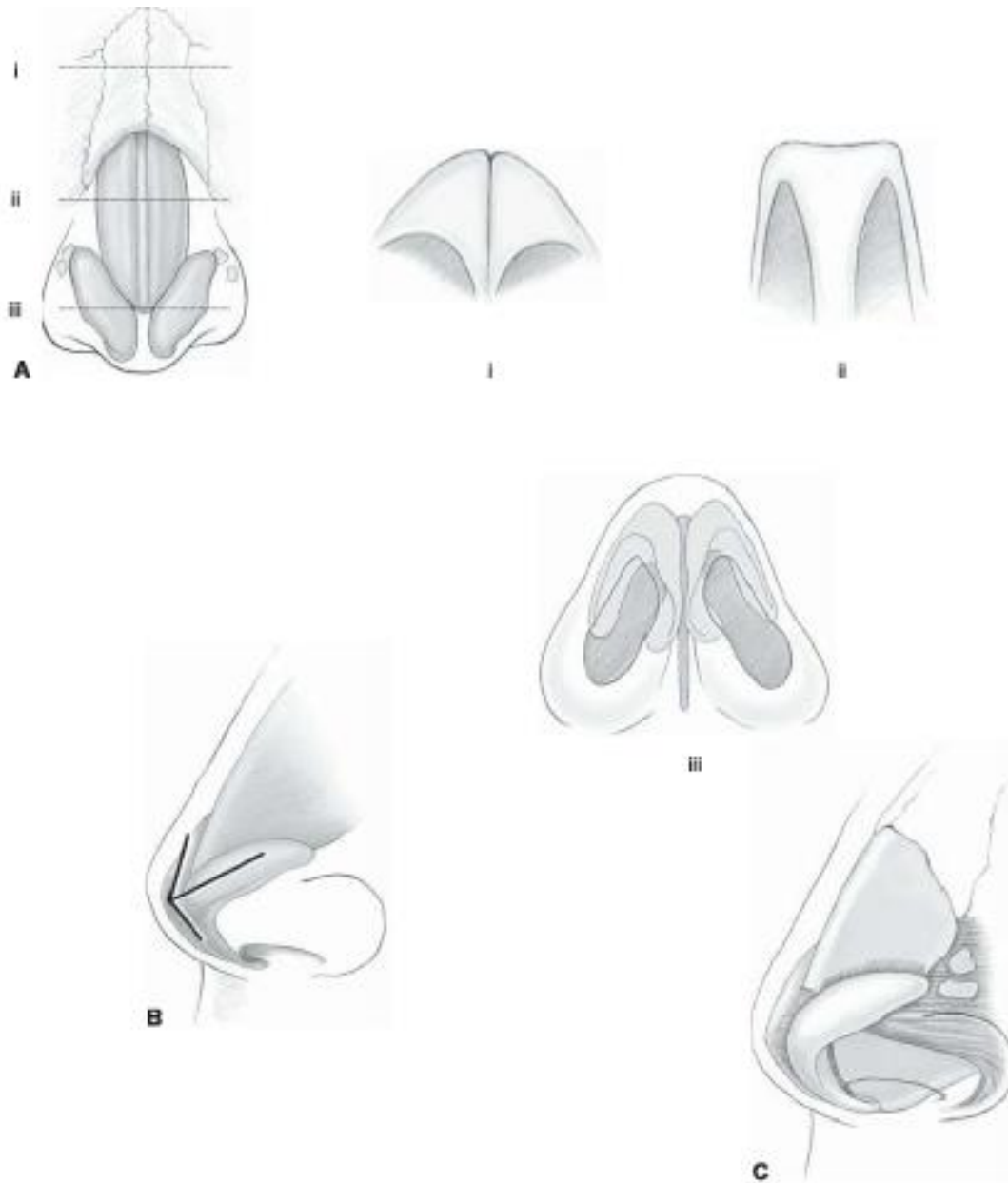


Figure 179.15 Various support mechanisms of the nose. **A:** The nasal septum functions as a supporting wall that reinforces the dorsal elements of the nose. Cross section through (i) osseous vault, (ii) upper cartilaginous vault, and (iii) nasal base. **B:** The two lateral crura and the conjoined medial crura create a nasal tip tripod, a model useful for understanding nasal tip dynamics. The degree to which the inherent strength of these structures contributes to nasal tip support varies significantly from individual to individual. **C:** The network of ligamentous attachments reinforce and interconnect the skeletal elements of the nose. The ULCs are connected firmly to the undersurface of the nasal bones, forming a cantilever-like support for the dorsum. The nasal tip and base are stabilized through ligamentous attachments between the lateral crura and ULC, the lateral crura and pyriform aperture, and between the two dorsal regions of each LLC.

Common presenting situations in tip:

- Ptotic Tip.
- Overrotated nose.
- Boulbus Tip.
- Pinch tip.

Ptotic Tip:

- Caudal tip rotation
- Acute nasolabial angle:
- <Male 90° to 95°
- <Female 95° to 110°
- Cause:
- Inherited
- Acquire causes:
- ✕ Nasal trauma
- ✕ Aging face
- ✕ Previous rhinoplasty

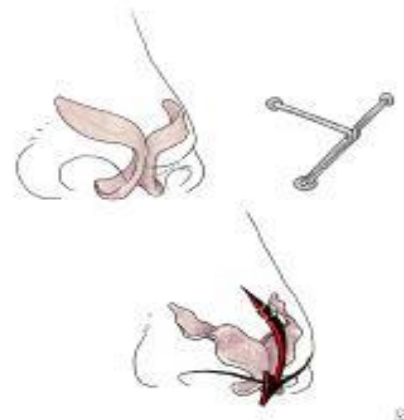


- **Loss of tip support mechanisms**

1. Loss of integrity of the medial and lateral crura
2. Loss of attachment of the medial crura to septum
3. Loss of attachment of upper lateral to lower laterals

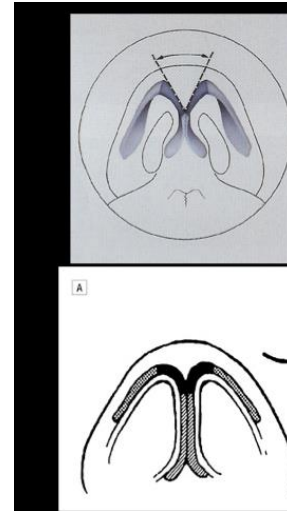
Treatment based on Tripod concept:

- ✕ Strengthening or elongating of the medial crura.
- ✕ Trimming of the lateral crura (cephalic trimming).
- ✕ Shortening caudal septum.
- ✕ Restore support mechanism.
- ✕ Medial crura (columellar) strut graft.



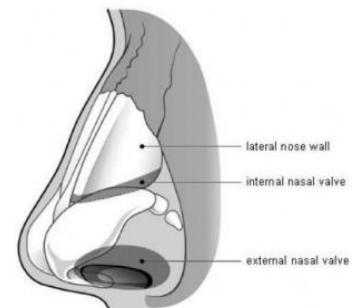
Boulbus Tip:

- Decrease nasal tip definition.
- Wide interdomal distance.
- Wide domes.
- Weak alar cartilage.
- Thick & inelastic skin.
- Management:
- Suture technique (trans domal, intra domal, medial crura).
- Cephalic trimming.



Functional Valvular Anatomy:

- Internal nasal valve:
 - Internal valve usually unchanged during inspiration; deep forced inspiration may cause some collapse in normal people.
- External nasal valve:
 - Usually, external valve dilates during deep inspiration.
 - **Bernoulli principle** → with increased airflow, the intraluminal pressure in the internal valve area decreases.
 - **Poiseuille's law** states that the rate of air flow is directly proportional to the fourth power of the radius of the conduit
 - **External valve collapse on deep inspiration can result from insufficient support of the alar rim and alar lobule.**
 - Things to watch for pre-op that predisposed to such collapse:
 - Short nasal bones and a long upper cartilaginous vault.
 - Narrow, projecting noses.
 - Slitlike nostrils.
 - Exaggerated supraalar creases.
 - Visible pinching of the lateral wall with inspiration.
 - Thin cartilages and thin skin.
 - Cephalically positioned lateral crura (minimal support to the alar margins).
- Areas of structural void:
 - Between lateral crura and piriform aperture prone to collapse (corresponds to internal valve area); alar batten grafts help; normally supported by dilator naris and nasalis muscles
 - Other area is alar margins and external valve



Ethnic Variations

**TABLE
179.3**

GENERAL NASAL CHARACTERISTICS BY MORPHOLOGY

	Platyrrhine	Mesorrhine	Leptorrhine
Skin type	Very thick	Moderately thick	Thin
Dorsum	Short, wide, concave	Short, wide	Long, narrow
Radix	Low	Low	High
Nasal bones	Short	Short	Long
Nasal tip	Bulbous, underprojected	Rounded, underprojected	Projected
Columella	Short	Short	Long
Nasal alar width	Wide	Intermediate	Relatively narrow
Ala	Prominent flaring	Variable	Modest flaring

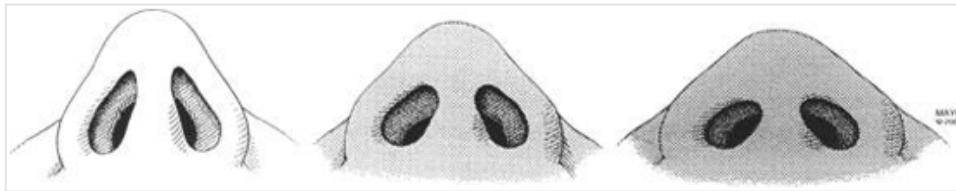
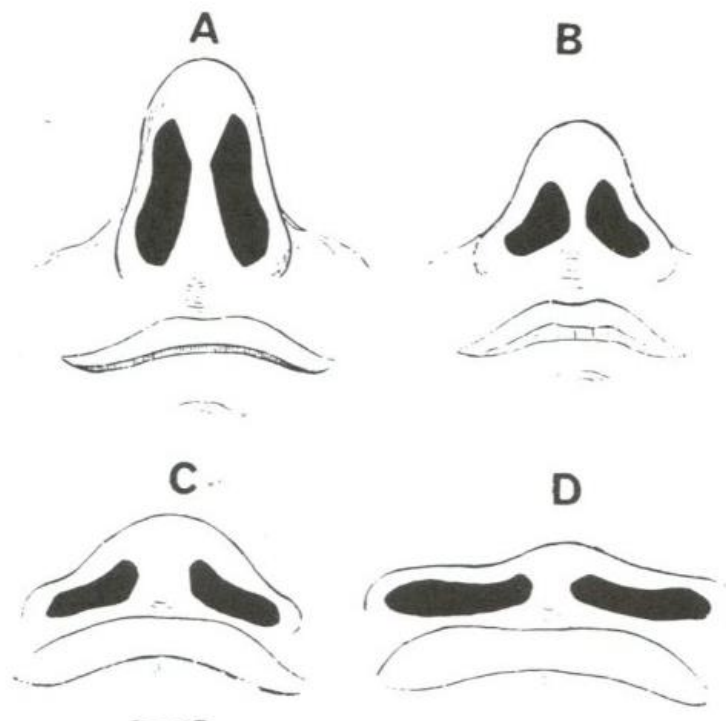


Figure 2. Variations in nasal shape based on race. The leptorrhine nose (A, Caucasian), when compared with the platyrrhine nose (C, African), has increased tip projection and more vertically oriented nostrils. The mesorrhine nose (B, Asian) has intermediate features. Nostril to infralobule ratios decrease when progressing from leptorrhine to platyrrhine. (By permission of Mayo Foundation.)



Rhinoplasty

Initial Evaluation and Consultation

- Ask the patient what she/he likes/dislikes in detail regarding shape and function.
- Must decide if desired outcome is achievable or not.

History:

- Chief complaint:
 - Obstruction, nasal appearance, or both
- Nasal obstruction:
 - Unilateral vs. bilateral
 - Continuous or intermittent
 - History of injury and trauma
 - Sinusitis, polyps
 - Rhinitis, allergy
 - Mouth breathing, snoring
- Medications:
 - Topical nasal medications
 - Systemic decongestants, antihistamines, steroids
 - Anticoagulants, ASA, other NSAIDs
 - **Accutane (Vit. A)**
 - Cocaine, other illicit drugs
- Medical and surgical History:
 - Prior nasal or sinus surgery
 - HTN, diabetes
 - Bleeding (personal and family history)
 - Immune deficiency
 - Connective tissue disease (Relapsing polychondritis)
 - Tobacco use, EtOH
 - Pregnancy (date of LMP) (MP is not a contraindication)
- Appearance:
 - Pt's perception of deformity
 - Helpful if patient was given a mirror.

Examination:

- Assess the nose using principles of facial analysis.

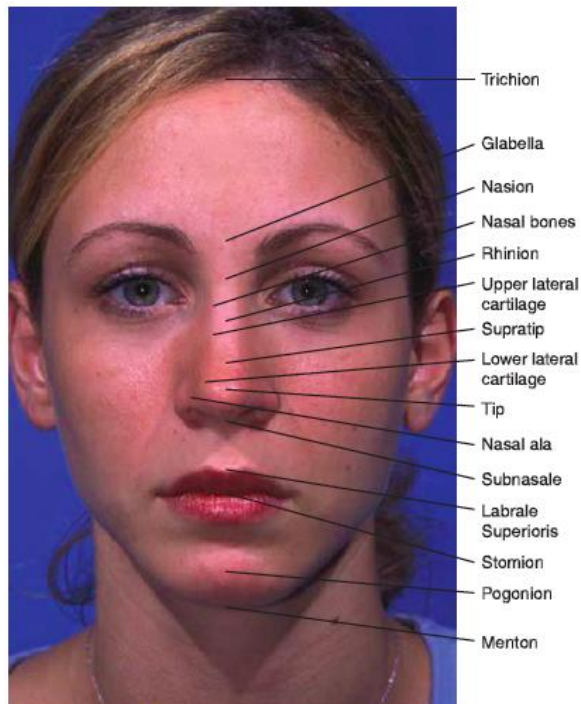


Figure 180.1 Surface anatomy nomenclature—frontal view.

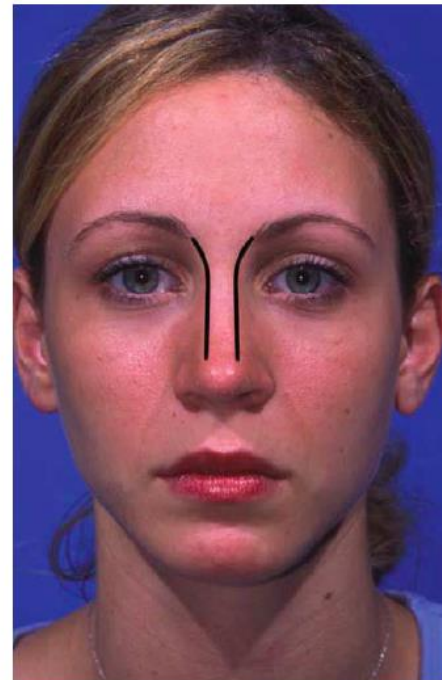


Figure 180.13 The brow-tip aesthetic lines outline the nasal dorsum.



Figure 180.2 Surface anatomy nomenclature—lateral view.

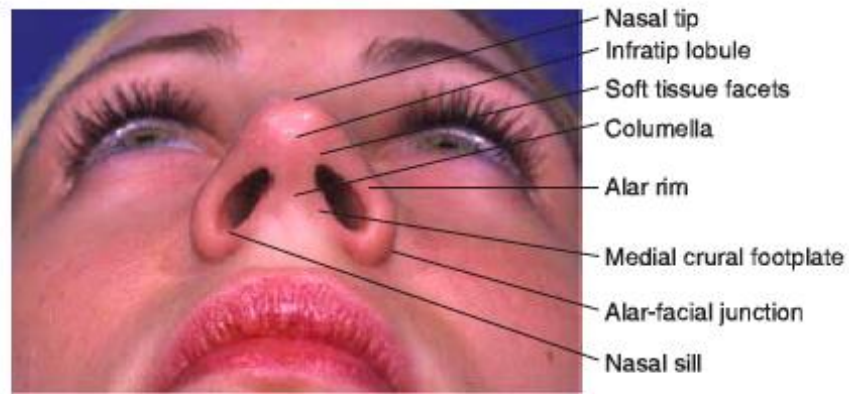


Figure 180.3 Surface anatomy nomenclature—base view.



Figure 180.5 Nasofacial angle—the normal angle measures 30 to 40 degrees.



Figure 180.6 Nasolabial angle—the normal angle measures 90 to 115 degrees.

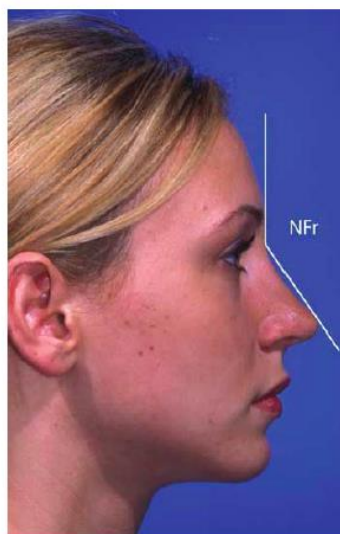


Figure 180.4 Nasofrontal angle—the normal angle measures 115 to 135 degrees.

- **External exam:**
 - Overall shape, proportionate for face, deviation, symmetry, length, width, profile, nasofrontal and nasolabial angles, rotation and projection of tip, **SSTE characteristics**.
- **Internal exam:**
 - Observe for natural inspiration, anterior rhinoscopy, endoscopy.
 - Internal valve.
 - External valve.
- **Informed consent:**
 - Bleeding, infection, intranasal scarring, inadequate corrections, palpable or visible irregularities, **tissue necrosis**, satisfactory cosmetic outcome and **CSF leak, meningitis, death**.
 - Post-op issues → bruising, swelling, black eyes; prepared for bleeding, nasal hygiene and wound management; possibility of packing, splints, and additional procedures (grafts... etc.).
- **Pre-op photos:**
 - Frontal, lateral, oblique and basal.

Surgical Planning (Treatment planning):

1. Determine specifically patient's goals and expectations for surgery
2. Explore patient's motivation for surgery
3. Photos and detailed anatomic analysis (digital if available)
4. Discuss financial arrangements
5. Decide on method of anesthesia
6. Obtain informed consent
7. Suitability for surgery (H&P)
8. Pre-op teaching

Anesthesia

- Can be done under GA or local with sedation; always infiltrate (mark area before).
- Lidocaine with epinephrine (most common); minimize volume to prevent distortion.
- 8-15 ml of 1% with epinephrine typically used; infiltrate wherever operating.
- Site of injections (clumella, vestibule, dorsum, side walls, septum).
- Topical oxymetazoline useful to prevent bleeding

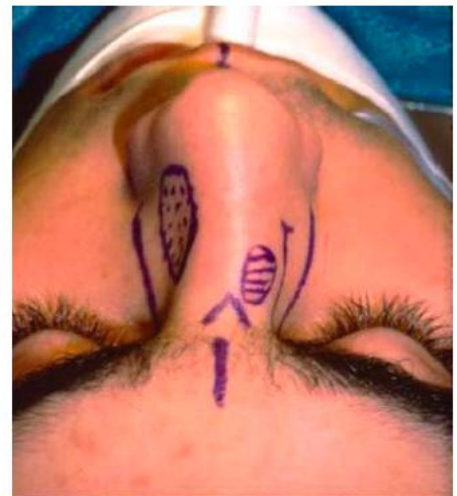


Figure 181.16 The surgical plan is marked on the skin prior to local anesthetic infiltration. Areas of grafting, reduction, and osteotomy are marked.

Overview of the Surgery:

- The typical order of surgery:
 1. Septoplasty
 2. Nasal dorsum
 3. Osteotomies
 4. Nasal tip
- **Incisions:**
 - All dissection should take place immediately above cartilage/bone.
- Septum:
 - Endo-nasal hemi-transfixion incision.
 - **Or** extension of external/anterior rhinotomy incision (complete transfixion incision disrupts fibrous attachments between base portion of the medial crura and septum → reduce projection (**identify anterior septal angle**)).

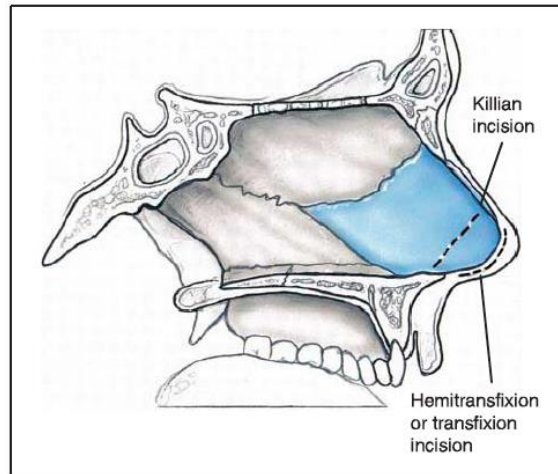
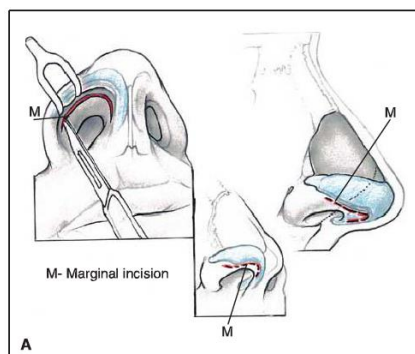
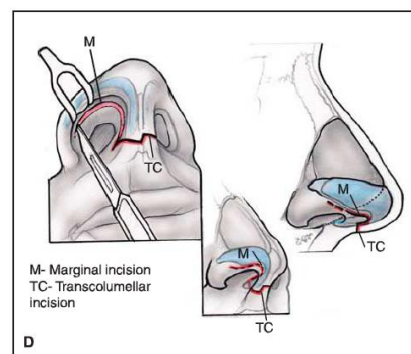


Figure 180.9 Nasal septal access incisions.

- Marginal (Infra-cartilaginous) incision:
 - Made along the caudal margin of the lower lateral cartilage.
- Trans-columellar incision:
 - Made transversely across the short axis (width) of the columella.
 - Joining bilateral marginal incisions at its lateral end to facilitate the external approach to the nose (**for degloving**).
 - Inverted V, V or step.



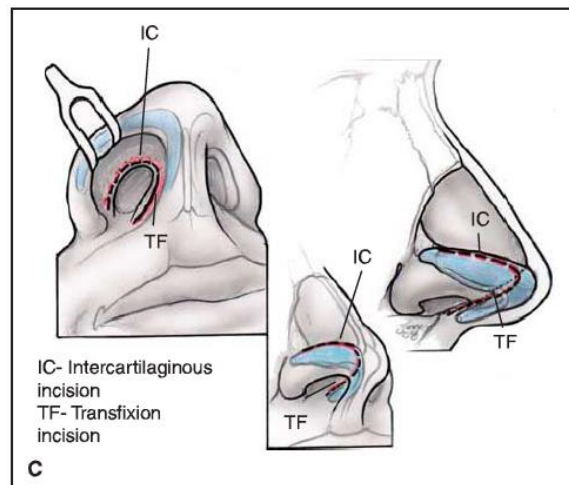
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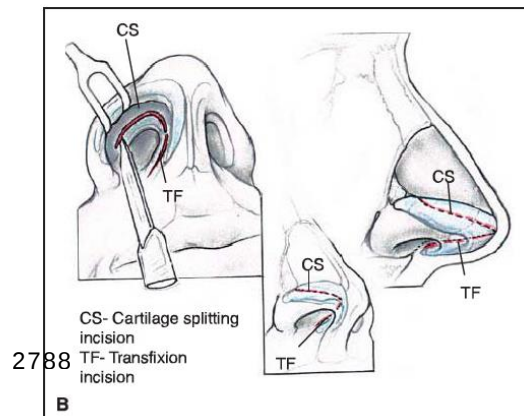
- Inter-cartilaginous incision:

- Made at the junction of the caudal border of the upper lateral cartilage and the cephalic border of the lower lateral cartilage
- Alone it can provide access to the upper two-thirds of the nasal vault (bony and cartilaginous dorsum, nasal sidewall)
- As well as retrograde access to the lateral crura of the lower lateral cartilages.
- When combined with marginal incisions, it allows the surgeon to release and deliver the lower lateral cartilage as a bipediced chondrocutaneous flap (enable tip contouring).



- Trans-cartilaginous (cartilage-splitting) incision:

- Made through the lateral crus of the lower lateral cartilage caudal to the junction of the upper and lower lateral cartilage and at least 5 to 6 mm above the caudal margin of the lateral crus of the lower lateral cartilage.
- Effectively this divides the lateral crus into a superior (cephalic) and inferior (caudal) segment enabling removal of the cephalic strip for volume reduction of the tip cartilage. (cephalic trimming)



- Rim incision:
 - Made within the nasal vestibule along the rim of the nostril margin.
 - Can cause (scar, visibility, retraction, notching, or irregularity along the alar margin).

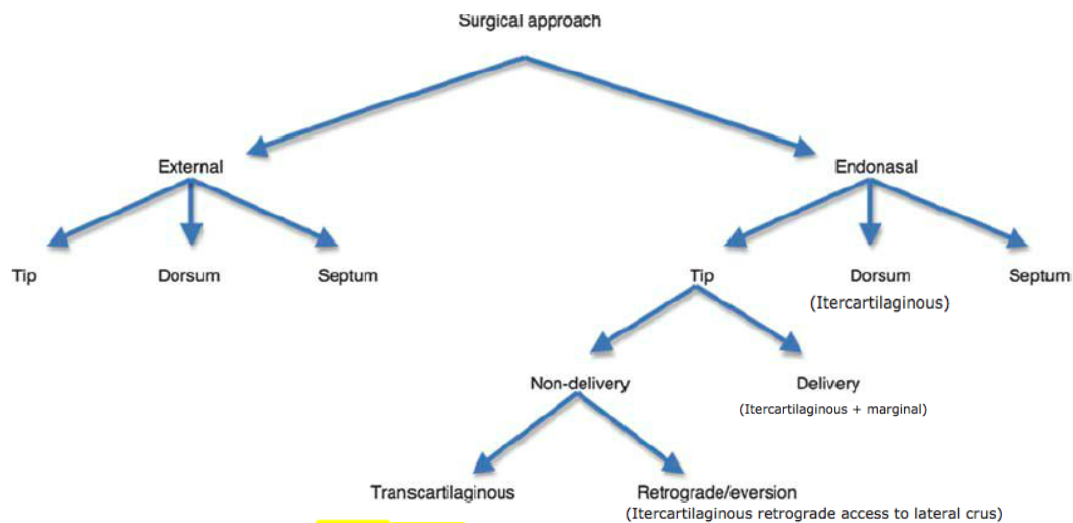
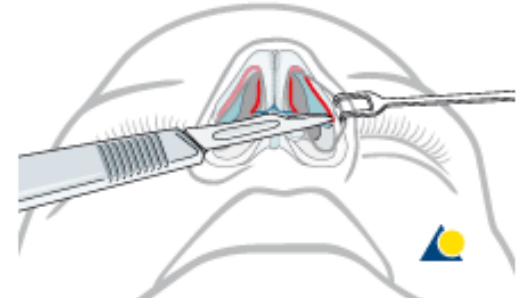


Figure 180.7 Surgical approaches to the nose.

**TABLE
180.1**

GUIDELINES FOR SELECTION OF SURGICAL APPROACH

Endonasal Rhinoplasty

- Dorsal reduction
 - Modest reduction, nasal bones of normal length (not short nasal bones with long upper lateral cartilages)
 - Normal width and alignment of middle third of nasal vault
- Tip surgery
 - Primary (non-revision) surgery
 - To modify tip definition (boxy, wide, bifid, broad/bulbous tip)
 - No gross asymmetry
 - Modest increase/decrease in tip projection
 - Limited tip revision surgery
- Linear deviation of nasal dorsum in need of osteotomies

External Approach Rhinoplasty

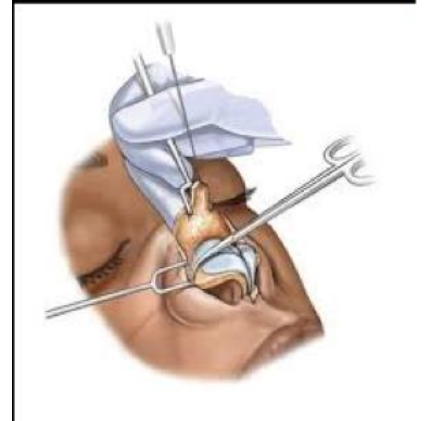
- Congenital nasal deformities, for example, cleft lip rhinoplasty
- Major dorsal reduction or dorsal reduction with narrow/pinched middle third of nasal vault
- Major change in tip projection
- Marked nasal septal deformities
- Twisted nose
- Need for sutured-in-place structural grafting (middle nasal vault or lower third)
- Very thick skin
- Large septal perforation for repair
- Major secondary (revision) surgery

- **Delivery:**
 - For more abnormal or asymmetric tip anatomy.
 - Allows presentation of the alar cartilages as bipediced chondrocutaneous flaps for surgical modification.
- **Non-delivery:**
 - For conservative/minimal tip refinement.
 - Disturbs little of the normal anatomy.
 - Cartilage splitting, or Retrograde.

External Approach

Indications:

1. Extensive tip work
2. Revision rhinoplasty
3. Major reconstruction
4. Cleft lip deformity
5. Crooked nose
6. Saddle nose
- 7. Closure of large septal perforation**
8. Teaching
9. Surgeon preference/experience
10. Exposure for diagnosis or cancer resection



Advantages:

- Maximized and undistorted visibility.
- Access for suture and graft placement.
- **Preserves 2/3 major tip support mechanisms.**

Disadvantages:

- External scar.
- Tip sensation altered.
- Increased post-op edema.
- Disrupts skin/soft tissue **envelope** attachment to framework.

Contraindications:

- Previous lateral rhinotomy incision.
- Previous extended bilateral alotomy (**at alar**).
- Previous excessive thinning of overlying skin.
- Significant medical problems that could potentially increase the surgical risk are a relative contraindication.
- History of significant bleeding problems or a family history of bleeding problems warrants a more extensive pre-op evaluation.

Complications:

- Bleeding.
- Infection.
- **Internal notching from transcolumellar incision.**
- Damage to caudal margin medial crura & domes.

Dissection Plane:

- Plane between the deep fatty layer under the SMAS and above the perichondrium because it is **avascular** and because it **heals with very little fibrosis; (protect neurovascular supply!)** but under periosteum to access skeletal subunit.
- Periosteum of the nasal bones in the midline It inserts into the intranasal suture line and required sharp dissection.

MANAGEMENT OF UPPER 2/3 OF THE NOSE:

Defects of the Bony Vaults:

- Most bony defects are from blunt trauma.
- If associated laceration, dermis can scar to periosteum causing SSTE deformity.
- Other causes: prior surgery; neoplasm; congenital deformity.
- Severe fractures as nasoorbital ethmoid (NOE) fractures can be disfiguring due to retrodisplacement of nasal bones, deprojection and flattening of the dorsum and shortening of the nose (telescopic defect); can be associated with telcanthus due to disruption of medial canthal tendon insertion on nasal bone.
- **Bony vault defects don't usually affect function.**

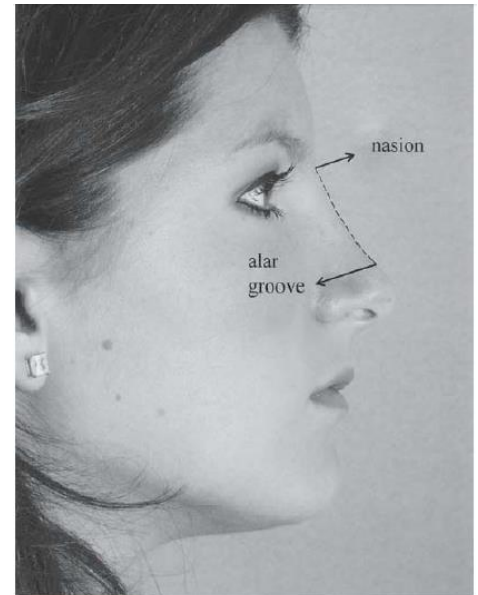


Figure 181.2 Upper nasal two-thirds: nasion to alar groove.

**TABLE
181.1**

NASAL VAULT DEFECTS

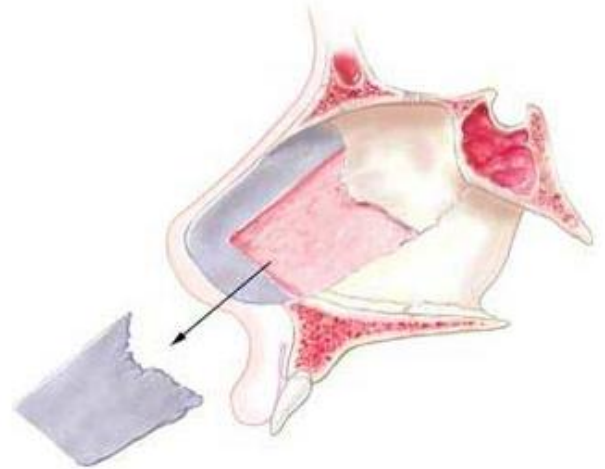
Dorsal hump	Curvatures
Deep nasion	Pollybeak
Saddle nose	Bony prominences
Asymmetric nasal bones	Rocker deformity
Short/long nasal bones	Open roof deformity
Wide/narrow bony pyramid	Inverted-V deformity
Depressions	
Twists	
Partial/total rhinectomy (nasal cancer, gunshot wound)	

Cartilaginous Vault Defects:

- Cartilaginous vault deformity frequently affects internal valve leading to airway compromise.
- Malleable, so susceptible to birth trauma and inflammatory changes and autoimmune

Management of the Cartilaginous Vault:

- Risk factors for poor outcomes:
 - Short nasal bones
 - Weak ULC
 - Thin skin
 - Narrow bony pyramid
 - Previous surgery or trauma
- Septoplasty is done first (open approach allows great exposure).
- Make sure to leave 1 cm dorsal and caudal struts (L-strut).



- **Spreader grafts:**

- Placed between the junctions of ULC and septum.
- Widen and stabilize the nasal valve (also widen dorsum, straighten upper septum).
- Usually 1.5-2.5 cm long; 1-3 mm wide.
- Start below bony-cartilaginous junction to anterior septal angle.
- Grafts can be layered to add more bulk.
- Lateralizes ULC and strengthens the middle vault to resist inward collapse on inspiration.
- For unilateral middle vault depression, spreader graft can lateralize and elevate the ULC.

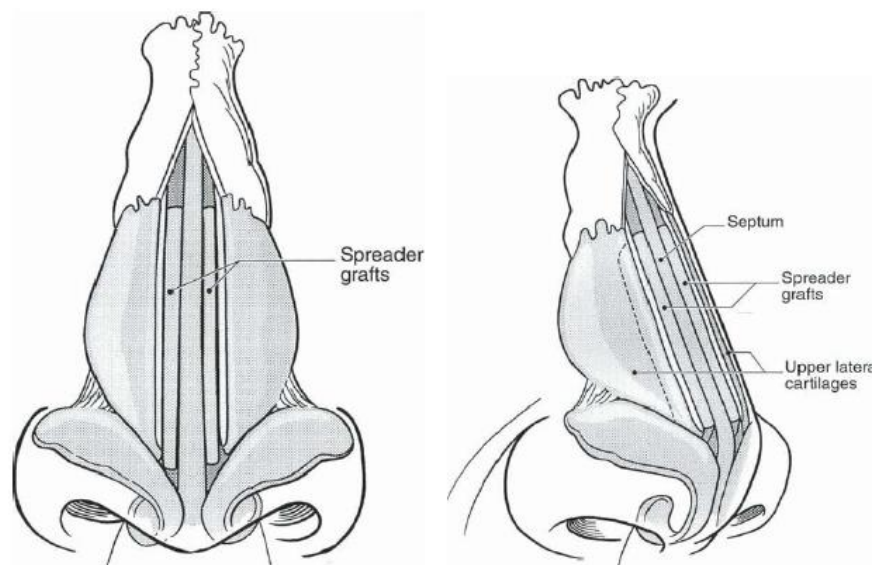


Figure 181.8 Placement of spreader grafts between septum and upper lateral cartilages. (From Papel ID. Management of the middle vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:409, with permission.)

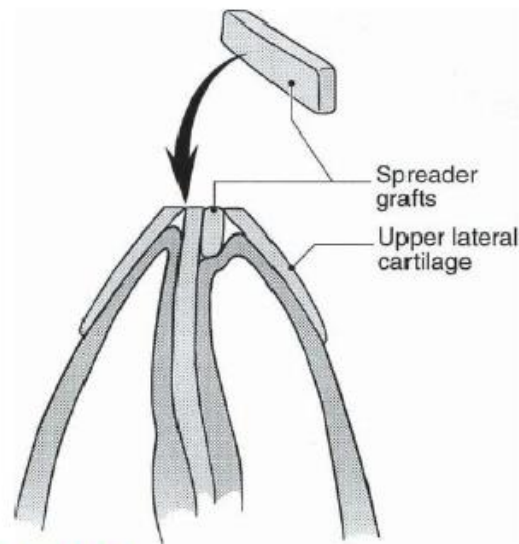


Figure 181.9 Placement of spreader graft lateralizes and elevates the upper lateral cartilage, widening the middle vault. (From Papel ID. Management of the middle vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:409, Fig. 35-5, with permission.)

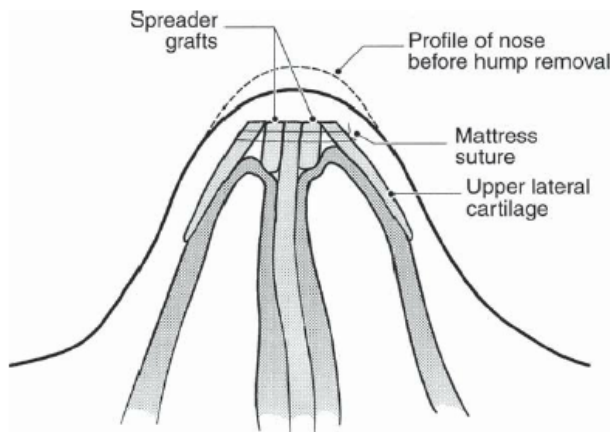
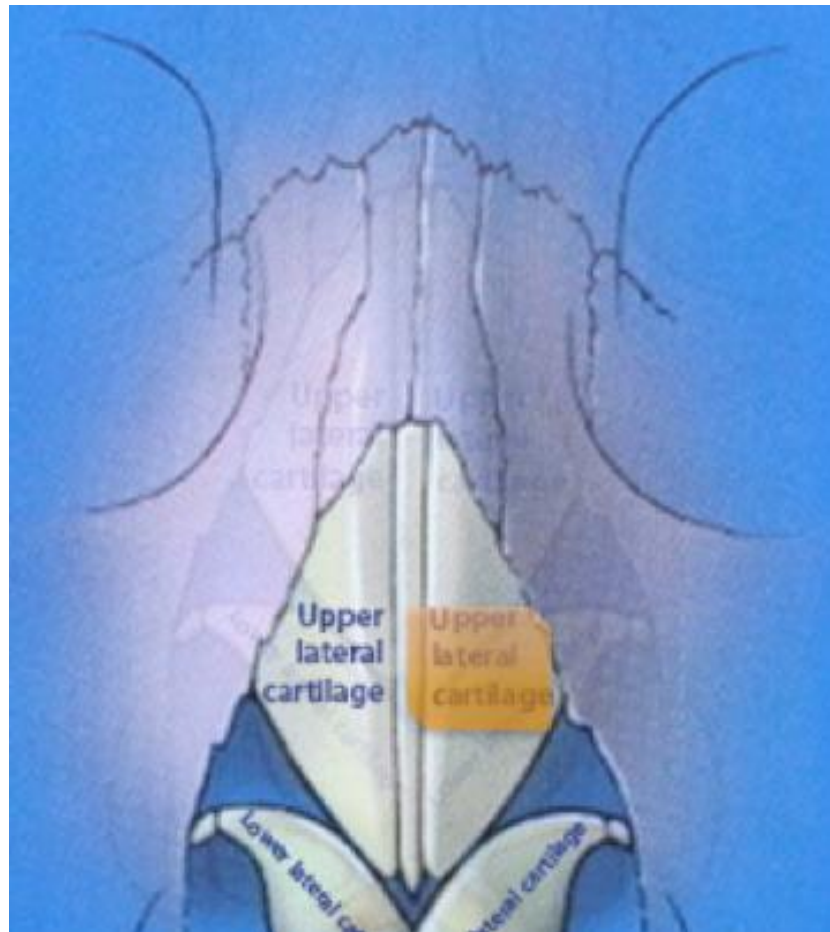
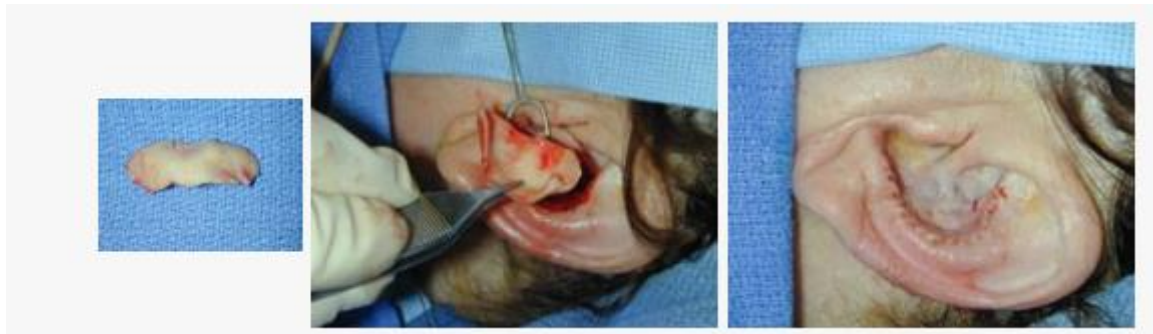


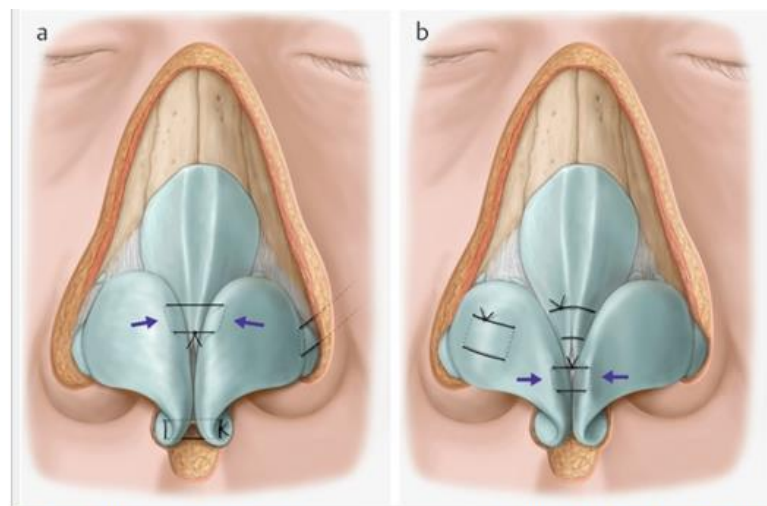
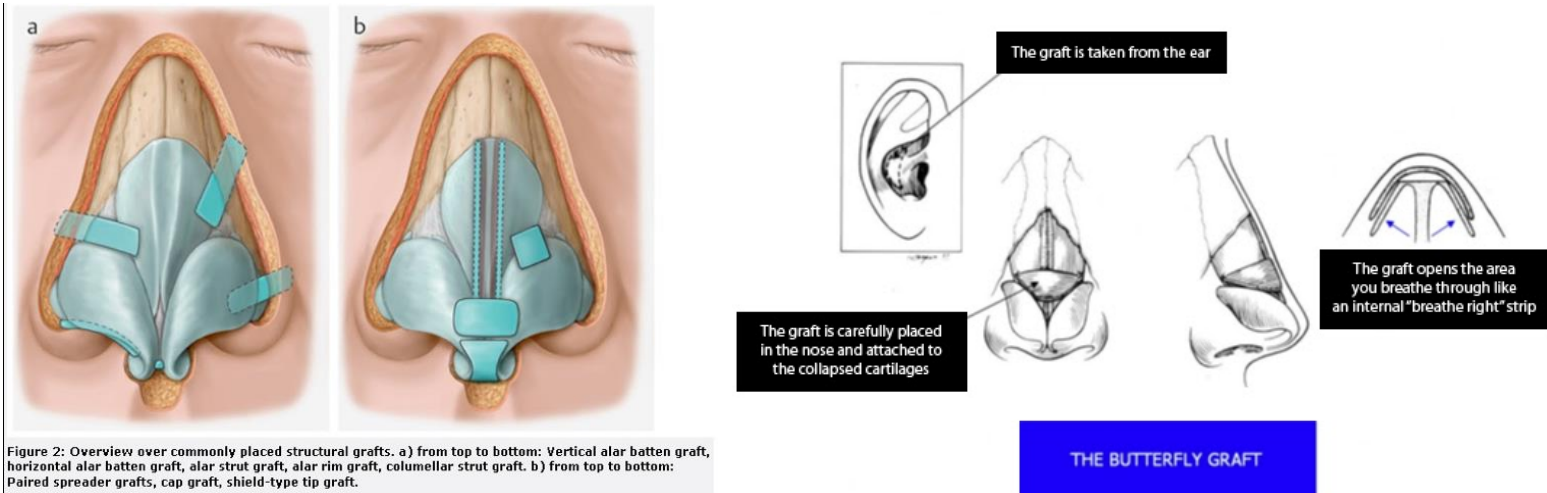
Figure 181.10 Spreader grafts in place and fixed with mattress sutures. The original dorsal height is identified. (From Papel ID. Management of the middle vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:409, with permission.)

- **Reverse spreader grafting:**
 - Reduction of the horizontal width of cartilaginous dorsum.
 - For overly broad middle nasal vault, minimal obstruction, no valve collapse.
- Revision cases:
 - Open approach is preferred.
 - If ULC has been resected, conchal cartilage **onlay grafts** in addition to spreader grafts can be used.
 - Grafts are fixed into position with **5-0 PDS** (polydioxanone) mattress sutures.



- **Other methods to augment valve function:**

- **Alar batten grafts**
- **Butterfly grafts**
- **Various suture methods (flaring sutures)**

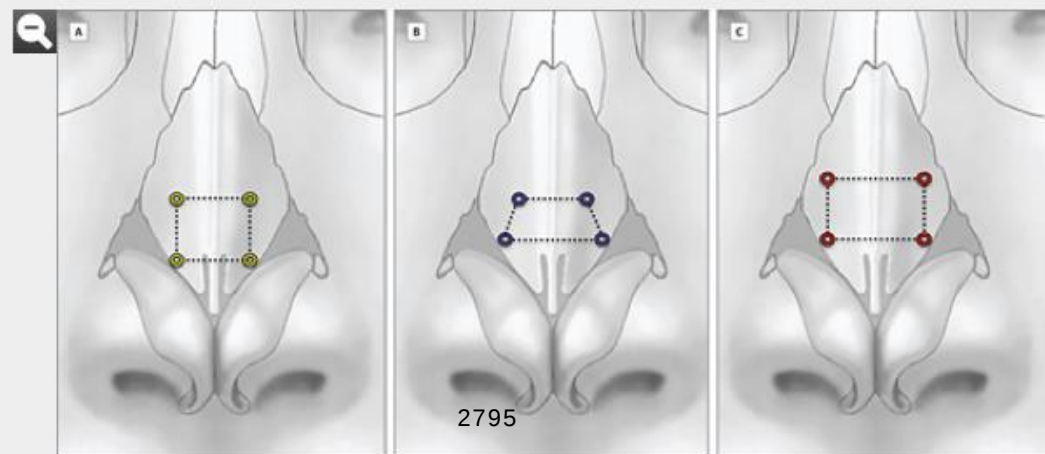


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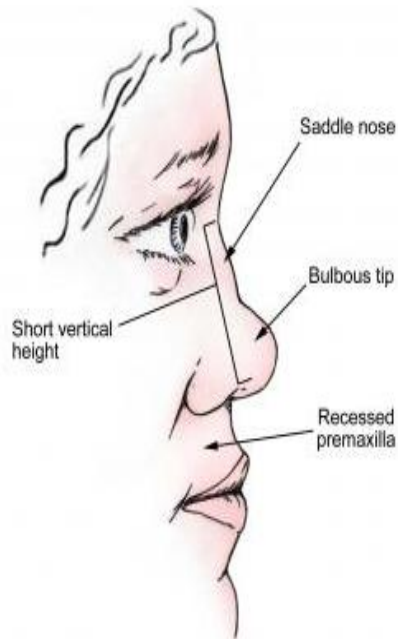
Figure 10: Frequently placed sutures. a) From top to bottom: High interdomal suture, flaring suture and middle crural suture. b) From top to bottom: Flaring suture, lateral crura suture, interdomal suture.

A, Medial (yellow square). B, Modified (blue trapezoid). C, Lateral (red rectangle).



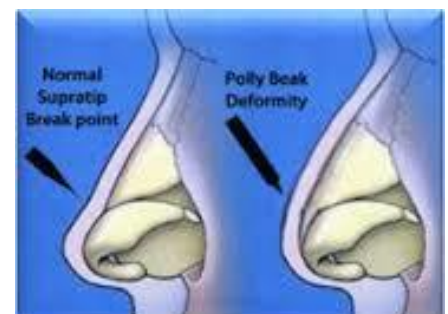
- **Saddle-nose deformity:**

- Often have middle vault collapse with large septal perforation.
- Graft types
 - **Autografts** - Auricular cartilage, rib, calvarial bone.
 - **Homografts** - Irradiated rib, pooled acellular dermis.
 - **Xenografts** - Leather, duck's sternum, bovine cartilage
 - **Precious metals** - Titanium, gold, silver, metal alloys
 - **Inert bioimplants** - Coral, ivory
 - **Synthetic compounds** - Silicone, polytetrafluoroethylenes, polyamide mes
- Dorsal graft fixation with **calvarial bone** (lag screw) or **rib cartilage** (sutures).
- Crushed cartilage or Alloderm can smooth irregular profile/
- **Dissected cartilage**



- **Pollybeak deformity:**

- Relative underprojection of the nasal tip with regard to the projection of the dorsum.
- Dorsal polybeak-normal tip projection; overprojected dorsum.
- Tip polybeak-normal dorsal projection; tip underprojection.



Management of Bony Vault:

- Goals:
 - Attain appropriate dorsal projection (reduction vs. augmentation).
 - Creation of smooth dorsal contour.
 - Correction of nasal width.
 - Straightening of the crooked nose (straighten).

Hump Reduction (illusion increase in projection):

- ULC insert deep to the caudal margin of nasal bones; care is taken not to disarticulate this union (elevate periosteum with a Joseph elevator 1-2 mm above the rhinion).
- Bone removal with a double-guarded osteotome or a rasp.
- Cartilage removal with knife.
- Many will carve down cartilage hump before targeting bony hump.
- Remember thin skin; advise to leave a slight skeletal convexity to avoid "scooped-out" appearance.
- Don't leave distal cartilaginous dorsum too high: **pollybeak deformity**.
- Can result in open roof deformity.

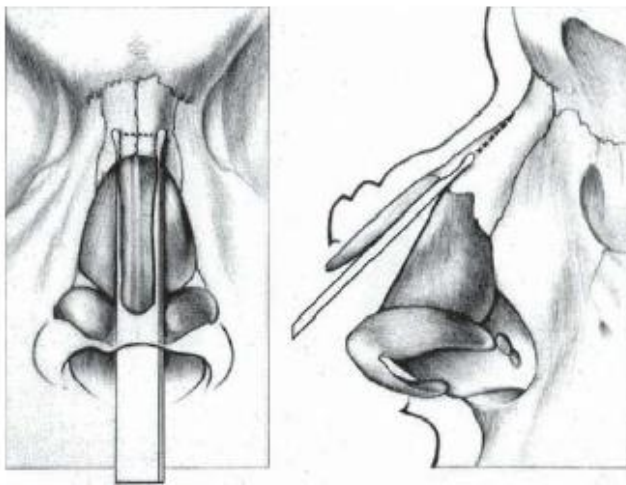
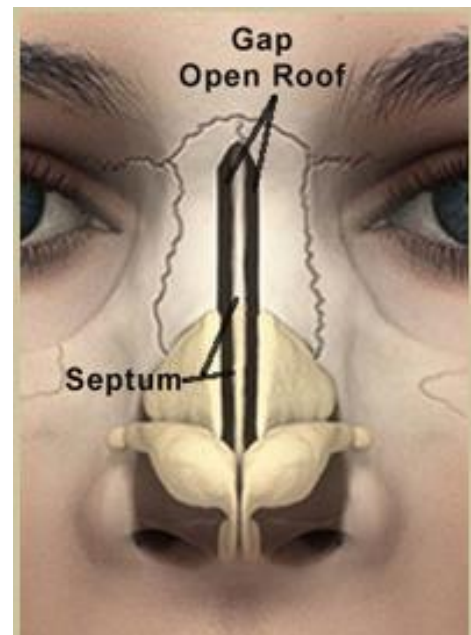
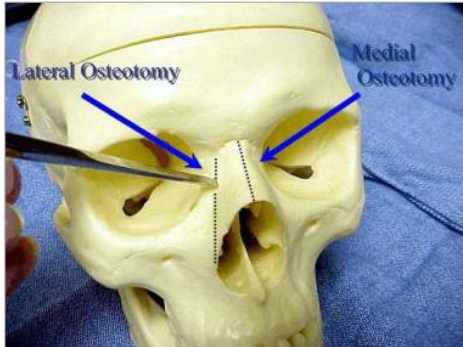


Figure 181.13 Removal of dorsal excess. (From Mostafour SP, Murakami CS, Larrabee WF Jr. Management of the bony nasal vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:403, with permission.)



Osteotomies:

- Osteotomies are commonly used to improve irregularities in the brow- tip aesthetic line and correct open roof deformities (diastasis of the nasal bones) associated with bony dorsum reduction.

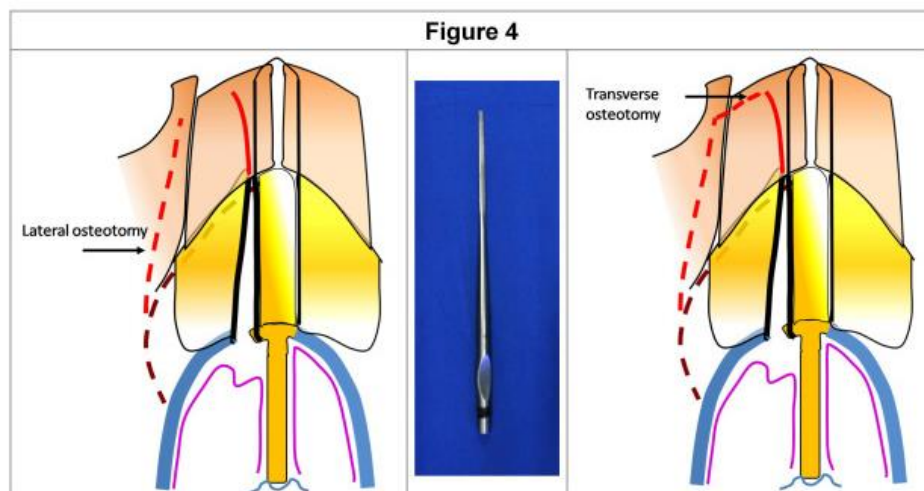


Osteotomy Indications:

- a. Straighten a deviated dorsum
- b. Narrow nasal sidewalls
- c. Close an open nasal vault

- **Lateral osteotomies:**

- Limited to the thin bone of pyriform aperture.
- **Linear** or **perforating**; internal or external.
- Mucosal stab incision is made just lateral to anterior face of the inferior turbinate.
- The ideal trajectory is described as “high-low-high” which indicates the anteroposterior position on the nasal pyramid.
- Curved guarded 4-mm osteotome is placed on the pyriform aperture (45 degree angle to the facial plane).
- Preservation of the inferior segment of the pyriform maintains the lateral suspensory ligaments (ensuring nasal patency).
- Osteotome is then transitioned to a plane parallel to the facial plane and is directed to the medial canthal area.
- Terminate at the medial canthus (where thicker frontal bone is encountered).



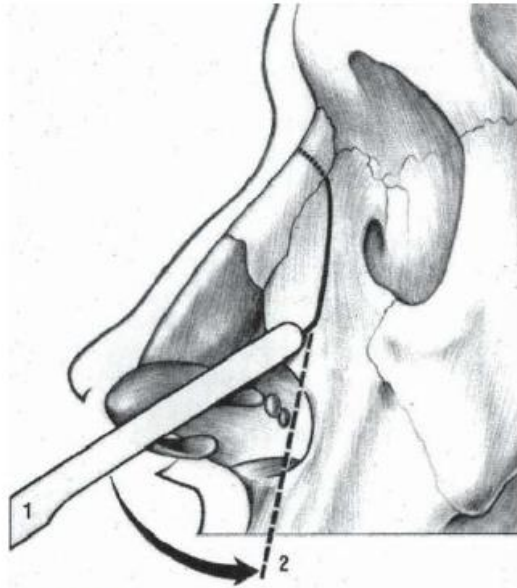


Figure 181.14 The lateral osteotomy is initiated at the anterior face of the inferior turbinate, initially perpendicular to the plane of the piriform aperture (position 1). The osteotomy is then transitioned (position 2) and carried to the medial canthal area. (From Mostafaei SP, Murakami CS, Larrabee WF Jr. Management of the bony nasal vault. In: Papel ID, ed. *Facial plastic and reconstructive surgery*, 2nd ed. Stuttgart, Germany: Thieme, 2002:404, with permission.)

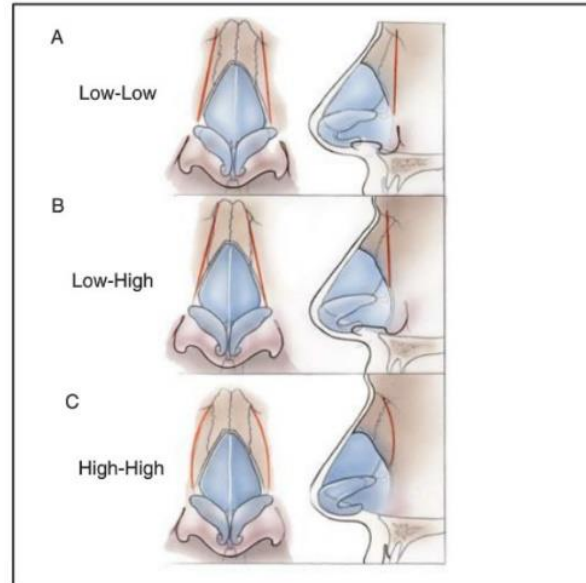


Figure 183.6 This figure shows three different types of osteotomies that can be performed. The upper image is that of a low-low osteotomies, where the osteotomy is closer to the maxilla (low) throughout its route. The middle image is that of an osteotomy that starts low and ends high (closer to the dorsum). The bottom image is that of a high-low-high route, which is the most commonly used type of osteotomy route. The midportion of the osteotomy dips closer to the maxilla (low).

- If lateral osteotomy begun too low along the piriform aperture, medialization of the bone can cause obstruction.
- If lateral osteotomy placed too medially, can result in "stair-step" deformity on the side of nose; chance of injury to lacrimal apparatus.

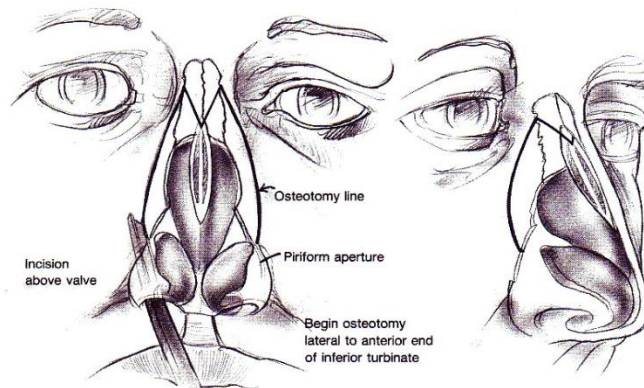


Figure 171.18 Illustration of medial and lateral osteotomies. Orientation of the medial osteotomy may need to be more sagittal from the cephalic portion of the hump reduction site to avoid creation of a "rocker" situation as the nasal bones are tilted medially.

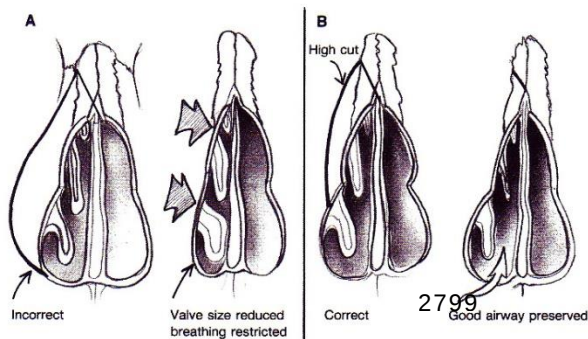


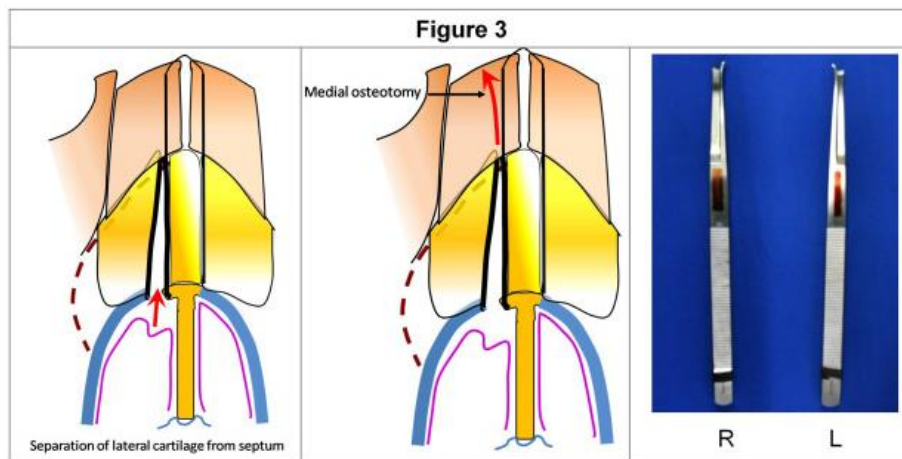
Figure 171.19 A: If the lateral osteotomy is begun too low along the piriform aperture, medialization of the bone can cause an obstruction of this portion of the nasal airway. B: Beginning the osteotomy at a higher point allows for medial tilting of the bone without clinical compromise to the airway.

- **Perforating later:**

- Internally (transnasally) or externally (percutaneously).
- Theory: intact periosteum preserves structure and adds stability; make series of perforations along fracture path, then fracture manually.
- Preferred technique when maintenance of support is critical (revision cases or short nasal bones).

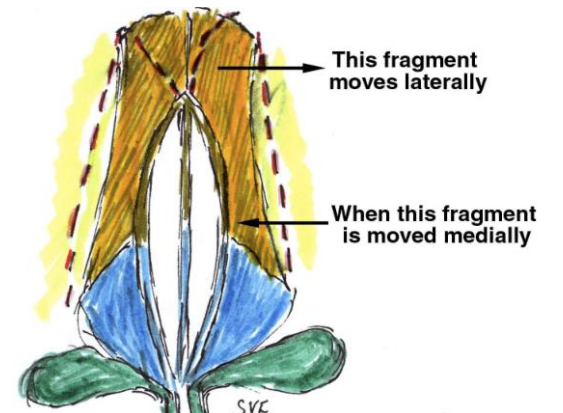
- **Medial osteotomies:**

- Start at the medial aspect of the caudal margin of nasal bones near the septum; superolateral trajectory, meeting the lateral osteotomies.
- If needed Medial done first.
- Must tilt bones inward to close open-roof deformity.
- Near the keystone area; care must be taken.
- Indications:
 - When mobilization of the entire nasal sidewall is required.
 - To help prevent uncontrolled or irregular back-fracture from the upper portion of a lateral osteotomy.
 - To widen an overly narrowed bony nasal vault.



Rocker deformity:

- Occurs when medial osteotomies are carried too high into the frontal bone (superior to medial canthus).
- The superior aspect of bone rocks laterally when the nasal bones are infractured, nasal bone inferior to the osteotomy sinks relative to the bone superior to the cut.
- Back fracture is performed at the end; if incomplete or inadequate, percutaneous osteotomy using a 2-mm instrument can help.



- **Intermediate Osteotomy:**

- If nasal bone has a very convex, concave, or irregular topography, an intermediate osteotomy may be necessary between the medial and lateral osteotomies (performed medial to lateral so that the cuts are always made on stable bone).
- If doing intermediate, do it before lateral.

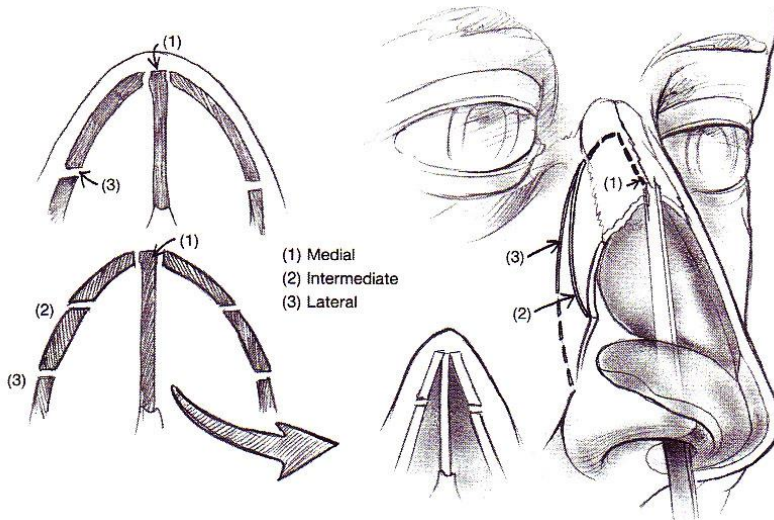


Figure 171.16 Illustration of sites for medial, intermediate, and lateral osteotomies. The caudal origin site for the lateral osteotomy must be placed too low so that the airway does not become obstructed as the bones are repositioned (Figs. 171.18 and 171.19).



Figure 181.15 An intermediate osteotomy is marked on the skin and the inferior edge of the bone is noted. A percutaneous method is used in a perforating manner.

- **Transverse Osteotomy:**

- Severe trauma result in excessive shift of the bony dorsum and bony pyramid.
- Indicated if there is need to move the entire bony dorsum as a unit, utilizing the transverse osteotomy in addition to traditional osteotomies.



- **Grafts:**

- Bony vault → radix grafts, onlay grafts, anchored bone grafts.
- **Radix grafts** increase the projection of nasofrontal trough (excessively deep nasion).
- **Alloderm** can be used as dorsal onlay graft to add height or camouflage irregularities.
- For significant bony vault defects, **calvarial bone graft**, cantilevered off the frontal bone can be used.

NASAL TIP SURGERY



Figure 182.1 Lateral view of the structures of the nasal tip.

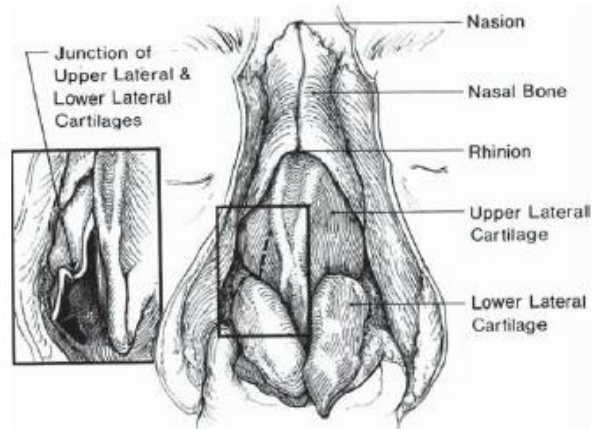


Figure 182.2 Frontal view of the underlying anatomic structures of the nasal tip.

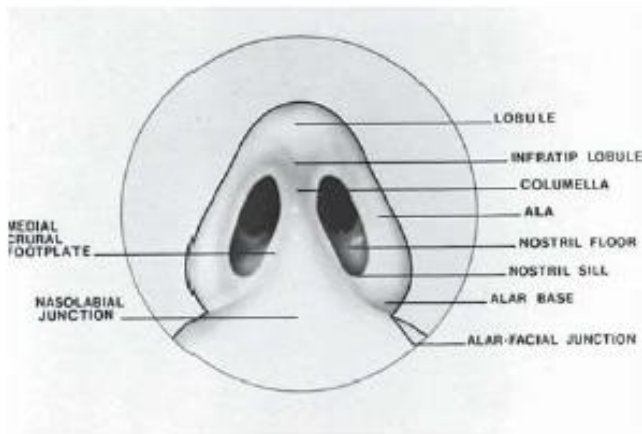


Figure 182.3 Anatomy and nomenclature of the nasal tip viewed from the basal aspect.

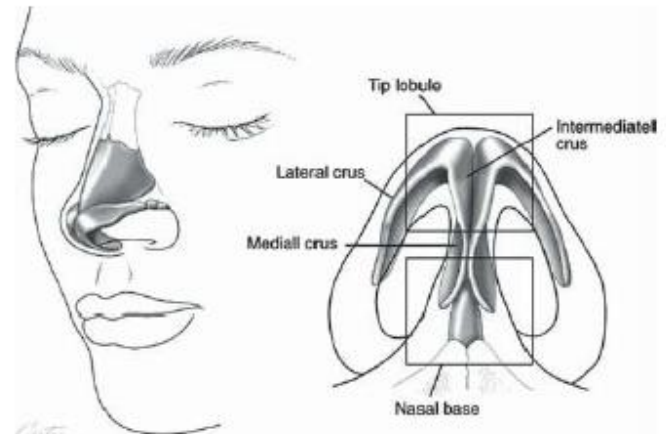


Figure 182.4 Underlying anatomic structures of the nasal base.

- Adjustments: projection or rotation, definition, support and symmetry.
- May also need to strengthen for A/W support.
- Avoid over-resection of cartilage; can't go back once cut!
- Poor outcome in tip surgery:
 1. Thick skin
 2. Many sebaceous units
 3. Poor elastin and collagen content

Analysis and Diagnosis:

- **Tip recoil** test to identify weak LLC or low anterior septal angle.
- Assess tip support (tripod model very useful here).
- Medial crura assessment on base view:
 - Short medial crura-little tip support and leads to deprojection.
 - Long medial crura- good support but prevents deprojection.
- Assess nasofrontal angle, depth of radix, chin.
- Assess for reasons for under or overprojections:
 - Overprojection:
 1. Long medial crura
 2. Tall domes
 3. Anteriorly positioned ant septal angle
 - Underprojection:
 1. Short medial/lateral crura
 2. Low anterior septal angle
 3. Flat domes
- **4. Hypoplastic premaxilla**
- Nasal obstruction and tip deformities also due to position and shape of LLC:
 - Cephalically positioned LLC → poor support of lat nasal wall; bulbous tip, pollybeak deformity.
 - Internally recurvate lateral crura (static obstruction).

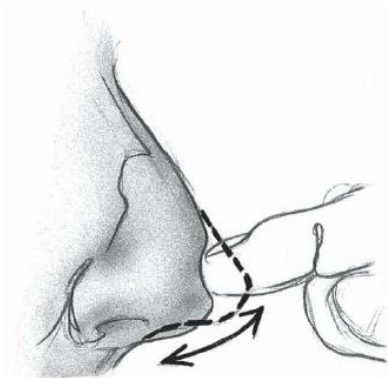


Figure 182.8 Demonstration of finger palpation of the recoil mechanism of the nasal tip. The relative resistance to deformity demonstrated by the nasal tip provides a useful indication of the integrity of nasal tip support mechanisms.



Figure 182.9 Tip position is a consequence of the combined forces of the three legs of the tripod as well as the support provided by the caudal septum.



Figure 182.7 This patient had a bulbous tip and thick skin. After several reductive procedures, the tip is poorly supported and unrefined.



Figure 182.14 When lateral crura are too long, they can recurve into the airway, creating a bulge on the internal vestibular surface of the lateral nasal wall and cause obstruction of the nasal valve.



Figure 182.13 Cephalic positioning of the lateral crura often appears as a bulbous tip with a "parentheses" appearance on frontal view.

Controlling Nasal Tip Projection:

- Hallmark of poorly supported tip is the pollybeak deformity (ideally concave supratip break becomes a convexity).
 - Reverse-"scooped out" appearance.
- Thick skin and bulbous tip:
 - Thick skin will not contract and redrape with excessive removal of underlying structures.
 - Structure must be "projected into" the thick soft tissue envelope.

Stabilizing the Nasal Base:

- To create a stable foundation at the nasal base.
- To stabilize medial crura:
 - Suture medial crura to caudal septum or caudal extension graft
 - Sutured columellar strut
 - Extended columellar strut



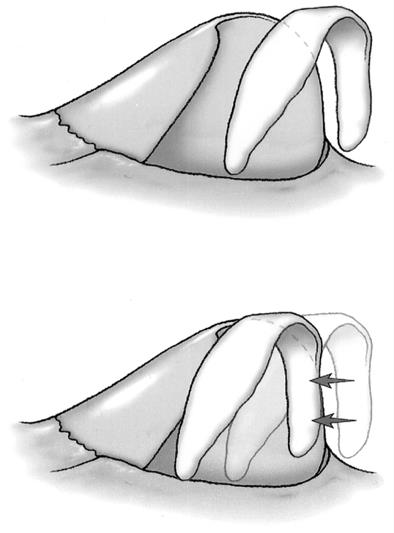
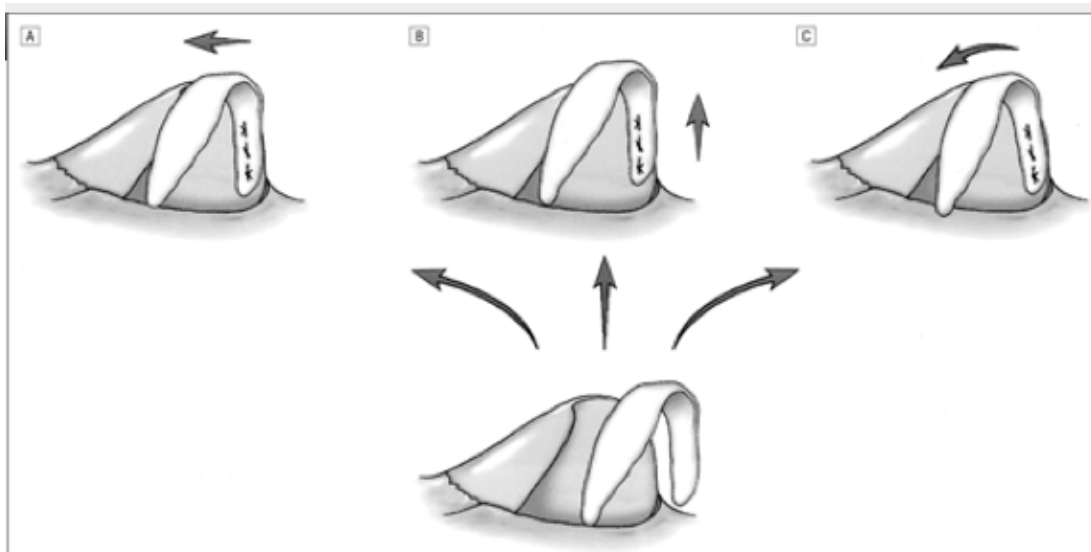
Figure 182.15 A columellar strut is placed by dissecting a pocket between the medial crura and suturing it in the midline.



Figure 182.16 The extended columellar strut usually sits on the premaxilla or is suture fixated into a notch made in the nasal spine. The strut is placed end to end with the septum and stabilized with extended spreader grafts.

- **Medial crura suture technique "Tongue-in-groove technique:**

- **Increase rotation and projection.**
- Medial crura repositioning on the caudal septum (4-chromic mattress suture; 5-0 polydioxanone).
- Excellent tip support and control of tip appearance.
- Indications:
 - Tension-nose deformity (A naturally-occurring cosmetic deformity characterized by dorsal overgrowth of the nasal septum. Hallmark features include "pollybeak" overgrowth deformity of the nasal bridge, overprojection of the nasal tip, and a wide nasal pedestal.)
 - Over or underprojected tip
 - Increase rotation
 - Hanging columella



- Avoid:
 - Retracted columella.
 - Obtuse columella-labial angle.
 - Over-rotated nose.
 - Can be avoided by selecting those with long caudal septum that would have been resected otherwise.

- **Caudal extension graft:**

- **Increase rotation and projection.**
- Overlap with septum, or end-to-end with splinting grafts
- Used to create a septum long enough to bind to the medial crura in the midline
- Similar to a columellar strut except it is more stable, as it is connected to the existing caudal septum

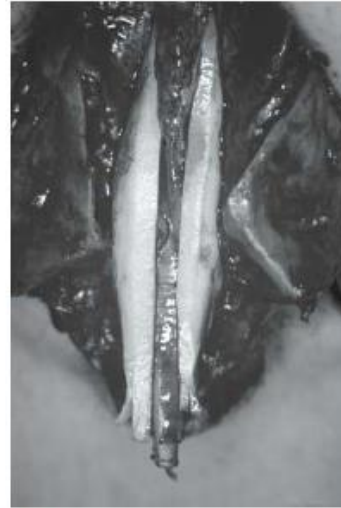
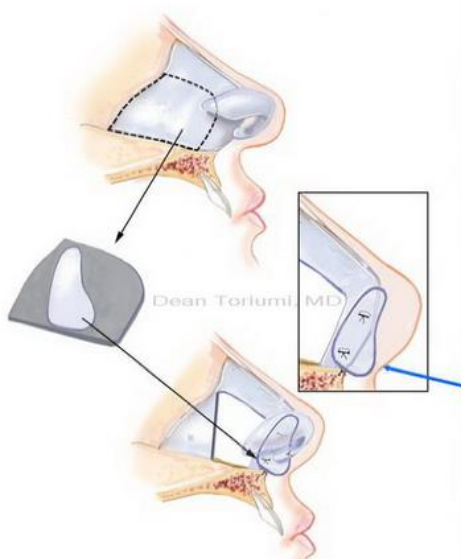


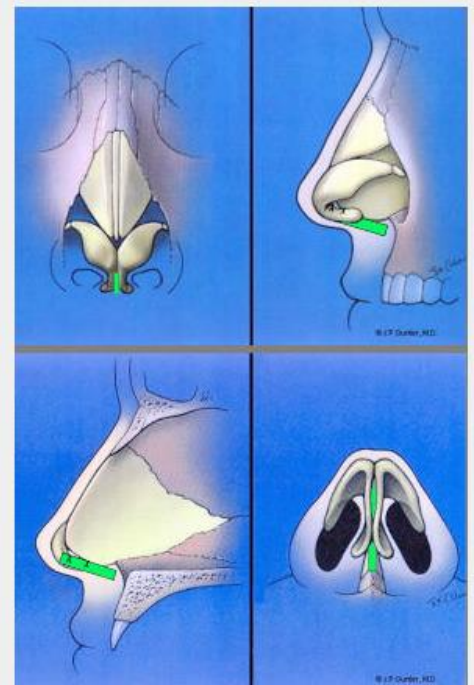
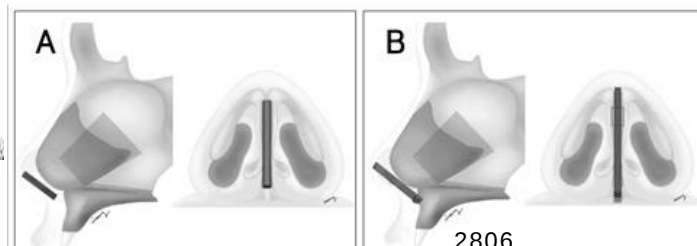
Figure 182.18 Inline caudal extension graft with splinting cartilage grafts.

- **Floating columellar strut:**

- **Increase rotation, tip thinning.**
- Use if good medial crura position but poor structure (buckling).
- 5-12 mm long and 3-6 mm wide and < 3 mm thick.
- Make pocket between medial crura.
- Don't extend to spine as it can cause clicking when moving.
- **Floating strut will not increase projection; just adds support.**



Figure 182.15 A columellar strut is placed by dissecting a pocket between the medial crura and suturing it in the midline.



- **Extended columellar strut:**

- **Increase rotation, tip thinning, increase projection.**
- Good if tip underprojected from poor support.
- Carved from costal cartilage and attached to ant nasal spine.
- Can be combined with premaxillary graft (cleft lip).

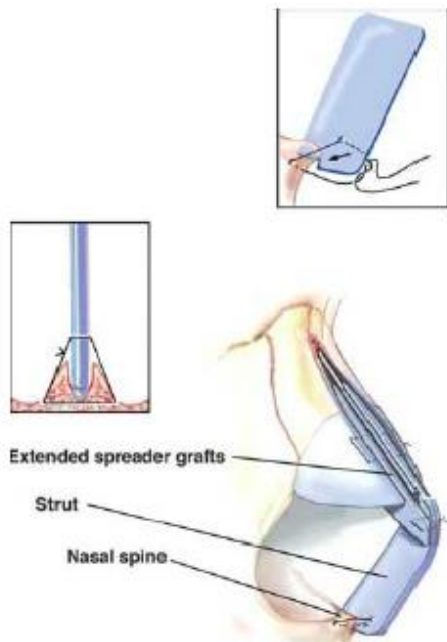
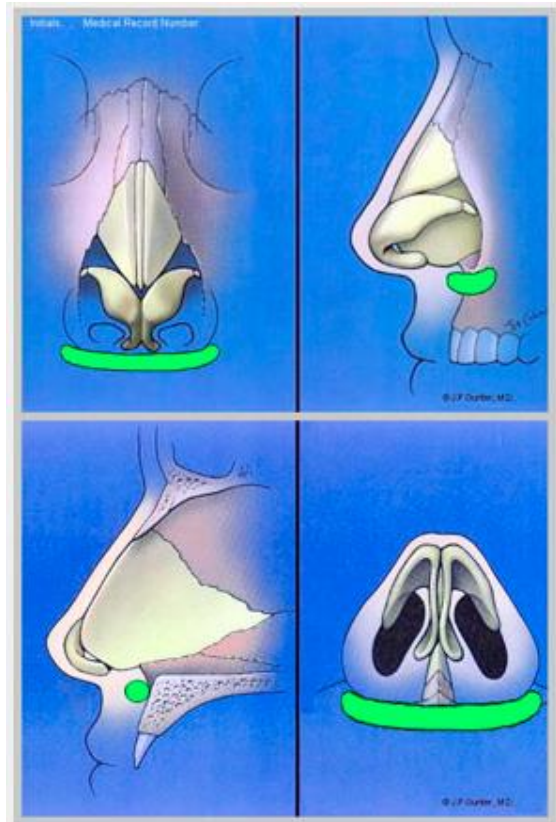


Figure 182.16 The extended columellar strut usually sits on the premaxilla or is suture fixated into a notch made in the nasal spine. The strut is placed end to end with the septum and stabilized with extended spreader grafts.



- **Plumping Graft:**

- A plumping graft is placed at the **nasolabial angle** and is used to augment the area in or around the nasal spine.
- These grafts can make a long nose look shorter by an optical illusion.



Nasal Tip Surgery:

- Tip thinning and increase projection.
- Tip work after solid foundation at the base has been established.
- Underprojected tip (both increases projection without affecting base):
 1. **Dome-binding suture** (intradomal, transdome or double dome).
 2. **Shield graft**.
- Dome-binding suture:
 - Narrows and projects the tip.
 - Usually combined with interdome {single dome}.
 - Lateral crura steal creates a new dome.
- Spanning suture:
 - When placed up in LLC (narrow tip, **increase flaring**).
- Shield graft:
 - Not used in thin skin.
 - Cover the leading edge with fascia, perichondrium, or **cap graft** in medium skin pts.
 - Attaches to medial crura in infratip lobule.

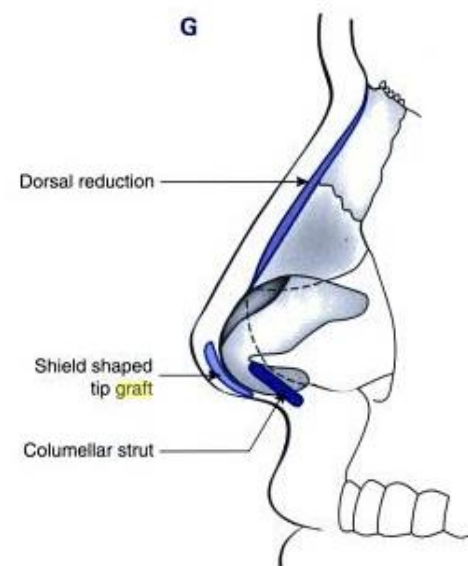
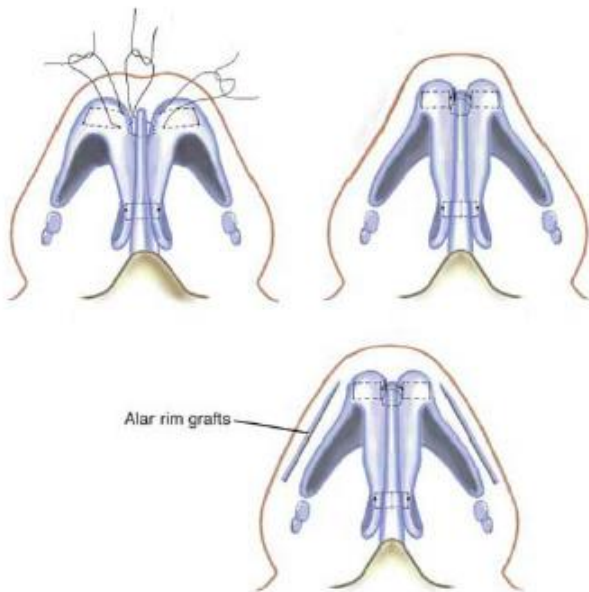
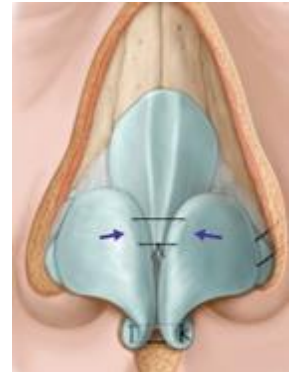


Figure 182.22 Separate dome-binding sutures are applied as shown with the knots tied between the domes. In many cases a separate interdome suture is placed to bring the domes closer together. If necessary alar rim grafts can be placed to eliminate any pinching of the nasal tip.





- **Lateral crural grafts:**
 - Overresected or misshapen lateral crura.
- **Lateral crural struts:**
 - Between lateral crura and vestibular skin for flattening lateral crura that are bulbous or internally recurvate; also to provide structural support.

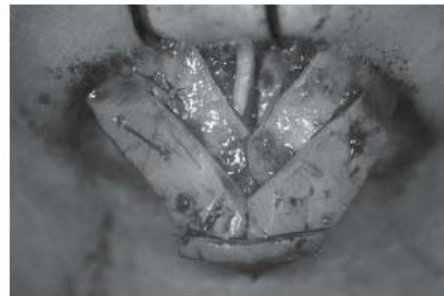
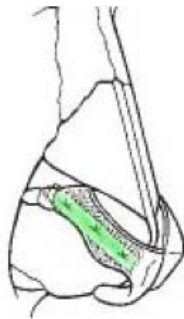


Figure 182.28 Lateral crural grafts conjoined with a shield graft.

- **Cephalic trim:**
 - **Increase rotation, definition.**
 - Often combined with lateral crural strut grafting.
 - Leaving 8-10 mm of the lateral crus.
 - Decreases supratip bulk if done medially.
 - Rotates tip cephalically by creating a gap between the LLC and ULC.
 - LLC scars upwards.
 - Trimming laterally is bad news; weakens sidewall.

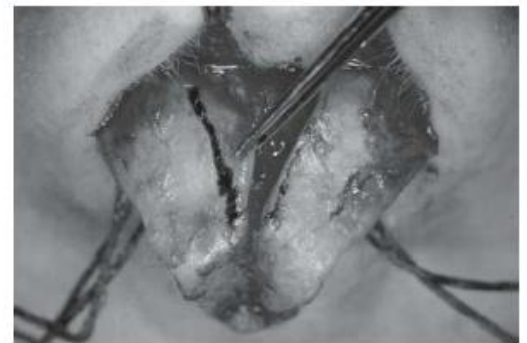


Figure 182.20 Medial-cephalic reduction of the alar cartilages, maintaining a generous residual complete strip.

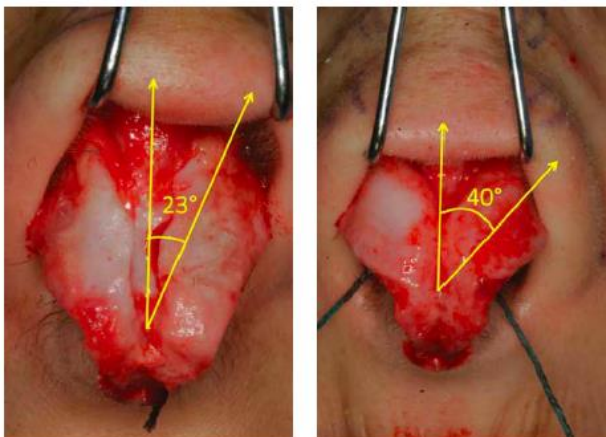
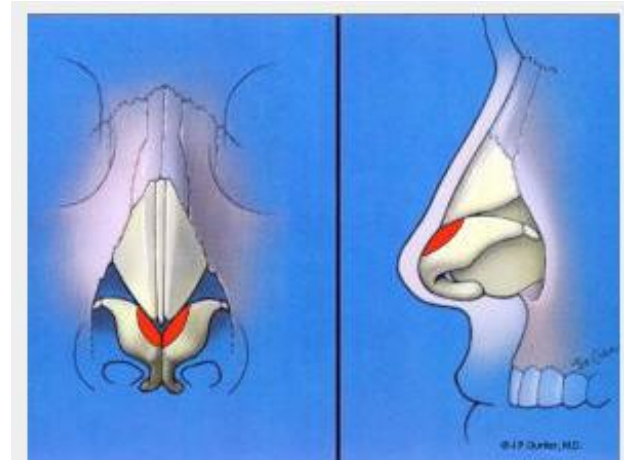
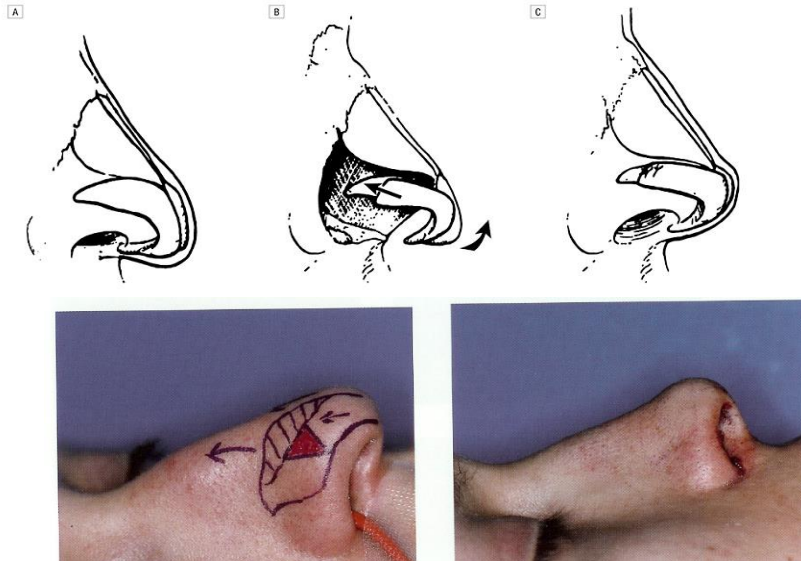


Figure 182.19 The ideal lateral crura are positioned so that they are pointing toward the lateral canthi (35 to 45 degrees off midline). Lateral crura that are oriented less than 30 degrees off midline will tend to create a fullness in the transition point between the tip and supratip and in some patients may present with a parentheses deformity.

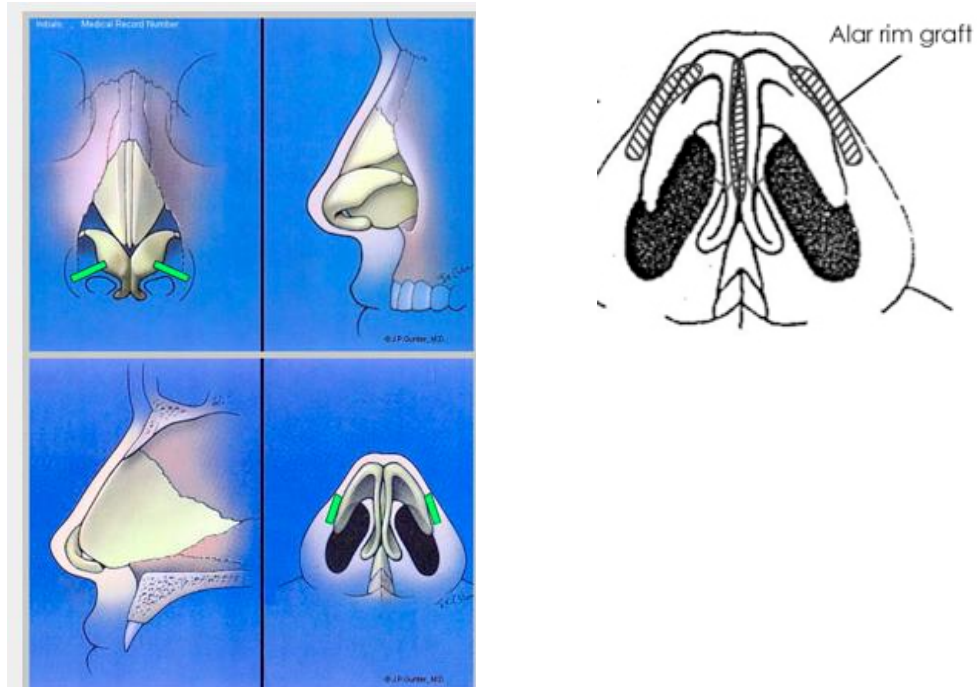


Lateral Crural Overlay:

- **Increases rotation and decreases projection.**



- **Alar rim and supraalar rim pinching due to weakening of the nose:**
 - Lack of support lateral to the tip in the alar margin.
 - Due to cephalic positioning of lateral crura or prior tip surgery.
 - Fix with **Alar rim grafts** (placed in the caudal aspect of marginal incisions).
 - Effective way to correct external valve collapse



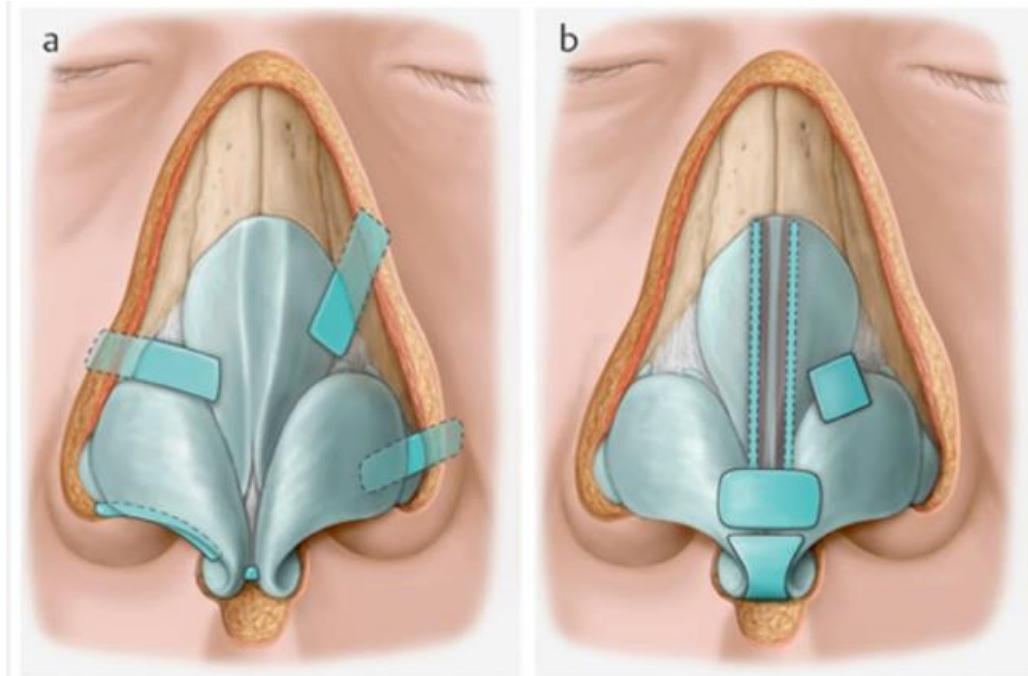


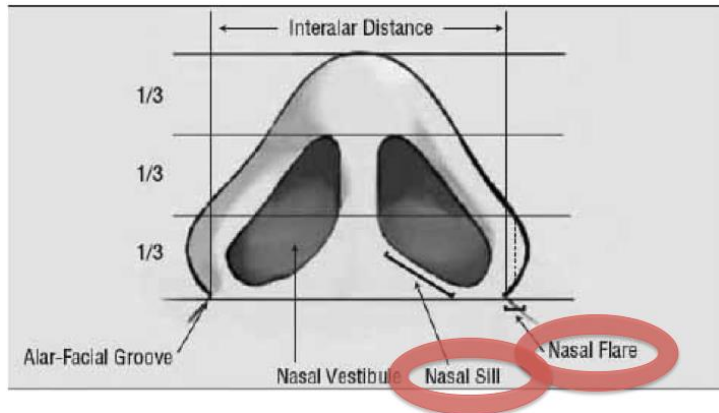
Figure 2: Overview over commonly placed structural grafts. a) from top to bottom: Vertical alar batten graft, horizontal alar batten graft, alar strut graft, alar rim graft, columellar strut graft. b) from top to bottom: Paired spreader grafts, cap graft, shield-type tip graft.

- **Tip cartilage camouflage is crucial:**
 - Perichondrium is excellent material for covering tip grafts

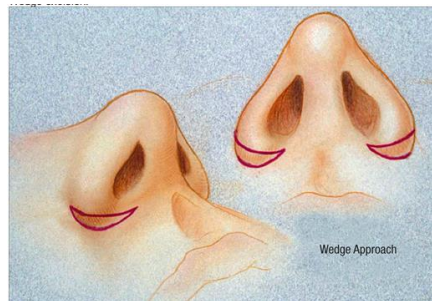
Increase Tip Projection	Decrease Tip Projection	Increase Tip Rotation	Decrease Tip Rotation
Shield graft	Complete transfixion (sacrifice major tip support)	Cephalic trim with interdomal suture	Extended spreader graft
Columellar strut	Appropriate caudal septum resection (if too long)	Columellar strut	Shield graft with dorsal augmentation and columellar strut (for very small nose)
Interdomal suture	Nasal spine reduction	Plumping graft	Caudal septal excision (wedge narrow at top)
Tongue in groove	Lipsett technique: divide medial crura at medial angle, overlap and resuture with 6-0 PDS	High septal transfixion	Shorten medial crura
Plumping graft	Vertical dome division	Caudal septal extension graft	Bilateral intercartilaginous incisions
Lateral crural steel	Tongue and groove	Lateral crural flap: divide lateral crura around the midpoint, overlap and resuture (vertical dome division)	
Caudal extension graft	Illusion of decrease tip Projection ✕Chin implant	Resect caudal edge of lower laterals	
Vertical dome division using lateral crural recruitment (Goldman tip)		Release LLC from caudal/dorsal septum and reposition	
Illusionary – Reduction of cartilaginous dorsum/enhancing supratip break			
Resection of Depressor Septi Nasi Muscle			

Basal width:

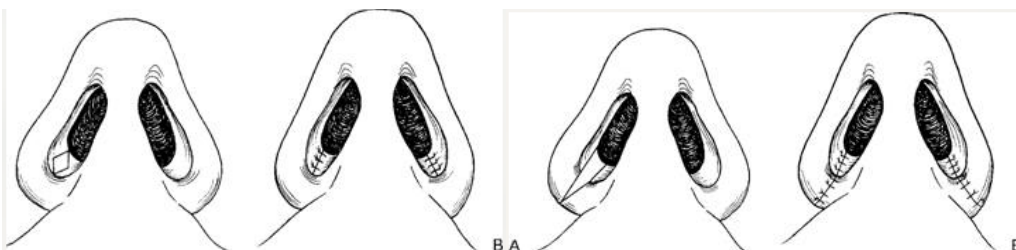
- Affected by:
 - Alar flare
 - Sill length



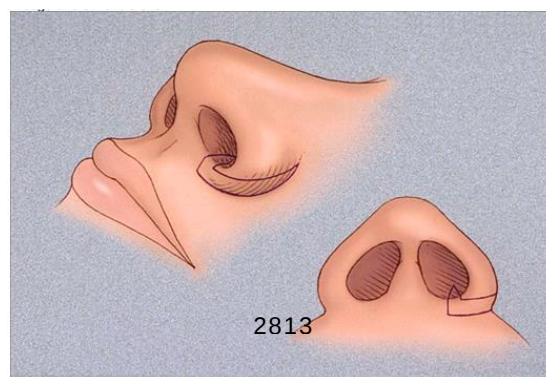
- Corrected by:
 1. **Alar wedge excision** (0.5-1 mm above alar groove).



2. Sill excision



3. Combined Alar and Sill excision



Closure and Splinting:

- Intranasal closed with 4-0 absorbable.
- External rhinotomy closed with 6-0 nylon; removed in 5 days.
- Occasionally can use a couple of buried stitches.
- Can use septal quilting sutures to hold graft and flaps in place.
- Silastic intranasal splints useful; avoid packing if possible; if splints in for long, see pt regularly to help minimize crusting.
- External splint: clean with tincture of benzoin; cover nose with steri-strips (taping can help support tip); plaster, foam lined aluminum or thermoplastic fiberglass can all be used.



Postoperative Care:

- 30 degrees head elevation.
- Cold compresses over the eyelids and paranasal region (24 hours as tolerated).
- Analgesic medication.
- +/- Prophylactic antibiotic.
- Avoid lifting and other strenuous activity for a period of at least 2 to 3 weeks.

CORRECTION IS **UNPREDICTABLE** BECAUSE IT DEPENDS ON BOTH SIDES HEALING IN **IDENTICAL WAY**, WITH SAME DEGREE OF

- SWELLING
- SCARRING
- CONTRACTURE

Otoplasty

Anatomy

- Normal ear-head angle is 15-30°
- **Muscles:**
- Intrinsic :
 - 1- Major and minor helices
 - 2- Tragus
 - 3- Antitragus
 - 4- Transverse
 - 5- Oblique
- Extrinsic
 - 1- Anterior auricularis
 - 2- Superior auricularis
 - 3- Posterior auricularis

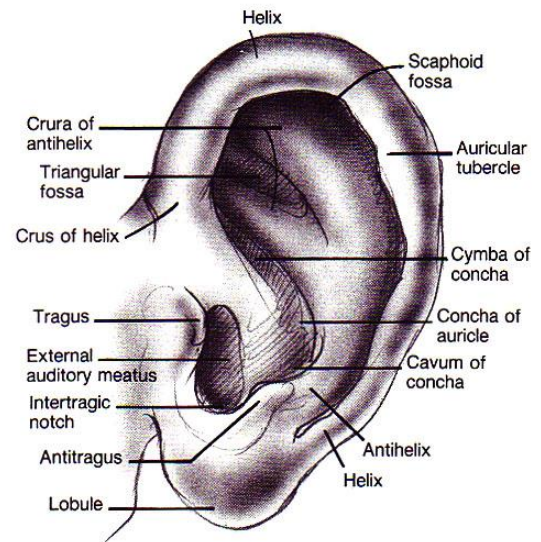


Figure 180.1 Anatomy of the auricle.

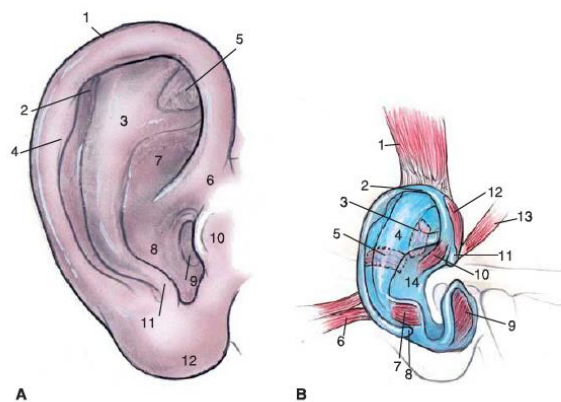


Figure 190.1 A: Normal external auricular anatomy. 1, Helix; 2, scaphoid fossa; 3, antihelix; 4, auricular (Darwin) tubercle; 5, triangular fossa; 6, crus helix; 7, cymba concha; 8, cavum concha; 9, external auditory meatus; 10, tragus; 11, antitragus; 12, lobule. B: Normal external and internal auricular musculature. 1, Auricularis superior; 2, helix; 3, obliquus auricularis; 4, antihelix; 5, transversus auricularis; 6, auricularis posterior; 7, antitragicus; 8, cauda helix; 9, tragus; 10, helix minor; 11, spina helix; 12, helix major; 13, auricularis anterior; 14, concha.

- Vertical axis inclined 20°
- Projection 20° (or 1.5-2 cm from skull).
- Height nearly equal to Distance from lateral orbital rim to root of helix (6 cm).
- Width ~ 55% of height (ratio 06:1).
- Superior aspect usually at level with eyebrow.

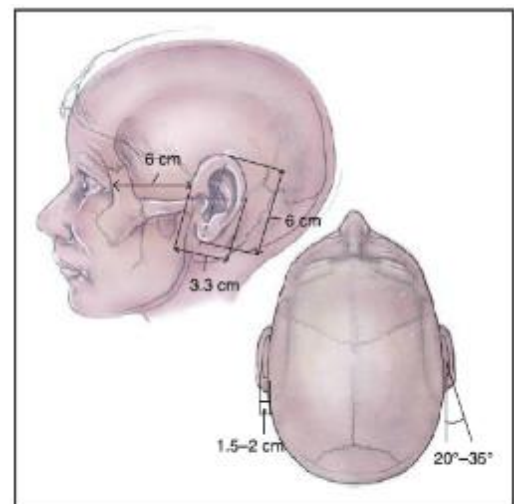


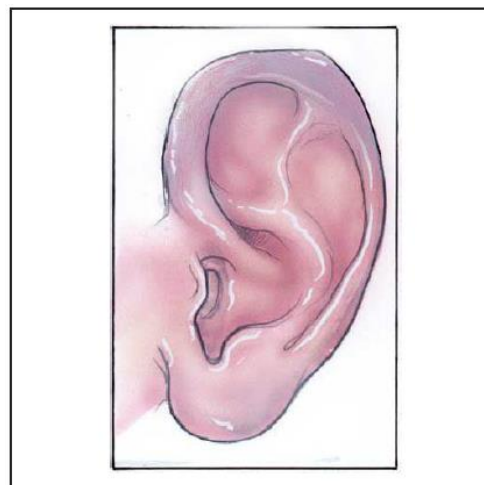
Figure 190.3 Normal ear dimensions.

AURICULAR ANOMALIES:**TABLE
190.1****AURICULAR DYSPLASIA
CLASSIFICATION SYSTEM
BY WEERDA**

	Anatomic Definition	Surgical Definition
First-degree dysplasia	Most structures of the normal auricle are recognizable Minor deformities	Reconstruction does not require the use of additional skin or cartilage
Second-degree dysplasia	Some structures of the normal auricle are recognizable Moderate deformities	Partial reconstruction requires the use of additional skin and cartilage
Third-degree dysplasia	None of the structures of the normal ear are recognizable Severe deformities	Total reconstruction requires the use of additional skin and large amounts of cartilage

Deformations**Stahl Ear:**

- Known as Satyr ear; Spack ear; and Vulcan ear
- Characterized by an abnormal transverse crus from the antihelix to the posterior superior helical rim and, an absent superior crus.
- Caused by external forces in utero or perhaps an abnormal course of the transversus auriculæ, one of the intrinsic muscles of the ear.
- Can be treated with molding techniques, suturing techniques, or excision of abnormal cartilage.

**TABLE 180.1****DIAGNOSIS****OTOPLASTY****Deformity criteria**

Angle between ear and head, 25 to 30 degrees or greater
 Dimensions different from normal auricle
 Vertical axis—about 20 degrees posteriorly
 Vertical height—about 6 cm
 Width—about 55% of length
 Helical rim—1- to 2-cm protrusion from skull
 Angle protrusion—21 to 30 degrees
 Superior aspect—usually level with brow
 Other anatomy that makes ear offensive to family or patient
 One or more significant auricular landmarks of deformity
 Very poor antihelical fold
 Overdeveloped concha
 Abnormally formed scapha
 Obvious lack of superior and inferior crus surrounding fossa triangularis

Comparative criteria

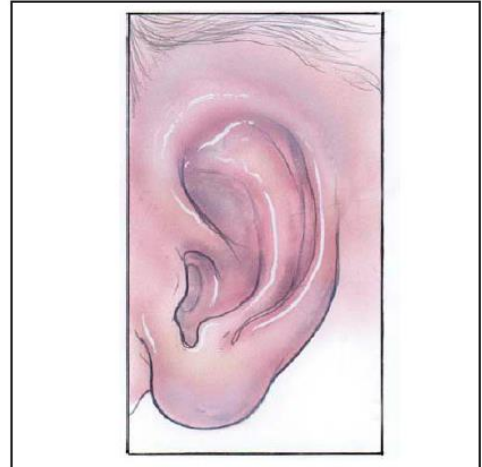
Thickness of scaphal cartilage
 Stiffness of scaphal cartilage
 Symmetry of two ears
 Darwinian tubercles
 Other preauricular tags

Documentary photographs

Frontal view
 Oblique view
 Rear view

Cryptotia:

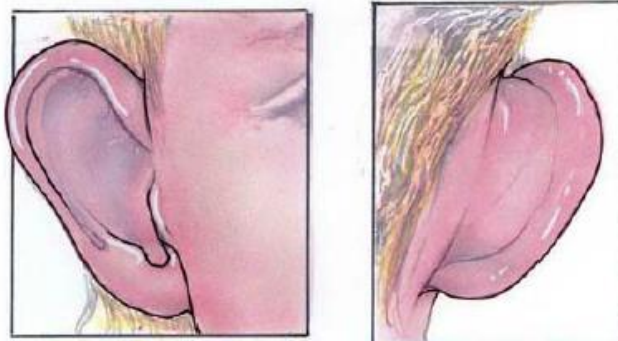
- Means hidden or pocket ear, is a condition in which the superior helical cartilage is buried under the skin.
- Caused by an abnormal attachment of the superior auricular muscle to the scapha rather than to the triangular fossa as well as a shortened transversus auriculae, effectively pulling the superior helix.
- Treated with molding or by releasing the superior helix from the scalp with skin grafting.



Protruding Ears:

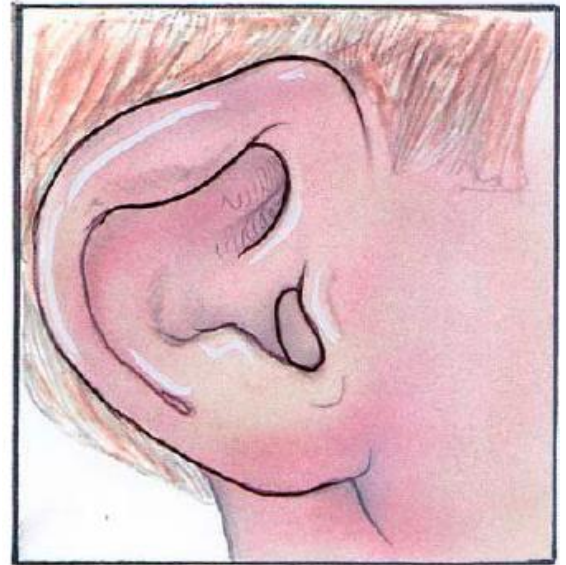
- Characterized by poor antihelical fold and overdeveloped concha.
- Some define ear prominence as an auriculocephalic angle greater than 40 degrees or a helix to scalp distance greater than 2.5 cm.
- Preoperative evaluation:
 - Three criteria:
 - 1- Cartilage thickness
 - 2- Cartilage stiffness
 - 3- Symmetry from one ear to the other
- **Best age of intervention is immediately before school (5-6 yrs).**
- Involve family and assess for other related issues (SNHL, syndromes, other medical conditions).
- In assessing ear protrusion, the distance from the mastoid to the helix is recorded at the superior aspect of the helix (**S**), Middle at the level of the external auditory canal(**M**), and lower at the lobule (**L**).
- The ideal values for these in the adult should be:
 - 1- (S) **16 to 18** mm
 - 2- (M) **14 to 16** mm
 - 3- (L) **16 to 18** mm

There is no absolute definition of prominent ear; an ear is prominent when a patient says it is.



Constricted Ear:

- Constricted ears are characterized by partial absence of cartilage at the upper third of the helical rim or sometimes the concha, hypoplastic helix or scapha.
- Variably labeled as cup, lop, and cockleshell ear.
- Can be found in a number of inherited syndromes but is usually sporadic when isolated.
- Classified as either:
 - 1- First-degree dysplasia (Mild constriction)
 - 2- Second-degree dysplasia (Moderate constriction and severe constriction)
- Corrected with molding techniques as a neonate or later with an otoplasty technique



Non-surgical Treatment:

- If a deformational auricular anomaly is identified in a neonate or, according to some, select older individuals, **molding techniques** should be considered.
- These techniques usually involve placing a bendable splint along the helical rim, antihelix, and conchal bowl and taping it in place to hold the ear in an appropriate position **for 2 to 12 weeks**.
- Molding is able to correct up to 90% of deformational auricular anomalies if started within the first week of life.
- Most authors only advocate molding for neonates.



BEFORE

DURING TREATMENT

AFTER

- **In general, after 3 weeks of age, molding is less successful**
- **Laser-assisted cartilage reshaping.** a new technique in which the ear is treated with an erbium/glass laser to reshape the cartilage without any anesthesia and molded into the desired shape with a silicone splint
- The splint is then worn at all times for 2 weeks and then at night only for 4 weeks.

Surgical Techniques:

• **CARTILAGE-SPARING TECHNIQUES:**

1. Incisionless otoplasty:

- Involves percutaneously scoring the anterior surface of the cartilage at the planned antihelical fold.
- Creating a small opening at the postauricular sulcus and removing soft tissue for a conchal setback if needed.
- Percutaneously placing horizontal mattress retention sutures from the posterior side of the pinna to create the antihelical fold and pull the concha posteriorly and burying the knots by pulling skin over them with a single-prong skin hook.
- The lobule is brought posteriorly by percutaneously dissecting the cauda helix from the pinna and using the same percutaneous suture technique to secure it to the posterior conchal bowl.

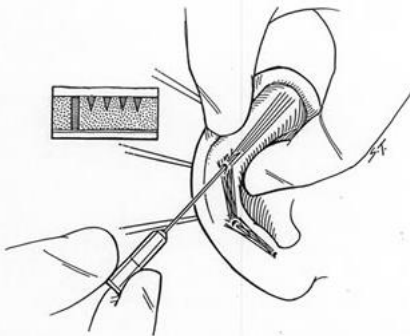


FIGURE 1 Cartilage scoring: Note several cuts made through each percutaneous needle insertion.

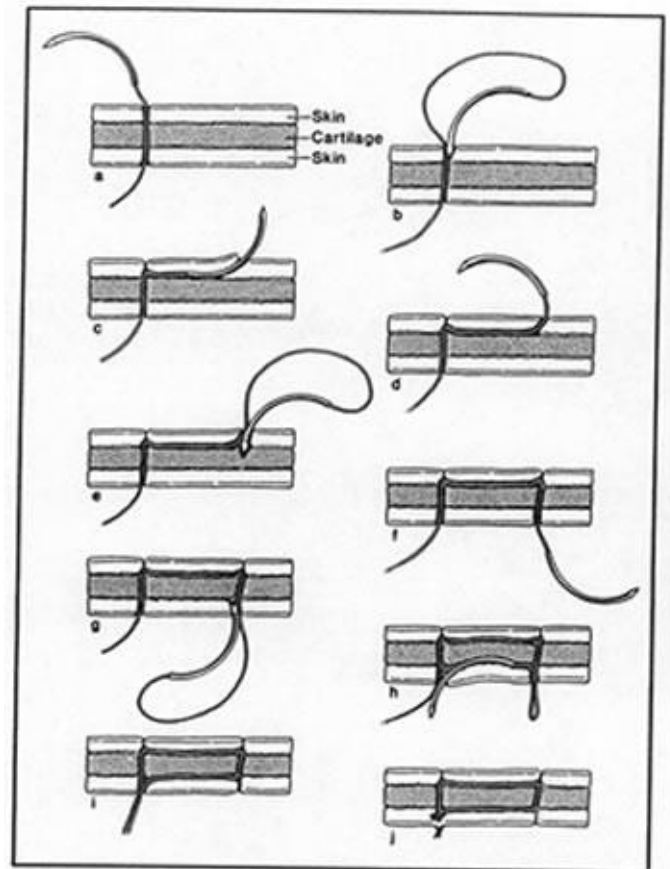
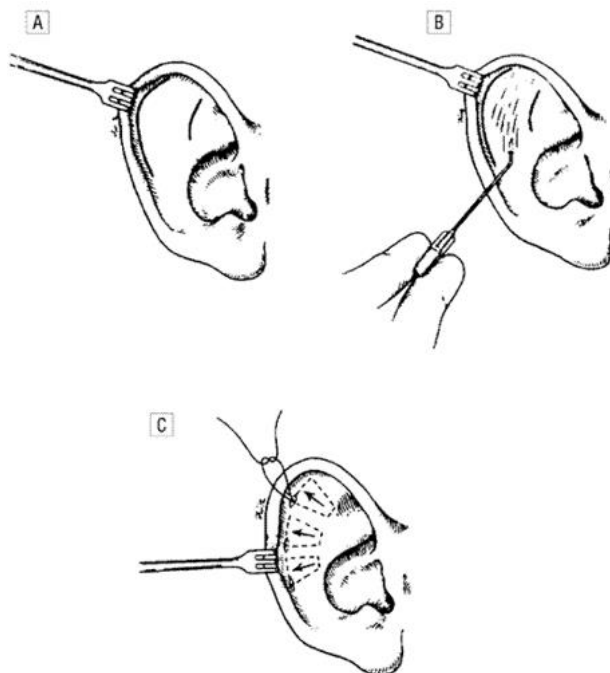
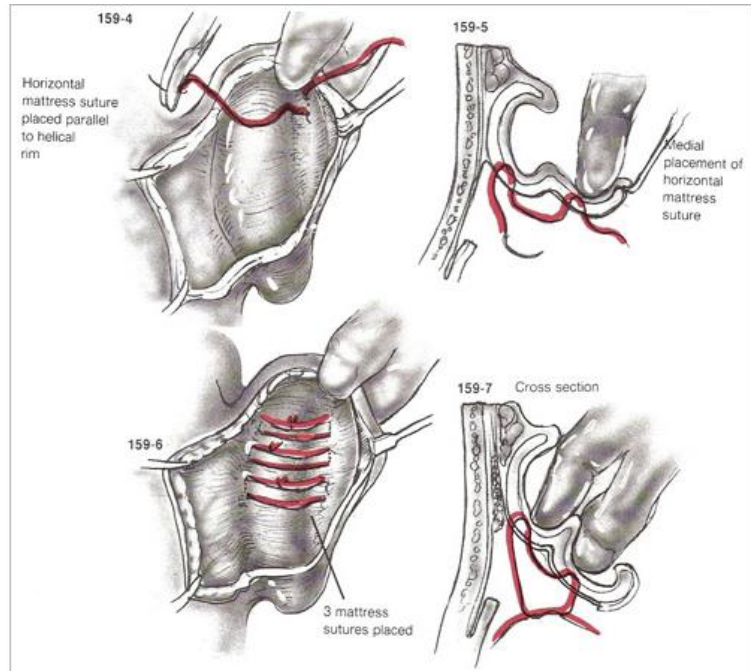
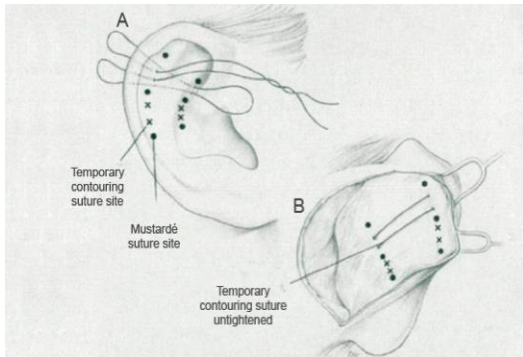


Fig. 1. The stitch: Incisionless technique for placing a retention suture. This is an example of how a complete buried suture loop is created.

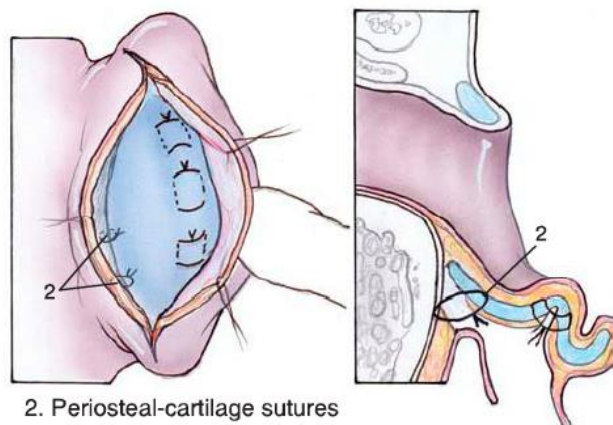
2. Mustarde:

- Less invasive technique of recreating the antihelical fold.
- Using three permanent horizontal mattress sutures to secure the auricular cartilage to itself without making any cartilage incisions.
- Advantages:
 - Normal AntiHelical fold, long-term hold and good results for even less-experienced hands.
- Disadvantages:
 - Suture erosion or release if not correctly placed, and the need for more work on conchal bowl



3. Furnas (conchal setback):

- Less invasive technique of conchal bowl reduction.
- Simply securing the conchal cartilage to the mastoid periosteum posteriorly.
- May help to remove posterior soft tissue and post auricular muscle.



2. Periosteal-cartilage sutures

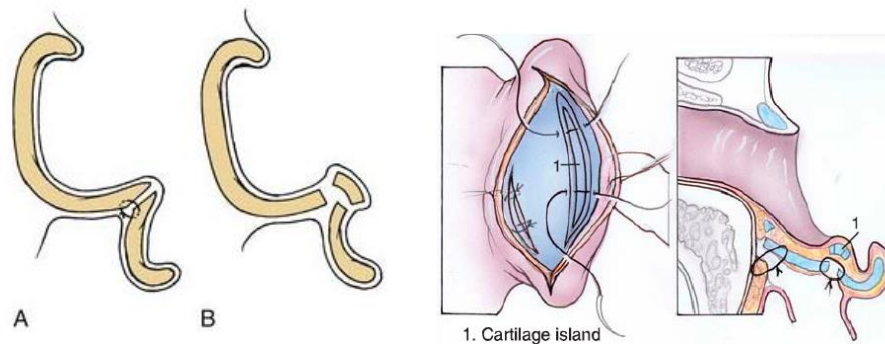
• **CARTILAGE-CUTTING TECHNIQUES:**

1. Converse technique:

- More complicated; requires experience; island of cartilage created that sit ant to rest of concha
- Advantages:
 - More permanent fix; gentle antihelix.
- Disadvantage:
 - Harder to do.

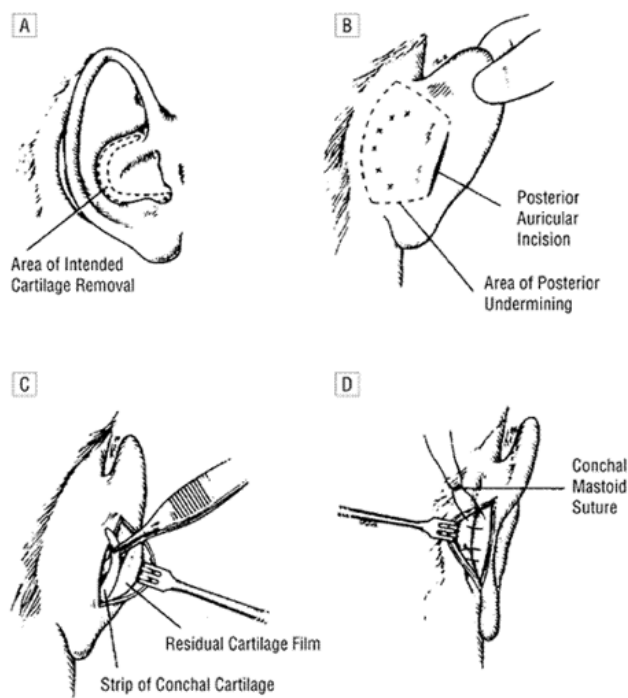
2. Pitanguy technique:

- Island flap again, only smaller.



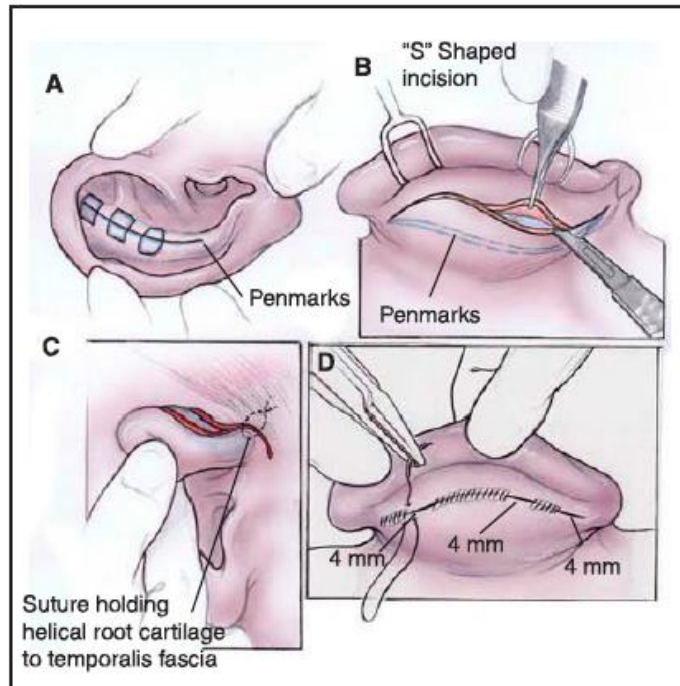
3. Farrar technique:

- Cartilage incision along conchal rim; longitudinal wedges taken from future sup crus and antihelix; also creates island, but gives more gentle bend than converse

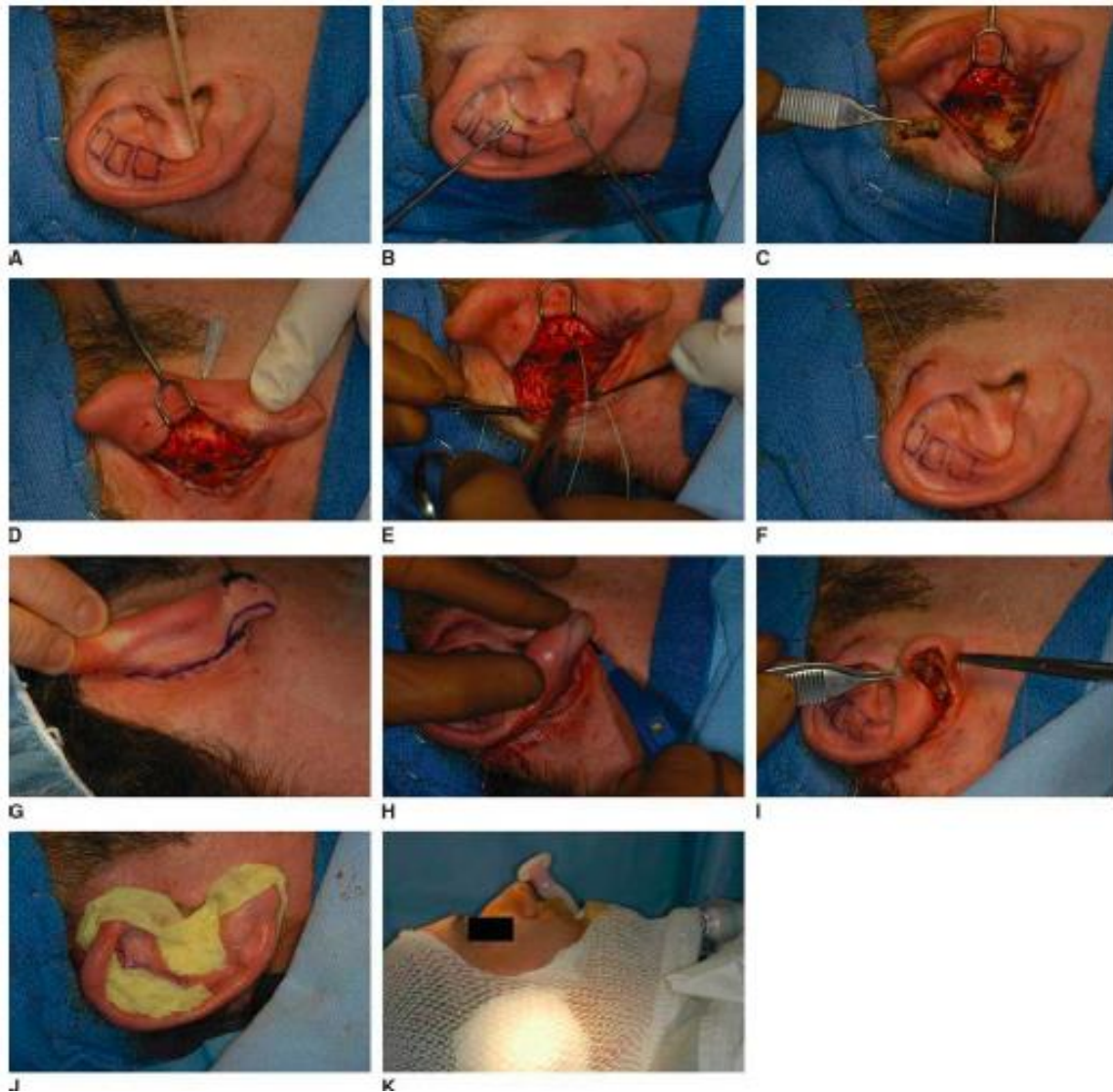


4. Erol:

- Described an anterior approach to otoplasty.
- Placing their incision along the conchal bowl rim, excising a portion of conchal cartilage, scoring the anterior surface of the cartilage to create an antihelical fold, and securing these changes with horizontal mattress sutures.



Might help to take strap of skin from postauricular incision



- Skin closed by simple mattress or continuous looked suture

Dressing:

- Petrolatum gauze (Xeroform) is cut into five 5 mm strips, two of which are packed in the postauricular sulcus, and three of which are packed along the anterior surface of the ear.
- 4 x 4-inch gauze sponges are placed over each ear and a size 8 elastic net bandage (BandNet) is placed over the face.
- Kept for 1 week.
- Then Tennis band at night for 1 month.

Otoplasty for the Constricted Ear:

- The goals are:
 - Provide adequate, symmetric ear height, projection, and shape.

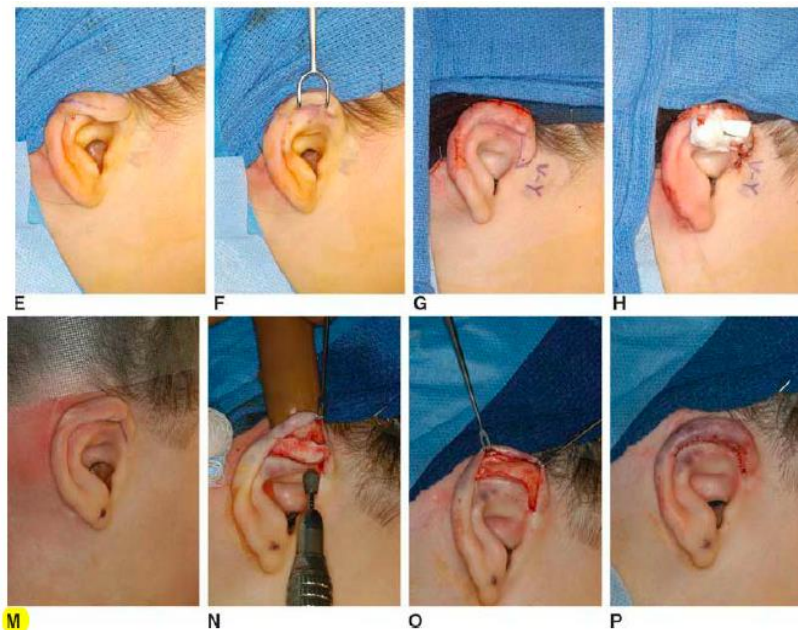
Mild Constricted Ear

- Can be treated with **molding** techniques.
- transcutaneous placement of through-and-through 4-0 silk sutures to create the antihelix and reduce helical prominence can be useful in determining if they alone can hold the ear in position for several days to a week preoperatively.
- Mustarde.
- Cartilage cutting.



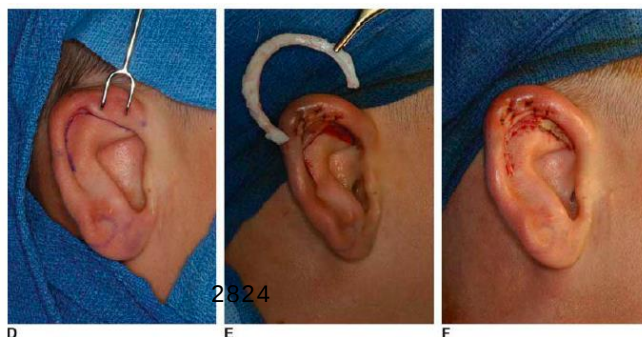
Moderately constricted Ear:

Often they will require a **V-Y** advancement of the helical root to release the helical rim into a more natural position and may require **cartilage grafting**
Or use **dermabrader**



Severely constricted Ear:

Always requires cartilage grafting and often requires an adjacent tissue transfer or skin graft.



- **Complications and emergencies of Otoplasty:**

1. Chondritis: most feared; rare
2. Inadequate correction: most common
3. Hematoma: easiest to detect and fix
4. **Telephone ear deformity**: too much antihelical flexion at midpoint and inadequate flexion at sup & inf poles; reverse happens too.
5. Infection: must be recognized and treated ASAP
6. Suture granuloma
7. Keloid
8. Skin necrosis

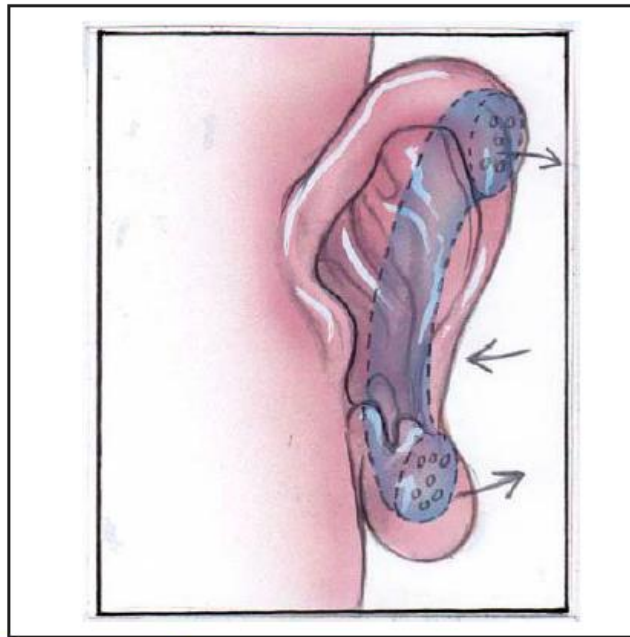


Figure 190.11 Telephone ear deformity is a postoperative result of adequate correction of ear projection in the middle one-third, but inadequate correction of ear projection in the superior and inferior one-third.

TABLE 180.2
OTOPLASTY



TREATMENT

Mustarde technique

Advantages

Very normal-appearing antihelical fold can be created, which sutures can hold indefinitely.

Sutures also can be used to create good superior or inferior crus.

Inexperienced surgeons can use this technique successfully.

Disadvantages

Incorrectly placed sutures will cause problems.

Technique is not applicable for work in conchal bowl area.

Converse technique

Advantages

Island of cartilage with normal-appearing fold can be created.

More permanent retraction of auricle is facilitated.

Gently curved and potentially more permanent correction of antihelix is permitted.

Disadvantage

Experienced surgeon must perform.

Farrior technique

Advantage

Bend of antihelix is gentle.

Disadvantage

Experienced surgeon must perform.

Pitanguy technique

Advantage

Patient can have small amount of antihelical cartilage.

Disadvantage

Experienced surgeon must perform.

Furna conchal mastoid suture technique

Advantage

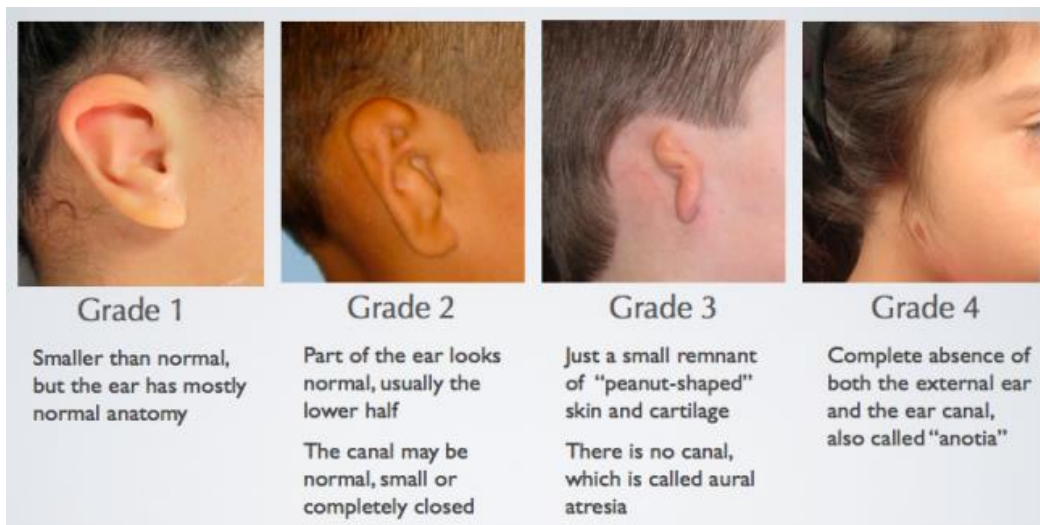
Permanent retraction of auricle is facilitated.

Disadvantage

Partial closure of os of external canal occurs.

- **Microtia:**

- Major auricular malformation.
- Often associated with EAC atresia.
- Degree of auricular malformation correlates with degree of middle ear deformity.
- Associated inner ear abnormalities are rare.
 - Vestibular dysplasia is the most common.
- Usually unilateral with right side is the most common.
- 25% of cases are bilateral.
- M:F ratio of 2.5:1.
- May have syndromic association (eg, hemifacial microsomia).
- Causes:
 - Genetic
 - Teratogens (vitamin A/isotretinoin, thalidomide)
 - Vascular insult
- Classification:
 - **Grade I:**
 - Small auricle with all subunits are present.
 - **Grade II:**
 - Small auricle with some subunits are severely underdeveloped or absent.
 - **Grade III:**
 - Small remnant of skin and cartilage (Peanut ear) associated with aural atresia.
 - **Grade IV (Anotia):**
 - Complete absence of auricle and lobule associated with aural atresia.
 - Usually forms part of 1st Arch Syndrome.

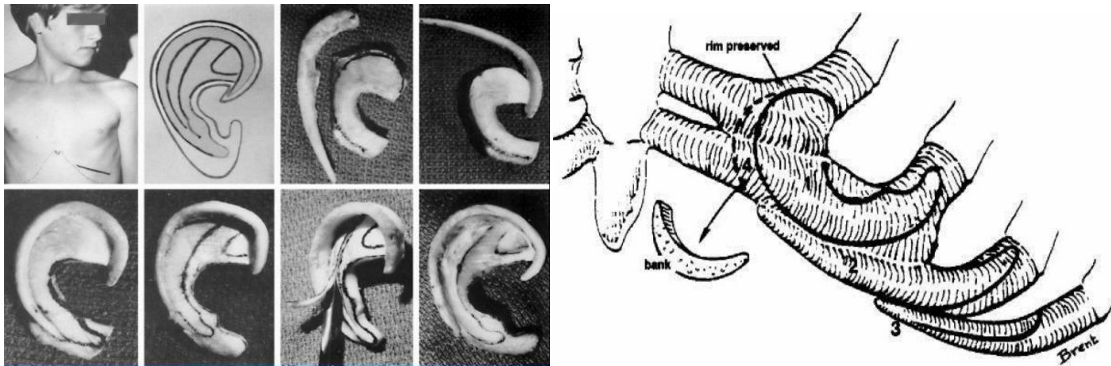


- Evaluation of patient with Microtia or Aural Atresia:
 - **H&P:**
 - Complete H&N examination looking for any other dysmorphic features.
 - Assess facial nerve function:
 - Most common anomaly of facial function is a congenital absence of depressor anguli oris muscle.
 - **Hearing Assessment:**
 - ABR:
 - Usually 40–60 dB CHL
 - 10–15% have SNHL
 - **CT Temporal bone:**
 - Evaluate presence of:
 - Middle and Inner ear anomalies.
 - Congenital cholestatoma.
 - No need to perform CT before age 4 years.
 - Should be performed near the time of operation and must be repeated if done early after birth before ultimate aural atresia repair.
 - Congenital cholesteatoma usually slow growing and not happen before this age.
- Treatment Options for Microtia:
 - 1. **Surgical Repair:**
 - Typically at age of 6 years old.
 - Timing also influenced by coexisting atresia.
 - Types of Grafts:
 - **Costal Cartilage Autograft:**
 - Rib is popular cartilage source due to suitable integrity, sufficient quantity, and minimal morbidity with harvest.
 - **Alloplastic Implant:**
 - Medpor (porous polyethylene) implant.
 - Technically easier, does not tolerate trauma well, risk of extrusion.
 - Techniques of Surgical Repair:
 - Brent Cartilage Autograft Technique.
 - Nagata Cartilage Autograft Technique.
 - 2. **Cosmetic Auricular Prosthesis:**
 - Prosthesis is anchored by magnets (previously clips) to titanium posts, which are connected to osseointegrated implants placed in the mastoid bone, often very realistic.

3. BAHA.

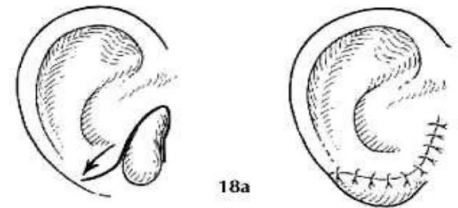
○ Brent Cartilage Autograft Technique:

- 4 stages separated by 3 months.
- Some stages may be combined.
- **1st Stage (Auricular Framework):**
 - Fabrication of the auricular framework from contralateral costal cartilage.
 - Placed in a postauricular subcutaneous pocket (thin overlying skin as much as possible).



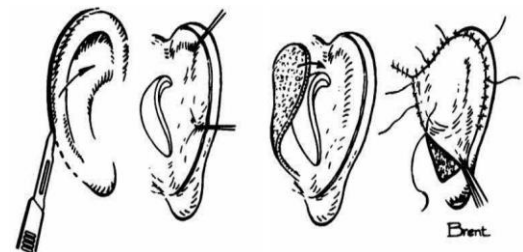
▪ **2nd Stage (Lobule Transposition):**

- Lobule is rotated and often filleted to receive the end of the cartilage framework



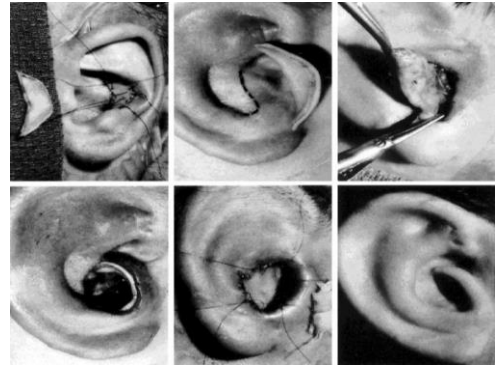
▪ **3rd Stage (Framework Elevation):**

- Framework is elevated to achieve projection of the helical rim.
- Retro-auricular scalp advancement flap and STSG used to close the postauricular defect.



▪ **4th Stage (Tragus Reconstruction):**

- Tragus construction, conchal excavation, symmetry adjustments.

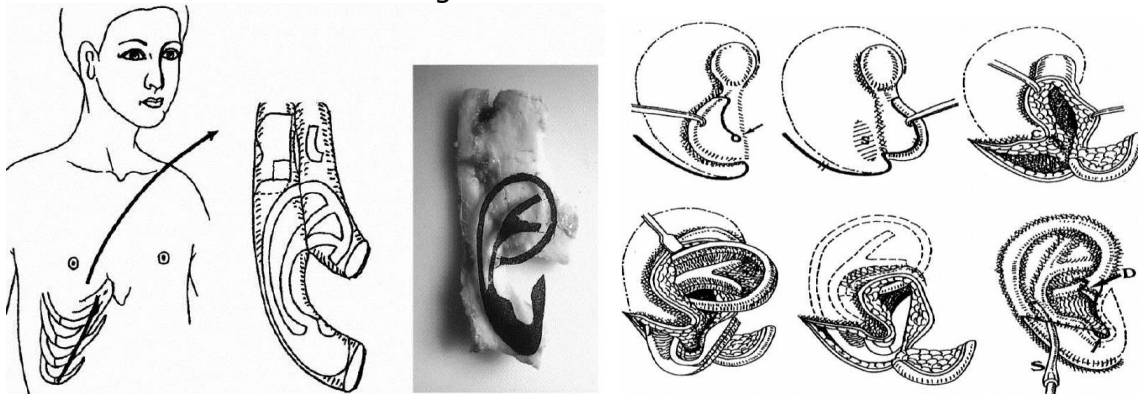


○ Nagata Cartilage Autograft Technique:

- 2 stages separated by 6 months.

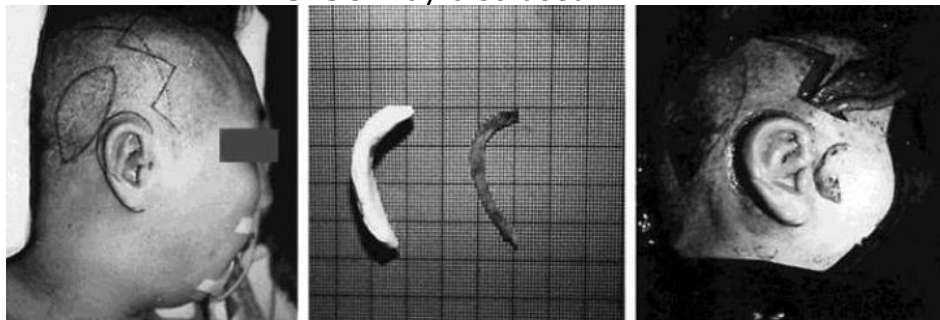
▪ **1st Stage:**

- Fabrication of the auricular framework from ipsilateral costal cartilage.
- Lobule Transposition.
- Tragus Reconstruction.



▪ **2nd Stage:**

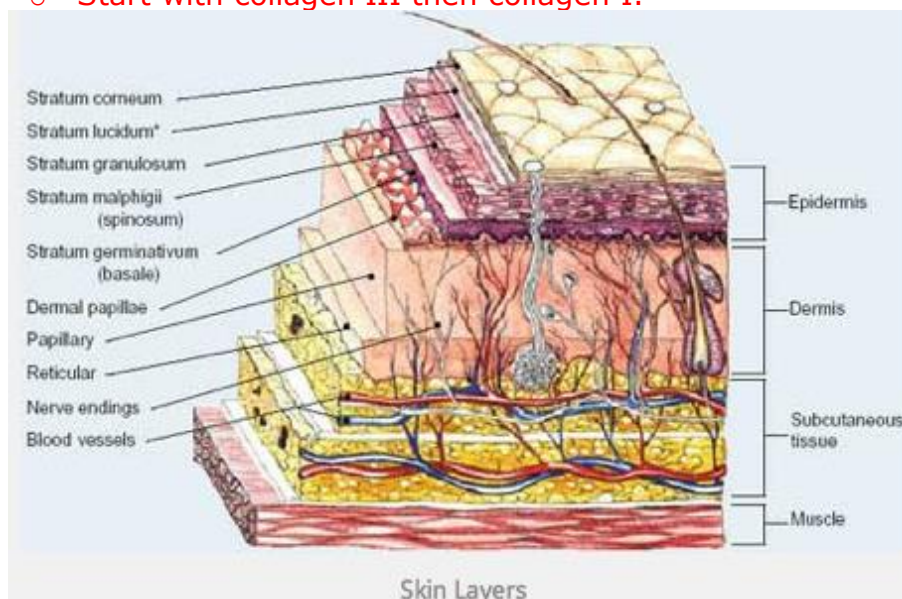
- Framework is elevated using a crescent-shaped piece of cartilage to achieve projection of the helical rim.
- Temporoparietofascial flap is elevated and tunneled subcutaneously to cover the posterior surface of the cartilage graft, reconstructed auricle, and the mastoid surface.
- STSG may also used.



Scar Revision

Skin Anatomy:

- Skin is composed of:
 1. **Epidermis** (Generally 5 layers, except at palms and soles):
 1. Stratum Corneum: most superficial
 2. Stratum Lucidum: absent in thin skin
 3. Stratum Granulosum
 4. Stratum Spinosum
 5. Stratum Basale: deepest
 2. **Dermis:**
 1. Papillary dermis:
 - Thin, loose collagen, blood vessels, fibrocytes.
 2. Reticular dermis:
 - Thick, compact collagen, sebaceous glands, and fibrocytes.
- Once a wound occurs, there are different phases of wound healing that occur:
 1. **Vascular Phase:**
 - Occurs immediately (inflammatory).
 - Early vasoconstriction (5 – 10 minutes).
 - Caused by platelet aggregation and fibrin.
 - Vasodilation (can occur over hours to days).
 2. **Proliferative Phase:**
 - Re-epithelialization.
 - Granulation tissue/fibroplasia.
 - Wound contraction.
 3. **Remodeling Phase:**
 - Start with collagen III then collagen I.



- Superficial injuries, most facial wounds will heal with little to no scar formation.
- Once the reticular dermis has been violated, some amount of residual scarring is destined to occur.
- Factors affecting the final appearance of the scar:

- **Beyond the surgeon's control:**

1. Mechanism of injury
2. Position of the wound
3. Health status of the patient
4. Patient skin type
5. The tendency to form robust scars

– Wound factors

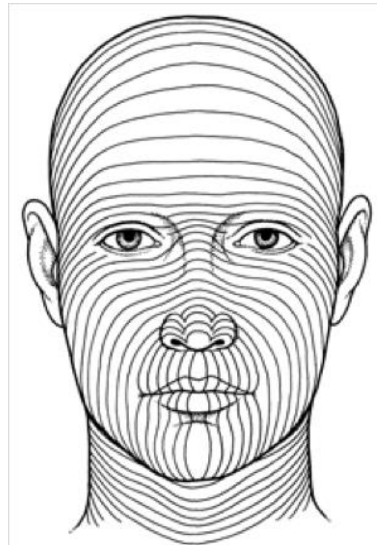
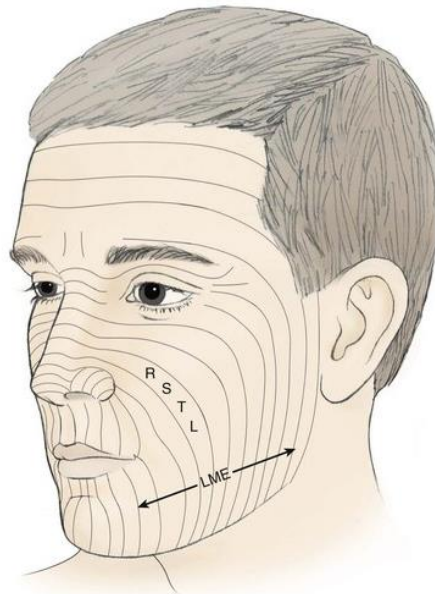
- infection
- tissue trauma
- tissue ischemia
- wound closure techniques
- wound dessication

- **Under the surgeon's control:**

1. Proper realignment of wound edges.
2. Conservative debridement of injured tissues.
3. Meticulous handling of tissues during primary repair.
4. Aesthetically favorable alignment of scars whenever possible.

- **Ideal scar:**

- Good color match and be flat and level with the surrounding skin; it should be narrow with No long unbroken segments and parallel to the relaxed skin tension lines (RSTLs).
- Lines that follow the furrows formed when skin is relaxed



Direction of force is along each line. Cuts perpendicular to these lines are thus under greatest tension and most likely to widen.

FIGURE 6-3 Relaxed skin tension lines (RSTLs) of face. Lines of maximal extensibility (LME) are perpendicular to RSTLs.

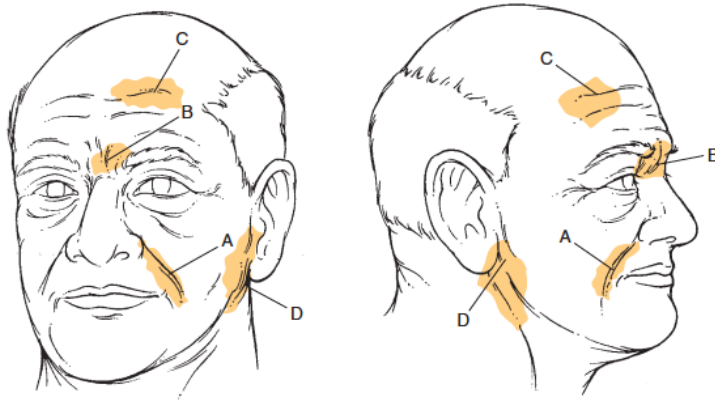


FIGURE 21-1. Relaxed skin tension lines and creases can aid in scar camouflage. A, Nasolabial. B, Glabellar furrows. C, Horizontal forehead rhytids. D, Subunit junctions (ear and cheek).

Scars with aesthetically unfavorable characteristics:

- Wide.
- Misaligned with RSTLs or aesthetic subunits.
- Those that are thickened, hypertrophied, or keloid.

Scar Revision:

- Begins with a precise analysis of both the scar and the patient's expectations.
- True realistic goal of surgery, which is to modify the scar to a point of maximized camouflage.
- Many narrow, well-positioned scars along aesthetic subunit borders or in parallel with RSTLs will continue to mature, improve, and become less noticeable over a period of 12 to 36 months.
- **Traditionally wait for 6-12 months before revision.**

Indications for scar revision:

1. Widened (>2-3 mm), or longer than 20 mm.
 2. Perpendicular to RSTLs.
 3. Webbed.
 4. Pincushioned.
 5. Long and linear and misaligned with RSTLs.
 6. Hypertrophied.
 7. Interrupting an aesthetic unit of the face.
 8. Adjacent to, but not lying within, a favorable site.
 9. Causing distortion of facial features or anatomic function.
- **Early revision with realignment of the scar may allow it to mature more rapidly, Many of these scars should be considered for revision after the first 60 to 90 days of scar maturation has occurred.**

Box 21-1. TECHNIQUES AND INTERVENTIONS FOR SCAR REVISION

Routine Scars

Primary closure
Serial excision and expansion
Irregularization, elongation, and reorientation
Topical therapies
Injectable therapies
Dermabrasion

Hypertrophic Scars and Keloids

Topical therapies
Injectable therapies
Lasers
Radiotherapy and cryotherapy

Hairstyling and Makeup

SCAR EXCISION:

- Cardinal principle of surgery is that the best treatment of any complication is primary prevention (proper alignment, follow RSTL, avoid tension).
- When presented with wounds that were not closed properly, reexcision with meticulous closure may be all that is needed.
- **Fusiform shape with 30-degree angled ends** positioned within RSTLs when possible.

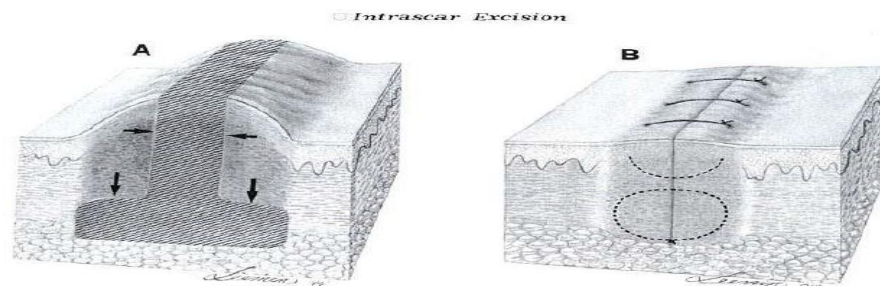


FIGURE 21-3. Examples of proper placement of fusiform incisions with 30-degree angled ends aligned with relaxed skin tension lines and aesthetic unit boundaries.



- A slight **vertical bevel** outward from the original scar will prepare the wound edges for proper everted closure, A scalpel pass just along the edge of the existing scar, This initial skin incision should be performed with a favorable bevel, about 10 degrees off of a true perpendicular cut.

- Epithelium of the scar is then excised with a sharp scissor or scalpel, the **dermis and subcutaneous tissue are left in the bed** of the wound to provide a more stable "platform" on which the revised scar can lie (bedrock principle).
- Routine **undermining of 1 to 2 cm** around the periphery of the wound allows for re-approximation of the skin edges with minimal tension.
- Use of **buried subcutaneous sutures** further decreases wound-edge tension.
- Final **eversion of the wound edges** is achieved with properly placed monofilament interrupted sutures, Vertical mattress sutures can also prove helpful when wound-edge eversion needs to be maximized.



EXPANSION WITH EXCISION

Serial Excision:

- Relies on the skin's biologic creep (ability to stretch over time).
- Limited by the given amount of stretch attainable during each excision.
- Older patients and those with increased skin laxity will require fewer excisions than younger patients with increased skin tone.
- Wide scars, birthmarks, and skin grafts with poor match to surrounding tissues can all be candidates for serial excisions.
- Sometimes scar can be moved to hair line

Shave excision:

- Best for small raised scars



Tissue Expansion:

- Same principle (ability to stretch over time).
- Larger scars.
- Expanders are available in a variety of shapes and sizes.
- Rectangular expanders provide the greatest expansion at 38%.
- Crescent-shaped expanders provide 32%.
- Round expanders provide only 25%.

- As a general rule, the base of an expander should be approximately 2.5 to 3 times as large as the area to be reconstructed.

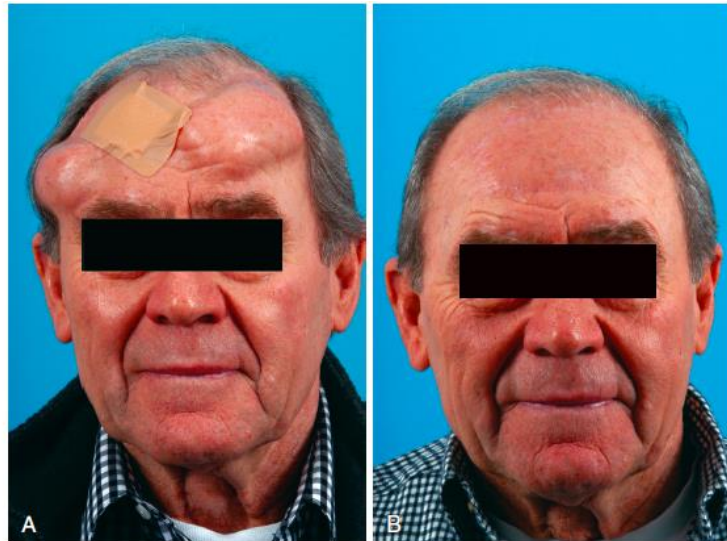
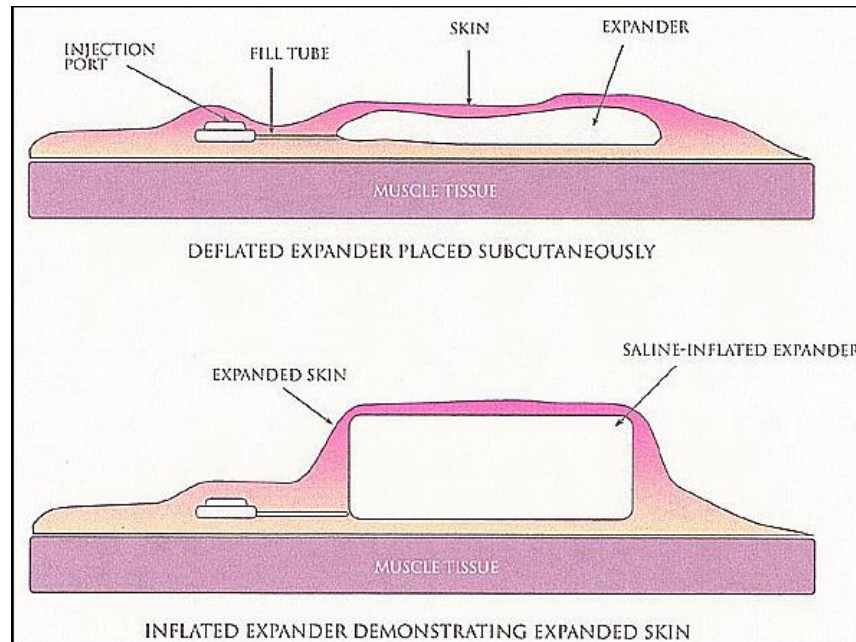


FIGURE 21-5. **A**, Patient midway through the process of bilateral forehead tissue expansion to close a Mohs defect in the forehead. Expanders were left in place for approximately 3 months. **B**, Eight months later.

- Expansion can proceed until the skin blanches or the patient complains of discomfort.
- Intervals between injections can be from 4 to 14 days, ideally two to three times per week.



IRREGULARIZATION, ELONGATION, AND REORIENTATION:

Z-Plasty:

- For scar **irregularization** with the additional benefits of scar **reorientation** and adjustable **elongation**.
- A classic Z-plasty involves the transposition of equilateral 60-degree triangles.

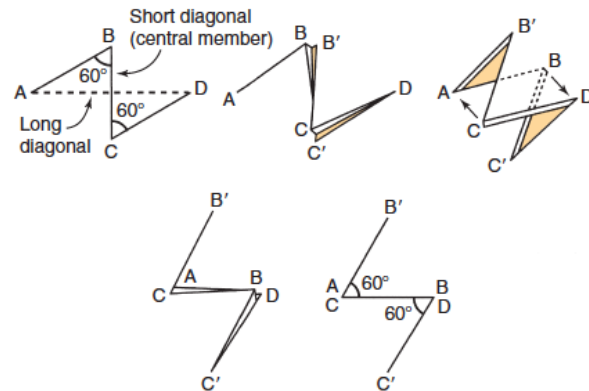


FIGURE 21-6. Classic equilateral triangle 60-degree Z-plasty.

- Original direction of the scar is rotated, and the scar is lengthened
- 75% when 60 degree Z
- 50% when 40 degree Z
- 25% when 30 degree Z
- Consecutive Z-plasties allow for the redistribution of forces more evenly along the entire length of the scar and help camouflage it into the surrounding RSTLs.

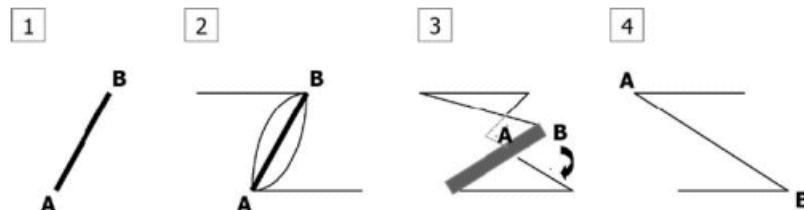


Figure 1: Z-Plasty

1=Scar oriented A-B;

2=Scar excised and creation of flaps with two limbs parallel to RSTLs;

3=Movement of the flaps;

4=Resultant scar with reorientation of A-B



FIGURE 21-7. A, Preoperative appearance of scar with webbing near medial canthus. B, Operative plan for Z-plasty technique. C, Postoperative appearance.

W-Plasty and Geometric Broken-Line Closure (GBLC):

- Useful irregularization techniques that often use shorter limbs compared with Z-plasty, and do not create lengthening of the scar (same concept though).
- The human eye will have a harder time following an irregular line versus a straight one.
- The key difference is that W-plasty creates "regular irregularization," and GBLC creates irregular irregularization of the final scar line.
- This technique begins by marking out a series of triangles, in the case of W-plasty, or consecutive shapes, in the case of GBLC.

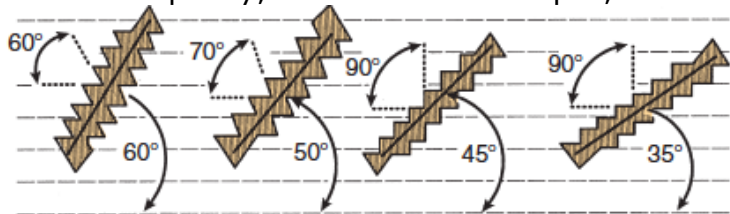
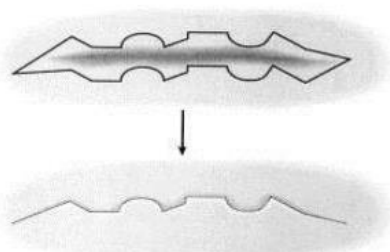
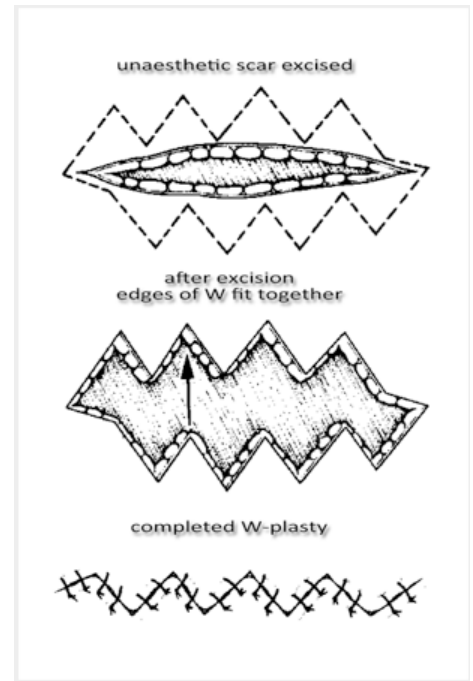


FIGURE 21-8. Running W-plasty with one area of triangle aligned in parallel with relaxed skin tension lines (RSTLs). As scar inclination decreases, the degree of the angles should be increased to keep one arm of the triangle aligned with RSTLs.

- Previously stated principles of wide undermining, low tension closure, and delicate skin-edge suturing are all followed.
- These wounds are also amenable to postoperative dermabrasion to further camouflage the wound.



1



2



3



Figure 2: W-Plasty

- 1=Straight line scar oriented perpendicularly to RSTLs;
- 2=Excision of scar with pattern of interdigitating W's on either side;
- 3=Resultant zig-zag line with interposition of the W's after scar excision

TOPICAL THERAPIES:

Intra-lesional Steroids:

- Adjunct in the treatment of healing wounds, hypertrophic scars, and keloids.
- Injections into areas of scar revision can be useful when persistent tissue edema,
- **Small doses of triamcinolone (10 mg/mL) injected into the dermis or the dermis-subcutaneous junction.**
- Steroids can cause :
 - o Hypopigmentation and telangiectasia when injected in higher concentrations into the dermis.
 - o Injection of steroids into the subcutaneous fat should be avoided, because this can lead to deformity from fat atrophy.

Silicone Sheeting:

- Application of silicone gel sheets to routine scars, as well as hypertrophic and keloid scars.
- Mechanism by which it may improve scar healing is not well characterized.
- **Wear the silicone for a minimum of 8 to 12 hours per day and to use it for 6 to 12 months.**

Onion Extract (*Allium cepa*):

- Anti-inflammatory, bacteriostatic, and collagen downregulatory properties.

DERMABRASION

- Useful for smoothing out surface contour irregularities and for softening the appearance of suture lines after primary closure or irregularization.
- Best performed at the 6- to 8-week interval.
- Diamond fraise bits are preferred.

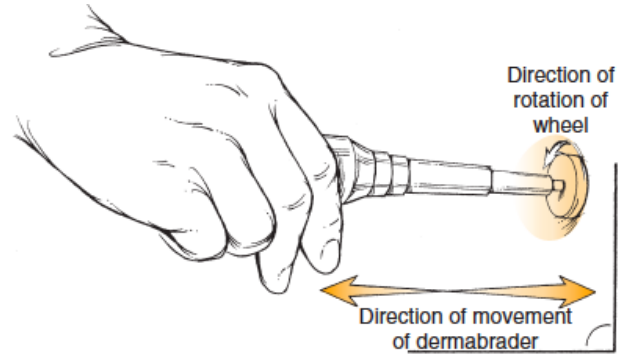
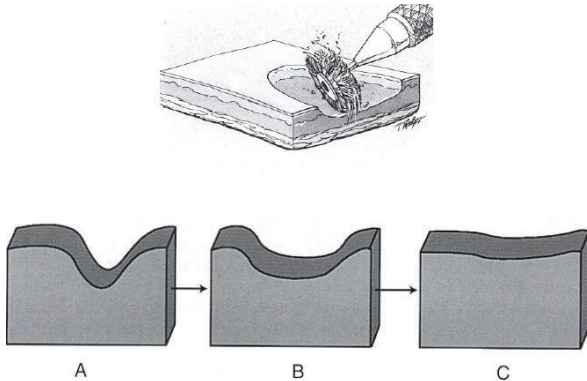
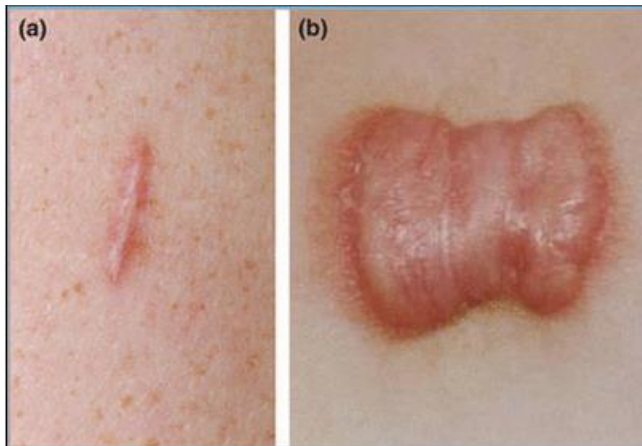


FIGURE 21-11. Dermabrader moved at right angles to the direction of wheel rotation.

- Preservation of the reticular dermis with its adnexal structures allows for the proliferation of undamaged epidermal cells across the abraded surface.
- Occlusive dressing is applied, such as polyethylene oxide hydrogel.
- This is left in place for 1 to 4 days, or until it becomes too soiled and needs to be removed for purposes of hygiene.
- The patient is then instructed to keep the area moist at all times with Fusidine or Bacitracin for the next 7 to 10 days.
- Following this, the patient can use a thick moisturizing lotion.
- Reepithelialization is usually accomplished after 5 to 7 days, but post-treatment erythema can often take 2 to 3 months to resolve.
- Avoid dermabrasion in patients with human immunodeficiency virus or hepatitis because of the risks to health care personnel from airborne pathogen.

HYPERTROPHIC SCARS AND KELOIDS

- Imbalance made up of the inflammatory, proliferative, and remodeling phases.
- Terms are oftentimes used interchangeably, it is important to understand the difference.
- **Hypertrophic** scars are raised, pigmented, do not extend beyond the boundaries of the original wound, and often regress with time.
- **Keloids**, on the other hand, migrate beyond the boundaries of the original wound and do not regress.



Hypertrophic scar	Keloid
Can regress	Does not regress
Oriented collagen	Random eosinophilic collagen
Confined to wound	Not confined
Scant mucin	Mucinous stroma
No myofibroblasts	Myofibroblasts

- In predisposed individuals, they can result from any degree of deep dermal injury such as piercings, traumatic lacerations, surgical incisions.
- Frequently associated with pain, pruritus, and paresthesia.
- Histologically, normal scar tissue displays organized collagen bundles parallel to the epithelial surface
 - o **hypertrophic** scars have a wavier pattern to the collagen fibers with the same epithelial orientation.
 - o **Keloids**, on the other hand, have disorganized collagen fibers with a random orientation to the epithelial surface.

TOPICAL THERAPY FOR HYPERTROPHIC SCARS AND KELOIDS

Pressure Therapy:

- Mechanism of action is unclear.
- However, it is believed to be related to thinning of the dermis, a decrease in edema, and reduction of blood flow that causes a hypoxic environment and decreased collagen synthesis.

Topical Mitomycin C:

- Antineoplastic antibiotic that inhibits fibroblast proliferation and is typically applied to wound beds after keloid excision.

Imiquimod:

- Immunomodulating agent that induces proinflammatory cytokines with an antifibrotic effect and induces apoptotic gene expression in keloidal tissue.

EXCISION AND INJECTABLES:

- Surgical excision allows for a fresh start to the healing process.
- Surgical excision alone has a keloid recurrence rate of **45% to 100%**.
- Combined with Intralesional corticosteroid injections due to their ability to decrease fibroblast proliferation, promote collagen degeneration, induce tissue hypoxia by vasoconstriction, and suppress proinflammatory mediators.
- This combination therapy can improve success rates to 72% to 92%.
- **Surgical excision followed by intralesional steroid therapy has become the mainstay.**

5-Fluorouracil Injections:

- Antimetabolic agent works by inhibiting cell proliferation of keloid fibroblasts by blocking DNA synthesis.
- Following surgical excision and 5-FU injection, keloids had a recurrence rate of 19% at 1-year.
- Side effects associated with 5-FU injection include pain, hyperpigmentation, and ulceration.

Bleomycin:

- Antineoplastic.
- Inhibit collagen synthesis in skin fibroblasts.
- Most feared complications of bleomycin use are pulmonary fibrosis and hepatotoxicity, but the dosages used for keloid treatments produce only local side effects in the form of hyperpigmentation and dermal atrophy.

Interferons

- Antiproliferative, antifibrotic cytokines that have been used as both monotherapy and adjuvant therapy for keloid treatment.

LASER THERAPY

- CO2 laser can be used to excise keloids at their base, much like scalpel excision, or to ablate the tissue.
- Studies have not shown it to be more successful than prior treatment options.
- When used as monotherapy, recurrence rates have been 90% or higher.
- Combined with intraoperative and/or serial postoperative steroid injections in compliant patients, success rates have been closer to 80%.
- Similar to the CO2 laser, the Nd:YAG laser.
- The argon laser has shown high recurrence rates.
- PDL, which targets oxyhemoglobin and traditionally has been used for vascular lesions, has been used for hypertrophic scars and keloids.

Cryotherapy

Radiotherapy

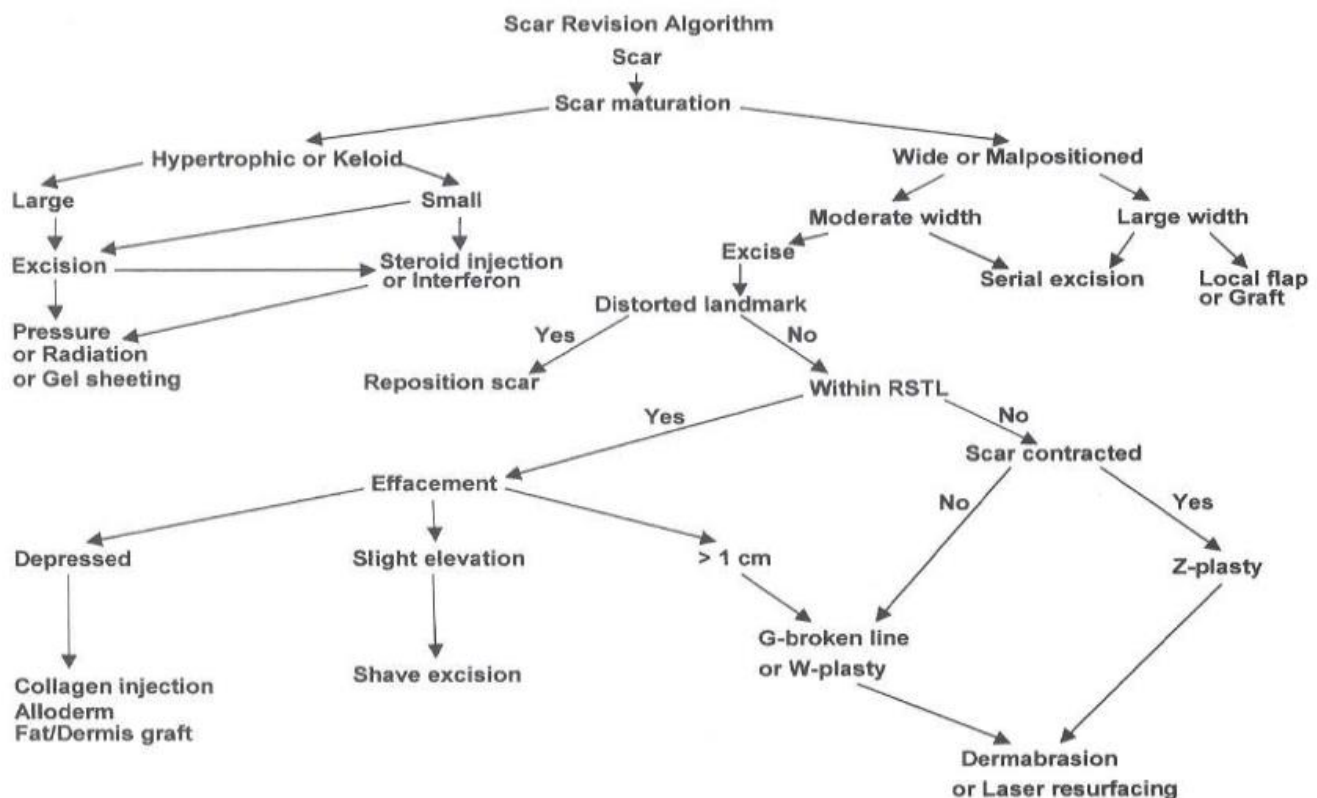
Adjunctive procedures (cosmetics and hairstyling)

Surgical Techniques

- Excision
- Z-plasty
- W-plasty
- Geometric broken line closure

Excisional Techniques

- Simple Excision
- Serial Excision
- Shave excision



Six considerations to minimize scar tissue formation when repairing facial laceration:

1. Tissue eversion
2. Patient's propensity for keloid/hypertrophic scar
3. Patient's natural complexion, eg. light vs. dark skin
4. Minimal tension on incision line
5. Clean wound, avoidance of infection
6. Absorbable vs. Nonabsorbable sutures, mono vs. multifilament